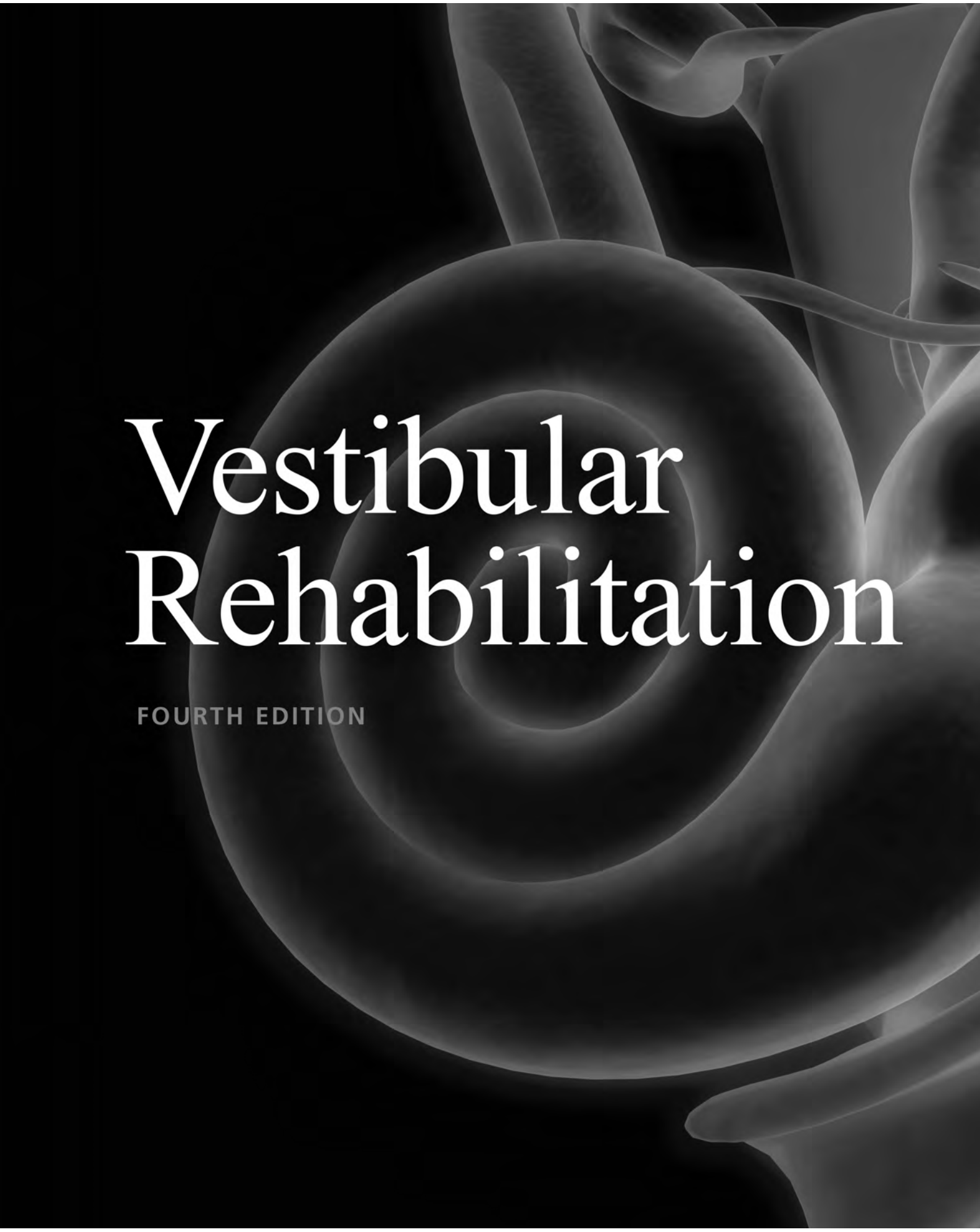
Susan J. Herdman

Richard A. Clendaniel

Vestibular Rehabilitation

FOURTH EDITION



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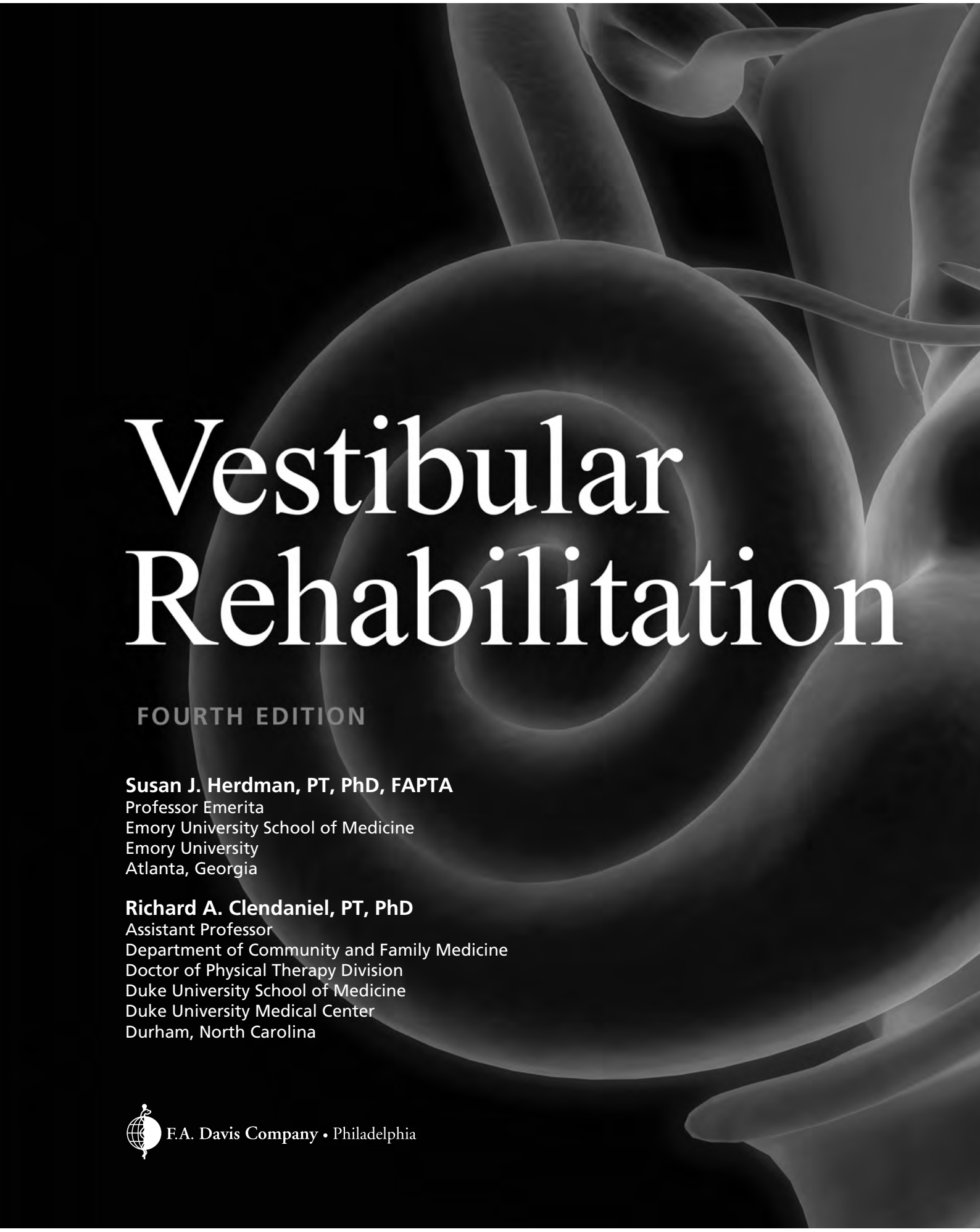

Vestibular Rehabilitation

FOURTH EDITION

Susan J. Herdman, PT, PhD, FAPTA
Professor Emerita
Emory University School of Medicine
Emory University
Atlanta, Georgia

Richard A. Clendaniel, PT, PhD
Assistant Professor
Department of Community and Family Medicine
Doctor of Physical Therapy Division
Duke University School of Medicine
Duke University Medical Center
Durham, North Carolina

F.A. Davis Company • Philadelphia

F. A. Davis Company
1915 Arch Street
Philadelphia, PA 19103
www.fadavis.com

Printed in the United States of America

Last digit indicates print number: 10 9 8 7 6 5 4 3 2 1

Senior Acquisitions Editor: Melissa A. Duffield
Manager of Content Development: George W. Lang
Developmental Editor: Peg Waltner
Art and Design Manager: Carolyn O'Brien

Library of Congress Control Number: 2014944299

This book is dedicated to "serendipity." The serendipity that led to me walking into a patient's room one weekend and finding there the person who would become my doctoral advisor. The serendipity that led to that surprising day when, after years of saying "no" to treating patients for their dizziness, I saw my first patient with BPPV and was forever "hooked" by that patient's gratitude. The serendipity that led to me working at Hopkins and NIH. The serendipity that led to four colleagues sitting down and planning a competency-based course in Vestibular Rehabilitation that we thought would last maybe 3 or 4 years. The serendipity that put so many wonderful opportunities in my life. The serendipity that put so many incredible people in my life. The grace that led me to take advantage of it all.
SJH

I would like to dedicate this book to my family who has supported me throughout, to my colleagues who have contributed to this book and to my education, and to my friend, colleague, mentor, and coeditor whose knowledge, guidance, and inspiration have been critical to my professional growth.
RAC

Foreword

Benjamin Franklin once said, "Life's tragedy is that we get old too soon and wise too late." While there is substantial wisdom in that statement, I can think of at least one exception that might set old Ben on his rear, or is it ear? It is hard to believe that 20 years have passed since the first edition of *Vestibular Rehabilitation* was published. Yet even at its birth there was considerable wisdom in its words. At that time the responsibility for evaluating and rehabilitating patients presenting with dizziness and vestibular compromise was restricted to a select few, and this area of rehabilitation had not emerged as a specialty. Susan Herdman recognized a need to train rehabilitation clinicians to better understand and treat the postural compromise generated through visual deficits, headache or other maladies that would adversely impact the quality of life of those unfortunate enough to have acquired vestibular pathologies. The extent to which this knowledge has grown over these two decades and with it, the interest of students and clinicians is truly extraordinary. Throughout this time *Vestibular Rehabilitation* has remained the "go to" text for specialists in this area while also serving as a critical reference source for all neurorehabilitationists.

All contributors to the *Contemporary Perspectives in Rehabilitation* series have always prided themselves on maintaining the infrastructural integrity of its foundation…….evidence based referencing, problem solving presentations and the latest and most novel data or treatment techniques. *Vestibular Rehabilitation* continues to adhere to these principles and undoubtedly the result has been a steadfast allegiance to its content from amongst its followers…….teachers, clinicians and students. The fact that the content has been updated consistently at 5 year intervals speaks to the dedication shown by its contributing authors, many of whom have persisted through several editions. Throughout this time, Dr. Herdman has amassed a larger international cohort of knowledgeable clinicians, many of whom have come to assist her in what arguably might be called the most popular (and intense) vestibular rehabilitation course in the United States, if not the world. Many of those once considered novices are now positioned to disseminate information themselves. Undoubtedly, Dr. Herdman has assembled the next generation of contributors. Amongst the 34 individuals whose collective efforts define the 4th edition of *Vestibular Rehabilitation*, 14 are first timers and all possess the skill and knowledge to become persistent contributors to subsequent editions. As one reads Chapters 3 (Laurie King), 6 (Anne Galgon), 7 (Natalia Ricci), 13 (Steve Benton), 14 (Rosalyn Schneider), 17 (Yew Ming Chan), 19 (Jeffrey Staab), 20 (Jeff Hoder), 24 (Jennifer Braswell Christy), 26 (Laura Morris and Kim Gottshall), 28 (Courtney Hall and Dara Meldrum) and 30 (Lisa Heusel-Gillig and Courtney Hall), please know that while these individuals might not be the sole author of those respective contributions, their effort to revise previous content was substantial and the resulting quality of their work is outstanding.

In this edition, all chapters have been updated and new cases permeate the management chapters to illustrate how patients with specific problems are treated. The thought-provoking and problem solving nature of the clinical chapters are supported by the necessity to foster evidence based references, a fundamental tenet of all volumes in the *Contemporary Perspectives in Rehabilitation* series. While the 3rd edition of Vestibular Rehabilitation was accompanied by 68 videos, this edition has over 100. The videos include normal and abnormal eye movements, assessment procedures, demonstrations of some exercises, and gait assessments. Chapters emphasizing tinnitus, novel approaches in the assessment and treatment of anterior and horizontal canal benign paroxysmal positional vertigo (BPPV), and data on outcomes in patients with unilateral and bilateral vestibular hypofunction are distinctly new to the 4th edition. The management of patients with chronic subjective dizziness represents another addition to the text. These are patients who present with chronic dizziness and motion sensitivity that often is accompanied by variable amounts of anxiety and phobic behavior.

The text has been further contemporized through the addition of a chapter on the management of patients with vestibular problems precipitated from head trauma

including etiologies from post-concussion syndrome caused by blast injuries. Regrettably there is ample reason to believe that the incidence of this problem may be profoundly underestimated. If so, then this category of patient will present unique challenges to vestibular rehabilitation clinicians and the treatment components may well incorporate a need to foster compliance because of concomitant behavioral changes. Additional information is contained in information regarding emerging technologies for the treatment of patients with vestibular disorders. Such rehabilitation includes novel biofeedback alternatives, use of virtual reality and gaming, such as the Wii. These advances, while challenging, provoke opportunities to discover and implement new approaches towards enlisting functional plasticity to achieve enhanced quality of life. We can be rest assured that advances in technologies and devices will continue to make their way into the armamentarium of tools to restore optimal balance, reduce dizziness or improve the consequences of migraine, as examples.

The contributions from vestibular neurorehabilitation therapists and specialty physicians are woven along a highly integrated network. Over each edition, the blend of input from these specialists becomes more tightly coupled. This fluidity may go unrecognized by students, but suffice to say, there are very few courses, let alone text books, in which the content amongst different specialists, both physician and non-physician, can be assembled and transmitted so smoothly. Herein rests an opportunity to learn from a panel of experts who would be very difficult to assemble collectively. As I concluded in a previous Foreword, the sum of these parts is truly greater than its whole……

Steven L. Wolf, Ph.D., PT, FAPTA, FAHA
Series Editor

Preface

There are several additions to the fourth edition of *Vestibular Rehabilitation*. One change is the addition of Richard A. Clendaniel as the coeditor. Rick is a clinician, educator, and researcher with many years of experience in vestibular rehabilitation, vestibular function testing, and vestibular physiology. His contributions to the field have been of benefit to many clinicians and researchers as well as to patients. The other changes in this edition of *Vestibular Rehabilitation* are the exploration of new areas of "vestibular" rehabilitation including current evidence that supports the use of the gaze stabilization exercises in patients with non-vestibular dizziness and the use of new technologies in exercise programs.

The practice of vestibular rehabilitation faces numerous challenges in the coming years, as does all health care. We must be more efficient in our assessments and treatments. We must provide evidence that the patient is improving through the application of functional outcome measures. We must be able to defend our choice of treatment based on research establishing the benefits of specific exercises or establishing that a specific exercise is not beneficial. We must be able to support our recommendation on the need for further treatment based on measures that suggest the patient has or has not reached the optimal level of recovery. We need to address the psychological state of the patient as well as the physical problems and their consequences.

As you should hope, the evidence supporting the treatment of patients with vestibular deficits has increased since the last edition 7 years ago. So once again, we have extended the material presented to include several new chapters to reflect and to challenge our understanding of the assessment and treatment of vestibular disorders. One new chapter is on management of patients with chronic subjective dizziness and is a nice complement to the chapter of psychological problems in patients with dizziness. Another chapter tackles a current "hot topic"—management of patients with vestibular dysfunction from concussion. Finally, a third new chapter explores the role of emerging technologies such as virtual reality, sensory substitution devices, and most excitedly, vestibular implants, which will undoubtedly require new treatment approaches. Of course, all chapters contain new material, from management of tinnitus to the newer treatments that have been proposed for anterior and horizontal semicircular canal BPPV. We have added "point and counterpoint" sections in some chapters that highlight the differing opinions about assessment and treatment. The number of videos has been increased to include more examples of eye movement abnormalities and to provide visual examples of some of the newer treatments.

The many chapters in this book are designed to provide you with a foundation for all these challenges, but more importantly, the chapters will hopefully give you the basis from which you can continue to apply new information in your practice for the betterment of all your patients.

Contributors

Steve Benton, AuD
Tinnitus Program Manager
Staff Audiologist
VA Medical Center
Decatur, GA

Thomas Brandt, MD, FRCP
Professor, Clinical Neurosciences
German Center for Vertigo and Balance Disorders
Ludwig-Maximilians-University
Munich, Germany

Yew Ming Chan, FRCS Edinburgh, FAMS
Senior Clinical Lecturer
Department of Otolaryngology
Yong Loo Lin Medical School,
Singapore Senior Otolaryngologist Consultant
Department of Otolaryngology
Singapore General Hospital

Jennifer Braswell Christy, PT, PhD
Associate Professor
Department of Physical Therapy
The University of Alabama at Birmingham
Birmingham, AL

Helen S. Cohen, EdD, OTR, FAOTA
Professor, Bobby R. Alford Department of Otolaryngology – Head and Neck Surgery
Baylor College of Medicine
Houston, TX

Ian S. Curthoys, PhD
Professor, School of Psychology
Director, Vestibular Research Laboratory
The University of Sydney, NSW
Sydney, Australia

Marianne Dieterich, MD, FANA
Professor, Department of Neurology
University Hospital
Ludwig-Maximilians University of Munich
Munich, Germany

Michael Fetters, MD
Associate Professor and Chair
Department of Neurology
SRH Klinikum Karlsbad-Langensteinbach
Teaching Hospital of University of Heidelberg
Karlsbad, Germany

Anne K. Galgon, PT, PhD, NCS
Assistant Professor, Department of Physical Therapy
College of Health Professions and Social Work
Temple University
Philadelphia, PA

Kim Gottshall, PT, PhD, ATC
Director of Vestibular Assessment and Rehabilitation
San Diego Naval Medical Center
Dan Diego, CA

Timothy C. Hain, MD
Professor (Emeritus)
Private Practice
Chicago Dizziness and Hearing
Chicago, IL

Courtney D. Hall, PT, PhD
Research Health Scientist
Audiology and Vestibular Research Enhancement Award Program
Mountain Home VA Medical Center
Mountain, Home, TN
and
Associate Professor
Department of Physical Therapy
East Tennessee State University
Johnson City, TN

G. Michael Halmagyi, MD, FRACP
Professor, The Department of Neurology
Director, Eye and Ear Research Unit
Royal Prince Alfred Hospital
Sydney, NSW, Australia

Janet Helminski, PT, PhD
Professor, Department of Physical Therapy
College of Health Sciences
Midwestern University
Downers Grove, IL

Lisa Heusel-Gillig, PT, DPT, NCS
Adjunct Faculty Member, DPT Program
Emory University School of Medicine
Senior Physical Therapist
Emory Dizziness and Balance Center
Emory University Healthcare
Atlanta, GA

Jeffrey M. Hoder, PT, DPT, NCS
Assistant Professor, Department of Neurology
Virginia Commonwealth University
Richmond, VA
and
Program Director of Rehabilitation and Wellness
VCU Parkinson's and Movement Disorders Center
Richmond, VA

Fay Bahling Horak, PT, PhD
Professor of Neurology
Adjunct Professor of Bioengineering
Oregon Health and Science University
Portland, OR

Emily A. Keshner, PT, EdD
Professor and Chair, Department of Physical Therapy
College of Health Professions and Social Work
Temple University
Philadelphia, PA

Laurie A. King, PT, PhD
Assistant Professor, Neurology
Balance Disorders Laboratory
Oregon Health and Science University
Portland, OR

Rob Landel, PT, DPT, OCS, CSCS, FAPTA
Professor, Clinical Physical Therapy
Director, Doctor of Physical Therapy Program
Director, Clinical Residency
Programs Division of Biokinesiology and Physical Therapy at
Herman Ostrow School of Dentistry
University of Southern California
Los Angeles, CA

R. John Leigh, MD
Blair-Daroff Professor of Neurology
Case Western Reserve University
Cleveland, OH

Gregory F. Marchetti, PT, PhD
Associate Professor
Department of Physical Therapy
Rangos School of the Health Professions
Duquesne University
Pittsburgh, PA

Douglas E. Mattox, MD
William Chester Warren Jr., MD Professor of Otolaryngology
Chair, Department of Otolaryngology – Head & Neck Surgery
Emory University School of Medicine
Atlanta, GA

Dara Meldrum, PT, MSc, PhD
Lecturer
School of Physiotherapy
Royal College of Surgeons in Ireland
Dublin, Ireland

Laura Morris, PT, NCS
Physical Therapist Specialist, Mild Brain Injury program
Sinai Hospital of Baltimore
Baltimore, MD

Sharon H. Polensek, MD, PhD
Chief, Audiology and Speech Pathology
Atlanta VA Medical Center
Decatur, GA
and
Assistant Professor of Neurology
Dizziness and Balance Center
Emory University Hospital Midtown
Atlanta, GA

Natalia A. Ricci, PT, PhD
Division of Otoneurology, Department of Otolaryngology-Head and Neck Surgery,
Federal University of Sao Paulo
Sao Paulo, SP, Brazil

Rose Marie Rine, PT, PhD
Associate Professor
Marshall University School of Physical Therapy
Huntington, WV
and
President, Specialty Therapy Source LLC
Jacksonville, FL

Rosalyn Schneider, BA
Research Assistant
Department of Neurology
Case Western Reserve School of Medicine
Cleveland, OH

Michael C. Schubert, PT, PhD
Associate Professor, Departments of Otolaryngology Head and Neck Surgery and Physical Medicine and Rehabilitation
Johns Hopkins University School of Medicine
Baltimore, MD

Neil T. Shepard, PhD
Professor of Audiology
Mayo Clinic College of Mediicne
Chair of Audiology and
Director of Dizziness and Balance Disorders Program
Mayo Clinic
Rochester, MN

Jeffrey P. Staab, MD, MS
Associate Professor of Psychiatry
Mayo Clinic College of Medicine
Department of Psychiatry and Psychology
Mayo Clinic
Rochester, MN

Ronald J. Tusa, MD, PhD
Professor, Departments of Neurology and Otolaryngology-Head and Neck Surgery, Emory University School of Medicine
Director, Dizziness and Balance Center, Emory Midtown Hospital, Atlanta, GA

Susan L. Whitney, DPT, PhD, NCS, ATC, FAPTA
Professor, Departments of Physical Therapy and Otolaryngology
University of Pittsburgh
Pittsburgh, PA

Reviewers

Heather D. Anderson, PT, DPT, NCS
Assistant Professor
Neumann University
Aston, PA

Susan Barker, PT, PhD
Professor
Misericordia University
Dallas, PA

Richard W. Bohannon, EdD, DPT, NCS, FAPTA, FAHA, FASNR, CEEAA
Professor
University of Connecticut
Storrs, CT

Yi-Po Chiu, PT, PhD
Assistant Professor
Louisiana State University Health Sciences Center
New Orleans, LA

Carolyn K. Craven, PT, DPT
Clinical Assistant Professor
University of Oklahoma Health Sciences Center
Oklahoma City, OK

Cheryl Ford-Smith, PT, DPT, MS, NCS
Associate Professor
Virginia Commonwealth University
Richmond, VA

Neeraj Kumar, MS, PT, PhD
Assistant Professor
Texas Tech University Health Sciences Center
Odessa, TX

Barbara S. Robinson, PT, DPT
Associate Professor
Missouri State University
Springfield, MO

Rachel D. Trommelen, PT, DPT, NCS
Assistant Professor
Louisiana State University Health Sciences Center
New Orleans, LA

Acknowledgments

I want to share with anyone who finds difficulty in mastering the information in this book and mastering how to treat people with "dizziness" that it's not unusual to start with a very real sense of inadequacy! I will always remember that during my early attempts to learn about "vestibular rehabilitation" I visited Fay Horak and was mystified as to how everyone knew that the patient had a central rather than a peripheral vestibular deficit. If it's any encouragement, I realize that I have come a "long way" since then but I would also add that I am still learning and I hope I always will be. And so, I would like to express my gratitude again to the many people it has been my good fortune to meet, work with, teach, treat, and learn from over my years as a physical therapist. These have included outstanding scientist-clinicians in the field of vestibular rehabilitation (many of whom have authored chapters in this book), teachers and clinicians from all over the world, students, and of course, patients. Thank you all so much.

SJH

I would like to express my sincere gratitude to the authors who shared their knowledge and expertise in this edition of *Vestibular Rehabilitation*; it is their contributions that make this text such a valuable resource for understanding and treating the patient with dizziness. I would like to also thank the patients, clinicians, and students who question and challenge what we do, forcing us to rethink our assumptions, to continue to learn, and continue to investigate the "best" treatments for individuals suffering from vestibular disorders. I have been blessed to learn from some of the very best, and recognizing all these individuals by name would fill many pages. So I would simply like to offer my thanks to all who have taught me over the years, to fortuitous meetings on Nantucket, and to an atypical insight that kept me from asking a really stupid question. Lastly, an immense 'thank you' to my family for their love, support and encouragement over the years.

RAC

Contents in Brief

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ONE

Fundamentals of Function

CHAPTER 1

Anatomy and Physiology of the Normal Vestibular System

Timothy C. Hain, MD ■ *Janet Helminski, PhD, PT*

Purpose of the Vestibular System

The human vestibular system estimates body position and motion. Motion inputs to the vestibular system include the inner ear signals ("vestibular" in Fig. 1.1), as well as position sensation, ("proprioception") visual signals, and intended movement ("motor commands"). These redundant inputs are integrated by the central processor, the "vestibular nuclear complex," which generates motor commands to drive the eyes and body. The system is normally very accurate. To maintain accuracy, the vestibular system is monitored and calibrated by the cerebellum.

The eye and body movement output of the central vestibular system is generally described in terms of three simple reflexes, the vestibulo-ocular reflex (VOR), the vestibulocollic reflex (VCR), and the vestibulospinal reflex (VSR). The VOR generates eye movements that enable clear vision while the head is in motion. The VCR acts on the neck musculature to stabilize the head. The VSR generates compensatory body movement to maintain head and postural stability and thereby prevent falls. Although the nomenclature of these reflexes, for example "vestibular ocular reflex," might make one think that these circuits are only concerned with inner ear input, the vestibular nucleus that drives all these reflexes also processes input from the other sources listed above.

After an acute loss of peripheral vestibular function, as for example after surgical removal of a tumor of the vestibular nerve, postural and oculomotor deficits appear. With the head still, spontaneous jumping of the eyes (nystagmus) and tilting of the body away from upright appear. When the head is moved, vision and balance are further impaired. Associated with these deficits are a reduced propensity to move the head as well as behavioral changes aimed at minimizing the risk of disorientation by avoiding visual input and minimizing the risk of falling by adopting a more cautious and stable stance.

Recovery from vestibular lesions has been studied for over 100 years.[1] Orientation in space and being able to walk upright are critical functions. It is understandable that the vestibular system is supported by multiple vestibular repair mechanisms. The capability for repair and adaptation is remarkable! Plasticity consists of neural adjustments that restore original function. This is supplemented by substitution of other sensory input or internal estimates. Finally, one may change one's behavior to "work around" problems presented by a vestibular lesion.

Given sufficient time, persons with up to approximately 50% loss of vestibular function adapt so well that a casual observer may find them indistinguishable from someone without a vestibular lesion. Nevertheless, such persons can rarely attain the same degree of performance as normal, and a sophisticated clinician can nearly always detect this situation.

In this chapter, we describe the anatomy and neurophysiology of the vestibular system, paying particular attention to aspects relevant to rehabilitation. We proceed

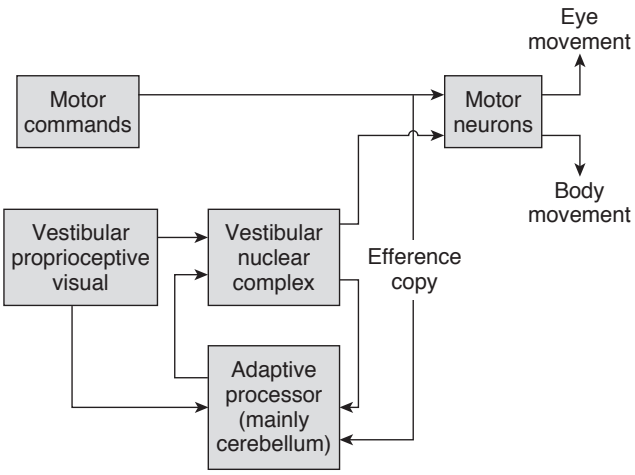

Figure 1.1 Block diagram illustrating the organization of the vestibular system. (Copyright Timothy C. Hain, MD.)

from the peripheral to central structures and conclude with a discussion of "higher-level" problems in vestibular neurophysiology, which are relevant to rehabilitation.

The Peripheral Sensory Apparatus

Figure 1.2 illustrates the peripheral vestibular system, which lies within the labyrinth of the inner ear. The labyrinth, subdivided into the bony and membranous labyrinths, contains the hair cells, which are the motion receptors of the vestibular system. The labyrinth is bordered laterally by the air-filled middle ear and medially by the temporal bone and is posterior to the cochlea.

Bony Labyrinth

The bony labyrinth consists of three semicircular canals (SCCs), the cochlea, and a chamber between the two called the vestibule (Fig. 1.3). The bony labyrinth is filled with perilymphatic fluid, which has chemistry similar to cerebrospinal fluid (high Na:K ratio). Perilymphatic fluid communicates via the cochlear aqueduct with cerebrospinal fluid. Because of this communication, disorders that affect spinal fluid pressure (such as lumbar puncture) can also affect inner ear function.[2]

Membranous Labyrinth

The membranous labyrinth (Fig. 1.3) is suspended within the bony labyrinth by perilymphatic fluid and supportive connective tissue. It contains five sensory organs: the membranous portions of the three semicircular canals and the two otolith organs, the utricle and saccule. Note that one end of each semicircular canal is widened in diameter to form an ampulla (bottom of Fig. 1.3). This widening will be relevant when we later

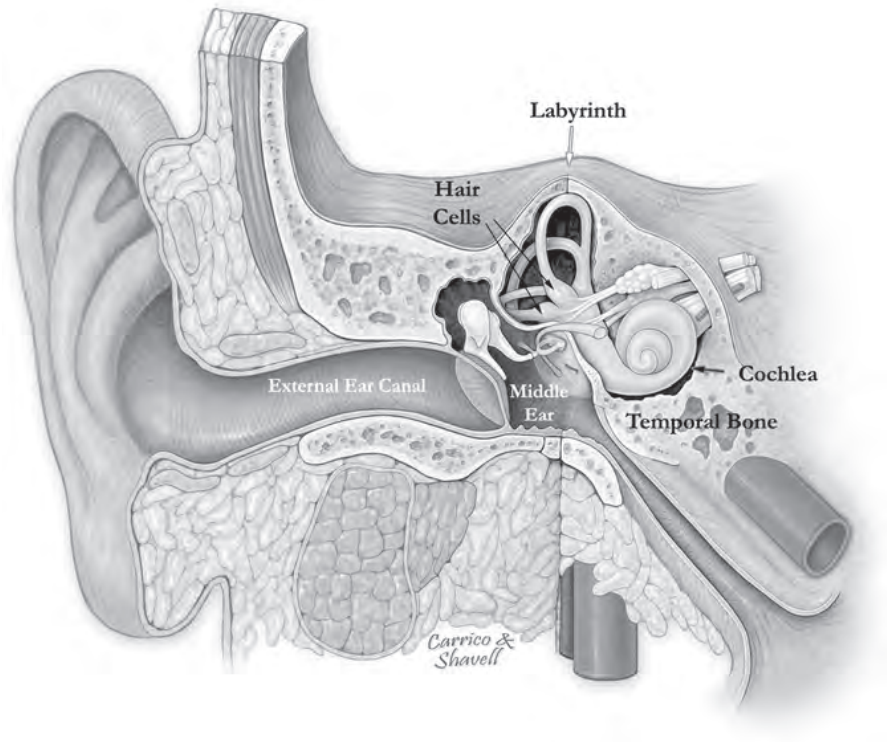

Figure 1.2 Vestibular and auditory apparatus in relation to skull. (Copyright Timothy C. Hain, MD.)

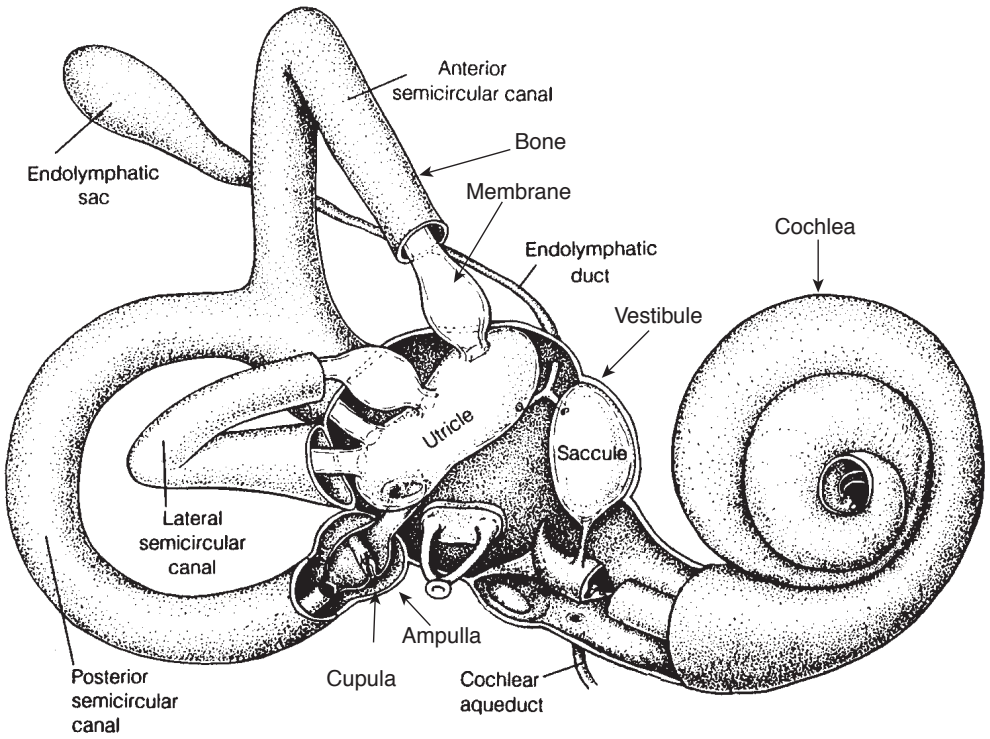

Figure 1.3 The membranous and bony labyrinths. (Adapted from an illustration by Mary Dersch from Pender, 1992.)[2]

discuss a common vestibular condition, benign paroxysmal positional vertigo.

The membranous labyrinth is filled with endolymphatic fluid. In contrast to perilymph, the electrolyte composition of endolymph resembles intracellular fluid (high K:Na ratio). Under normal circumstances, no direct communication exists between the endolymph and perilymph compartments.

Hair Cells

Hair cells contained in each ampulla and otolith organ convert displacement due to head motion into neural firing (Fig. 1.4). The hair cells of the ampullae rest on a tuft of blood vessels, nerve fibers, and supporting tissue called the crista ampullaris. The hair cells of the saccule and utricle, the maculae, are located on the medial wall

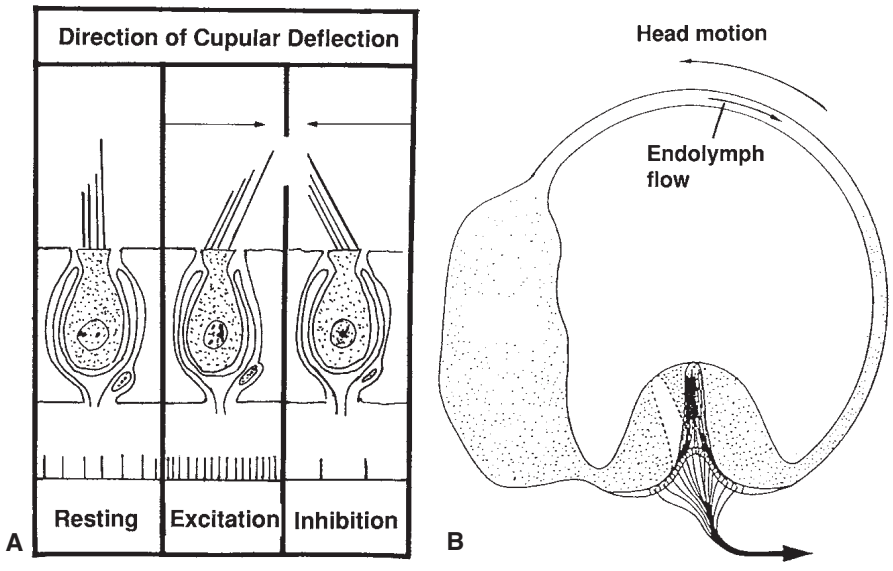

Figure 1.4 Effects of head rotation on the canals. **(A)** The direction from which hair cells are deflected determines whether hair-cell discharge frequency increases or decreases.[3] **(B)** Cross section of the membranous labyrinth illustrating endolymph flow and cupular deflection in response to head motion.[4]

of the saccule and the floor of the utricle. Each hair cell is innervated by an afferent neuron whose cell body lies in the vestibular (Scarpa's) ganglion, which is located close to the ampulla. When hairs are bent toward or away from the longest process of the hair cell, firing rate increases or decreases in the vestibular nerve (see Fig. 1.4*A*). A flexible, diaphragmatic membrane called the cupula overlies each crista and completely seals the ampulla from the adjacent vestibule. With angular head motion, endolymphatic pressure differs across the cupula and causes the cupula to bend back and forth, which stimulates the hair cells (see Fig. 1.4*B*).[4]

The otolithic membranes are structures similar to the cupulae but are weighted. They contain calcium carbonate (limestone) crystals called otoconia and have substantially more mass than the cupulae (Fig. 1.5). The mass of the otolithic membrane causes the maculae to be sensitive to gravity and linear acceleration. In contrast, the cupulae normally have the same density as the surrounding endolymphatic fluid and are insensitive to gravity.[3]

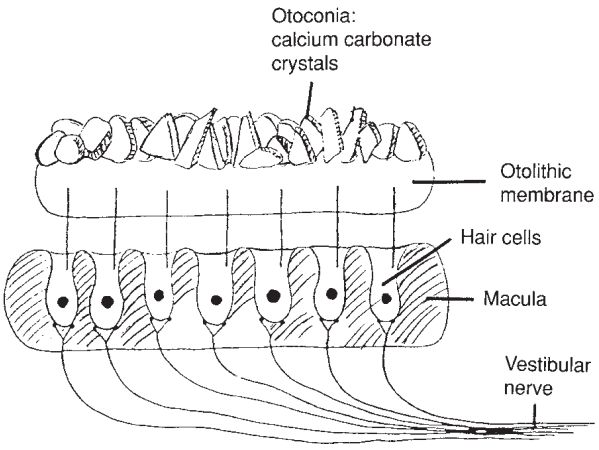

Figure 1.5 The otolithic macula and its overlying membrane.[3]

Vascular Supply

The labyrinthine artery supplies the peripheral vestibular system (Fig. 1.6; see also Fig. 1.11).The labyrinthine artery has a variable origin. Most often it is a branch of the anterior inferior cerebellar artery (AICA—top of 1.6), but occasionally it is a direct branch of the basilar artery. Upon entering the inner ear, the labyrinthine artery divides into the anterior vestibular artery and the common cochlear artery. The anterior vestibular artery supplies the vestibular nerve, most of the utricle, and the ampullae of the lateral and anterior SCCs. The common cochlear artery divides into a main branch, the main cochlear artery, and the vestibulocochlear artery. The main cochlear artery supplies the cochlea. The vestibulocochlear artery supplies part of the cochlea, the ampulla of the posterior semicircular canal, and the inferior part of the saccule.[5]

The labyrinth has no collateral anastomotic network and is highly susceptible to ischemia. Only 15 seconds of

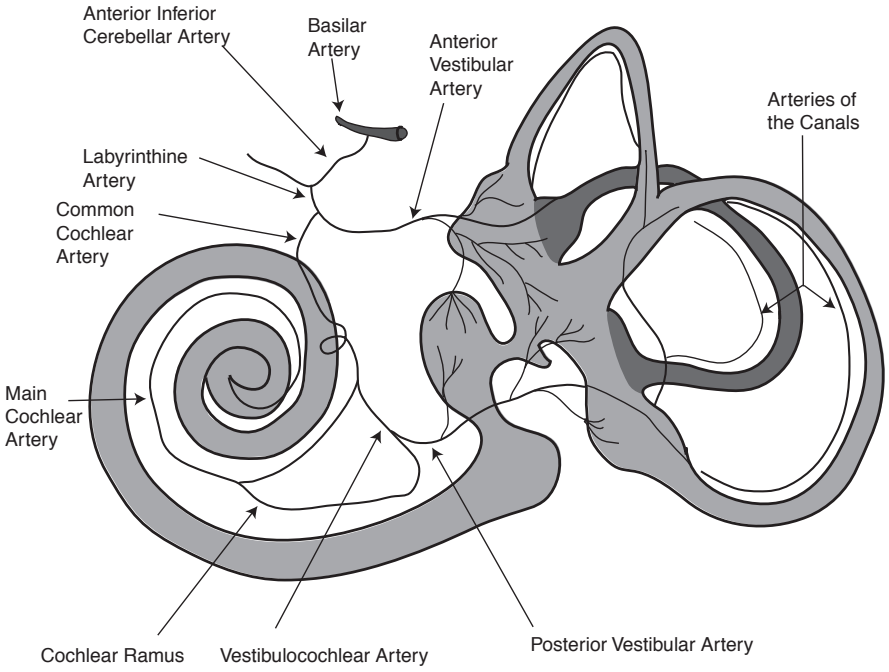

Figure 1.6 The arterial supply of the labyrinth.[5]

selective blood flow cessation is needed to abolish auditory nerve excitability.[6]

Physiology of the Periphery

The hair cells of the canals and otoliths convert the mechanical energy generated by head motion into neural discharges directed to specific areas of the brainstem and the cerebellum. By virtue of their orientation, the canals and otolith organs are able to respond selectively to head motion in particular directions. Because of differences in their fluid mechanics, the canals respond to angular velocity, and the otoliths to linear acceleration.

Semicircular Canals

The SCCs provide sensory input about head velocity, which enables the VOR to generate an eye movement that matches the velocity of the head movement. The desired result is that the eyes remain stationary in space during head motion, enabling clear vision. Neural firing in the vestibular nerve is proportional to head velocity over the range of frequencies in which the head commonly moves (0.5 to 7 Hz). In engineering terms, the canals are "rate sensors."

This fact poses a significant problem: How do the hair cells of the SCCs, which are activated by displacement, produce sensory input proportional to velocity? The labyrinth must have a method of converting head velocity into displacement. Biophysical properties of the semicircular canals' loops accomplish the conversion.[7] The membranous canal loops have very thin walls and a small lumen diameter relative to the radius of the loop curvature. These characteristics make viscous drag on the endolymph very powerful. Viscosity, or fluidic friction, slows down endolymph flow in a way similar to how honey slowly runs down the side of a jar. In a frictionless system with a freely moving cupula, for a step of constant rotational velocity, endolymph displacement would be proportional to velocity times time, or rotational position. The viscosity creates resistance to endolymph movement so that trans-cupular pressure and displacement become more closely proportional to head velocity. Because of these considerations, over the usual frequencies of head movement, endolymph displacement is proportional to angular head velocity, and the SCCs transmit a velocity signal to the brain.

A second important dynamic characteristic of the canals has to do with their response to prolonged rotation at constant velocity. Instead of producing a signal proportional to velocity, the canals respond reasonably well only in the first second or so, because output decays exponentially with a time constant of about 7 seconds. This behavior is due to a springlike action of the cupula that tends to restore it to its resting position.[7]

Three important spatial arrangements characterize the alignment of the SCC's loops. First, each canal plane within each labyrinth is perpendicular to the other canal planes, analogous to the spatial relationship between two walls and the floor of a rectangular room (Fig. 1.7). Second, paired planes of the SCCs between the labyrinths conform very closely to each other. The six individual semicircular canals become three *coplanar pairs*: (1) right and left lateral, (2) left anterior and right posterior, and (3) left posterior and right anterior. Third, the planes of the canals are close to the planes of the extraocular muscles, thus allowing relatively simple connections between sensory neurons (related to individual canals) and motor output neurons (related to individual ocular muscles).

The coplanar pairing of canals is associated with a *push-pull* change in the quantity of SCC output. When angular head motion occurs within their shared plane, the endolymph of the coplanar pair is displaced in opposite directions with respect to their ampullae, and neural firing increases in one vestibular nerve and decreases on the opposite side. For the lateral canals, displacement of the cupula toward the ampulla (ampullopetal flow) is excitatory, whereas for the vertical canals, displacement of the cupula away from the ampulla (ampullofugal flow) is excitatory.

There are three advantages to the push-pull arrangement of coplanar pairing. First, pairing provides sensory

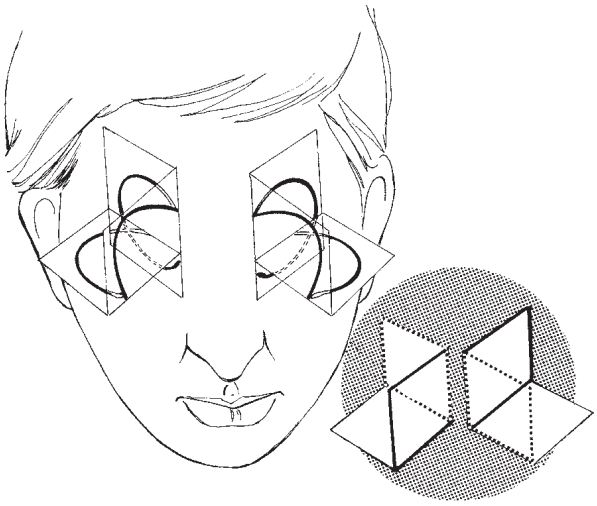

Figure 1.7 The spatial arrangement of the semicircular canals. The canals on each side are mutually perpendicular, are paired with conjugate canals on the opposite side of the head, and also are closely aligned with the optimal pulling directions of the extraocular muscles.

redundancy. If disease affects the SCC input from one member of a pair (e.g., as in vestibular neuritis), the central nervous system will still receive vestibular information about head velocity within that plane from the contralateral member of the coplanar pair.

Second, such a pairing allows the brain to ignore changes in neural firing that occur on both sides simultaneously, such as might occur as a result of changes in body temperature or neurochemistry. These common changes in firing in both nerves are not related to head motion and are called common-mode noise. The engineering term for the desirable feature of ignoring this type of noise is called *common-mode rejection*. Third, as is discussed in a later section, a push-pull configuration assists in compensation for overload.

Otoliths

The otoliths register forces related to linear acceleration (Fig. 1.8). They respond to both linear head motion and static tilt with respect to the gravitational axis. The function of the otoliths is illustrated by the situation of a passenger in a commercial jet. During flight at a constant velocity, we have no sense that we are traveling at 300 miles per hour. However, in the process of taking off and ascending to cruising altitude, we sense the change in velocity (acceleration) as well as the tilt of the plane on ascent. The otoliths therefore differ from the SCC in two basic ways: They respond to linear motion instead of angular motion, and their output is mainly proportional to acceleration rather than velocity.[7]

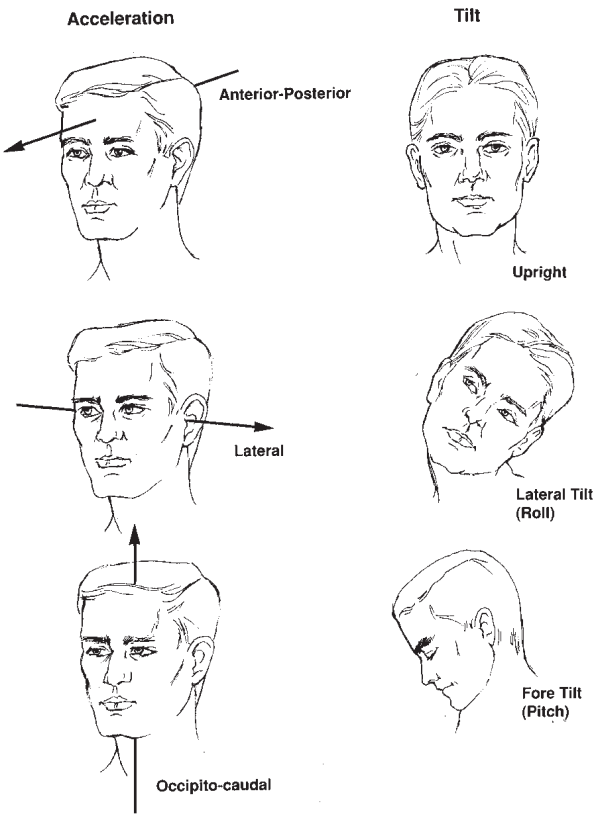

Figure 1.8 The otoliths register linear acceleration and static tilt.

The otoliths have a simpler task to perform than the canals. Unlike the canals, which convert head velocity into cupular displacement through hydrodynamic viscosity, the otoliths need no special hydrodynamic system to do their job. Exquisite sensitivity to gravity and linear acceleration is obtained by incorporating crystals of calcium carbonate (limestone), the otoconia, into the otolithic membrane (see Fig. 1.5). Because force is equal to mass times acceleration, by incorporating a large mass, a given acceleration produces enough shearing force to make the otoliths extremely sensitive. (Shearing force refers to force that is directed perpendicularly to the processes of the hair cells.)

Like the canals, the otoliths respond to motion in all three dimensions (Fig. 1.9). However, unlike the SCC of one ear, which have one sensory organ for each axis of angular motion, there are two sensory organs for three axes of linear motion. In an upright individual, the saccule is vertical (parasagittal), whereas the utricle is horizontally oriented (near the plane of the lateral SCC). The saccule senses linear acceleration in the sagittal plane, such as might be associated with a forward pitch of the head. The utricle senses acceleration in its predominantly horizontal plane, such as might be provoked by a roll (lateral tilt) of the head.[8] The two organs together can encode all possible vectors of linear acceleration.

Because earth's gravitational field is a linear acceleration field, on earth the otoliths register tilt. For example, as the head is tilted laterally (which is also called roll; see Fig. 1.9), shear force is exerted on the utricle, while shear force is lessened on the saccule. Similar changes occur when the head is tilted forward or backward (pitch). Because linear acceleration can come from two sources—earth's gravitational field and linear motion—there is an ambiguity problem. We discuss strategies that the central nervous system might use to solve this problem in our section on higher-level vestibular processing.

For the otoliths, as in the canals, there is redundancy, because there are organs similar in orientation and function located on both sides of the head. Push-pull processing for the otoliths is also incorporated into the geometry of each of the otolithic membranes. Within each otolithic macula, a curving zone, the striola, separates the direction of hair cell polarization on each side. Consequently, head

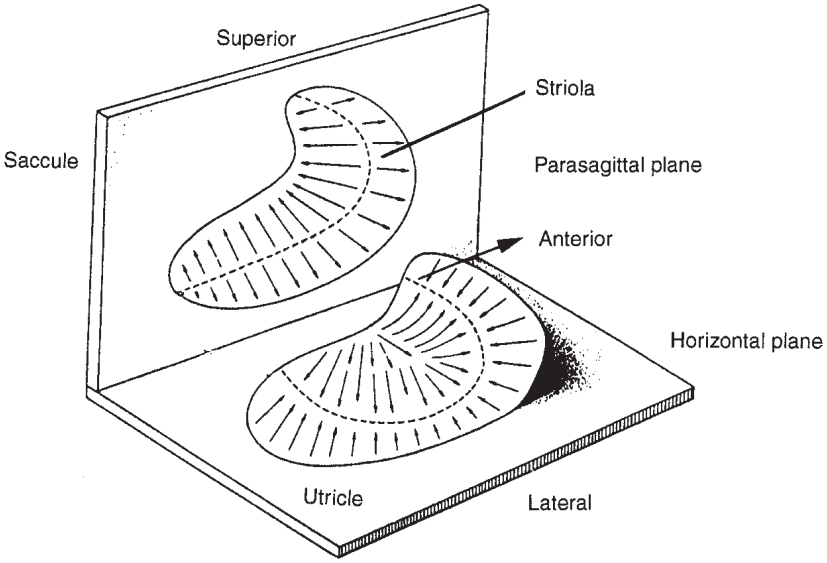

Figure 1.9 Geometry of the otoliths.[8]

tilt results in increased afferent discharge from one part of a macula, while reducing the afferent discharge from another portion of the same macula. This extra redundancy (compared to the SCC) probably makes the otoliths less vulnerable to unilateral vestibular lesions.

The Vestibular Nerve

Vestibular nerve fibers are the afferent projections from the bipolar neurons of Scarpa's (vestibular) ganglion. The vestibular nerve transmits afferent signals from the labyrinths along its course through the internal auditory canal (IAC). In addition to the vestibular nerve, the IAC contains the cochlear nerve (hearing), the facial nerve, the nervus intermedius (a branch of the facial nerve, which carries facial sensation), and the labyrinthine artery. The IAC travels through the dense petrous portion of the temporal bone to open into the posterior fossa at the level of the pons. Note that "petrous" is derived from the Latin word *petrosus*, meaning "stone-like, hard," and thus the inner ear is very well protected! The vestibular nerve enters the brainstem at the pontomedullary junction. Because the vestibular nerve is interposed between the labyrinth and the brainstem, some authorities consider this nerve a peripheral structure, whereas others consider it a central structure. We consider it a peripheral structure.

There are two patterns of firing in vestibular afferent neurons. Regular afferents usually have a tonic rate and little variability in interspike intervals. Irregular afferents often show no firing at rest and when stimulated by head motion develop highly variable interspike intervals.[9] Regular afferents appear to be the most important type for the VOR; irregular afferents can be turned off with electrical stimulation without much change in the VOR of monkeys.[10] Irregular afferents may be important for the VSR and in coordinating responses between the otoliths and canals.

Regular afferents of the monkey have tonic firing rates of about 90 spikes per second and sensitivity to head velocity of about 0.5 spikes per degree per second.[11,12] We can speculate about what happens immediately after a sudden change in head velocity. Humans can easily move their heads at velocities exceeding 300 deg/sec. As noted previously, the SCC are connected in push-pull, so that one side is always being inhibited while the other is being excited. Given the sensitivity and tonic rate noted earlier, the vestibular nerve that is being inhibited should be driven to a firing rate of 0 spikes per second for head velocities of only 180° per second! In other words, head velocities greater than 180° per second may be unquantifiable by half of the vestibular system. This cutoff behavior has been advanced as the explanation for *Ewald's second law*, which says that responses to rotations that excite a canal are greater than responses for rotation that inhibits a canal.[13,14] Cutoff behavior explains why patients with unilateral vestibular loss avoid head motion toward the side of their lesion. More is said about this when we discuss how the central nervous system may compensate for overload.

Central Processing of Vestibular Input

There are two main targets for vestibular input from primary afferents: the vestibular nuclear complex and the cerebellum (see Fig. 1.1). The vestibular nuclear complex

is the primary processor of v estibular input and implements direct, fast connections between incoming afferent information and motor output neurons. The cerebellum is the main adaptive processor—it monitors vestibular performance and readjusts central v estibular processing if necessary. At both locations, vestibular sensory input is processed in association with somatosensory and visual sensory input.

Vestibular Nucleus

The vestibular nuclear complex consists of four major nuclei (superior, medial, lateral, and descending) and at least seven minor nuclei (Fig. 1.10). This large structure, located primarily within the pons, also extends caudally into the medulla. The superior and medial vestibular nuclei are relays for the VOR. The medial vestibular nucleus is also involved in vestibulospinal reflexes and coordinates head and eye movements that occur together . The lateral vestibular nucleus is the principal nucleus for the vestibulospinal reflex. The descending nucleus is connected to all the other nuclei and the cerebellum b ut has no primary outflow of its own. The vestibular nuclei between the two sides of the brainstem are laced together via a system of commissures, which are mutually inhibitory. The commissures allow information to be shared between the two sides of the brainstem and implement the push-pull pairing of canals discussed earlier.[15]

In the vestibular nuclear complex, processing of the vestibular sensory input occurs concurrently with the processing of extravestibular sensory information (proprioceptive, visual, and efferent (see Fig. 1.1). Extensive connections between the vestibular nuclear complex, cerebellum, ocular motor nuclei, and brainstem reticular activating systems are required to formulate appropriately oriented and timed signals to the VOR and VSR effector organs, the extraocular and skeletal muscles.

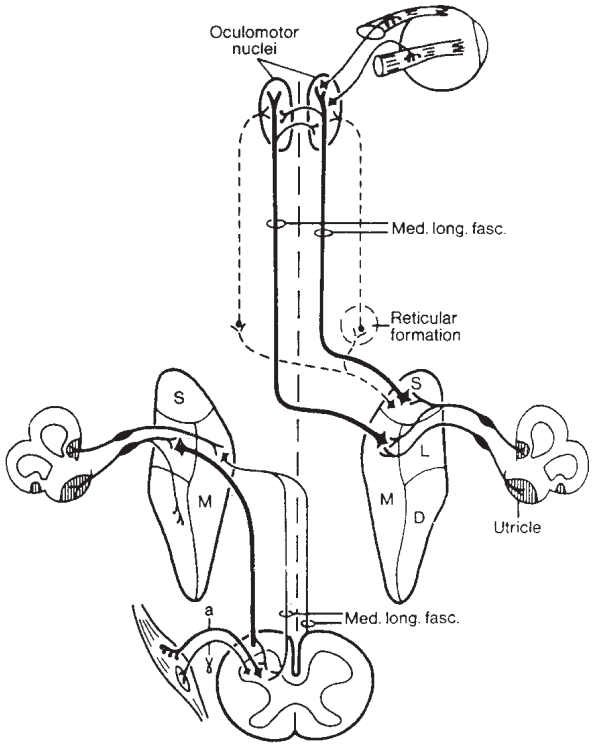

Figure 1.10 The vestibulo-ocular reflex (VOR) and vestibulospinal reflex (VSR) arcs. S, L, M, and D indicate the superior, lateral, medial, and descending vestibular nuclei, respectively. The lateral vestibulospinal and medial vestibulospinal tracts are shown as heavy lines and light lines, beginning in the lateral vestibular nucleus and medial vestibular nucleus, respectively.[15]

Vascular Supply

The vertebral-basilar arterial system supplies blood to the peripheral and central vestibular system (Fig. 1.11). The posterior-inferior cerebellar arteries (PICAs) branch of f the vertebral arteries. The two PICAs are the most important arteries for the central vestibular system. They supply the surface of the inferior portions of the cerebellar hemispheres as well as the dorsolateral medulla, which includes the inferior aspects of the vestibular nuclear complex. The basilar artery is the principal artery of the pons.The basilar artery supplies central vestibular structures via perforator branches (which penetrate the medial pons), short circumferential branches (which supply the anterolateral aspect of the pons), and long circumferential branches (which supply the dorsolateral pons). The AICA is an important branch of the basilar artery because it is the sole blood supply for the peripheral v estibular system via the labyrinthine artery. The AICA also supplies blood to the ventrolateral cerebellum and the lateral te gmentum of the lower two-thirds of the pons.

Recognizable clinical syndromes with v estibular components may appear after occlusions of the basilar artery, labyrinthine artery, AICA, or PICA. PICA territory strokes, also called "lateral medullary syndrome," cause purely central balance symptoms as a result of damage to the vestibular nucleus and inferior cerebellum. AICA territory strokes cause a mixed peripheral/cerebellar pattern because AICA supplies both the labyrinth and part of the cerebellum.

Cerebellum

The cerebellum (Fig 1.12) is a major recipient of outflow from the vestibular nucleus complex and is also a major

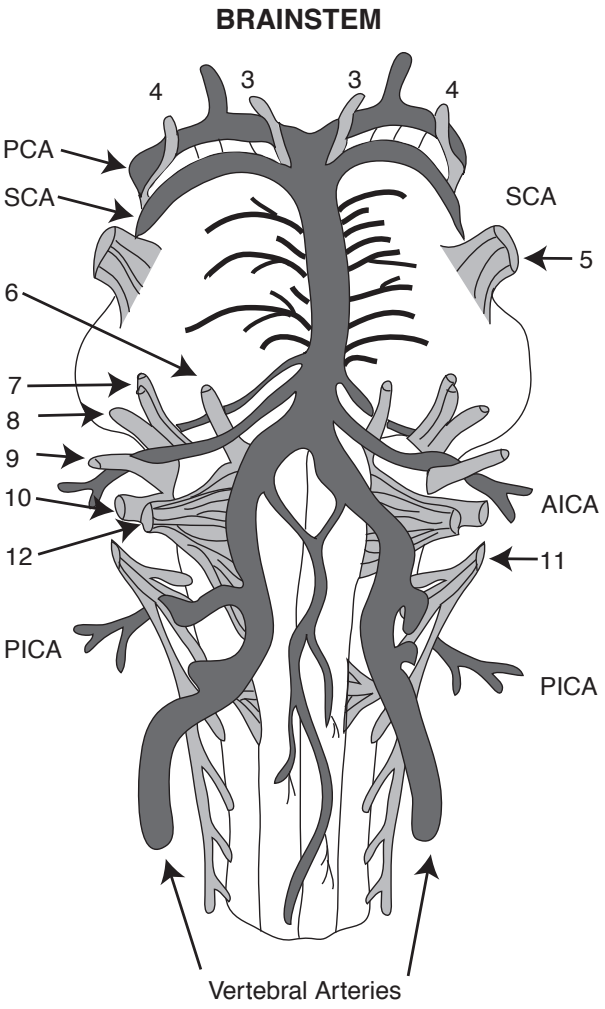

Figure 1.11 The vertebral-basilar system. Numerals indicate individual cranial nerve roots. (All nerves are paired, but, for clarity, both sides are not always labeled here.) AICA = anterior inferior cerebellar artery; PCA = posterior cerebellar artery; PICA = posterior inferior cerebellar artery; SCA = superior cerebellar artery. (Copyright Timothy C. Hain, MD.)

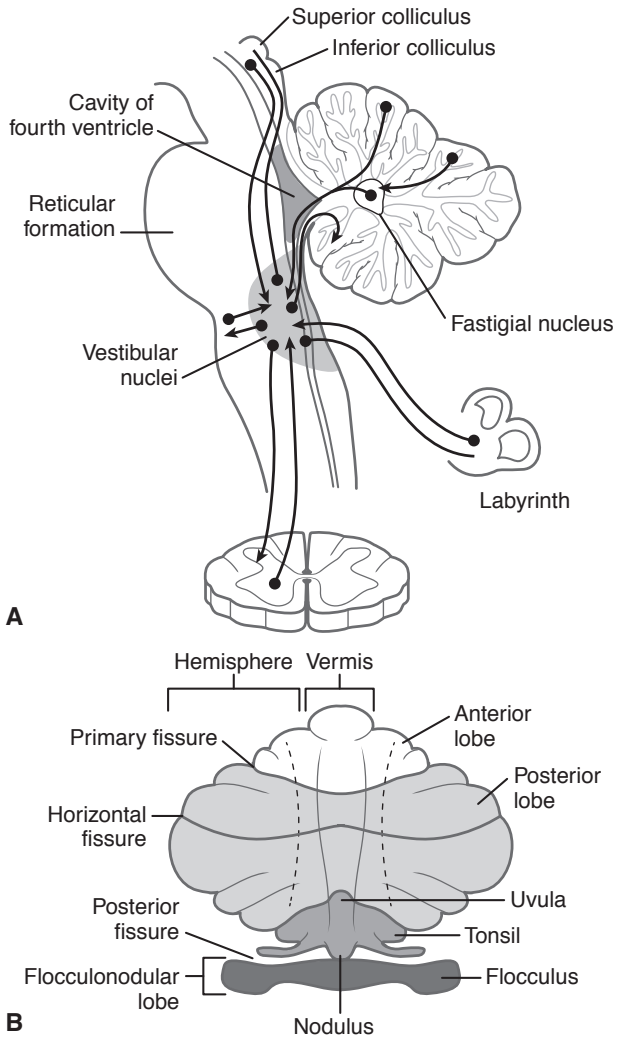

Figure 1.12 The cerebellum and its relationship and connections to vestibular structures. **(A)** Sagittal view. The vestibular nuclei are connected to the cerebellar vermis, the labyrinth, the superior colliculus and spinal cord. **(B)** Posterior view. Most vestibular connections are to the midline structures (vermis, nodulus).

source of input itself. Although not required for vestibular reflexes, vestibular reflexes become uncalibrated and ineffective when the cerebellum is removed. Figure 1.12*A* and *B* shows the relationship of the cerebellum to the vestibular nucleus, labyrinth, spinal cord, and ascending pathways.

The cerebellar flocculus is required to adapt the gain of the VOR.[16] Adaptation is initially learned in the flocculus, maintained in "software," and, after a delay, is downloaded to "hardware"—brainstem flocculus target neurons.[17] Lesions of the flocculus reduce the ability of experimental animals to adapt to disorders that reduce or increase the gain of the VOR. Flocculus disorders are found typically in patients with the Arnold-Chiari Malformation or other cerebellum disorders.

The cerebellar nodulus adjusts the duration of VOR responses and is also involved with processing of otolith input. Patients with lesions of the cerebellar nodulus, such as patients with medulloblastoma, show gait ataxia and often have nystagmus, which is strongly affected by the position of the head with respect to the gravitational axis. The nodulus is required for motion sickness.[18]

Lesions of the anterior-superior vermis of the cerebellum affect the VSR and cause a profound gait ataxia with truncal instability. These patients are unable to use sensory input from their lower extremities to stabilize their

posture. These lesions are often related to excessive alcohol intake and thiamine deficiency.

Neural Integrator

Thus far, we have discussed processing of velocity signals from the canals and acceleration signals from the otoliths. These signals are not suitable for driving the ocular motor neurons, which need a neural signal encoding eye position. The transformation of velocity to position is accomplished by a brainstem structure called the neural integrator. The nucleus prepositus hypoglossi, located just below the medial vestibular nucleus, appears to provide this function for the horizontal oculomotor system.[19] Although a similar structure must exist for the connections with the vestibulospinal system,[20] the location of the VSR neural integrator is presently unknown. Clinically, poor function of the oculomotor neural integrator causes gaze-evoked nystagmus.

Motor Output of the Vestibular System Neurons

Output for the Vestibulo-ocular Reflex

The output neurons of the VOR are the motor neurons of the ocular motor nuclei, which drive the extraocular muscles. The extraocular muscles are arranged in pairs, which are oriented in planes very close to those of the canals (see Fig. 1.7). This geometrical arrangement enables a single pair of canals to be connected predominantly to a single pair of extraocular muscles. The result is conjugate movements of the eyes in the same plane as head motion.

There are two white matter tracts that carry output from the vestibular nuclear complex to the ocular motor nuclei. The ascending tract of Deiters carries output from the vestibular nucleus to the ipsilateral abducens nucleus (lateral rectus) during the horizontal VOR. All other VOR-related output to the ocular motor nuclei is transmitted by the medial longitudinal fasciculus (MLF) (see Fig. 1.12). Because the MLF is often injured in multiple sclerosis, this connection may account for central vestibular symptoms in these patients (see Fig. 1.12).[15]

Output for the Vestibulospinal Reflex

The output neurons of the VSR are the anterior horn cells of the spinal cord gray matter, which drive skeletal muscle. However, the connection between the vestibular nuclear complex and the motor neurons is more complicated than for the VOR. The VSR has a much more difficult task than the VOR because there are multiple strategies to prevent falls, which involve different motor synergies. For example, when shoved from behind, one's center of gravity might become displaced anteriorly. To restore "balance," one might (1) plantarflex at the ankles, (2) take a step, (3) grab for support, or (4) use some combination of all three activities. The VSR also has to adjust limb motion appropriately for the position of the head on the body (see the frame of reference problem discussed in the section on higher-level problems in vestibular processing). The VSR must also use otolith input, reflecting linear motion, to a greater extent than the VOR. Although the eyes can only rotate and thus can do little to compensate for linear motion, the body can both rotate and translate.

There are three major white matter pathways that connect the vestibular nucleus to the anterior horn cells of the spinal cord. The lateral vestibulospinal tract originates from the ipsilateral lateral vestibular nucleus, which receives the majority of its input from the otoliths and the cerebellum (see Fig. 1.10). This pathway generates antigravity postural motor activity or protective extension, primarily in the lower extremities, in response to the head position changes, which occur with respect to gravity. The medial vestibulospinal tract originates from the contralateral medial, superior, and descending vestibular nuclei (see Fig. 1.10) and mediates ongoing postural changes or head righting in response to semicircular canal sensory input (angular head motion). The medial vestibulospinal tract descends only through the cervical spinal cord in the medial longitudinal fasciculus and activates cervical axial musculature.

The reticulospinal tract receives sensory input from all the vestibular nuclei as well as all the other sensory and motor systems involved with maintaining balance. This projection has both crossed and uncrossed components and is very highly collateralized. As a result, the reticulospinal tract through the entire extent of the spinal cord is poorly defined but is probably involved in most balance reflex motor actions, including postural adjustments made to extravestibular sensory input (auditory, visual, and tactile stimuli).

Vestibular Reflexes

The sensory, central, and motor output components of the vestibular system have been described. We now discuss their integration into reflexes called the VOR, VSR, and VCR. Additionally, we include brief descriptions of cervical, visual, and somatosensory reflexes. Although not directly mediated by the vestibular apparatus, these reflexes have a close interaction with vestibular reflexes.

The Vestibulo-ocular Reflex

The VOR normally acts to maintain stable vision during head motion. The VOR has two components. The angular

VOR, mediated by the SCC, compensates for rotation. The linear VOR, mediated by the otoliths, compensates for translation. The angular VOR is primarily responsible for gaze stabilization. The linear VOR is most important in situations where near targets are being viewed and the head is being moved at relatively high frequencies. An example of how the horizontal canal VOR is orchestrated is the following:

1. When the head turns to the right, endolymphatic flow deflects the cupulae to the left (see Fig. 1.4*B*).
2. The discharge rate from hair cells in the right crista increases in proportion to the velocity of the head motion, while the discharge rate from hair cells in the left lateral crista decreases (see Fig. 1.4*A*).
3. These changes in firing rate are transmitted along the vestibular nerve and influence the discharge of the neurons of the medial and superior vestibular nuclei and cerebellum.
4. Excitatory impulses are transmitted via white matter tracts in the brainstem to the oculomotor nuclei, which activate the right (ipsilateral) medial rectus and the left (contralateral) lateral rectus. Inhibitory impulses are also transmitted to their antagonists.
5. Simultaneous contraction of the left lateral rectus and right medial rectus muscles and relaxation of the left medial rectus and right lateral rectus occur resulting in lateral compensatory eye movements toward the left.
6. If the eye velocity is not adequate for the given head velocity and retina image motion is greater than 2° per second, the cerebellar projection to the vestibular nuclei (see Fig. 1.12) will modify the firing rate of the neurons within the vestibular nuclei to reduce the error.

The Vestibulospinal Reflex

The purpose of the VSR is to stabilize the body. The VSR actually consists of an assemblage of several reflexes named according to the timing (dynamic vs. static or tonic) and sensory input (canal vs. otolith). An example of a vestibulospinal reflex is the sequence of events involved in generating a labyrinthine reflex.

1. When the head is tilted to one side, both the canals and otoliths are stimulated. Endolymphatic flow deflects the cupula, and shear force deflects hair cells within the otoliths.
2. The vestibular nerve and vestibular nucleus are activated.
3. Impulses are transmitted via the lateral and medial vestibulospinal tracts to the spinal cord.
4. Extensor activity is induced on the side to which the head is inclined, and flexor activity is induced on the opposite side. The head movement opposes the movement registered by the vestibular system.

The Vestibulocollic Reflex

The vestibulocollic reflex (VCR) acts on the neck musculature to stabilize the head. The reflex head movement produced counters the movement sensed by the otolithic or semicircular canal organs. The precise pathways mediating this reflex have yet to be detailed.

Cervical Reflexes

The Cervico-ocular Reflex

The cervico-ocular reflex (COR) interacts with the VOR. The COR consists of eye movements driven by neck proprioceptors that can supplement the VOR under certain circumstances. Normally, the gain of the COR is very low.[21] The COR is facilitated when the vestibular apparatus is injured.[22,23] Although the COR or lack thereof is generally not recognized as a source of clinical disturbance, it is well known that vibration of the neck muscles in persons with unilateral vestibular loss induces a very strong nystagmus, often accompanied by vertigo.[24]

The Cervicospinal Reflex

The cervicospinal reflex (CSR) is defined as changes in limb position driven by neck afferent activity. Analogous to the COR, which supplements the VOR under certain circumstances, the CSR can supplement the VSR by altering motor tone in the body. Like the VSR, the CSR consists of an assemblage of several reflexes. Two pathways are thought to mediate these reflex signals—an excitatory pathway from the lateral vestibular nucleus and an inhibitory pathway from the medial part of the medullary reticular formation. When the body is rotated with head stable, neurons of the excitatory vestibulospinal system increase their rate of firing on the side to which the chin is pointed. At the same time, neurons thought to be in the inhibitory reticulospinal system show a reduced rate of firing. This activity leads to extension of the limb on the side to which the chain is pointed and flexion of the limb on the contralateral side. Vestibular receptors influence both these systems by modulating the firing of medullary neurons in a pattern opposite to that elicited by neck receptors. The interaction between the effects on the body of vestibular and neck inputs tends to cancel one

another when the head moves freely on the body so that posture remains stable.[25]

The Cervicocollic Reflex

The cervicocollic reflex (CCR) is a cervical reflex that stabilizes the head on the body. The afferent sensory changes caused by changes in neck position create opposition to that stretch by way of reflexive contractions of appropriate neck muscles.[21] The degree to which the CCR contributes to head stabilization in normal humans is presently uncertain, but it seems likely that it is useful primarily to stabilize head movement in the vertical plane and it may also be facilitated after labyrinthine loss. Experimental studies, mainly in animals, document that cervical blocks can cause imbalance and nystagmus.[26]

Visual Reflexes

The visual system is a capable and sophisticated sensory system that influences vestibular central circuitry and drives visual following responses (i.e., smooth pursuit) and postural reactions. Because of intrinsic delays in multisynaptic visual mechanisms, the earliest visual responses occur at a substantially longer latency (about 80 msec) compared to vestibular responses (about 14 msec) and are much less suited to tracking at frequencies above about 0.5 Hz.[27] Visual tracking responses may be facilitated after vestibular loss.

Somatosensory Reflexes

Somatosensory mechanisms are involved in postural stability as well. Bles and associates documented somatosensory-induced nystagmus ("stepping around nystagmus").[28] Interestingly, the subjects with bilateral vestibular loss developed a more pronounced nystagmus than did normal subjects. This implies that subjects with bilateral vestibular loss use somatosensory information to a greater extent than normal subjects.

On the other side, individuals who have "two hits"—somatosensory disturbance and vestibular injuries—naturally fare more poorly than persons who have well-functioning sensory input. For example, patients with diabetic peripheral neuropathy recover from vestibular deficits less well than do persons without neuropathy.[29]

Neurophysiology of Benign Paroxysmal Positional Vertigo

Although most vestibular disorders can be described in terms of imbalance between the ears or loss of function, benign paroxysmal positional vertigo (BPPV) has an entirely different mechanism. BPPV is caused by movement of detached otoconia within the inner ear (canalithiasis) or otoconia adherent to the cupula (cupulolithiasis) (Fig. 1.13). Great progress has been made in our understanding of BPPV.

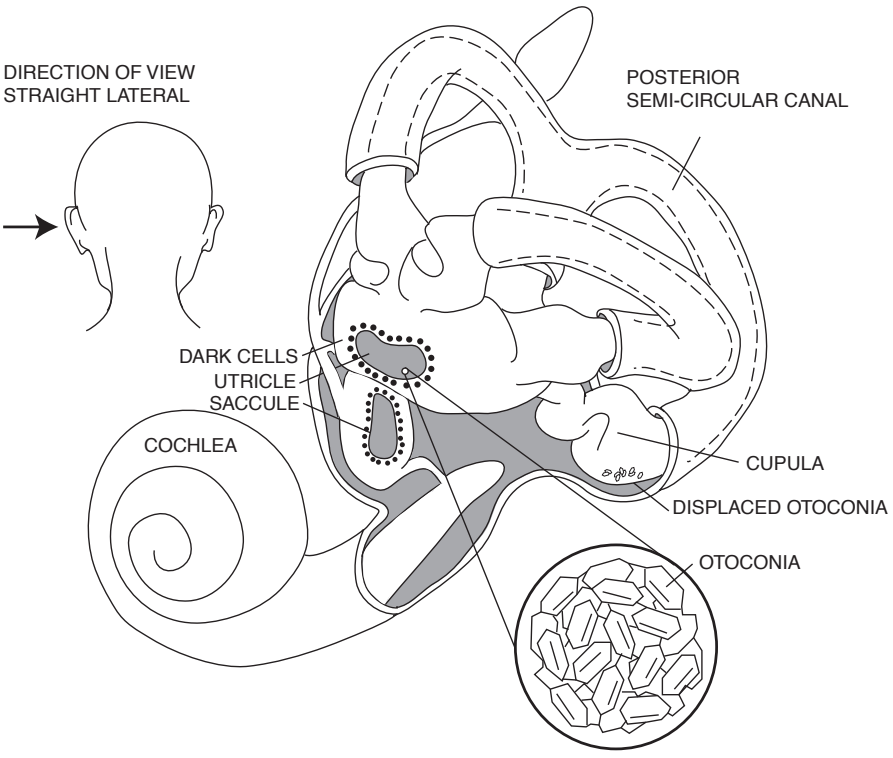

Figure 1.13 Physiology of benign paroxysmal positional vertigo. Otoconia become displaced from the utricle and relocate to the bottom of the posterior semicircular canal, which is the lowest part of the inner ear. (Copyright Timothy C.Hain, MD.)

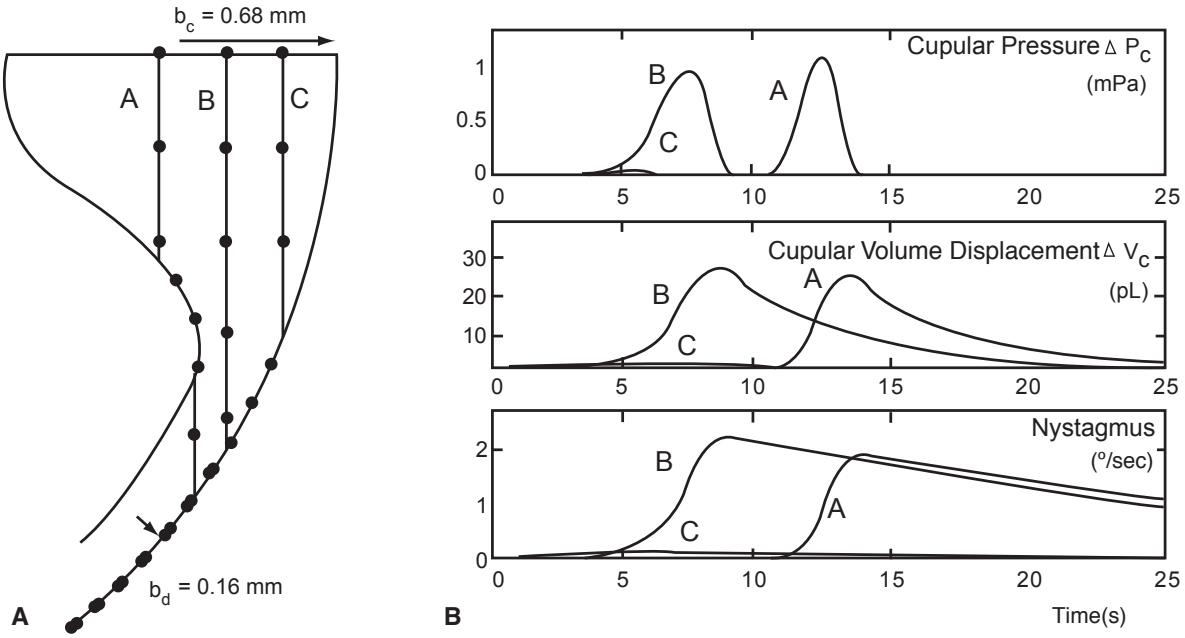

Figure 1.14 Fluid mechanics of benign paroxysmal positional vertigo. **(A)** Trajectories of three otoconia after a sudden change of head position that makes the posterior canal vertical. Otoconia begin close to the cupula, fall through the ampulla with radius b_c, and then enter the duct with radius b_d. **(B)** Simulated pressure, displacement, and nystagmus due to otoconia falling with the trajectories of *A*.[30]

Figure 1.14, from Squires and colleagues,[30] illustrates the fluid mechanics of BPPV. In this disorder, vertigo and nystagmus begin after a characteristic latency of about 5 seconds. The delay in onset of symptoms is caused by movement of detached otoconia through the ampulla, because pressure caused by moving otoconia is negligible until otoconia enter the narrow duct of the SCC. Figure 1.14 also shows that particle-wall interactions can account for variability in duration and latency of BPPV.[30]

Other results from fluid mechanics have direct bearing on our understanding of treatment maneuvers for BPPV. Under the influence of a full 1 g of gravity, typical otoconia move at a rate of 0.2 mm/sec, or only about 1% of the circumference of the canal each second. It follows that inertial effects of treatment maneuvers can cause negligible movement of otoconia and that, practically, sudden jerks of the head or maneuvers that incorporate eccentric moments (such as the Semont maneuver) are unlikely to have a substantial additional effect in comparison with maneuvers that rely on gravity to accomplish canalith repositioning (such as the Epley maneuver).[30]

Higher-Level Vestibular Processing

In this section we discuss some of the more sophisticated aspects of central vestibular processing, which are not reflexes but rather require much more processing, are generally much more accurate, and often are at least partially under conscious control. Because these mechanisms are more modifiable than vestibular reflexes, they are especially relevant to rehabilitation. Most of these mechanisms process multiple sensory inputs.

Velocity Storage

How good does the VOR have to be? To keep the eye still in space while the head is moving, the velocity of eyes should be exactly opposite to head movement. When this happens, the ratio of eye movement to head movement velocity, called the gain, equals –1.0. To maintain normal vision, retinal image motion must be less than 2° per second. In other words, for a head velocity of 100° per second, which is easily produced by an ordinary head movement, the gain of the VOR must be 98% accurate, because any greater error would cause vision to be obscured.

The normal VOR can deliver this high standard of performance only for brief head movements. In other words, the VOR is compensatory for high-frequency head motion but is not compensatory for low-frequency head motion. This fact can be most easily demonstrated by the response of the SCC to a sustained head movement, which has a constant velocity. The canals respond by producing an exponentially decaying change in neural firing in the vestibular nerve. The time constant of the exponential is about 7 seconds, or,

in other words, the firing rate decays to 32% of the initial amount in 7 seconds. Ideally, the time constant should be infinite, which would be associated with no response decline. Apparently, a time constant of 7 seconds is not long enough, because the central nervous system goes to the trouble to perseverate the response and replace the peripheral time constant of 7 seconds with a central time constant of about 20 seconds. The perseveration is provided via a brainstem structure called the velocity storage mechanism.[31]

The velocity storage mechanism is a repository for information about head velocity derived from several kinds of motion receptors. During rotation in the light, the vestibular nucleus is supplied with retinal slip information. Retinal slip is the difference between eye velocity and head velocity. Retinal slip can drive the velocity storage mechanism and keep vestibular-related responses going even after vestibular afferent information decays. The vestibular system also uses somatosensory and otolithic information to drive the velocity storage mechanism.[32] This example shows how the vestibular system resolves multiple, partially redundant sensory inputs.

Internal Estimation—Going Beyond Reflexes

Reflexes are simple sensory processors that rapidly convert sensory input into motor outflow. What happens when sensory input is not available (such as when the eyes are closed) or inaccurate (such as when a person with positional vertigo tilts the head) or is noisy? A mechanism that combines sensory inputs, weights them according to their relevance, and provides a reasonable estimate of orientation in space, even without any recent sensory input, is needed.

In engineering terms, we are discussing an "estimator" Navigating the space shuttle involves similar problems. There are hundreds of sensors and many actuators (motor outputs). Some sensors respond quickly and some slowly. They may differ in accuracy, scaling, coordinate frame, timing, and noise characteristics. A mechanism is needed to integrate sensors and develop an internal estimate of the state of the system (i.e., position, velocity, acceleration) to keep the shuttle from crashing.

The engineering solution to this problem developed out of work done by Kalman at MIT, and is often called a "Kalman filter." It is also called an "optimal estimator," or an "internal model." We prefer the term "optimal estimator." The essentials of an optimal estimator are shown in Figure 1.15. There is very strong evidence that mechanisms similar to optimal estimators are used for human sensorimotor processing [33] and vestibular processing.[33,34]

Estimators are far more powerful than simple reflexes. There are several key concepts that need to be introduced to understand how they are different. First, internal models of transducers and motor output are used to

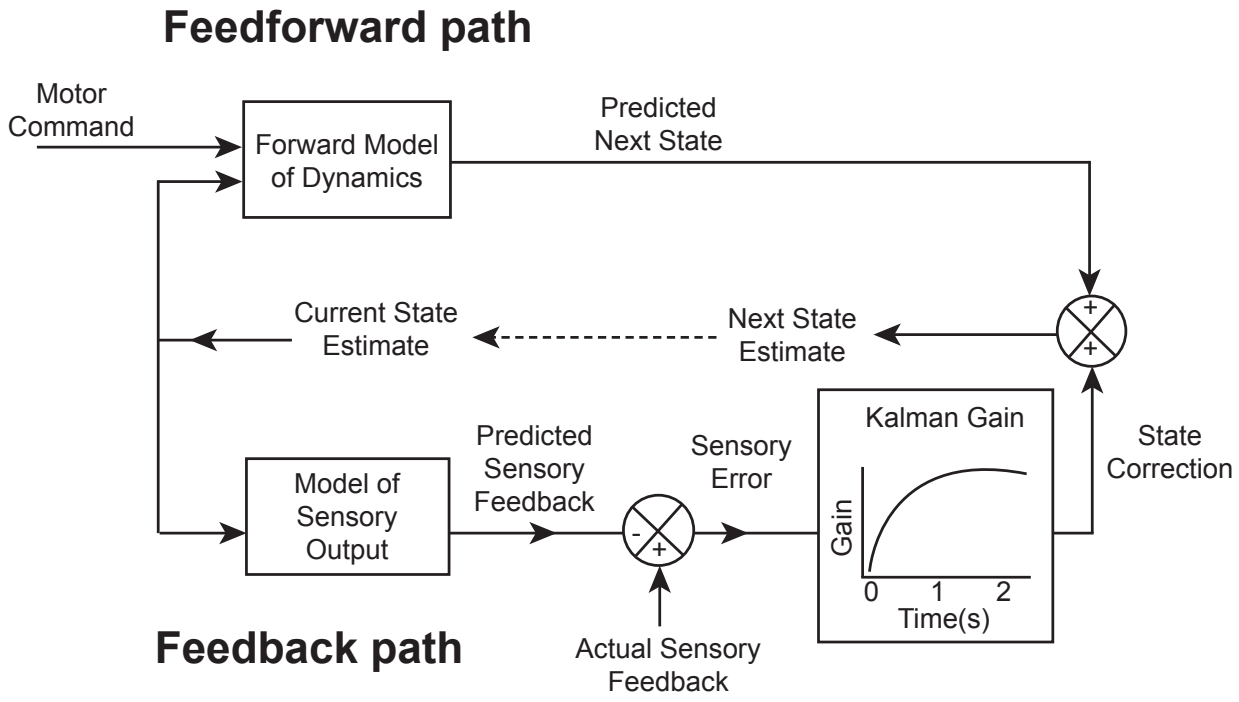

Figure 1.15 Block diagram showing an optimal estimator such as may be used by the human body for sensorimotor integration. Sensory inflow and motor outflow are used to estimate the current state.[33] Compare to Figure 1.11.

develop an estimate of the current state. These internal models are adjusted according to experience and track changes in bodily function. It seems highly likely that vestibular rehabilitation improves internal models.

Second, body state is not computed from sensory input directly, but rather the *difference* between sensory input and predicted sensory input is used to update an *estimate* of body state. This design makes it possible for the estimator to continue to work even in the absence of sensory input, which is clearly a desirable feature.

Third, a "Kalman gain" weights the degree to which a sensory input affects the ongoing state estimate. This provides a method of adjusting for the salience and reliability of sensory streams. Again, it seems highly likely that vestibular rehabilitation adjusts something similar to the Kalman gain to teach persons with vestibular deficits to compensate for unreliable sensory streams.

Overall, estimators are fundamentally superior to vestibular reflexes—they are intrinsically far more accurate. Estimators function even in the absence of sensory input. Estimators are modifiable to a greater extent than reflexes by experience. Estimators are just not as fast as reflexes because they are not "hard wired."

Higher Level Problems of the Vestibular System

Compensation for Overload

Humans can move their heads at velocities exceeding 300° per second. For example, while driving in the car, when one hears a horn to the side, the head may rapidly rotate to visualize the problem and potentially to avoid an impending collision. Similarly, during certain sports (e.g., racquetball), head velocity and acceleration reach high levels. One must be able to see during these sorts of activities, but the vestibular nerve is not well suited to transmission of such high-velocity signals. The reason is the cutoff behavior discussed in the section on the motor output of the vestibular system. High-velocity head movement may cause the nerve on the inhibited side to be driven to a firing rate of 0.

In this instance, the vestibular system must depend on the excited side, which is wired in "push-pull" with the inhibited side. Whereas the inhibited side can only be driven to 0 spikes per second, the side being excited can be driven to much higher levels. Thus, the push-pull arrangement takes care of part of the overload problem. Note, however, that patients with unilateral vestibular loss do not have this mechanism available to deal with the overload problem and are disturbed by rapid head motion toward the side of their lesion.

Vestibular Ambiguity

Sensory input from the otoliths is intrinsically ambiguous, because the same pattern of otolith activation can be produced by either a linear acceleration or a tilt. In other words, in the absence of other information, we have no method of deciding whether we are being whisked off along an axis or if the whole room just tilted. Canal information may not be that useful in resolving the ambiguity because one might be rotating and tilting at the same time. These sorts of problems are demonstrated in subway cars and airplanes, which can both tilt and/or translate briskly.

Outside of moving vehicles, vision and tactile sensation can be used to decide what is happening, perhaps using an optimal estimator as discussed earlier. As long as one does not have to make a quick decision, these senses may be perfectly adequate. However, remember that visual input takes 80 ms to get to the vestibular nucleus and that tactile input must be considered in the context of joint position and intrinsic neural transmission delays between the point of contact and the vestibular nuclear complex.

Another strategy that the brain can use to separate tilt from linear acceleration is filtering. In most instances, tilts are prolonged, whereas linear accelerations are brief. Neural filters that pass low and high frequencies can be used to distinguish one from the other. Nevertheless, in humans, evolution apparently has decided that the ambiguity problem is not worth solving. Otolith-ocular reflexes appropriate to compensate for linear acceleration or tilt do exist in darkness but are extremely weak in normal humans.[35] Stronger otolith-ocular reflexes are generally seen only in the light, when vision is available to solve the ambiguity problem. Sensory ambiguity becomes most problematic for patients who have multiple sensory deficits because they cannot use other senses to formulate appropriate vestibulospinal responses.

Motion Sickness

An instructive illustration of how the brain routinely processes multiple channels of sensory information simultaneously is found in the motion sickness syndrome. The motion sickness syndrome consists of dizziness, nausea or emesis, and malaise following motion. It is thought to be caused by a conflict between movement information in related sensory channels, such as visual-vestibular conflicts or conflict between an actual and an anticipated sensory input. For example, motion sickness is often triggered by reading a book while riding in a car. In this instance, the vestibular and proprioceptive systems signal movement but the visual system signals relative stability.

The vestibular apparatus provides partially redundant information, and this allows for the possibility of intralabyrinthine conflict. Space motion sickness is thought to be caused by intralabyrinthine conflict. About 50% of space shuttle astronauts experience motion sickness during the initial 24 to 72 hours of orbital flight. It is currently thought that space motion sickness is due to a disturbance in "otolith-tilt translation."[36] The otoliths normally function in the context of a gravitational field, so that at any moment the total force acting on the otoliths is the vector sum of that due to gravity and that due to linear acceleration of the head. The central nervous system expects linear acceleration to be mainly related to tilt because linear acceleration due to gravity is usually much greater than that due to acceleration of the head. When outside of earth's gravitational field, such as is the situation for astronauts in outer space, the only source of otolith stimulation is linear acceleration of the head. In susceptible individuals, the central nervous system continues to interpret linear acceleration as being primarily related to tilt, which is untrue in this situation, causing the motion sickness syndrome.[36,37]

Structures that are generally required for motion sickness include (1) intact labyrinth and central vestibular connections, (2) cerebellar nodulus and uvula that coordinate labyrinthine stimuli,[18] (3) the chemoreceptive trigger zone located in the area postrema, and (4) the medullary vomiting center.[38] Why certain subjects are more prone to motion sickness than others is not completely understood.

Repair

Thus far, we have described some of the problems posed by the limitations of the vestibular apparatus and the constraints of physics. In normal individuals, these problems can be resolved by relying on redundancy of sensory input and central signal processing. In addition to these intrinsic problems, there are also extrinsic problems that are related to ongoing changes in sensory apparatus, central processing capabilities, and motor output channels. Because being able to see while one's head is moving and avoiding falls are so important to survival, the repair facility of the vestibular system must be considered as an integral part of its physiology. For this reason, it is our final topic.

Adaptive plasticity for peripheral vestibular lesions is amazingly competent, even enabling the vestibular system to adapt to peculiar sensory situations requiring a reversal of the VOR (see Chapter 2 for more details on adaptation).[39] Adjustments of internal models and weighting of sensory inputs (e.g., Kalman gain) are likely at least as important as readjustment of reflexes, because internal models provide many important features that reflexes cannot (such as functioning in the absence of sensory input).

Although most people are capable of abstract thought and can generalize from one context to another, this is not the case for vestibular adaptation—there is a high degree of context dependency in the repair of peripheral vestibular lesions. In other words, adaptations learned within one sensory context may not work within another. For example, a patient who can stabilize gaze on a target with the head upright may not be able to do so when making the same head movements from a supine posture. Experimentally, in the cat, VOR gain adaptations can be produced that depend on the orientation of the head.[40] Similarly, when the VOR of cats is trained using head movements of low frequency, no training effect is seen at high frequencies.[41]

Another type of context dependency relates to the vestibulospinal reflexes and has to do with the difference in reference frames between the head and body. Because the head can move on the body, information about how the head is moving may be rotated with respect to the body. For example, consider the situation when the head is turned 90° to the right. In this situation, the coronal plane of the head is aligned with the sagittal plane of the body, and motor synergies intended to prevent a fall for a given vestibular input must also be rotated by 90°. For example, patients with vestibular impairment who undergo gait training in which all procedures are performed only in a particular head posture (such as upright) may show little improvement in natural situations when the head assumes other postures, such as looking down at one's feet. Little is understood about the physiology of context dependency.

Repair of central lesions is much more limited than that available for peripheral lesions; this is the "Achilles' heel" of the vestibular apparatus. Symptoms due to central lesions last much longer than symptoms due to peripheral vestibular problems. The reason for this vulnerability is not difficult to understand. To use a commonplace analogy, if your car breaks down, you can take it to the repair shop and get it fixed. If, however, both your car and the repair shop are broken, you have a much bigger problem. The cerebellum fulfills the role of the repair shop for the vestibular system. When there are cerebellar lesions, or lesions in the pathways to and from the cerebellum, symptoms of vestibular dysfunction can be profound and permanent. Clinicians use this reasoning when they attempt to separate peripheral from central vestibular lesions. A spontaneous nystagmus, which persists over several weeks, is generally due to a central lesion; a peripheral nystagmus can be repaired by an intact brainstem and cerebellum.

Summary

The vestibular system is an old and sophisticated human control system. Accurate processing of sensory input

about rapid head and postural motion is dif ficult as well as critical to survi val. Not surprisingly, the body uses multiple, partially redundant sensory inputs and motor outputs, combined with a competent central repair capability. The system as a whole can withstand and adapt to major amounts of peripheral v estibular dysfunction. The Achilles' heel of the vestibular system is a relative inability to repair central vestibular dysfunction.

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CHAPTER 2

Vestibulo-ocular Reflex Adaptation

Michael C. Schubert, PT, PhD

The vestibulo-ocular reflex (VOR) has been created with an incredible plasticity for the purpose of enabling change inevitable with disease and aging. In fact, without this adaptation, the normal effects of age or the constant attacks from disease would render us with significant functional impairments—namely gaze and gait instability. Some examples of the critical and extreme plasticity within the VOR include altered VOR gain with newly fitted glasses and complete reversal of the VOR when fitted with lenses that demand it. The focus of this chapter is on evidence for VOR adaptation from animal and human studies. In particular, I focus on the robustness of VOR plasticity, evidence for location sites of such plasticity, and how this adaptation occurs. For information concerning oculomotor (e.g., compensatory saccade) and vestibulospinal compensatory strategies that may assist a deficient VOR, please see Chapters 6 and 9.

Role of Vision and Head Motion in Adaptation—Overview

Two sensory stimuli are required for significant adaptation of the VOR: vision and head motion. Traditionally, adaptation paradigms designed to enhance the gain of the VOR have been done with short-term (less than 1 hour) or long-term (greater than 1 day) time exposure. A number of VOR adaptation studies have demonstrated a robust capability for changing the normal VOR by coupling head motion with target motion. When the viewed targets move in such a manner that the image slips on the retina, the VOR is recalibrated. The direction of the recalibrated VOR (increased or decreased) depends on the direction of the target motion in relation with the head motion. During head rotation, visual following of targets that move at a velocity different from the head rotation will create an adaptation (change) in the gain of the VOR. This is commonly achieved by one of two means: (1) targets are viewed wearing lenses that maximize the visual world (increases target velocity) or minimize the visual world (decreases target velocity) or (2) targets are viewed that move in a direction opposite the head rotation (increases target velocity) or move in the same direction of head rotation (decreases target velocity). The difference between the target velocity and the eye velocity is termed retinal slip and can be quantified. Retinal slip is therefore a velocity error signal, thought to be the most effective means for changing VOR behavior (e.g., VOR gain).[1,2]

Eye rotation—angular motion of the eye along a specified axis. For example, torsion refers to a clockwise or counterclockwise *rotation* around a cephalocaudal axis (the line of sight) when the eye is at its centered and primary position.

Critical Evidence of VOR Adaptation in Animals

Early and seminal studies, critical for establishing therapeutic principles of gaze and gait stability, investigated the role of vision and motion in animals that underwent a unilateral vestibular lesion.[3-5] Data suggest when animals with a lesioned VOR are restrained to a darkened environment, the VOR does not recover, and imbalance persists. Once those same animals are released into the light, their VOR and balance begin to recover. This has been demonstrated across canal plugging, labyrinthectomy, and neurectomy lesions, suggesting that it is a universal principle of VOR adaptation after vestibular hypofunction. It was hypothesized that light exposure was critical for VOR recovery because it revealed retinal slip, which would occur when the visual environment blurred during head motion. This was confirmed when investigators conducted lesion studies removing the visual cortex (occipital lobes) in monkeys, and VOR gain adaptation was lost.[4]

Adaptation within the VOR appears to be most robust for sinusoid head rotations at frequencies less than 4 Hz whether the VOR gain is increased or decreased.[6-8] Additionally, VOR gain tends to be greatest at the frequency of rotation that was used to change it (e.g., if 1 Hz is used as the frequency of head rotation, then the gain change will be greatest at 1 Hz and less at head rotations of 2 Hz or 0.5 Hz). Data also suggest that increasing the VOR gain is more prone to frequency specificity than is reducing VOR gain. The difference in VOR gain for up versus down adaptation paradigms may be related to differences in afferent physiology. In primates, primary vestibular afferents of the healthy vestibular system have a resting firing rate that is typically 70 to 100 spikes per second.[9,10] The discharge regularity of the afferent is determined by the spacing of the interspike intervals between action potentials (Fig. 2.1). This enables classification of afferents into irregularly and regularly discharging groups. The information carried by irregular and regular afferents varies over the spectral range of frequency and acceleration of natural head movements. Irregular afferents tend to be more sensitive to rotations during large head accelerations.[10] The increased sensitivity of the irregular afferents may be more critical for the rapid detection of head movements as well as initiation of the VOR.[10,11] Regular afferents, in contrast, provide a signal that is proportional to head velocity over a wide spectral range.[10] In addition, the regular afferents may be the primary source of input to the VOR for low-frequency and small head accelerations (as measured from steady-state responses to sinusoidal rotations) because temporarily silencing the irregular afferents had no effect on the VOR.[12]

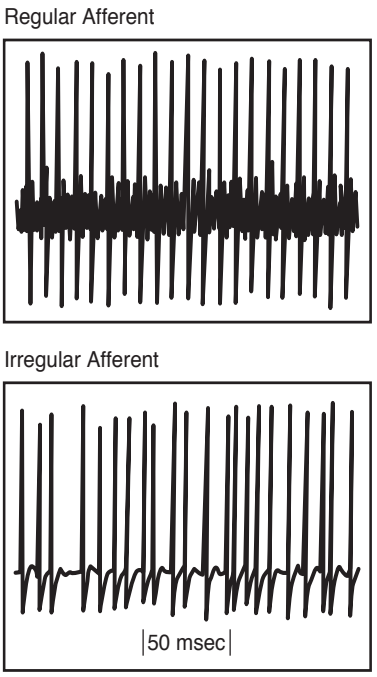

Figure 2.1 Classification of the peripheral vestibular afferents is based on the discharge regularity of the afferent. The discharge regularity is determined by the spacing of the interspike intervals between action potentials. The resting discharge rate is 97 spikes per second for regular afferents and 98 spikes per second for irregular afferents, though their periodicity is different. (Reprinted with permission from Goldberg JM, Fernandez C. Physiology of peripheral neurons innervating semicircular canals of the squirrel monkey, I: resting discharge and response to constant angular accelerations. *J Neurophysiol* 1971;34:635-660.[9])

The VOR has been tested across multiple frequencies and velocities and shows velocity-dependent nonlinearities that may correlate with unique afferent physiology.[11] The gain of the VOR remains constant (*linear*) across multiple frequencies of sinusoidal rotation and peak velocities of ±20 deg/sec.[11] For rotations at higher frequencies and velocities, the VOR gain rises with increases in stimulus velocity (*nonlinear*) and steps of acceleration. Therefore, it may be that the output of the VOR is the combined result of linear and nonlinear components.

Clendaniel et al demonstrated differences in magnitude of VOR gain adaptation dependent on the acceleration or velocity portion of a step rotation.[8] A step rotation is named based on its graphed profile and refers to a sudden acceleration that then settles at a constant velocity (see Chapter 11 on vestibular function for description). VOR gain changes during the acceleration component (~86%) were nearly double the magnitude of VOR gain changes during the velocity component (~46%). Based on these properties, the authors proposed that both a linear pathway (tonic) and nonlinear pathway (phasic) mediate the VOR. The

nonlinear pathway (irregular afferents) appears more pliable than the linear pathway of the VOR. In a later study, the same authors were able to preferentially adapt the linear and nonlinear pathways of the VOR using unique frequency and velocity profiles, supporting their hypothesis that differential adaptive control exists within the VOR.[13]

Consolidation of VOR Learning

Our understanding of consolidation of new motor learning in the VOR is limited. Consolidation refers to a specific duration of exposure that then enables long-term memory (retention). A recent study of VOR gain retention showed that head movements were required for the return to normal of a VOR gain that had been adaptively decreased.[14] In this study, investigators adaptively increased (high gain) or decreased (low gain) the VOR in rhesus monkeys for several days (long-term experiment). The animals were then placed in the dark (no visual stimulation) and either head restrained or allowed to make head movements. Regardless of head motion, animals that had their VOR gain adaptively increased showed little reduction in the magnitude of the gain. However, monkeys that had the VOR gain decreased required head movements for the VOR gain to return to baseline. This suggests that head movements are required for the VOR gain to increase when it is low (i.e., patients with vestibular hypofunction) but not required for the VOR gain to decrease when it is high. An interesting finding, unresearched in humans, is the idea that consolidation of VOR motor learning can be enhanced with a break during the training. In studies in which monkeys are exposed to VOR adaptation for a period of time (i.e., 1 hour) and then are not allowed to move or see light for a period of time, the learning (adaptation) is more robust and not as susceptible to disruption. The prevention of disruption in learning appears more robust when the gain is adapted down.[15] This "consolidation rest" benefit appears independent of the frequency of training, meaning once the adaptation and rest period have occurred, the retained learning persisted across all tested frequencies—not just the ones that were adapted.[16] This may also explain earlier prior long-term adaptation studies that showed that retention of the adapted VOR gain is possible when animals are kept in the dark or not allowed to move their head[17,18]—conditions that may have enabled consolidation of learning.

At least two non-error signal principles also affect VOR adaptation: context specificity and duration of exposure. Context specificity implies a unique condition that would not normally be expected to affect the performance of a motor task. We have demonstrated that VOR adaptation in squirrel monkeys can be retained for times (days) much longer than the adaptive exposure (hours), provided that the context of adaptation is unique to the animal.[19] Squirrel monkeys were exposed to a sum of sines rotation for 3 hours in both heads up (common experience) and either left or right ear-down position (unique experience). Retention only occurred in the ear-down condition. Others have shown similar results for short-term adaptation.[20] Head position relative to gravity is the context in these paradigms. Vertical eye position,[21] vergence angle,[22] and head position[19,20,23] are all examples of contexts for which the vestibular system can be uniquely adapted. For example, the VOR gain can be driven up when looking up and down when looking down. Context specificity has given insight into plasticity of the VOR, establishing that the brain can be trained to generate VOR gains uniquely related to the context it was exposed to during that training. A second principle important in VOR adaptation involves the duration of exposure time to an adapting stimulus. As expected, a longer time exposure leads to a greater magnitude of VOR gain change and usually requires more time to recover.[24]

Role of the Peripheral End Organ in VOR Adaptation

Although the cerebellum and brainstem will remain atop the list of regions critical for VOR adaptation, evidence suggests that the peripheral end-organ anatomy also has a role. Scarpa's ganglion has been shown to have a change in neuronal proteins after unilateral labyrinthectomy (UL) in rat, presumed to provide neuroprotection against oxidative damage and thermal signaling for modulation within the vestibular nerve.[25] The proportion of regular and irregular vestibular afferents also appears affected after UL. After UL in monkey, an increase in irregular afferents and a decrease in regular afferents occur on the contralesional side.[26] This may represent an attempt of the brain to improve the afferent sensitivity for higher acceleration and brief head rotations, believed to be mediated primarily from the irregular afferents.[27] Finally, as mentioned previously, there appears to be a unique contribution from the peripheral afferents to carry different velocity and frequency content of information concerning the VOR adaptation. However, we are not certain if this is related to primary afferent function (peripheral) or to something happening after synapse occurs in the vestibular nuclei. These cellular changes may represent an attempt to stabilize the ganglionic input to the vestibular nuclei, which in turn may initiate the adaptation process.

Brainstem versus Cerebellar Site of Adaptation

The error signals that drive VOR adaptation are sent to brainstem nuclei (primarily the vestibular Nu) and the cerebellum (specifically the floccular and paraflocculars

lobes).[28-31] Unit recordings from both brainstem and cerebellar regions demonstrate that response modulation is dependent on the magnitude of VOR gain adaptation. In the brainstem, neurons that synapse with the flocculus (floccular target neurons [FTNs]) show an increased firing rate when the gain of the horizontal VOR is adapted up (≥1.6) compared to when the gain of the horizontal VOR is adapted down (≤0.4). Similar evidence exists in the vertical VOR.[30] The firing rate of these individual neurons can even be reversed (Fig. 2.2). These data suggest that motor learning in the VOR is at least partially due to f iring rate changes of individual FTNs within the vestibular nuclei.

Purkinje cells in the flocculus of the cerebellum have been shown to consistently change their f iring rate responsiveness in parallel with the adaptive gain demand placed on the VOR. These changes persist even after the horizontal VOR had been extinguished by lesioning of the vestibular nuclei.[31] Ablation of the flocculus prevents VOR gain adaptation in rabbit [32] and monkey.[14] More specifically, lidocaine injections into the flocculus of macaque monkeys led to an immediate loss *only* of the adapted VOR, whereas pre-adapted VOR gains were not affected (Fig. 2.3).[33] These results support the hypothesis that the flocculus is critical for both adaptive modification and maintenance of VOR adaptation. In contrast, VOR gain is unaffected by nodulouvulectomy.

Cerebellar-deficient mice show less recovery of VOR gain after UL than do normal mice (wild type). In wild type, ipsilesional VOR gain recovery is approximately 80% by postoperative day 10 and 100% for contralesional

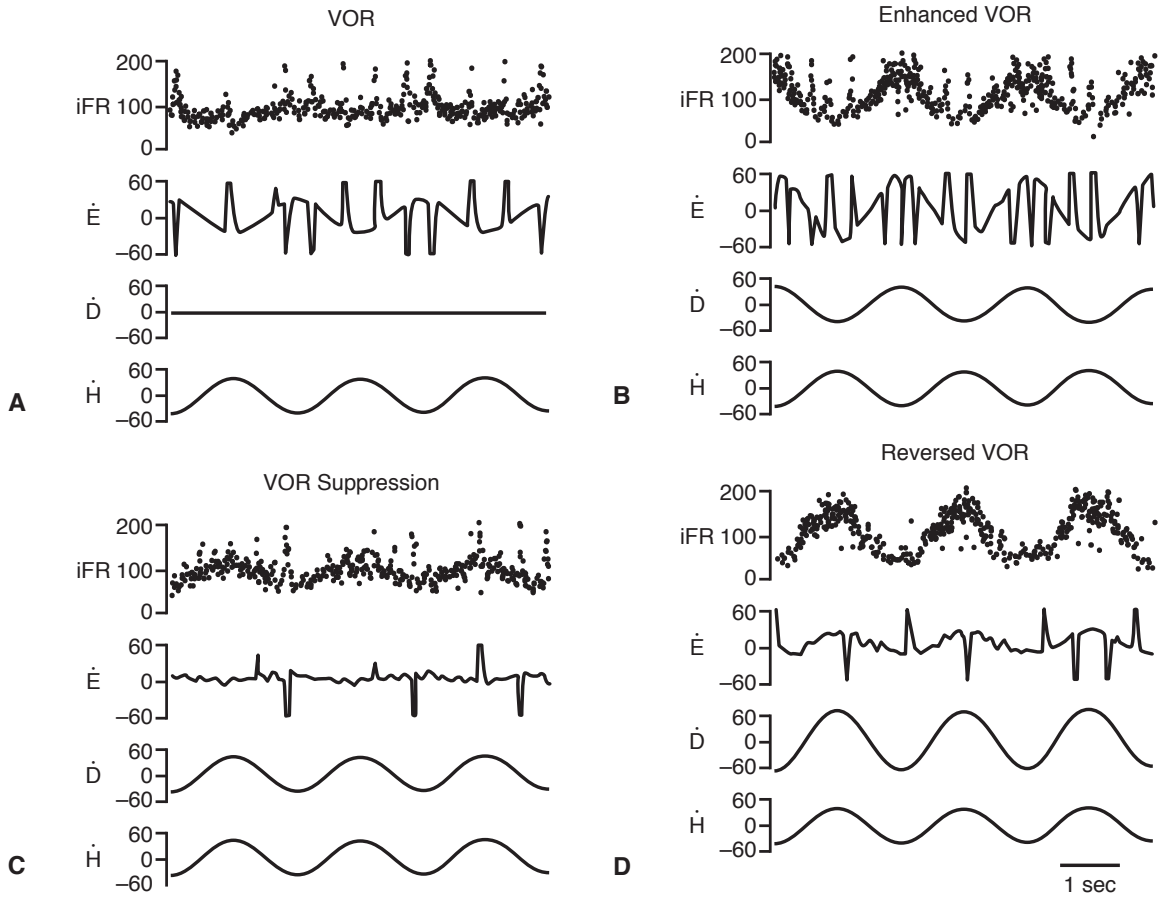

Figure 2.2 Behavior and firing rate of a VOR neuron during vertical head rotation under four unique conditions: **(A)** Normal VOR with no moving visual target. Note that the vertical head (Ḣ) and vertical eye (Ė) velocity rotation is opposite each other. **(B)** Enhanced VOR—visual target (Ḋ) moving opposite direction of head (as in the X2 VOR exercise). Note the increased eye velocity (Ė) and increased instantaneous firing rate (iFR). **(C)** VOR suppression with the visual target moving with the head. Note the reduced iFR and eye velocity. **(D)** Reversed VOR with the drum moving at a velocity twice that of head velocity. Note that the direction of the iFR has reversed! iFR = instantaneous firing rate; Ė = vertical eye velocity in d/s; Ḣ = head velocity in d/s; Ḋ = the optokinetic drum velocity in d/s. Stimulation parameters are 0.5 Hz, 35 d/s. (Reprinted with permission from Partsalis et al *J Neurophysiol*. 1995;73:615-631.[30])

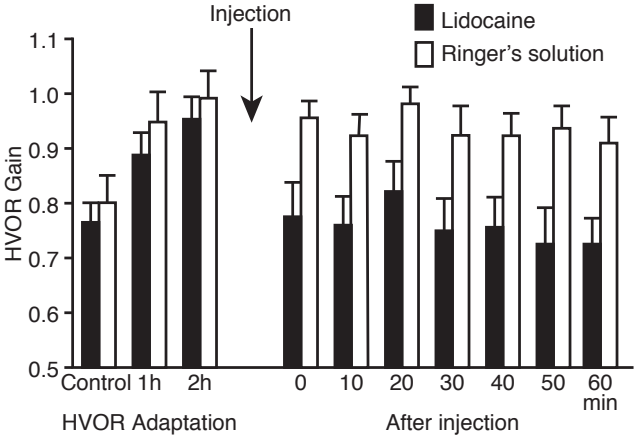

Figure 2.3 The injection of lidocaine into the flocculus of macaque monkeys. After the injection (*arrow*), the prior VOR gain adaptation is lost, though the VOR gain returns back to pre-adaptation values. (Reprinted with permission from Nagao and Kitazawa, 2003.[33])

head rotations. In contrast, the cerebellar -deficient mice plateau at a VOR gain near 0.6 for ipsilesional and 0.8 for contralesional rotations.[34] The fact that compensation still occurred suggests that VOR gain adaptation is not an isolated cerebellar process.

Unique Neuroplasticity Dependent on Lesion Type

Reactive neurogenesis within the vestibular nuclei refers to the type of neuronal birth (plasticity) that occurs in the vestibular nuclei when damaged. Studies suggest that reactive neurogenesis is dependent on the type of lesion that has damaged the vestibular system. In the cat model, reactive neurogenesis in the v estibular nuclei occurs on the ipsilesional side only after vestibular deafferentation, not after UL or induced transient hypofunction (e.g., transtympanic tetrodotoxin [TTX] injection). Even though UL and TTX cause similar post-assault nystagmus and postural instability, only the v estibular nuclei of the unilateral vestibular deafferentation (vestibular nerve section) preparation becomes a neurogenic zone where cells are differentiated into GABA neurons (gamma-aminobutyric acid is an important inhibitory neurotransmitter in the VOR pathway), astrocytes, and microglial cells.[35] When cell mitosis (and proliferation) is prevented by injection of v arious drugs into the vestibular nuclei, the postural reco very is similarly delayed.[36] This was important to establish that the neurogenic cells were of functional rele vance. Similar increased cell proliferation has been sho wn in bilateral vestibular deafferentation in rat.[37]

Critical Evidence of VOR Adaptation in Human

Short-term VOR Adaptation in Normal Function

Short-term VOR adaptation studies using v elocity error signals for human e xperiments have demonstrated increases in the VOR gain ranging from 10% to 35% compared with preadaptation levels.[21,38-41] The overall amount of adaptation, however, varies across individuals. Though few studies exist, long-term experiments in humans show that VOR gain changes nearly 66% of the preadapti ve level—though this is incomplete relati ve to the adapti ve demand.[42-43]

In addition to velocity error signals being employed to change the gain of theVOR, position error signals have also been used. Jones and Mandl adaptedVOR gain using a strobe light stimulus that eliminated retinal slip.[44] In this case, it was presumed that the brain was able to extrapolate data of changing position and then string those unique position signals together to create an error signal.[44] Preliminary evidence from our laboratory also suggests partial adaptation of the VOR using a position error signal during passive, low-velocity (i.e., 43 de g/sec) whole body head rotation. In this study, a target was extinguished during head movement, which then reappeared at the end of each ipsilateral half-cycle whole body rotation, thereby exposing subjects to a 10-de gree position error signal.[2] Using this paradigm, a 5% to 16% increase in VOR gain occurred that varied depending on the individual. Interestingly, compensatory saccades were also elicited by this paradigm and accounted for more of the impro vement in gaze stability than did increased slo w component e ye velocity (VOR gain adaptation). Recently , VOR gain change with a position error signal during rapid head rotations was examined in healthy controls.[45] Although significant, a paltry 2 ± 5% (range –18% to 12%) change in VOR gain in response to a position error signal occurred during rapid head rotations.[45] It appears that using a position error signal to adapt the VOR gain during rapid head rotations leads to less o verall adaptive change than a velocity error signal. Position error signals, however, may recruit saccades in addition to changing theVOR gain. The role of VOR adaptation with a position error signal is unknown in patients with vestibular lesions.

Other inputs have also been identified as adapting the VOR, including visual input alone. Shelhamer et al showed the VOR gain can be increased (~11%, Pre 1.02 ± .03; Post 1.13 ± .07) simply by ha ving subjects view a rotating drum, without any head rotation.[21] Das and colleagues established that asking subjects to make horizontal smooth pursuit (~11%) or horizontal saccades (~13%)

prior to measuring the VOR with horizontal head rotations always led to an increase in the gain of the VOR, even though the "priming" eye movements occurred without head motion.[46] These priming eye rotations were applied in both directions. In contrast, neither vertical pursuit nor vertical saccades had a similar effect on the horizontal VOR gain.

VOR Gain Retention

Few studies have investigated retention of VOR gain in humans. Recently, new information has elucidated the retention of an adapted VOR gain using short-term experiments and has exciting implications for vestibular rehabilitation. In healthy humans, Yakushin et al documented that only *one* 1-hour session of VOR adaptation in a unique position (ear-down, horizontal rotation for pitch stimulus) was needed to cause a VOR gain change to be retained for 2 days (tested in the adapting position).[40] This study suggests that the VOR adaptation can persist when the exposure of the training stimulus is unique.

VOR Adaptation in Unilateral Vestibular Hypofunction

The VOR retains its adaptive capabilities in the presence of unilateral hypofunction and can be enhanced using visual and vestibular stimuli that create retinal image slip.[47-49] However, improvements in VOR gain after vestibular rehabilitation (vestibular physical therapy [VPT])—though function improves—have been difficult to verify.[50,51]

Three human studies have demonstrated that the VOR retains its adaptive capacity in the presence of unilateral vestibular hypofunction (UVH).[3,41,48] Two of these studies measured VOR gain change using passive whole body rotation and reported a mean 15% increased.[3,41] Viirre reported that the greatest amount of adaptation occurred at the two highest frequencies at which the authors tested the VOR (0.32 Hz and 0.64 Hz, 50 deg/sec).[48]

Studies of motor control reveal that retention of newly learned motor behavior is most robust when the training (adaptation) is presented in an incremental manner.[52,53] Referred to as "credit assignment," the brain has the difficult task of determining where blame resides when motor performance is impaired. Smaller error signals may "deceive" the brain into believing that the perceived error signal is intrinsic and warrants efforts toward plasticity. Larger error signals may be interpreted by the brain as extrinsic and transient and thus disregarded as invalid.[54] Thus, the brain attempts a more enduring modification for smaller error signals and ignores large error signals. Incremental velocity error (IVE) refers to a progressively changing visual error signal used to increase VOR gain. We investigated how using an IVE coupled with rapid active head impulses might change VOR gain.[41] We compared IVE against a large VOR gain demand (×2 viewing). For IVE adaptation, a ×1.1 stimulus was used to boost the VOR by 10%. After a brief rest, a ×1.2 stimulus demanded greater adaptation by 10% (20% total), and this was repeated until the ×2 stimulus was reached. The total adaptation time was 15 minutes. Both passive and active VOR gain was measured in all subjects before and after training. For both subject groups, traditional all-at-once adaptation resulted in no VOR gain increase. IVE adaptation resulted in significant increases in both normal (17.3% active; 14.2% passive) and UVH subjects (18.2% active). In all the normal and several of the UVH subjects, the active IVE training also increased the passive VOR gain (Fig. 2.4).

Evidence for VOR adaptation exercises:

- Two sensory stimuli are required for significant adaptation of the VOR: vision and head motion.
- The VOR retains its adaptive capabilities in the presence of unilateral hypofunction.
- Retinal slip is thought to be the most effective means for changing VOR gain.
- Small errors in retinal slip enable larger VOR gain adaptation than larger errors.
- The error signals that drive VOR adaptation are sent to brainstem nuclei (primarily the vestibular Nu) and the cerebellum (specifically the floccular and parafloccular lobes).

Summary

When individuals with vestibular hypofunction (unilateral or bilateral) are able to predict timing, direction, and amplitude of an imposed head movement, gaze stability improves relative to an unpredictable head movement.[55-60] Additionally, the gain of the VOR is greater during ipsilesional predictable head motions compared with ipsilesional unpredictable head motion in UVH.[57,58,60,61] Functionally, visual acuity during self-generated head rotation is better than it is during unpredictable head rotations.[62,63] Although it is believed that the enhancement of gaze stability and visual acuity during predictable head movement is due to central preprogramming or efference copy of the motor command, we do not know whether the augmenting effects of prediction can be deliberately increased. Although we know that VOR gain improvement is possible in hypofunction, less is known about whether such VOR gain adaptation can be retained. Long-term VOR adaptation studies have not been conducted in humans with

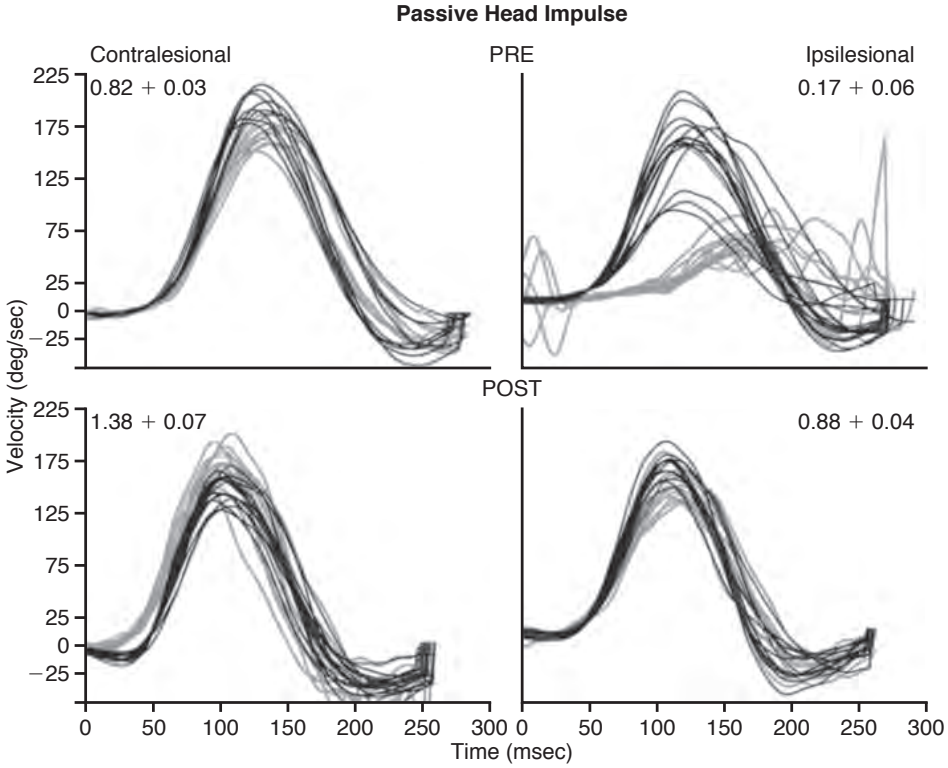

Figure 2.4 VOR adaptation in a subject with unilateral vestibular hypofunction using an incremental velocity error signal (IVE). Active VOR training for head rotations to both sides was done by gradually increasing the retinal slip error signal over a 15-minute period. Passive head impulse testing revealed that VOR gain increased for contralesional and ipsilesional head rotations. Black traces represent head velocity, and grey traces (inverted for ease of comparison) represent eye velocity. VOR gains are listed in the corner of each plot. Note that the Post ipsilesional VOR gain is restored, whereas the Post contralesional gain is greater than 1.0.

vestibular hypofunction. Much room exists for important research to be answered: Can we create a behavioral paradigm to improve the VOR gain and keep that improvement over time? Can VOR adaptation be unilaterally applied in the case of UVH? Is there a difference in VOR gain adaptation in unilateral versus bilateral vestibular hypofunction? Because of both existing knowledge of the neuronal synapses mediating the VOR and the latter's tremendous plasticity, it is expected that motor control research in general will continue to rely on the VOR as a model of study for insight.

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CHAPTER 3

The Role of the Vestibular System in Postural Control

Laurie A. King, PT, PhD ■ *Fay B. Horak, PT, PhD*

One of the most important tasks of the human postural control system is that of balancing the body over the small base of support provided by the feet. As a sensor of gravity and head acceleration, the vestibular system is one of the nervous system's most important tools in controlling posture. The vestibular system is both a sensory and a motor system. As a sensory system, the vestibular information is closely integrated with somatosensory and visual information so that the central nervous system (CNS) can estimate the position and movement of the entire body as well as the surrounding environment. In addition to providing sensory information, the vestibular system also contributes directly to motor control. Descending motor pathways such as the vestibulospinal tracts receive vestibular and other types of information to control eye, head, and trunk orientation and to coordinate postural movements.

Because the vestibular system is both a sensory and a motor system, it plays many different roles in postural control. In this chapter, we explore the four most important roles (Fig. 3.1). First, we discuss the role of the vestibular system in the perception of body position and self-motion. Second, we discuss its role in orienting the trunk to vertical using sensory orientation and weighting-appropriate sensory cues under different sensory environments. Third, we discuss the role of the vestibular system in controlling the position of the body's center of mass (COM), both for static positions and dynamic movements via postural responses. Fourth, we discuss its role in stabilizing the head during postural movements.

Perceiving Position and Self-Motion

The vestibular system provides information about the movement of the head and its position with respect to gravity and other inertial forces (like those generated by moving vehicles). Therefore, this system contributes important information to the sensation and perception of the motion and position of the body. Graviceptive information from the vestibular system is combined with information from the somatosensory system to perceive gravitoinertial forces and use them to orient the body. For example, the skier in Figure 3.1 maintains equilibrium by integrating gravitational and centripetal forces from the vestibular and somatosensory systems together with orientation information from vision. The vestibular system provides information about the position and motion of the head. Because the largest head motions during quiet stance sway and while walking or running usually occur in the sagittal (forward-backward) and frontal (left-right) planes, the vertical canals and otoliths are more critical for postural control than are the horizontal semicircular canals. However, the powerful influence of the horizontal canals on gaze stability during horizontal head motions (as in shaking the head "no") also contributes to control of posture because a stable visual reference is important for spatial orientation and balance.

In contrast to the canals, which sense rotational motion, the otoliths sense linear accelerations. Vertical linear

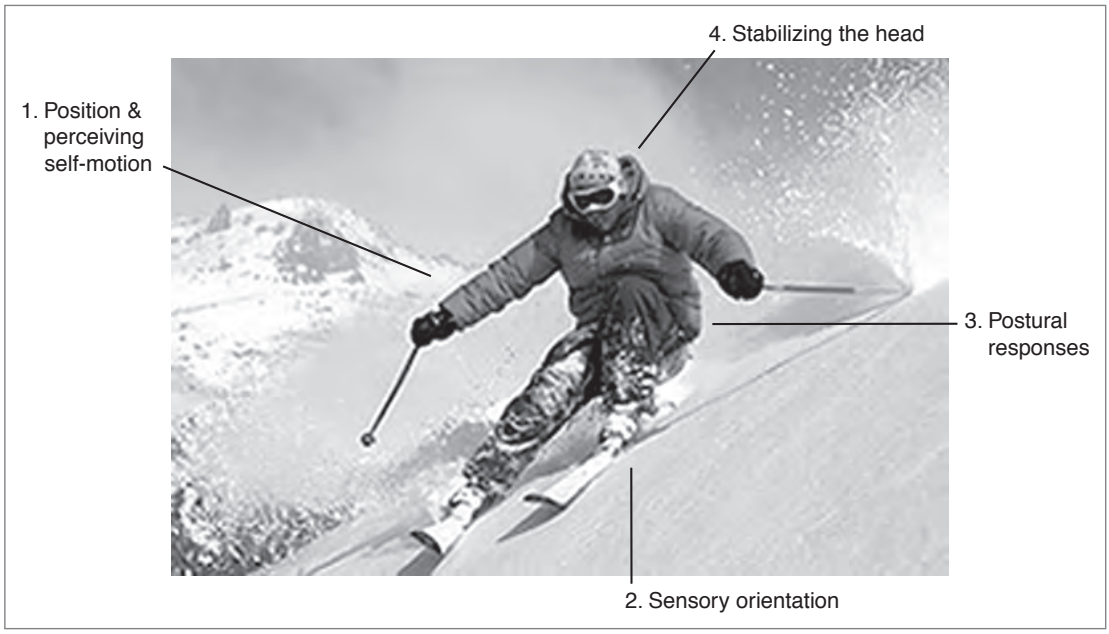

Figure 3.1 Four important roles of the vestibular system in postural control. As the skier leans into a curve, he (1) perceives his body orientation with respect to both gravity and the mountain, (2) orients his upper body to gravity (sensory orientation), (3) controls his COM (postural reactions), and (4) stabilizes his head in space.

accelerations of the head, like the head translations generated during heel strike in gait, are sensed by the saccular otoliths. Horizontal linear accelerations, like the translations of the head generated during walking, are sensed by the utricular otoliths. The otolith organs also provide information about the direction of gravity. Gravity, which is also a linear acceleration, produces an otolith signal that changes systematically as the head is tilted. The CNS uses this signal to determine head and trunk alignment with respect to gravitational vertical.

The importance of vestibular otolith function in perception of orientation in space can be seen by studies of perceived vertical and straight ahead in patients with bilateral and unilateral vestibular loss.[1] Whereas healthy subjects can accurately indicate the direction of gravity and straight ahead to within 1 degree, patients with bilateral vestibular loss make large errors and patients with unilateral vestibular loss often perceive up and straight ahead as tilted to the side of the lesion.[2-4] These asymmetrical perceptions of spatial orientation in patients with unilateral vestibular loss are reflected in the asymmetrical alignment of their eyes, head, and trunk before compensation for loss of vestibular function on one side.[5]

Vestibular information must be closely integrated with somatosensory and visual inputs to perceive position and motion of the body and environment because vestibular inputs can provide ambiguous information about body motion. First, the vestibular system only provides information about head movements and not the position or movement of the head on the trunk or any of the other body segments. Thus, the vestibular system alone cannot distinguish between head tilt on a stationary trunk versus whole body tilt over the feet, both of which activate the semicircular canals and otolith receptor organs similarly. Second, the otolith receptor organs cannot distinguish between the acceleration because of gravity and linear acceleration of the head in space. For example, tilting the head to the left can produce the same vestibular stimulation as a linear acceleration to the right. Resolution of the ambiguity between tilt and linear motion is important because the postural response necessary to maintain equilibrium and orientation may be opposite for these two postural perturbations. Additional information from somatosensory and visual systems is required to resolve these ambiguities.

Each sensory system contributes a different, important kind of information about body position and motion to the CNS, and each sensory system is most sensitive to particular types of motion.[6-12] The visual system signals the position and movement of the head with respect to surrounding objects. The visual system can provide the CNS with the information necessary to determine whether

a signal from the otoliths corresponds to a tilt with respect to gravity or a linear translation of the head. The visual system also provides information about the direction of vertical, because walls and doorframes are typically aligned vertically, parallel to gravity. The visual system provides good information about slow movements or static tilts of the head with respect to the visual environment.[7,13-15]

In contrast to vision, the somatosensory system provides information about the position and motion of the body with respect to its support surface and about the position and motion of body segments with respect to each other. For example, somatosensory information can help the CNS distinguish whether a head rotation signal from the vertical canals is a result of motion of the head on the neck or because of falling. The somatosensory system can also provide information about how body segments are aligned with respect to each other and the support surface by providing information about muscle stretch and joint position. The somatosensory system is particularly sensitive to fast movements, like those generated by sudden, external perturbations of the body.[16,17]

The contribution of each sensory system to the sensation of self-motion has been demonstrated experimentally by stimulating the individual sensory systems. Electrical stimulation of the vestibular nerve with direct current through electrodes placed on the skin over the mastoid bones produces sensations of body tilt by mimicking the vestibular signals that would be generated by an actual head movement.[15,18,19] Similar sensations of body motion can be achieved by presenting subjects with large moving visual scenes, such as when a bus or train moves alongside the stationary car in which you are sitting.[7,14,20,21] Figure 3.2 shows a person standing in front of a rotating disc. This person perceives himself to be tilting in the direction opposite the rotation of the disc and therefore compensates for this illusion of movement by tilting and orienting his body in the direction of the movement. Whenever the head is moved, the image of the entire visual scene moves in the opposite direction. Thus, when subjects watch a large, moving visual scene, the CNS often misinterprets the visual stimulus as self-motion in the opposite direction. Vibrating muscles at 60 to 100 Hz, which stimulate muscle proprioceptors, can also give rise to sensations of body motion because it is interpreted by the CNS as muscle lengthening.[6,22,23]

The perception of self-motion and orientation depends on more than sensory cues alone, however. What the subject predicts and knows about the sensory environment or what the subject has experienced in the past (sometimes called the subject's "central set" or "body schema") can contribute powerfully to how sensory signals are interpreted.[24-26] For example, imagine two cars stopped next to each other at a traffic light. If one car moves forward slightly, the driver of the second car may step on the brake, mistakenly believing that his car has rolled backward. This illusion is very powerful, because cars often do roll backward when stopped, as most drivers know. The "central set" of the driver is to expect the car to move, and so the driver accepts the visual motion cue, despite the fact that both the driver's vestibular and somatosensory systems indicate that the driver has not moved. This illusion of self-motion from vision has also been demonstrated in the laboratory. Subjects seated in a stationary chair who observe a large moving visual scene may perceive either chair motion or visual scene motion, depending on whether they are asked to concentrate on visual or somatosensory cues.[28] This observation is particularly interesting because these illusions of motion occur despite the fact that the vestibular system is signaling lack of motion.

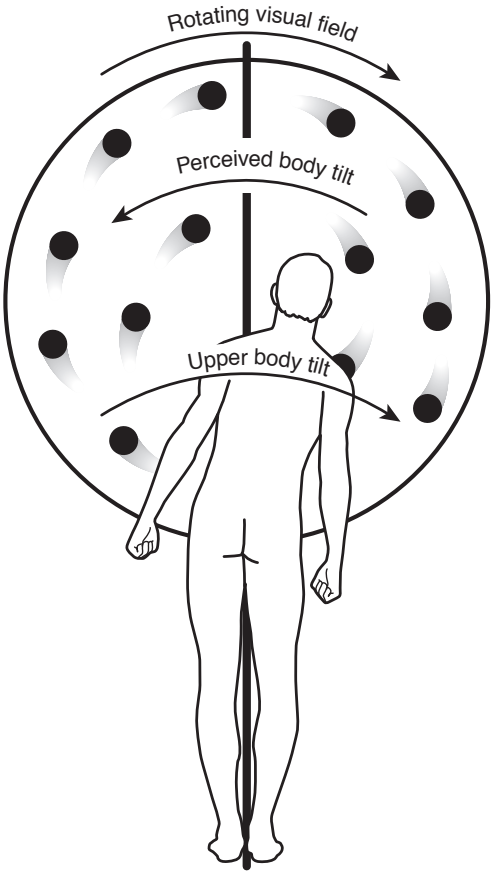

Figure 3.2 A subject standing in front of a rotating visual field perceives body tilting in the opposite direction as the visual field, so the upper body tilts in the same direction as the rotating visual field. (Adapted from Kandel & Schwartz, 2013.[27])

Given the vestibular system's important role in the sensation of self-motion, it is not surprising that patients with vestibular disorders often have abnormal perceptions of self-motion. Patients may report that they feel themselves spinning, dropping, or rocking or that the room appears to spin around them. These sensations may be associated with leaning head and trunk postures as they attempt to compensate for perceived body tilt or motion. Patients with profound loss of vestibular function also have difficulty perceiving how they are aligned or moving in environments lacking good visual and somatosensory orientation cues, such as walking at night on a sandy beach or swimming in murky water.

In summary, the vestibular system, along with other sensory systems, provides the CNS with the information about body motion and position with respect to vertical, which is critical for sensing and perceiving self-motion. No sensory system alone provides all the necessary information for sensing position and motion of the whole body; sensory information from multiple systems must be integrated and interpreted before being used to effect body orientation and equilibrium. In the next section, we explore how sensory information is used by the CNS to align the body to vertical, and how the CNS selects sensory information for body orientation in different environments.

Sensory Orientation; Orienting the Body to Vertical

Keeping the body properly aligned, parallel to gravity, and over the base of foot support is one of the most important goals of the postural control system. The vestibular system, which can detect the direction of gravity, plays a very important role in maintaining the orientation of the whole body to vertical. Because the term "orientation" also includes the alignment of body segments other than the head with respect to each other and with respect to vertical, other sensory systems contribute to body orientation as well. In this section, we discuss the role of the vestibular system in the alignment of the head and body to vertical, and how the nervous system selects appropriate sensory information for orientation in different sensory environments.

Postural Alignment

Spinal x-rays and fluoroscopy have revealed that most vertebrates hold the vertical spine parallel to gravitational vertical.[29] The vestibular system, which signals the direction of gravity, plays an important, but not exclusive, role in the head and trunk alignment in animals. Unilateral vestibular lesions result in head and body tilts toward the lesioned side.[29-31] The amount of asymmetrical posturing gradually diminishes over time, and the return to normal postural alignment is considered a sign of vestibular compensation.[32,33] In humans, the vestibular system also plays an important role in the alignment of the head and body with respect to gravity, although the effect of unilateral vestibular lesions on postural alignment is more variable and short-lived than in lower species.[30,31,34] Humans with sudden loss of vestibular function on one side can also show lateral flexion of the head to the side of the loss during the acute phase of the lesion.[32-34] However, within weeks following total unilateral vestibular loss in humans, postural alignment and control can be indistinguishable from that of a normal person.[35] Bilateral loss of vestibular function may be associated with a forward head position.[36-38] Altered postural alignment, sometimes associated with excessive muscle tension and pain, especially in the neck, is a familiar problem for patients with vestibular dysfunction.[39]

In addition to head tilts, the entire body seems to shift, temporarily, to the side of vestibular loss. Patients with unilateral vestibular lesions shift their weight to the side of their lesions and then regain normal weight distribution over the course of several weeks.[40,41] Fukuda developed a stepping-in-place test to document the asymmetry and gradual compensation that follow unilateral vestibular loss.[42] In this test, subjects attempt to step in place with eyes closed, and patients with unilateral losses typically rotate slowly toward the side of the lesion, although the effect is quite variable among patients.

Another way to investigate the role of the vestibular system in aligning the body to gravity is to stimulate the vestibular system electrically by delivering low-level (<2 mA) direct currents through electrodes on the mastoid processes, with an anode placed on one mastoid and a cathode placed on the other.[18,19,22,43-49] Cathodal currents increase the tonic firing in the vestibular nerve, and anodal currents decrease the firing rate.[50] With the head facing forward, sway induced by galvanic current is lateral and toward the anode, because the galvanic current simulates the vestibular nerve signal, which would result if the body was tilted toward the side of the cathode. Subjects sway toward the anode to correct the apparent tilt induced by the galvanic stimulation. Although galvanic stimulation reliably results in tonic head tilts and weight shifts in normal humans,[18,19,43-49] these responses are typically absent or abnormal in patients with vestibular nerve sections. However, these responses can be normal in patients with loss of peripheral hair cell receptors, which confirms that the galvanic current directly stimulates the vestibular nerve.[51-53] Postural responses to electrical stimulation can be enhanced when subjects stand on a sway-referenced

surface or have peripheral neuropathy that provides poor somatosensory feedback for orientation or when galvanic current is delivered during responses to a platform movement.[46,51,54,55] These findings suggest that the role of the vestibular system in automatic postural alignment is increased when somatosensory information for postural control is unreliable.[18] When visual spatial references are available with eyes open, postural responses to galvanic vestibular stimulation are greatly suppressed, suggesting that the importance of vestibular inputs for posture are also increased when visual information is unavailable. Figure 3.3 shows a person with bilateral vestibular loss attempting to stand on a platform that is rotated up 5 degrees. With her eyes closed, she is unable to maintain her balance because she continued to align her trunk with the surface, rather than with gravity.

Studies of postural responses to electrical stimulation of the vestibular system also show that the nervous system takes both vestibular and somatosensory information into account when organizing these responses. The direction of body sway and the corresponding muscle activations induced by galvanic stimulation are modulated by the position of the head on the trunk. With the head facing forward, galvanically induced sway is lateral and toward the anode.

In contrast, when the head is turned on the trunk so that the ear with the anode is turned forward, the galvanically induced sway is forward.[18] Thus, the same vestibular stimulation results in a very different and appropriate postural response because it is interpreted together with somatosensory signals about body orientation.[18,56] Thus, equilibrium control centers use information about body position and motion derived from proprioceptive afferents from many body segments, not just the neck, in combination with vestibular information to produce an accurate picture of body sway to trigger appropriate postural responses.[57,58]

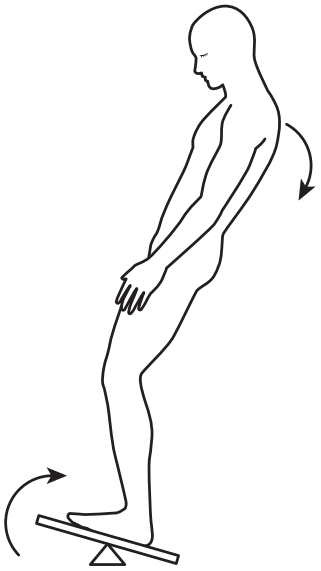

Figure 3.3 This figure represents a patient with bilateral vestibular loss attempting to stand on a tilted surface with eyes closed. Arrows indicate upward rotation of the surface and his postural alignment with surface tilt. Note he aligns his trunk in relation to the tilted surface rather than to gravity.

One hypothetical explanation for the altered postural alignment of patients with vestibular deficits is that the vestibular lesion has resulted in an altered internal map of body orientation in space. Gurfinkel and colleagues[8] have suggested that the CNS constructs a model or internal map of the direction of gravity based on vestibular and other sensory information, and that the CNS aligns the body according to this map. This hypothetical explanation could also account for the body realignments that result from galvanic stimulation. The galvanic current, which simulates the pattern of vestibular nerve firing that results when the body is tilted toward the cathode, may result in a change of the central nervous system's estimate of the position of gravity—that is, that it has shifted toward the anode.[54] Subjects sway toward the anode to realign their bodies to the new estimate of the position of gravity. The tonic body and/or head misalignment in patients with vestibular disorders may also result from a faulty internal map based on abnormal information from their malfunctioning vestibular systems.

Vestibular loss patients appear to misinterpret the movement of external objects as self-motion in the opposite direction. As a result, they may throw themselves into disequilibrium as they attempt to maintain a constant orientation with reference to the moving visual object. Thus, vestibular patients may either align themselves with a faulty vestibular estimate of the direction of gravity, or align themselves with an estimate of the direction of gravity from another sensory system.

Vestibular information also contributes to another important internal map for postural control, the map of "stability limits." Vestibular pathology may lead to defects in this map as well. A human standing with feet planted on the ground may sway forward or backward a small amount (about 4 deg backward and about 8 deg forward) without losing balance or taking a step. The boundaries of the area over which an individual may safely sway are called the stability limits.[59,60] The actual stability limits for any individual in any situation are determined by biomechanical constraints, such as range of motion, the firmness and size of the base of support, and by neuromuscular constraints, such as strength and swiftness of muscle responses.[61,62]

Vestibular pathology might result in a poor match between a patient's actual stability limits and the internal map of those limits. The internal map could be smaller or larger than the actual stability limits, or the map could be poorly aligned with respect to gravity. As a result, patients may align themselves near the edges of their actual stability limits. Because visual and somatosensory information may substitute for vestibular information, alignment may be normal in patients with well-compensated vestibular losses, but may be very abnormal in patients with deficits in multiple sensory systems or in patients who cannot compensate.

Weighting Sensory Information

Whether and how vestibular or other sensory information is used for postural orientation depends in part on the sensory information available in the environment. Under normal conditions (i.e., a stable support surface and a well-lit visual environment), postural orientation information from all three sensory modalities is available and is congruent; that is, all three modalities yield similar estimates of body position and motion. Although a person could theoretically rely equally on vestibular, vision, and somatosensory inputs for postural orientation, studies suggest they rely primarily on somatosensory information from the support surface in these conditions.[1,63,64] There are, however, many environmental conditions in which the sensory orientation references are not congruent. For example, when the support surface is compliant (like mud, sand, or a raft floating on water) or uneven (like a ramp or rocky ground), the position of the ankle joints and other somatosensory and proprioceptive information from the feet and legs bears little relationship to the orientation of the rest of the body; that is, the body's COM could be aligned well within the stability limits despite large amounts of ankle motion. Figure 3.4 *A* shows a person standing on an unstable surface, without vision, having to rely on vestibular information for orientation. Under such circumstances, the CNS down-weights dependence on the unreliable somatosensory inputs and unavailable visual information, and increases dependence on the vestibular inputs for spatial orientation.

The relative dependence on, or weighting of, each sensory system changes predictably with changes in environmental conditions.[27] A change in sensory weighting on somatosensory, vestibular, and visual information for postural orientation was shown experimentally by Robert Peterka (Fig. 3.4*A* to *C*).[65] For example, when blindfolded subjects are exposed to surface tilts of various amplitudes, they align themselves to the surface for very small amplitudes of rotation but orient their posture more with respect to gravity with larger amplitudes of rotation, because they rely more on vestibular information. In contrast, people with bilateral loss of vestibular information standing with eyes closed align their posture with the rotating surface until they fall. He calculated that healthy subjects standing on a firm surface with vision available normally rely 70% on somatosensory information from the surface, 20% on vestibular information, and 10% on vision for postural orientation. However, as surface rotation increases from

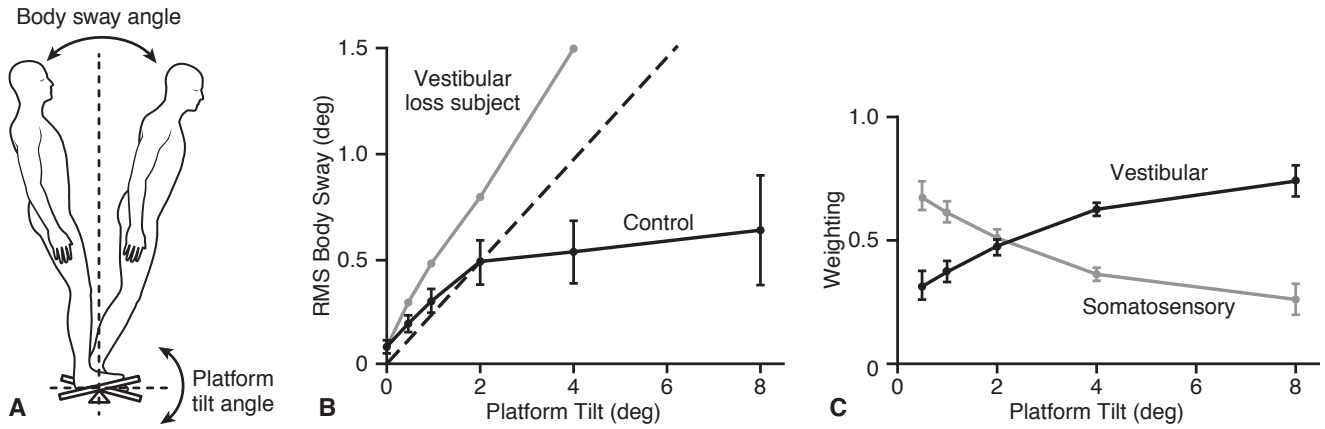

Figure 3.4 **(A)** Body sway changing in response to platform tilt angle when standing with eyes closed. **(B)** As platform tilt increases up to +/–2 degrees, all subjects increase body sway as they orient their body to the surface. Larger than +/–2 degrees surface tilt, control subjects stabilize body sway by orienting their bodies to gravity, whereas vestibular loss subjects continue to orient their bodies to the surface and fall. **(C)** Normal sensory reweighting as platform tilt angle increases. Healthy adults gradually increase dependence on vestibular inputs and decrease dependence on somatosensory inputs. (From Peterka, 2002.[65])

1 to 8 degrees, sensory weighting changes such that subjects now rely 70% on vestibular information, 20% on vision, and only 10% on somatosensory information. Because our nervous system normally depends primarily on somatosensory information for postural orientation, it can be difficult to identify postural problems in vestibular patients when they are standing on a firm support surface with eyes open. However, they may have severe balance problems on an unstable surface with eyes closed.

A computerized "Sensory Organization Testing" paradigm was developed by Nashner to measure the extent to which people can reweight sensory information for body orientation[66-68] and adapted to the clinic by Shumway-Cook and Horak.[69] In this paradigm, postural sway in stance is measured when subjects stand in six different sensory environments. In the first environment, the subject's support surface and visual surround are fixed to the earth and the subject stands with eyes open; the second is the same, but subject stands with eyes closed. This part of the test is equivalent to the standard Romberg test.[70] In the remaining four environments, the support surface, the visual surround, or both are moved in proportion to the subject's postural sway. This type of stimulation is referred to as "sway-referencing." By sway-referencing the support surface and/or the visual surround, the normal sensory feedback relationships between the different sensory systems can be disrupted. For example, when the support surface is sway-referenced, somatosensory information from the feet correlates poorly with the position of the body's COM. Support surface sway-referencing can be mimicked by placing the subject on compliant foam, and visual sway-referencing can be mimicked by placing a striped dome over the subject's head.[69] Vestibular information gives a more accurate estimate of body position and motion under these circumstances, and the CNS should rely more heavily on vestibular information for orientation.

Figure 3.5*A* shows how normal subjects respond when exposed to such altered conditions as eyes closed and stance on foam. Subjects with a healthy CNS and normal vestibular system sway a small amount during ordinary stance with eyes open. They increase sway slightly when they stand on foam with eyes open. However, when both the support surface is sway-referenced and visual information is eliminated by eye closure, normal subjects are able to use vestibular cues to orient the body, albeit with about 50% more sway than when somatosensory and visual information is available. This suggests that vestibular information is not normally our first choice of sensory input for postural control, although it is critical in some environments, such as when in the cabin of a ship at sea. Figure 3.5B shows a person with bilateral vestibular loss attempting to maintain balance during altered sensory

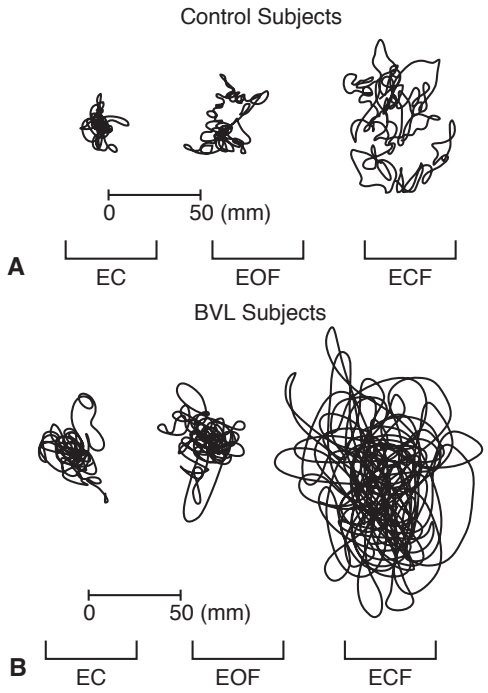

Figure 3.5 **(A)** Postural sway from surface center of pressure displacement in a control subject under three sensory conditions: EC= eyes closes, EOF = eyes open, standing on foam, and ECF = eyes closed, standing on foam. **(B)** Postural sway in a subject with bilateral vestibular loss (BVL) can be normal when standing on a firm surface with eyes open or closed, or on a foam surface with eyes open. However, sway in subjects with BVL is much larger than normal when standing on foam with eyes closed. (Dozza et al, 2007.[71])

condition. Note that this person appears to have normal sway with eyes open and eyes open on foam because they can still use the visual information. When this visual information is removed and the surface is unreliable, the sway dramatically increases as they attempt to maintain the orientation of their body.

Patients with clinically diagnosed vestibular disorders in the sensory organization test[66-69] do not all respond in the same way. Patients identified as having a vestibular loss pattern lose all sense of orientation and fall in conditions in which orientation information from both the surface and vision have been altered and the subject is forced to rely primarily on vestibular information (as in the example in Fig. 3.5*B*). Nevertheless, these patients can perform as well as normal in conditions in which at least one unaltered source of sensory information is available to them. In conditions in which they are forced to use vestibular information, however, these patients lose balance as though that information is unavailable. This pattern of results is typical of patients with long-standing,

well-compensated bilateral losses of peripheral vestibular function (although this pattern can also be seen in any patient whose nervous system does not use vestibular information for postural orientation, even if they may have some existing peripheral vestibular function). The fact that patients with well-compensated vestibular losses can use either visual or somatosensory information to orient the body limits the sensitivity and specificity of the standard Romberg test as a test of vestibular function.[37]

In contrast to the vestibular loss pattern, patients who have uncompensated vestibular disorders may show increased sway in conditions in which somatosensory or visual information is altered or not available. Their nervous system appears to orient their bodies to visual and support surface references even when these inputs are not reliably indicating motion of their body's COM in relation to the environment. Investigators have hypothesized that even healthy subjects vary in how much they "weigh" (rely more heavily on) sensory information from a particular source, like vision or somatosensation, so that may affect how they respond to a vestibular lesion.[10,28] Incomplete CNS adaptation to a vestibular lesion, particularly in the acute stages, is associated with excessive sway or falls in many sensory conditions.[72] However, as patients recover and the CNS adapts to the vestibular loss, patients with profound bilateral vestibular loss show the vestibular loss pattern, and many patients with total unilateral loss eventually return to normal sensory orientation for postural control.[35]

A recent study shows that patients with unilateral vestibular loss vary quite a bit on how much they use their remaining vestibular function system for postural orientation, with a group average of about 50% of normal (Fig. 3.6*A*).[73] Some patients with unilateral vestibular loss are able to rely on their one remaining vestibular system similar to normal when both vision and the somatosensory inputs are unavailable. In contrast, other patients with unilateral vestibular loss behave as if they cannot use their remaining vestibular function. The patients who were able to use their remaining vestibular function when standing on the sway-referenced surface with eyes closes showed better perceived activities of daily living than patients who increased weighting on somatosensory inputs (Fig. 3.6*B*).[74]

Some vestibular patients who have distorted, but not absent, vestibular function because of pathologies such as acute hydrops that alters, but does not eliminate, vestibular input may show a Visually Dependent Pattern of sensory organization.[66] These patients show excessive sway whenever the visual surround is sway-referenced but show normal sway with eyes closed. It is as if the nervous system relies on visual information whenever the eyes are open,

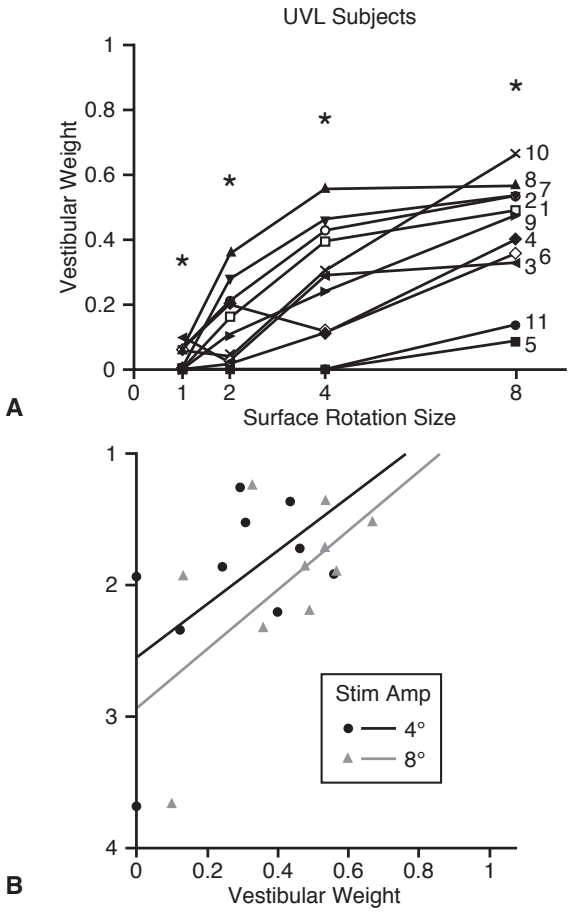

Figure 3.6 **(A)** People with unilateral vestibular loss (UVL) vary in their sensory reweighting strategies. Solid lines show vestibular weight as surface tilt angle increases in 11 individual patients with UVL. Asterisks show normal vestibular weighting. **(B)** Correlation between Vestibular Dysfunction ADL by Cohen[73] (VDADL) scores and vestibular weighting. Patients with UVL who had highest vestibular weighting for 4 degrees and 8 degrees surface rotations had the best ADL scores. (Adapted from Peterka et al, 2011, with permission.[73])

even when vision is not providing accurate information about body sway. However, when the eyes are closed, they are able to rely on surface information when standing on a firm surface and on vestibular information when standing on a sway-referenced surface.

People with bilateral or unilateral loss of vestibular function can readily substitute alternative sensory inputs to improve their balance. For example, studies have shown that light touch as minimal as that needed to read braille is even more effective than vision in stabilizing the postural sway of people with bilateral vestibular loss.[75,76] People with bilateral or unilateral vestibular loss can also use therapeutic audio-, visual-, or vibrotactile-biofeedback about trunk motion to control stance posture, even when

standing on a compliant foam surface with eyes closed.[71,77] In fact, patients with unilateral loss of vestibular function can substitute audio-biofeedback about direction and extent of lateral body sway, sensed with an accelerometer on the trunk, to successfully walk tandem with eyes closed, a task normally impossible for them. Figure 3.7*A* illustrates reduced sway during stance with eyes closed in a patient with bilateral vestibular loss using audio-biofeedback.[78] Figure 3.7*A* shows improvement of COM stabilization during tandem walking trials with and without biofeedback in a group of nine patients with unilateral vestibular loss.[79] All patients with unilateral vestibular loss significantly improved their balance with practice and retained this improvement the next day. Biofeedback immediately improves balance performance but does not speed motor learning (Fig. 3.7*B*).

Even simple sensory feedback such as an alarm indicating that the trunk has tilted beyond a particular limit can significantly reduce postural sway.[80] A study of healthy young adults standing on a rotating surface showed that all modes of biofeedback reduced improved multisegmental control of posture and stabilized the trunk in space, although spontaneous postural motor learning also improved multisegmental postural control (Fig. 3.7*C*). However, the more information provided to the nervous system about the direction and magnitude of postural sway, the more sway can be reduced (Fig. 3.7*C*). Animal studies have recently shown that galvanic vestibular stimulation scaled to head rotation applied directly to the 8th nerves of guinea pigs and monkeys could improve their equilibrium control, so this type of implanted biofeedback may one day be applied to patients with vestibular loss.[81] These results suggest that vestibular loss patients who complain of balance problems will benefit from use of a cane or other augmented sensory feedback to provide sensory information about trunk sway with respect to earth to substitute for their missing vestibular function, particularly when balancing on an unstable surface.

In summary, these studies show that vestibular information for body orientation is most important in environments that lack good somatosensory and/or visual cues for orientation. Patients attempting to rely on faulty or missing vestibular information in these environments may align themselves poorly and fall. However, they may also choose another source of orientation information when vestibular information is missing.

Controlling Center of Mass—Postural Responses

The previous sections described how the vestibular system detects the position and motion of the head, and how this sensory information is used for postural orientation. This section will describe how motor output from the vestibular system contributes to static body positions and dynamic postural movements, which help serve the postural goal of maintaining equilibrium, that is, controlling the center of body mass within its limits of stability. We know from

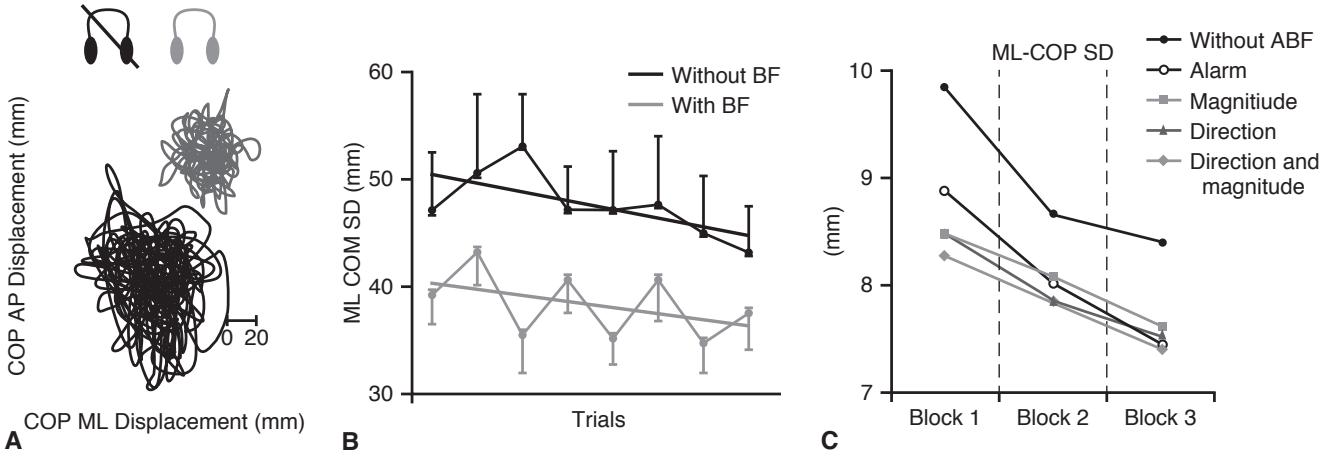

Figure 3.7 Biofeedback can reduce postural sway in subjects with BVL **(A)** Postural sway is reduced in a subject with audio-biofeedback (light gray) as compared with no biofeedback (dark gray) in the same subject. (Adapted from Dozza et al, 2005.)[78] **(B)** Effect of practicing tandem gait across trials with and without audio-biofeedback (BF). Each value represents the average among the nine subjects with UVL on three consecutive trials. Error bars represent standard errors. (Adapted from Dozza et al, 2007.)[79] **(C)** Effects of different types of information provided by biofeedback. Averaged SD of ML-COP over the three blocks of trials in all conditions with and without ABF in 13 healthy controls. (Adapted from Dozza et al, 2011.[80])

anatomic studies that motor output pathways leave the central vestibular nuclei and descend in the spinal cord, where they terminate on the neurons that activate primarily neck and trunk but also limb muscles.[82] However, the functional significance of the descending vestibular system for the control of orientation and equilibrium in alert, intact humans and animals is still poorly understood. Nevertheless, there is evidence that vestibular signals probably play a variety of roles, including stabilizing the head and trunk in space, scaling postural responses, and contributing to the selection of appropriate postural strategies for the environmental conditions.

Orientation and equilibrium represent two distinct postural goals. To accomplish some tasks, greater priority must be placed on achieving a specific postural orientation, at the cost of postural equilibrium. For example, an experienced soccer or volleyball player may make contact with a ball even though making contact requires falling to the ground. Other tasks require equilibrium at the cost of postural orientation. For example, balancing across a wire may require rapid hip flexion and extensions to maintain equilibrium. In this task, trunk orientation with respect to vertical is sacrificed to achieve the goal of equilibrium. The way in which the CNS achieves the trade-off between control of orientation and control of equilibrium in postural tasks is not well understood. Both static positions and dynamic movements require a system that prioritizes behavioral goals and uses all the sensory information available to effectively and efficiently control the limbs and trunk to achieve both orientation and equilibrium.

Triggering Automatic Postural Responses

If balance is disturbed in a standing human, limb muscles are activated at short latencies to restore equilibrium. Because the latencies of these muscle activations are shorter than a voluntary reaction time, and because they act to restore equilibrium, they are called "automatic postural responses." Although the most important sensory trigger for automatic postural responses is somatosensory inputs, vestibular inputs may also play a role. Both cats and humans respond to sudden drops from a height with short latency ankle extensor activations (50 to 100 ms in cat; 80 to 200 ms in humans).These muscle responses are present with eyes closed, so they can be triggered without visual stimulation. These responses are missing in patients with absent vestibular function, but survive procedures in animals that eliminate the canals but spare the otoliths. The magnitude of the responses is also proportional to head acceleration, all of which suggests a vestibular and, more specifically, an otolith origin for fast, automatic postural responses when surface and visual inputs are not available.[7,83-87] Quick displacements directly to the head during stance also result in muscle activations in the neck (45 ms), trunk (85 ms), thigh (90 ms), and ankle (70 ms).[88-90] These responses are absent in patients with adult-onset vestibular loss, suggesting a vestibular origin. However, adults who lost vestibular function as infants may show normal patterns of response to these head perturbations, suggesting that cervicospinal responses may adaptively compensate for the loss of vestibular input early in life.[89,90]

Because limb muscles are activated in response to sudden drops, perturbations in head position, and galvanic stimulation,[43,44,46] investigators have hypothesized that vestibulospinal mechanisms may also play a role in automatic postural responses to stance perturbations.[91] Two types of surface perturbations have been used to test this hypothesis: horizontal translations, which induce body sway such as from a slip or trip, and platform rotations, which induce ankle dorsiflexion or plantar flexion, as when standing on a boat or pier. Surface translations result in activation of the stretched ankle muscles, which occur at 80 to 100 ms following the onset of platform movement and act to restore the body to initial position. It does not appear, however, that vestibular inputs contribute a great deal to these responses. Human subjects and cats with complete absence of vestibular function can respond to surface translations using automatic postural responses with normal latencies and patterns, even when vision is not present.[72] Proprioceptive information from stretched muscles appears to be sufficient for recovery from surface translations. This conclusion is supported by the finding that automatic postural responses to surface translations are delayed when proprioceptive inputs are disrupted but not when vestibular inputs are disrupted.[92-96] Finally, relatively large head accelerations are required to produce relatively weak responses in the limbs, and the head accelerations that occur in response to platform translations are quite small.[88-90] Therefore, vestibulospinal responses probably do not play a large role in the recovery of equilibrium to slips or trips.

In contrast to responses to translations, responses to surface rotations rely much more heavily on the vestibulospinal system. Surface rotations stretch muscles in the lower leg, but do not produce corresponding forward or backward body sway. Responses to platform rotations consist of two parts. First to occur is a response in the stretched ankle muscle at 70 to 100 ms, which is probably triggered by proprioceptive inputs.[88,91,95,97-99] and which could, if unopposed, actually destabilize the body. Slightly later, a stabilizing response occurs in the shortened ankle muscle (at 100 to 120 ms), and this corrective response is

probably more dependent on vestibular (and visual) inputs. Patients with bilateral and unilateral loss of vestibular system have reduced magnitudes and delayed latencies of this late, stabilizing response, particularly if the velocity of the surface tilt is not so fast that quick, somatosensory responses are triggered, nor too slow such that orientation to the surface is sufficient for equilibrium.[97] The ability to adaptively reduce the magnitude of the destabilizing response to repeated surface tilts is also impaired in some patients with loss of vestibular function.[68] The cause of this failure to adapt could either be that vestibular information is required to trigger the adaptive process or because patients with absent vestibular function become dependent on proprioceptive information.[100,101]

Surprisingly, complete vestibular loss does not result in reduced or absent equilibrium responses. In fact, postural responses to surface translations are larger than normal in animals and humans with complete, bilateral loss of vestibular information, consistent with vestibular ataxia.[102] Figure 3.8 shows the center of pressure change in response to a surface perturbation. The person with vestibular loss has a response that is too large for the given stimuli compared with a control subject. Thus, postural instability and falls in patients with loss of vestibular function can be a result of responding too much to inappropriate sensory cues and consequently throwing themselves into disequilibrium.

Selection of Postural Strategies

Not all postural tasks require the same type of movement for the recovery of equilibrium, and some automatic postural responses require more vestibular involvement than others. These different postural responses have different muscle activation patterns (called postural synergies) as well as different body movements and joint forces, called "postural strategies."[99] Two very different strategies for moving the body's COM without moving the feet have been identified: the "ankle strategy" and the "hip strategy" (Fig. 3.9).[103] An ankle strategy is used to recover from a small postural disturbance when standing on a firm, flat support surface. The body sways roughly as an inverted pendulum by exerting force around the ankle joints. A hip strategy, which consists of rapid body motions about the hip joints, is typically used on narrow support surfaces (beams), compliant or tilting support surfaces (foam, tilt boards), when stance is narrow (one-foot standing or tandem stance), or when COM position must be corrected quickly. There is evidence that these postural control strategies are centrally programmed and can be combined depending on biomechanical conditions, subject expectations, and prior experience.[36,103,104] A stepping (or grabbing) strategy that increases the base of support over the falling center of body mass usually occurs later, after an ankle or hip strategy is insufficient to recover equilibrium.[105,106]

Vestibular information does not seem to be essential for initiation or execution of a normal ankle or stepping strategy, because subjects with complete absence of vestibular function show normal kinematic and EMG patterns, with the exception of neck muscles.[36,72,107] This suggests that proprioceptive information is sufficient to control the ankle and stepping strategies, albeit with abnormal activation of neck muscles resulting in larger than normal head accelerations.[108] In contrast, vestibular information appears to be useful in controlling the hip strategy. Patients with bilateral vestibular loss show poor performance in tasks such as one-foot standing, heel-toe walking, and beam balancing, all tasks requiring hip strategy for good balance control.[72,107,109,110] Also, subjects with loss of somatosensory information from the feet because of ankle ischemia, who presumably must increase their reliance on the vestibular system, use a hip strategy when an ankle strategy would be more efficient.[109] All of these findings suggest a critical link between the vestibular system and the use of hip strategy to control posture.

Although vestibular loss patients appear to be able to initiate hip strategy responses to fast surface translations when standing on a firm, flat surface, they take compensatory steps to maintain balance in response to these faster translations more frequently than normal subjects.[35] They may resort to stepping because hip strategy may require efficient control of the trunk and good stabilization of the head with respect to gravity.

Whereas vestibular loss is associated with a failure to use hip strategy in some conditions, some patients with pathology of the vestibular system habitually use hip movements to control COM position.[66,107,111] These

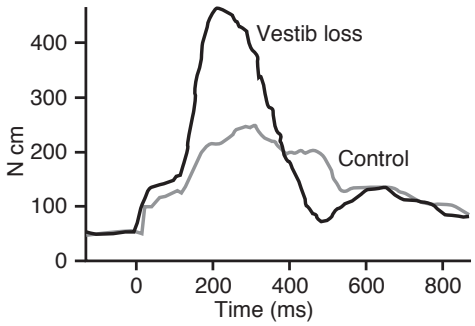

Figure 3.8 Postural responses to perturbations are too large in patients with bilateral vestibular loss (BVL). Graph shows center of pressure changes from plantar flexion in response to a forward fall in a subject with BVL and a control subject.

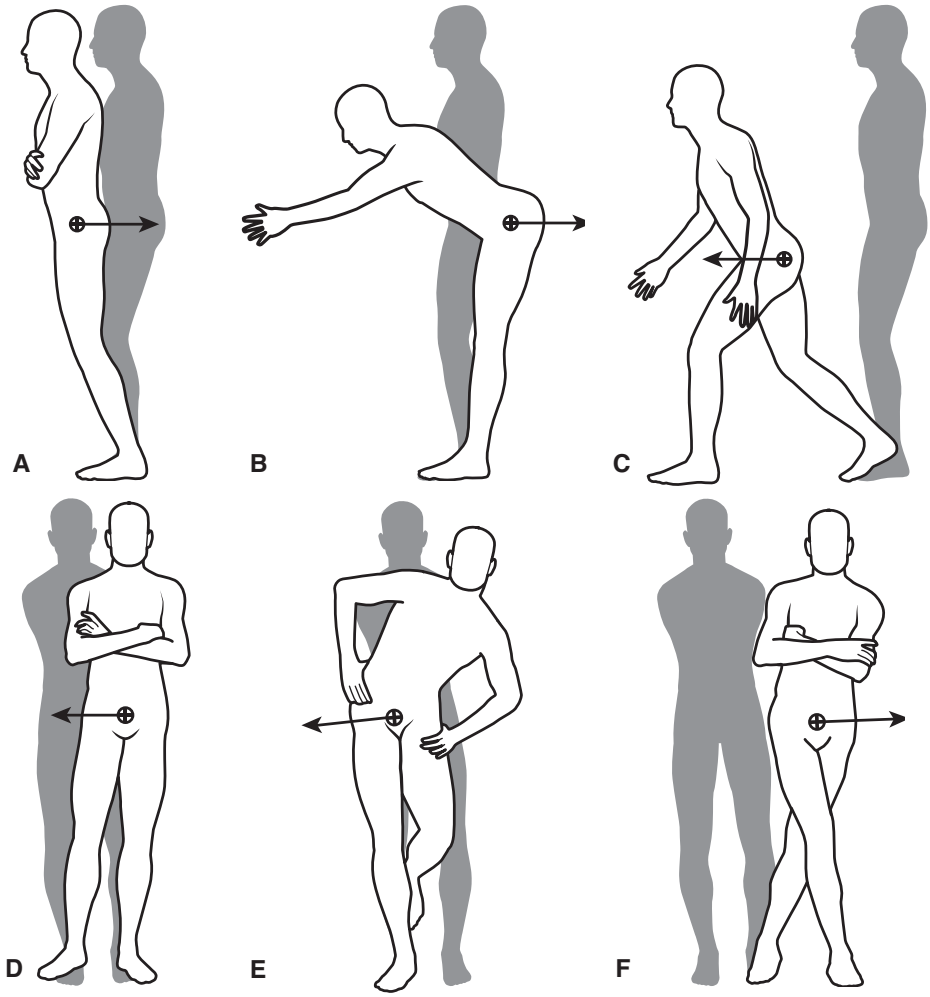

Figure 3.9 Three main postural response strategies used to maintain COM within base of support for balance control in the sagittal plane **(A, B, C)** and in the frontal plane **(C, D, E)**. Ankle strategy can be utilized for small perturbations **(A, D),** hip strategy is required for larger perturbations or more challenging stance conditions **(B, E)**, and finally a step is required if perturbation is large enough to displace COM the body COM beyond the base of support **(C, F)**.

patients typically report visual motion sensitivity, vertigo, and ataxia consistent with vestibular dysfunction, and have histories consistent with vestibular pathology, but often show normal horizontal vestibulo-ocular reflex (VOR) function during rotation testing; these findings suggest that these patients have sustained damage to their vestibular systems, but have not lost a significant amount of horizontal canal vestibular function.[66,111,112] Coordination of head and trunk motions are abnormal in vestibular patients who use excessive hip strategy.

Patients who perceive themselves to be in a different relation to their stability limits than they actually are may show inappropriate postural movement strategies in response to destabilization.[39,112] For example, some patients may not take a step necessary to recover equilibrium in response to a displacement of center of body mass outside their limits of stability because they perceive themselves to be well within their stability limits. In contrast, other patients may make exaggerated postural responses to small perturbations well within their limits of stability because they perceive themselves to be at their limits of stability and therefore at risk for a fall. The type of response may depend both on the type of vestibular dysfunction and the requirements of the task.

In summary, vestibular information is used, with information from other senses, to construct internal maps of the limits of stability, which affect body alignment and recovery from postural disturbances. The information available from the vestibular system and its relationship to information from the other senses changes depending

on the movement strategy used in controlling equilibrium. We suggest that postural strategies are specific prescriptions for mapping interactions among sensory and motor elements of postural control; these maps are, in essence, a method for solving sensorimotor equilibrium and orientation problems. Individuals need a variety of different movement strategies to choose from depending on current, past, and expected environmental conditions. Although the vestibular system may not be prescribing the details of the coordinated motor pattern for postural movements, it seems to be intimately involved in the appropriate selection of movement strategies and in coordination of the head with the rest of the moving body.

Stabilizing the Head and the Trunk

The use of visual and vestibular information for the control of posture is complicated by the fact that these sense organs are located in the inertially unstable head. Because the center of gravity of the head is located above its axis of rotation, any movement of the body will result in head motion. Uncontrolled head motion complicates the use of vestibular information to make estimates of body motion and position. Also, if the range of head motions exceeds that which can be compensated for by the VOR, blurred vision could result. For these reasons, investigators have suggested that the nervous system stabilizes the head with respect to gravity during postural control to simplify the interpretation of vestibular information and to facilitate gaze stabilization.[113-116] In the absence of reliable information about gravity from the vestibular system, or in an attempt to simplify the control of head on trunk movements, the nervous system might stabilize the head with respect to the trunk.[113]

Although there is some movement of the head in space during most locomotor tasks, the position of the head with respect to gravity is often held relatively constant, despite the large movements of the body that can occur during tasks like hopping or running.[114,117,118] For example, neck muscle activations are observed in normal subjects using a hip strategy.[36,38,107,118] In these studies, the neck muscle activations occurred before any large change in head position, and so the activations appear to be a result of an anticipatory control strategy. In other words, these muscle activations serve to prevent the large tilts of the head with respect to gravity that could occur during the large trunk movements, characteristic of a hip strategy.

During tasks like walking and running, patients with profound vestibular loss show increased variability in head position with respect to gravity.[107,119-122] When standing on an oscillating surface with eyes closed, patients with vestibular loss show large oscillations of their heads and trunks in space, unlike healthy control subjects who stabilize their trunks with respect to gravity more and more as the surface oscillations increase in frequency.[123] Figure 3.10 compares stable control of the trunk in a control subject compared with large forward and backward drift of the trunk in a patient with vestibular loss during forward and backward translations of their support surface. Some well-compensated vestibular loss patients, however, can stabilize their trunks in space when their eyes are open, whereas poorly compensated vestibular loss patients cannot use vision to compensate for loss of vestibular function. Despite abnormal top-down coordination of the head and the trunk, vestibular loss subjects showed normal coordination of their legs at all frequencies of surface oscillation, even with eyes closed, consistent with a bottom-up somatosensory control of postural coordination of the legs.[124]

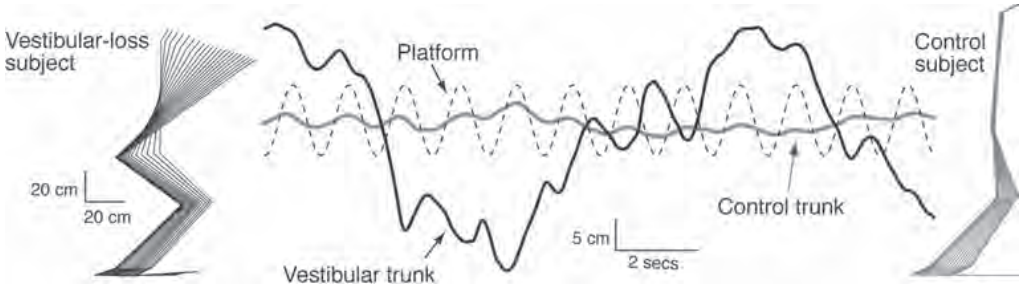

Figure 3.10 Comparison of stable trunk in space in healthy control subject versus large trunk instability in a subject with bilateral vestibular loss during fast, anterior-posterior surface translations. Stick figures show lateral view of trunk, thigh, and legs during surface oscillations. Solid lines show anterior (up) and posterior (down) trunk displacements in a representative control subject (gray line) and vestibular loss subject (darker line). Dashed lines show platform surface oscillations.

In summary, the vestibular system plays an important role in head and trunk stabilization with respect to gravity. In contrast, the vestibular system appears to be less important for the control of the position of the body's COM or postural coordination of the legs, especially when good somatosensory cues about body position are available. Vestibular loss subjects are very successful in substituting light fingertip touch on a stable surface, such as a cane, for loss of vestibular function to stabilize posture.

Clinical Implications

The vestibular system plays many potential roles in postural control. The role it plays in any given postural task will depend both on the nature of the task and on the environmental conditions. When the stabilization of the head or trunk is critical for performance, the vestibular system assumes a very important role. Likewise, when somatosensory (and, to a lesser degree, visual) information is not available, vestibular information for postural control assumes a dominant role. Table 3-1 suggests some tasks and conditions in which vestibular information for postural control is important, and some balance abnormalities that suggest a vestibular disorder. It is also important to note that although this table and this chapter have been devoted to the role of the vestibular system in the control of standing balance, the vestibular system is equally important during locomotion, and problems with sensing movement, orienting to vertical, controlling the position of the COM, and stabilizing the head result in impairments in gait as well as standing balance.

Consider, for example, a task and a set of environmental conditions that occur frequently in clinical examinations of postural control: the patient is asked to maintain verticality in a condition of inaccurate proprioception and absent or inappropriate visual information. This task is appropriate to test the ability of the patient to use vestibular information for the control of posture because visual and somatosensory cues for orientation are poor, and normal subjects typically stabilize their heads with respect to gravity while executing such tasks. When asked to perform this task, patients who have recently lost vestibular function will demonstrate abnormalities in the use of vestibular information in each of its four roles. First, they will have difficulty perceiving and reporting when their bodies or heads are properly oriented to gravity; that is, they will have difficulty identifying when they are upright. Second, they will orient head and body position poorly to gravity, showing tilts rather than upright orientations when asked to right themselves. Third, if the board is tipped suddenly, they may not be able to recover balance, and fourth, head position with respect to gravity will vary a great deal as they attempt to recover from the perturbation.

Although the vestibular system has a prominent role in postural control, some aspects of balance do not rely on the vestibular system or may be compensated for by other systems. Figure 3.11 outlines six underlying systems that contribute to postural control; only Anticipatory Postural Adjustments are unaffected by vestibular pathology. These include *Biomechanical* constraints (such as neck pain and stiffness), *Verticality and Limits of Stability* (impaired and may be difficult to detect because of compensation), *Anticipatory Postural Adjustments* (preserved in people with

■ Table 3-1 **ROLES OF THE VESTIBULAR SYSTEM IN POSTURAL CONTROL**

Role of the Vestibular System	Clinical Assessment	Findings Suggestive of Vestibular Disorder
Sensing and perceiving self-motion	Patient performs head motions and/or positions in different planes	Patient reports abnormal sensations of motion and/or vertigo
Orienting to vertical	Patient stands on inclined surface or on foam with eyes closed	Trunk oriented to support surface instead of gravity; patient falls or sway increases markedly
Controlling center of mass (COM)	Patient stands or walks on a beam	Patient is not able to use hip strategy to control COM and falls
Stabilizing the head	Patient leans or is tilted	Head not stabilized with respect to vertical

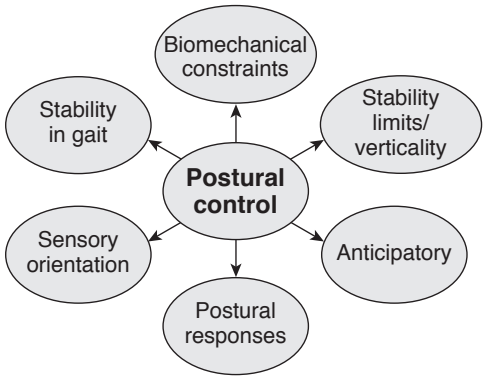

Figure 3.11 This model summarizes the multiple systems underlying postural control. The vestibular system impacts postural control of many, but not all, systems. (Adapted from Horak et al, 2009.[125])

vestibular dysfunction), *Postural Responses* (may be too large causing further loss of balance), *Sensory Orientation* (impaired and may be compensated so need to test in a variety of conditions), and *Stability in Gait* (trunk ataxia but need to test in a variety of conditions such as walking with eyes closed, on unstable surface, or with head turns). These subsystems of postural control comprise the postural systems tested in the Balance Evaluation Systems Test[125] which borrowed items from previously published tests such as the Berg Balance, Functional Reach, Dynamic Gait Index, and CTSIB.[69,126-128] Given the complex nature of the role the vestibular system plays in postural control, one single test may be insufficient to detect the true underlying problem. For example, the BESTest does not include items to address perception of movement (vertigo) or orientation of the head and trunk.

Unfortunately, the assessment of vestibular function in a clinical setting is not so straightforward with patients whose vestibular losses are well compensated. What one observes in these patients is often not the primary action of the vestibular system, but the result of the body's attempt to compensate for the loss. For example, vestibular patients often have difficulty with stiffness in the neck and shoulders. Assuming, however, that this neck stiffness is a primary result of vestibular activation of tonic neck reflexes would be a mistake. The stiffness may be the result of an increase in gain in the cervicocollic system, or a change in strategy; if the head can no longer be aligned to gravity because the direction of gravity cannot be detected, the CNS may choose to stabilize the head to the trunk. Alternatively, the neck stiffness could be a result of voluntary attempts to stabilize head position to limit vertigo or oscillopsia.

Whereas we once assumed that the abnormal balance of patients with vestibular disorders was the simple and necessary consequence of the loss of vestibular reflexes, we now know that the role of the vestibular system in the control of posture is much more complex. In addition to providing (together with vision and somatosensation) the sensory information necessary for orientation and balance, the vestibular system also interacts with the parts of the CNS responsible for expectation and learning. Although automatic and rapid, postural control is also flexible, capable of adaptation to many different sensory environments and musculoskeletal constraints. The role of the vestibular system in postural control will not be fully appreciated until we better understand the complex and multifaceted nature of postural control itself.

CASE STUDY 3-1

A 16-year-old young woman suffered a closed head injury, resulting in loss of vestibular function and hearing in the left ear 1 year ago. She no longer complains of dizziness, but balance problems prevent participation in athletic activities.

Assessment

Perception of Motion: Asymmetrical limits of stability and verticality tilted to the left with eyes closed in sitting and standing. She had complaints of unstable vision and dysequilibrium with head movements and during walking and riding in a car but no dizziness.

Postural Responses: The patient was unable to balance in tasks requiring use of the hip strategy such as standing across a beam, on one foot or tandem.

Sensory Orientation: Sway in stance on a firm surface was normal with eyes open or closed or with sway-referenced vision. However, the patient free fell when attempting to stand on compliant foam with eyes closed or with sway-referenced surface with a vestibular loss pattern.

Head stabilization: Her head was actively fixed on her trunk during gait and she complains of neck stiffness and pain.

Continued

CASE STUDY 3-1

Discussion

Although she had a unilateral loss of vestibular function, she does not seem to be using her remaining vestibular function for postural control. For example, she had a vestibular loss pattern when attempting to stand without visual or surface orientation information and could not use a hip strategy in tasks with compromised surface somatosensory information (i.e., running on a field outdoors on uneven surface). In addition, she fixed her head on her trunk, which can lead to cervical symptoms. The oscillopsia associated with head movements also increased her imbalance.

Physical therapy goals focused on increasing use of remaining vestibular function and increasing use of a hip strategy when tasks require it. If sufficient vestibular function were remaining, treatment of neck symptoms and practice turning the head on the trunk while stabilizing gaze on words during stance and locomotion would allow vision to help recalibrate vestibular function. Gaze stabilization may also improve her oscillopsia with secondary improvements in balance. The patient may temporarily benefit from a cane as a sensory substitution device.

CASE STUDY 3-2

A 50-year-old man was thrown from a horse 3 months ago. He reported severe disorientation when riding in a car, especially near moving traffic. Any busy visual environment, such as crowds, stores, patterns, and windshield wipers is aversive, and he fell when trying to walk on the beach near the ocean waves. He also reports spinning vertigo when he looks up.

Assessment

Perception of motion: Dizziness with sagittal head motion.
Postural responses: Excessive hip strategy and complaints of disorientation when attempting to stop suddenly or turn while walking.
Sensory Orientation: The patient had normal sway when standing on a firm or foam surface with eyes open and closed, even with tandem stance. Trunk sway was excessive, dizziness increased, and he could not stand independently for 30 s when the visual surround was stabilized with sway-referencing (Visual Dependent Pattern).
Head stabilization: Large head and trunk motion when balancing on a firm surface.

Discussion

The Visual Dependent Pattern of sensory organization for posture associated with increased dizziness symptoms with visual motion in the environment is consistent with an abnormally increased weighting to visual input for postural control. His normal sway with eyes closed on a foam surface suggests that he still has adequate vestibular information for postural orientation, but he relies on vision whenever his eyes are open, even when visual inputs poorly reflect body sway. The patient's excessive use of the hip strategy suggests he is not using somatosensory information from the support surface to control his postural sway strategy, resulting in larger than normal head accelerations that may be contributing to his vertigo.

Physical therapy goals included reduction in visual dependence by systematic exposure to moving or stabilized visual environments while increasing use of somatosensory cues from a firm support surface during standing and walking. The patient also practiced use of the ankle strategy by swaying forward and backward slowly at the ankles and when stopping during gait.

Summary

As a sensor of gravitational and head acceleration, the vestibular system plays many important roles in postural control. The role it plays in any given postural task depends both on the nature of the task and on the environmental conditions. Furthermore, because the vestibular system is both a sensory and a motor system, it plays many different roles in postural control. In this chapter, we explore four important roles of the vestibular system in postural control. First, we discuss the role of the vestibular system in the perception of body position and self-motion. Second, we

discuss its role in orienting the trunk to vertical using sensory orientation and weighting-appropriate sensory cues under different sensory environments. Third, we discuss the role of the vestibular system in controlling the position of the body's COM, both for static positions and dynamic movements via postural responses. Fourth, we discuss its role in the vestibular system in the control of standing balance. The vestibular system is equally important during locomotion so problems with sensing movement, orienting to vertical, controlling the position of the COM, and stabilizing the head results in similar impairments in gait as in standing balance.

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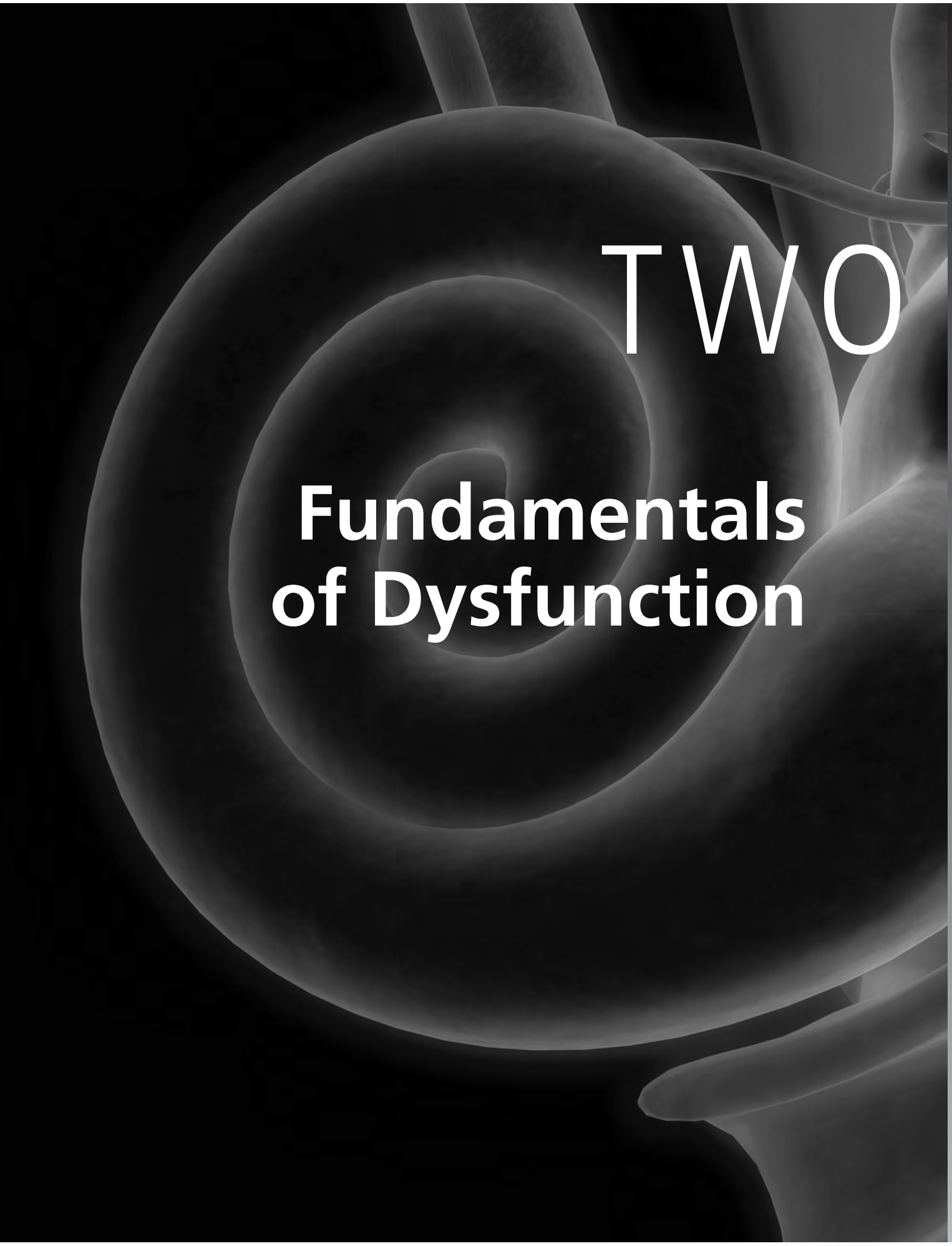

TWO

Fundamentals of Dysfunction

CHAPTER 4

Vestibular System Disorders

Michael Fetter, MD

Peripheral vestibular dysfunction, which involves the vestibular end organs and/or the vestibular nerve, can produce a variety of signs and symptoms. A thorough evaluation by a physician is needed to identify the specific pathology behind the patient's complaints of vertigo or disequilibrium. Patient history is the main key for diagnosis, supported by a careful otoneurologic examination. Determining whether vestibular rehabilitation is appropriate and, if it is, which approach should be used is based in part on the patient's diagnosis. This chapter describes the clinical presentation of the more common peripheral vestibular disorders. The results of diagnostic tests, and the medical, surgical, and rehabilitative management of each of these disorders is presented as an overview only, because this material is covered in detail in other chapters.

Benign Paroxysmal Positional Vertigo

Benign paroxysmal positional vertigo (BPPV) is the most common cause of vertigo. Typically, a patient with BPPV complains of brief episodes of vertigo precipitated by rapid changes of head posture. Sometimes symptoms are brought about by assuming very specific head positions. Most commonly these head positions involve rapid extension of the neck, often with the head turned to one side (as when looking up to a high shelf or backing a car out of a garage) or lateral head tilts toward the affected ear. The symptoms often appear when a patient rolls from side to side in bed. Patients can usually identify the offending head position, which they often studiously avoid. Many patients also complain of mild postural instability between attacks. The vertigo lasts only 30 seconds to 2 minutes (usually less than 1 minute) and disappears even if the precipitating position is maintained. Hearing loss, aural fullness, and tinnitus are not seen in this condition, which most commonly occurs spontaneously in the elderly population but can be seen in any age group after even mild head trauma. Women are more commonly affected than men (2:1) and the right labyrinth is slightly more often involved than the left (1.4:1). Bilateral involvement can be found in 7.5%; 90% of these are traumatic cases.[1] Spontaneous remissions are common, but recurrences can occur, and the condition may trouble the patient intermittently for years with a high recurrence rate of up to 50% within 10 years.[2]

Evaluation should include a careful otoneurologic examination, the most important part being the history. In the case of the most common posterior canal BPPV, a key diagnostic maneuver is the Dix-Hallpike positioning test[3] while the examiner observes the patient's eyes with a pair of Frenzel lenses or in combination with videooculography monitoring. Electronystagmography (ENG) can also be used, but because the typical response in posterior BPPV is a combined vertical-torsional nystagmus, recordings might be hard to interpret, because this technique cannot measure torsional eye movements and in the vertical dimension recordings might be hampered by artifacts.

A typical response is induced by rapid position changes from the sitting to the head-hanging right or left position. Vertigo and nystagmus begin with a latency of 1 or more seconds after the head is tilted toward the affected ear and increase in severity within about 10 seconds to a maximum accompanied by a sensation of discomfort and apprehension that sometimes causes the patient to cry out and attempt to sit up. The symptoms diminish gradually after 10 to 40 seconds and ultimately abate, even if the precipitating head position is maintained. The nystagmus is mixed upbeat and torsional with a slight horizontal component: The direction corresponds very closely to the plane of the offending semicircular canal, very similar to experimental stimulation of the afferents of the posterior semicircular canal of the dependent ear.[4] Because the rotation axis of the eye is fixed and parallel to the axis of the offending canal, the nystagmus changes with the direction of gaze, becoming more torsional as the patient looks toward the dependent ear with the gaze line parallel to the axis of the offending canal and more vertical as the patient looks toward the higher ear with the gaze line orthogonal to the axis of the offending canal.

Sometimes, a low-amplitude, secondary nystagmus, directed in the opposite direction, may occur If the patient then quickly sits up, a similar but usually milder recurrence of these symptoms occurs, the nystagmus being directed opposite to the initial nystagmus. Repeating this procedure several times will decrease the symptoms. This adaptation of the response is of diagnostic value, because a clinical picture similar to that of BPPV can be created by cerebellar tumors. In the latter, though, there is no habituation of the response with repetitive testing. Further diagnostic criteria indicating a central positional nystagmus are as follows: (1) the condition does not subside with maintenance of the head in the precipitating position, (2) the nystagmus may change direction when different head positions are assumed, (3) the nystagmus may occur as downbeat nystagmus only in the head-hanging position, or (4) the nystagmus direction does not coincide with the previous stimulation axis. BPPV must also be differentiated from positional nystagmus in Ménière's disease, perilymph fistulas, and alcohol intoxication (positional alcohol nystagmus [PAN]) and can also be seen as a transient phenomenon in vestibular migraine.

A few patients do not display the typical torsional upbeat nystagmus but show a strong horizontal nystagmus, which nevertheless follows a similar pattern of buildup and decline but often over a longer period (posterior canal BPPV: duration <30 s, latency 2–15 s; lateral canal BPPV: duration >30 s, latency <3 s). This horizontal nystagmus indicates a lateral canal variant of BPPV and is best provoked when repositioning takes place in the plane of the lateral semicircular canals. For this maneuver (Pagnini-McClure) the patient lies on his back and the head is moved either to right or left ear-down position about the long body axis. Two types of lateral canal variant BPPV can be observed. The canalolithiasis type is usually geotropic (80% of the cases with lateral canal variant) with the nystagmus beating toward the lowermost ear being more vigorous when lying on the offending ear. Sometimes one can also see an ageotropic (the nystagmus beating to the uppermost ear) nystagmus, that is transient in canalolithiasis or more long lasting without onset latency in the case of cupulolithiasis. In this case the offending ear is the one where the nystagmus is less vigorous when lying on that ear.

The classic explanation of the underlying pathophysiology (cupulolithiasis) was first described by Schuknecht in 1969.[5] His study of the temporal bones of two patients afflicted with this disorder showed deposition of otoconial material at the cupula of the posterior semicircular canal. The cupulolithiasis theory suggests that the debris adheres to the cupula, making it denser than the surrounding endolymph and thereby susceptible to the pull of gravity. This theory, however, implies that a positioning maneuver should result in an enhanced positioning response with a nystagmus initially beating in the direction of an ampullopetal stimulation. This nystagmus should occur immediately after the positioning maneuver, and should change direction as gravity drags the cupula down. The nystagmus, however, should not subside as long as the head-down position is maintained; that is, one would expect positional instead of positioning nystagmus. None of these features are typically seen in posterior canal BPPV but sometimes are seen in ageotropic lateral canal BPPV.

Brandt and Steddin emphasized a second theory, canalithiasis, which better explains the typical features of BPPV.[6] It suggests that the debris of a higher density than the endolymph is free-floating in the canal. When the head is moved in the plane of that canal, the debris sinks to the lowest point in the canal, causing the endolymph to move and deflecting the cupula by suction or pressure (like a plunger), depending on the direction it moves. This theory accords with the direction of the nystagmus and also allows for a latency.

If symptoms persist longer than expected or if the positional nystagmus shows atypical features, further investigation, such as magnetic resonance imaging (MRI), should be made to assess for unusual causes of positional vertigo, such as acoustic neuroma or tumors of the fourth ventricle.

BPPV is usually a self-limiting disorder and commonly resolves spontaneously within weeks to months. Simple vestibular exercises or maneuvers aimed at dispersing the otolithic debris from the cupula or to remove the

debris from the canal can significantly speed recovery[7]; antivertiginous drugs are usually not helpful, unless used to ease the vertiginous symptoms while performing the liberatory maneuvers. In 1988, Semont and colleagues [8] introduced a single liberatory maneuver, and Epley,[9] in 1992, proposed a variation, later modified by Herdman and associates[10] (see also Chapter 20). For the lateral canal variant, there also exist meanwhile helpful liberatory maneuvers (Lempert, Gufoni). With these newer liberatory maneuvers, most patients can be treated successfully within less than a week. In many cases, immediate recovery after one repositioning maneuver can be seen. Symptoms persist in only a few patients in spite of compliance with vestibular exercises. For more severe symptoms unresponsive to exercises, three surgical options are available for relief. The first is transmeatal posterior ampullary nerve section (also known as singular neurectomy). The other two options are partitioning of the labyrinth using a laser technique and nonampullary plugging of the posterior semicircular canal. Nonampullary plugging seems to be a safe and effective alternative to singular neurectomy for the small group of patients with physically intractable BPPV.

Vestibular Neuritis

Acute unilateral (idiopathic) vestibulopathy, also known as vestibular neuritis, is the second most common cause of vertigo. Although in most cases a definitive etiology is never proved, evidence to support a viral etiology (similar to Bell's palsy or sudden hearing loss) comes from histopathological changes of branches of the vestibular nerve in patients who have suffered such an illness and the sometimes epidemic occurrence of the condition. [11] Onset is often preceded by the presence of a viral infection of the upper respiratory or gastrointestinal tracts. The associated viral infection may be coincident with the vestibular neuritis or may have preceded it by as long as 2 weeks. The chief symptom is the acute onset of prolonged severe rotational vertigo that is exacerbated by movement of the head, associated with spontaneous horizontal-rotatory nystagmus beating toward the good ear, postural imbalance with a tendency to fall toward the affected side, and nausea. Hearing loss is not usually present, but when it is then mumps, measles, and infectious mononucleosis, among other infections, have been implicated. The latter condition should also alert the physician to consider other diagnoses (i.e., ischemia of labyrinth artery, Ménière's disease, acoustic neuroma, herpes zoster oticus, Lyme disease, or neurosyphilis). The condition mainly affects those aged between 30 and 60 years, with a peak for women in the fourth decade and men in the sixth decade.

If examined early, the patient may manifest an irritative nystagmus from the acute phases of the inflammation. Usually the patient is examined after these initial findings have given way to a more paralytic, or hypofunctional, pattern. Caloric testing invariably shows ipsilateral hyporesponsiveness or nonresponsiveness (horizontal canal paresis). The possibility that the three semicircular canals and the otoliths (utricle and saccule) may be separately involved in partial labyrinthine lesions is suggested by the occasional observation (about 10% of the patients) of an acute unilateral vestibulopathy and a BPPV simultaneously in the same ear of a patient (Lindsay-Hemenway-syndrome).[12] With three-dimensional measurements of the vestibulo-ocular reflex in patients with vestibular neuritis, this notion could be confirmed. Patients with this condition most often showed a partial involvement of only the superior vestibular nerve portion (subserving the anterior and lateral semicircular canals, the utricle, and a small part of the saccule) leaving part of the saccule and the posterior semicircular canal afferents intact.[13] The symptoms usually abate after a period of 48 to 72 hours, and gradual return to normal balance occurs over approximately 6 weeks. Rapid head movements toward the lesioned side, however, can still cause slight oscillopsia of the visual scene and impaired balance for a short moment. Recovery is produced by the combination of central compensation of the vestibular tone imbalance, aided by physical exercise, and peripheral restoration of labyrinthine function. The latter is found in about two-thirds of the patients.

The differential diagnosis should initially include other causes of acute vertigo, and careful history taking, physical examination, and an audiogram are required. Physical examination should include a neurological examination with attention to cranial nerve findings and cerebellar testing. Careful otoscopy is performed to rule out the presence of a potential otologic infectious process as the source of a toxic serous labyrinthitis. Fever in the presence of chronic ear disease and labyrinthitis suggests suppuration and meningitis. Commonly, a toxic labyrinthitis is the result of a well-defined event such as surgery or trauma.

More recently, the so-called acute vestibular syndrome in elderly patients with at least one risk factor for vascular disease has been carefully reevaluated. Kattah and coworkers found that in this situation more than ¾ of the patients had a central disorder (ischemia, bleeding) rather than vestibular neuritis.[14] They further found, that with careful examination for vertical skew, direction changing nystagmus in different directions of gaze (gaze evoked nystagmus) and normal head impulses, a central cause could be detected with a sensitivity of 100% and a specificity of 96%. This important finding for emergency room evaluation has been summarized with the acronym

HINTS (**H**ead **I**mpulse (normal), **N**ystagmus (in different directions), **T**est for **S**kew).

Initial treatment of vestibular neuritis is accomplished with the use of vestibular suppressants such as the antihistamine dimenhydrinate or the anticholinergic scopolamine. In addition, bed rest is very helpful early on in the course of the disease. After the most severe vertigo and nausea have passed (after 24 to 72 hours), then ambulation may resume with assistance; independent ambulation may be achieved over the next few days. At the same time, the administration of vestibular suppressants should be greatly diminished or, even better, stopped completely because they prolong the time required to achieve central compensation. To further speed up the process of recuperation, vestibular exercises challenge the compensatory mechanisms of the central nervous system (CNS), stimulating adaptation. These exercises are designed to improve both gaze stability and postural stability (see Chapter 22).[15]

Animal experiments have shown that alcohol, phenobarbital, chlorpromazine, diazepam, and ACTH antagonists retard compensation; caffeine, amphetamines, and ACTH accelerate compensation.[16] Recently Strupp and coworkers performed, in 141 patients with vestibular neuritis, a prospective randomized trial of methylprednisolone, valacyclovir, and the two in combination within 3 days after the onset of symptoms.[17] The authors showed that methylprednisolone significantly improves the recovery of peripheral vestibular function (from 39% in the placebo group to 62% in the methylprednisolone group), whereas valacyclovir does not. The study shows that steroids significantly improve the recovery of peripheral vestibular function in humans with vestibular neuritis. The recurrency rate of vestibular neuritis is very low (2% within 5 to 10 years) and mostly then the other side is involved. In the rare cases of recurrence on the same side, this usually happens within 6 months, but symptoms are milder.[18]

Ménière's Disease and Endolymphatic Hydrops

Ménière's disease is a disorder of inner ear function that can cause devastating hearing and vestibular symptoms. The typical attack is experienced as an initial sensation of fullness of the ear, a reduction in hearing, and tinnitus, followed by rotational vertigo, postural imbalance, nystagmus, and nausea and vomiting after a few minutes. This severe disequilibrium (vertigo) will persist anywhere from approximately 30 minutes to 24 hours. Gradually, the severe symptoms will abate, and the patient is generally ambulatory within 72 hours. Some sensation of postural unsteadiness will persist for days or weeks, and then normal balance will return. During this recuperation time, hearing gradually returns. Hearing may return to the preattack baseline or there may be residual permanent sensorineural hearing loss, most commonly in the lower frequencies. The rare transient improvement of hearing during the attack is known as the Lermoyez phenomenon. Tinnitus will also usually diminish as hearing returns. As the disease progresses, hearing fails to return after the attack, and after many years, the symptoms of vertigo may gradually diminish in frequency and severity. Some patients may suddenly fall without warning; these events, which may occur in later stages of the disease, are referred to as Tumarkin's otolithic crisis and should be differentiated from other forms of drop attack.

The typical form of Ménière's disease is sometimes not complete and is called vestibular Ménière's disease, if only vestibular symptoms and aural pressure are present, or cochlear Ménière's disease, if only cochlear symptoms and aural pressure are encountered.[19]

The disease is about equally distributed between the sexes and usually has its onset in the fourth to sixth decades of life. However, there are reports of children as young as 6 years of age with classical Ménière's disease.[20] About 15 percent of the patients have blood relatives with the same disease, suggesting genetic factors. The incidence of bilateral involvement ranges between 33% after 10 years and 50% after 20 years.[21]

A phenomenon fundamental to the development of Ménière's disease is endolymphatic hydrops. Whether endolymphatic hydrops itself is the cause of the symptoms characteristic of Ménière's disease or whether it is a pathological change seen in the disease is still unclear. The development of hydrops is generally a function of malabsorption of endolymph in the endolymphatic duct and sac. Malabsorption may itself be a result of disturbed function of components comprising the endolymphatic duct and sac, mechanical obstruction of these structures, or altered anatomy in the temporal bone. Endolymph is produced primarily by the stria vascularis and flows both longitudinally (along the axis of the endolymphatic duct toward the endolymphatic sac) and radially (across the membrane of the endolymphatic space into the perilymph system). Ménière's disease is generally a consequence of altered longitudinal flow, usually evolving over a course of many years. Experimental obstruction of the endolymphatic duct will routinely result in endolymphatic hydrops in many animal models.[22] Lesions in the temporal bone that have been associated with the development of hydrops include fractures of the temporal bone, perisaccular fibrosis, atrophy of the sac, narrowing of the lumen in the endolymphatic duct, otitis media, otosclerotic foci enveloping the vestibular aqueduct, lack of vascularity surrounding the

endolymphatic sac, syphilitic osteitis of the otic capsule, and leukemic infiltrations, to name just a few. Anatomically, ears affected by Ménière's disease are likely to demonstrate hypodevelopment of the endolymphatic duct and sac, periaqueductal cells, and mastoid air cells. Therefore, one can postulate a cause-and-effect relationship between constricted anatomy in the temporal bone and malabsorption of endolymph.

Any explanation of the clinical symptoms of Ménière's disease should account for all the symptoms, including rapid or prolonged attacks of vertigo, disequilibrium, positional vertigo during and between attacks, fluctuating progressive sensorineural hearing loss, tinnitus, aural pressure, inability to tolerate loudness, and diplacusis. These symptoms probably result from both chemical and physical mechanisms. Physical factors can tamponade the cochlear duct, contributing to fluctuating progressive sensorineural hearing loss and other cochlear symptoms, whereas distension of the otolithic organs can physically affect the crista ampullaris, resulting in vestibular symptoms. The prolonged nystagmus and vertigo are commonly believed to be caused by periodic membrane ruptures with subsequent transient potassium palsy of vestibular nerve fibers.

Useful diagnostic tests include the audiogram and ENG. Typically, the audiogram displays an ipsilateral sensorineural hearing loss involving the lower frequencies. Fluctuation in discrimination scores is often seen, with a long-term trend toward poor scores. ENG may demonstrate a unilateral vestibular weakness on caloric testing, again involving the ear symptomatic for pressure, hearing loss, and tinnitus. Electrocochleography is useful in cases that are unclear. The finding of enlarged summating potentials in the suspected ear is diagnostic of endolymphatic hydrops. Furthermore, Taylor and colleagues found, in 33% of patients with Ménière's disease, pathological cervical vestibular-evoked myogenic potentials (cVEMP) indicating a specific involvement of the saccule in this disease.[23]

A brainstem-evoked acoustic response (BAR) must be done in those cases with findings of retrocochlear pathology on routine audiometry to screen for cochlear nerve or brainstem pathology. If the BAR is found to be positive, then MRI scanning with the use of intravenous gadolinium should be done to assess for central nervous system pathology or eighth-nerve schwannoma.

Treatment in the remission phase aims to reduce the frequency of the attacks and preserve hearing without distressing tinnitus. Dietetic programs, including restriction of salt, water, alcohol, nicotine, and caffeine, are as valueless in treating the disease as are physical exercise or avoidance of exposure to low temperatures. Stellate ganglion blocks, diuretics, vasoactive agents, tranquilizers, neuroleptics, and lithium have been employed under the mistaken assumption that diminishing endolymphatic hydrops is possible by changing inner-ear blood flow, osmotic diuresis, or central sedation. There has never been prospective proof of the efficiency of these therapies. The histamine derivative betahistine has been advocated as the drug of first choice. Findings from a 1-year prospective double-blind study showed that this treatment is preferable to leaving the disease untreated.[24] Recently, the recommended dosage has been continuously increased up to 3*48 mg/day for at least 1 year.[25] The action is attributed to improvement of microcirculation of the stria vascularis, but betahistine also has inhibitory effects on polysynaptic vestibular neurons. Adjunct medications in the form of vestibular suppressants other than betahistine are to be used primarily during the acute episodes of vertigo and should be discouraged as a chronic daily medication. If the betahistine treatment does not result in a significant reduction of vertigo attacks within 6 months, intratympanic application of steroids can be tried.[26] This procedure proved to be effective; the evidence level, however, is low.[27]

In addition to pharmacologic therapies, many patients with Ménière's disease require psychological support to help cope with the frustrations and changes brought about by their medical condition. Those patients in whom the vertigo becomes disabling by virtue of increased severity or frequency of attacks despite maximal medical therapy would be considered candidates for surgical intervention. Only about 1% to 3% of patients ultimately require surgical treatment, because the success of regular endolymphatic sac shunt operations has been shown to be a placebo effect.[28]

Sacculotomy has been proposed by a variety of authors as a method of relieving the pressure buildup in the endolymphatic chamber. Long-term success rates for this procedure are not yet available, but significant hearing loss is observed in 50% of patients undergoing cochleosacculotomy. Advantages are ease of performance, utility in elderly patients as a first procedure under local anesthesia, and little risk other than hearing loss.

Intratympanic treatment with ototoxic antibiotics such as gentamicin sulfate, instilled via a plastic tube inserted behind the annulus via the transmeatal approach, is obviously able to damage selectively the secretory epithelium (and thereby improve endolymphatic hydrops) before significantly affecting vestibular and cochlear function.[29]

The current treatment most successful is vestibular nerve section. This procedure is indicated in individuals with serviceable hearing in whom maximal medical therapy has been unsuccessful in controlling vertigo. Success rates in the range of 90% to 95% have been reported by numerous authors. The newer technique of focused ultrasound seems to have an advantage over open surgery in

that partial ablation of vestibular function (with preservation of hearing) can be performed without invading the labyrinth.

In patients with hearing loss, destructive procedures are also possible, such as transmeatal, transmastoid, or translabyrinthine labyrinthectomy. The success rate is 95%. An extension of this surgery is the translabyrinthine vestibular nerve section, shown to eliminate vertigo in 98% of cases. However, particularly in elderly patients, ablative surgical procedures may cause long-lasting postural imbalance because of the reduced ability of central mechanisms to compensate for the postoperative vestibular tone imbalance.

Vestibular exercises are not appropriate in patients with Ménière's disease unless there is permanent loss of vestibular function. Vestibular exercises are designed to induce long-term changes in the remaining vestibular system or to foster the substitution of other strategies to compensate for the loss of vestibular function. In Ménière's disease, the vestibular dysfunction is episodic, and between episodes the system usually returns to normal function. Some patients developed a loss of vestibular function at the end stages of the disease, and for those patients, vestibular rehabilitation may be appropriate. Vestibular exercises are also beneficial in those patients who have surgical destruction of the inner ear.

Perilymphatic Fistula

Perilymphatic fistula may lead to episodic vertigo and sensorineural hearing loss owing to the pathological elasticity of the bony labyrinth. Most commonly, these fistulas occur at the round and oval windows of the middle ear. Classically, a history of (often minor) head trauma, barotrauma, mastoid or stapes surgery, penetrating injury to the tympanic membrane, or vigorous straining precedes the onset of sudden vertigo, hearing loss, and loud tinnitus. The patients often report a "pop" in the ear during the precipitating event. Later on, patients with fistula may complain of imbalance, positional vertigo, and nystagmus as well as hearing loss. Tullio phenomenon—vestibular symptoms that include vertigo, oscillopsia, nystagmus, ocular tilt reaction, and postural imbalance induced by auditory stimuli—is usually the result of a perilymph fistula, in most cases because of superior canal dehiscence, but subluxation of the stapes foot plate and other ear pathology may be responsible. The symptoms will often subside while at rest only to resume with activity. Sneezing, straining, nose blowing, and other such maneuvers can elicit the symptoms after the initial event. Perilymph fistulas probably account for a considerable proportion of those patients presenting with vertigo of unknown origin. Diagnosing perilymph fistula is difficult because of the great variability of signs and symptoms and the lack of a pathognomonic test. In the acute phase, medical treatment is universally recommended because these fistulas usually heal spontaneously, and the results of surgical interventions are not encouraging.[30]

Physical examination, particularly otoscopy, is important. In the cases of head trauma and barotrauma, hemotympanum is often seen as an early finding. In cases of penetrating injury to the ear, a tympanic membrane perforation makes the likelihood of ossicular discontinuity with fistula very high. A useful clinical test consists of applying manual pressure over the tragus or applying pressure to the tympanic membrane with the pneumatic otoscope; a positive test is indicated by the evocation or exacerbation of vertigo (Hennebert's sign) or the elicitation of nystagmus. Audiometric findings usually demonstrate a mixed or sensorineural hearing loss, depending on the mechanism of injury. This loss may be quite severe and usually involves the high frequencies more than the low frequencies. ENG with caloric testing may be normal or show a unilateral weakness in the affected ear. The specificity of the clinical fistula tests can be augmented by recording eye movements or measuring body sway as pressure on the tympanic membrane is increased. Despite refinements, these tests remain unreliable in detecting all fistulas. The diagnosis remains essentially a historical one, and in those patients with a suggestive history and symptoms, treatment is indicated. Often the only manner in which the diagnosis is made definitively is at the time of surgical exploration by tympanoscopy as the patient performs Valsalva maneuvers.

Medical treatment consists of absolute bed rest with the head elevated for 5 to 10 days. Mild sedation with tranquilizers; avoidance of straining, sneezing, coughing, or head-hanging positions; and the use of stool softeners is important for reduction of further explosive and implosive forces that may activate perilymph leakage.[31]

When symptoms persist for longer than 4 weeks, or if hearing loss worsens, exploratory tympanotomy is indicated. Considerable controversy persists surrounding the frequency with which perilymph fistulas are found at surgery. Surgical management consists of middle ear exploration and packing of the oval and round window areas with fat, Gelfoam, and areolar and/or fibrous tissue. These areas are packed whether or not a clear-cut fistula is demonstrated. Reported success rates for this treatment vary between 50% and 70% and likely reflect some element of variable patient selection.

More recently, a new variant of perilymphatic fistula causing episodic vertigo has been described by Minor and coworkers.[32] The disease is a result of dehiscence of the superior semicircular canal. It is probably the most frequent form of a fistula and probably most often overlooked.

Hallmarks are vertigo spells induced by pressure increase like coughing, sneezing, or loud noises (Tullio phenomenon). Vertical-torsional eye movements in the plane of the defective superior semicircular canal can be observed in precipitating conditions. Pfammater and coworkers showed in a study with 27 patients that cVEMPs are typically altered in this disease.[33] Depending on the size of the dehiscence (≥2.5 mm) the stimulation threshold was significantly reduced and the amplitude of the cVEMPs was increased. In more than half of the cases symptoms start after even mild head- or barotrauma. The diagnosis can be made by high-resolution CT of the temporal bone showing a dehiscence of the apical part of the superior semicircular canal. Very recently, posterior canal dehiscence and bilateral and combined involvement of anterior and posterior canals have also been reported.[34]

Vestibular Paroxysmia (Disabling Positional Vertigo)

Neurovascular cross-compression of the root entry zone of the vestibular nerve can elicit disabling positional vertigo.[35] The term describes a heterogeneous collection of signs and symptoms rather than a reliable diagnosable disease entity Brandt and Dieterich[36] proposed the following criteria: (1) short and frequent attacks of rotational or to-and-fro vertigo lasting from seconds to minutes, (2) attacks frequently dependent on particular head positions and modification of the duration of the attack by changing head position, (3) hypacusis and/or tinnitus permanent or during the attack, (4) measurable auditory or vestibular deficits by neurophysiological methods, and (5) positive response to antiepileptic drugs (carbamazepine). With a special MRI technique (CISS-3D), vascular loops around the most proximal part of the vestibular nerve (first 1.5 mm) can often be detected in this disorder, but there is a considerable amount of false positive and false negative results.

Neurovascular cross-compression can cause local demyelinization of the root entry zone of the eighth nerve. Ephaptic transmission between bare axons or central hyperactivity initiated and maintained by the peripheral compression are the suggested mechanisms. Analogous to trigeminal neuralgia, antiepileptic drugs are the first choice of medical treatment of the condition before surgical microvascular decompression is contemplated.

Bilateral Vestibular Disorders

Bilateral vestibulopathy may occur secondary to meningitis, labyrinthine infection, otosclerosis, Paget's disease, polyneuropathy, bilateral tumors (acoustic neuromas in neurofibromatosis), endolymphatic hydrops, bilateral sequential vestibular neuritis, cerebral hemosiderosis, ototoxic drugs, inner-ear autoimmune disease, or congenital malformations, but in many cases no clear cause can be found (idiopathic bilateral vestibulopathy). Autoimmune conditions affecting the inner ear are rare but distinct clinical entities,[37] characterized by a progressive, bilateral sensorineural hearing loss often accompanied by a bilateral loss of vestibular function. Other autoimmune-mediated disease is often present in the afflicted patients; examples include rheumatoid arthritis, psoriasis, ulcerative colitis, and Cogan's syndrome (iritis accompanied by vertigo and sensorineural hearing loss). The history is the most useful diagnostic tool. Support for the diagnosis can be obtained by blood testing for complete blood count, erythrocyte sedimentation rate, rheumatoid factor, and antinuclear antibodies. Western-blot precipitation studies to look for anticochlear antibodies can be done in some research centers and may be the future definitive test of choice in these cases.

Little is known about how autoimmune disorders cause otologic symptoms. As with other autoimmune conditions, the otologic symptoms may occur as a direct assault by the immune system in the form of humoral and cellular immunity directed at the inner ear. Another mechanism of injury may be related to the deposition of antibody-antigen complex in capillaries or basement membranes of inner ear structures. Further immunologic studies of temporal bones harvested from deceased patients who had clinical evidence of autoimmune inner ear involvement may shed some light on the underlying process.

Because autoimmune vestibulopathy usually affects both ears, therapy is almost exclusively medical. Vestibular suppressants are most useful in controlling the more severe exacerbations of vertigo. The use of corticosteroids and some cytotoxic agents (cytoxan, methotrexate) has been shown to provide relief in some patients. There is some newer evidence to suggest that serum plasmapheresis may play a more prominent role in controlling this disease in the future. The natural history of the disease leads to eventual bilateral vestibular ablation. This end result is almost inevitable unless the underlying process can be arrested with treatment or arrests spontaneously.

The most common toxic cause of acute vertigo is ethyl alcohol. We know that positional changes exacerbate the vertigo of a hangover. The reason may be that alcohol diffuses into the cupula and endolymph at different rates and so creates a density gradient, making the cupula sensitive to gravity.[38] Other agents that may produce vertigo include organic compounds of heavy metals and aminoglycosides. The aminoglycosides are notorious for causing irreversible failure of vestibular function without vertiginous warning or hearing loss. Thus, monitoring of vestibular function may be necessary during such therapy.

Independent of vestibulopathies produced by ototoxins, single cases of "progressive vestibular degeneration" of unknown origin have been described, with the following factors in common: repeated episodes of dizziness relatively early in life, bilateral loss of vestibular function with retention of hearing, and freedom from other neurological disturbances.[39]

Alport's (inherited sensorineural deafness associated with interstitial nephritis), Usher's (inherited sensorineural deafness associated with retinitis pigmentosa), and Waardenburg's syndromes (inherited deafness associated with facial dysplasia) usually cause bilateral labyrinthine deficiency when they affect the vestibular system. Congenital vestibular loss is secondary to either abnormal genetic or intrauterine factors including infection (most commonly rubella and cytomegalovirus), intoxication (thalidomide), or anoxia.

Controlled physical exercises can improve the condition in patients with permanent bilateral vestibulopathy by recruiting nonvestibular sensory capacities such as the cervico-ocular reflex and proprioceptive and visual control of stance and gait (see Chapters 9 and 23).

Summary

This chapter describes the clinical presentation of the more common peripheral vestibular disorders and the differential diagnosis to central origins of vertigo. Although the symptomatology of a certain peripheral vestibular disorder might be rather specific, as in acute unilateral vestibular loss, the cause can be rather different, ranging from infection to ischemia to traumatic lesions. A thorough evaluation, therefore, should, in addition to the specific otoneurological investigation, always include a detailed history and a general physical examination. For a quick review, Table 4-1 summarizes the hallmarks of the peripheral vestibular disorders treated in this chapter.

■ Table 4-1 **SUMMARY OF VESTIBULAR SYSTEM DISORDERS**

	BPPV	Vestibular Neuritis	Ménière's Disease	Fistula	Nerve Compression	Bilateral Vestibular Disorder
Vertigo	+	+	+	+	+	–
Type	Rotational	Rotational	Rotational	Rotational/ linear	Rotational/ linear	–
Nystagmus	+	+	+	+	+	–
Duration	30 sec–2 min	48–72 hr	20 min–24 hr	Seconds	Seconds to minutes	Permanent
Nausea	–/(+)	+	+	–	+	–
Postural Ataxia	–/(+)	+	+	+	+	++
Specific symptoms	Onset latency, adaptation	Acute onset	Fullness of ear, hearing loss, tinnitus	Loud tinnitus, Tullio sign, Hennebert's sign	Frequent attacks, tinnitus, hypacusis	Gait ataxia
Precipitating action	Positioning, turning in bed	–	–	Head trauma, ear surgery, sneezing, straining, nose blowing	Changing head position	–

Key: – absent, + present, ++ very strong.

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CHAPTER 5

Vestibular Lesions of the Central Vestibular Pathways

Marianne Dieterich, MD ■ *Thomas Brandt, MD, FRCP*

Vestibular pathways run from the eighth nerve and the vestibular nuclei through ascending fibers, such as the ipsilateral or contralateral medial longitudinal fasciculus (MLF), the brachium conjunctivum, or the ventral tegmental tract to the ocular motor nuclei, the supranuclear integration centers in the rostral midbrain, and the vestibular thalamic subnuclei. From there they reach several cortex areas through the thalamic projection. Another relevant ascending projection reaches the cortex from vestibular nuclei via vestibular cerebellum structures.

In the majority of cases, central vestibular vertigo syndromes are caused by dysfunction or a deficit of sensory input induced by a lesion. In a small proportion of cases, they are a result of pathological excitation of various structures, extending from the peripheral vestibular organ to the vestibular cortex. Because peripheral vestibular disorders are always characterized by a combination of perceptual, ocular motor, and postural signs and symptoms, central vestibular disorders may manifest as "a complete syndrome" or with only single components. The ocular motor aspect, for example, predominates in the syndromes of upbeat or downbeat nystagmus. Lateral falls may occur without vertigo in vestibular thalamic lesions (thalamic astasia) or as lateropulsion in Wallenberg's syndrome.[1,2]

Clinical Classification of Central Vestibular Disorders

The "elementary" neuronal network of the vestibular system is the di- or trisynaptic vestibulo-ocular reflex (VOR). A useful simple clinical classification of central vestibular syndromes is based on the three major planes of action of the VOR (Fig. 5.1): yaw, roll, and pitch.[2-4]

The plane-specific vestibular syndromes are determined by ocular motor, postural, and perceptual signs as follows:

- Yaw plane signs are horizontal nystagmus, past pointing, rotational and lateral body falls, and horizontal deviation of perceived straight-ahead.
- Roll plane signs are torsional nystagmus, skew deviation, ocular torsion, and tilts of head, body, and perceived vertical.
- Pitch plane signs are upbeat/downbeat nystagmus, forward/backward tilts and falls, and vertical deviations of perceived straight-ahead.

The defined VOR syndromes allow for a precise topographic diagnosis of brainstem lesions as to their level and side, as follows (Fig. 5.2):

A tone imbalance in *yaw* indicates lesions of the lateral medulla, including the root entry

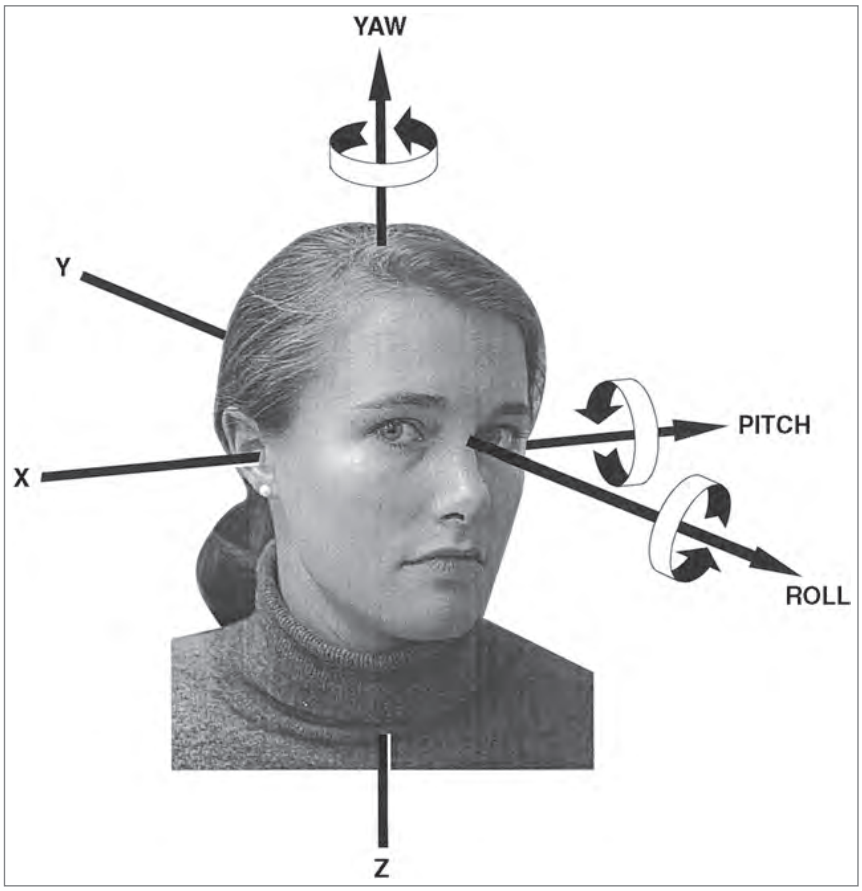

Figure 5.1 Schematic representation of the three major planes of action of the vestibulo-ocular reflex. yaw = horizontal rotation about the vertical z axis; pitch = vertical rotation about the binaural y axis; roll = vertical rotation about the x axis ("line of sight"). (Courtesy of Alice Kniehase.)

zone of the eighth cranial nerve and/or the vestibular nuclei.

A tone imbalance in *roll* indicates unilateral lesions (ipsiversive at pontomedullary level, contraversive at pontomesencephalic level).

A tone imbalance in *pitch* indicates bilateral (paramedian) lesions or bilateral dysfunction of the cerebellum, especially the flocculus.

Some vestibular disorders are characterized by a simultaneous peripheral and central vestibular involvement. Examples are large acoustic neurinomas, infarctions of the anterior inferior cerebellar artery, head trauma, and syndromes induced by alcohol intoxication. Others may affect the vestibular nerve root in the brainstem, where the transition between the peripheral and central nervous systems has been defined as the *Redlich-Oberstein zone* (lacunar infarction or focal demyelination in multiple sclerosis [MS] mimicking vestibular neuritis).

Cortical vestibular syndromes include vestibular seizures and lesional dysfunction with tilt of the perceived vertical, lateropulsion, and, rarely, rotational vertigo. There is no primary vestibular cortex, but the parietoinsular vestibular cortex (PIVC)[5] seems to act as a kind of main integration center within a network of multisensory cortical areas in the temporoparietal cortex. Dysfunction of this multisensory and sensorimotor cortex for spatial orientation and self-motion perception may be also involved in spatial hemineglect and rare paroxysmal room-tilt illusions, which belong to disorders of higher vestibular function.[6]

Most central vertigo syndromes have a specific locus (Table 5-1) but not a specific etiology. The etiology may, for example, be vascular, autoimmunologic as in MS, inflammatory, neoplastic, toxic, traumatic, or degenerative.

Vestibular Disorders in (Frontal) Roll Plane

The "graviceptive" input from the otoliths converges with that from the vertical semicircular canals (SCCs) at the level of the vestibular nuclei[7] and the ocular motor nuclei[8,9] to subserve static and dynamic vestibular function in pitch (up and down in the sagittal plane) and roll

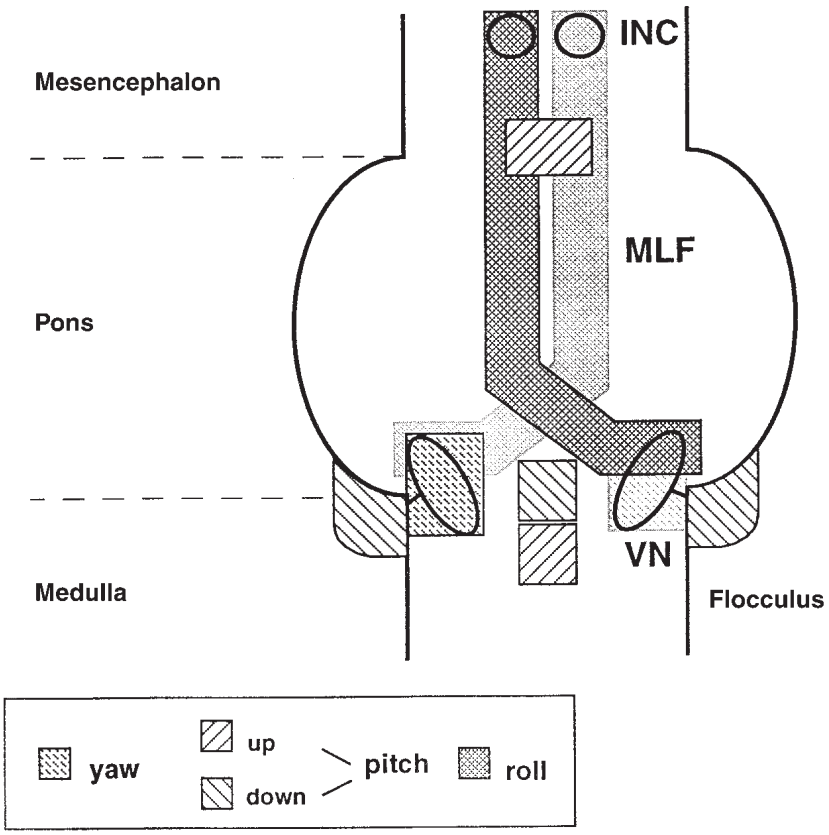

Figure 5.2 Vestibular syndromes in roll, pitch, and yaw planes: critical areas are schematically represented on the basis of our current knowledge of vestibular and ocular motor structures and pathways, a lesion of which causes a vestibular tone imbalance in one of the three major planes of action. The mere clinical sign of a vertical, torsional, or horizontal nystagmus—if central-vestibular—allows a topographic diagnosis of the lesion, although the particular vestibular structures involved are still under discussion. Whereas a vestibular tone imbalance in the roll plane indicates unilateral brainstem lesions (a crossing in the pons), vertical nystagmus indicates bilateral lesions. Two separate causative loci are known for upbeat nystagmus, medullary and pontomesencephalic. Downbeat nystagmus indicates a bilateral paramedian lesion of the commissural fibers between the vestibular nuclei or a bilateral flocculus lesion. Horizontal nystagmus indicates unilateral pontomedullary lesions involving the vestibular nuclei. The differentiation of vestibulo-ocular motor signs according to the three major planes of action of the vestibulo-ocular reflex (VOR) and their mapping to distinct and separate areas in the brainstem are helpful for topographic diagnosis and for avoiding incorrect assignment of clinical signs to brainstem lesions identified with imaging techniques. INC = interstitial nucleus of Cajal; MLF = medial longitudinal fasciculus; VN = vestibular nucleus.[3]

■ Table 5-1 CENTRAL VESTIBULAR SYNDROMES

Site	Syndrome	Mechanism/Etiology
Vestibular cortex (multisensory)	Vestibular epilepsy	Vestibular seizures are auras (simple or complex partial multisensory seizures)
	Volvular epilepsy	Sensorimotor "vestibular" rotatory seizures with walking in small circles
	Nonepileptic cortical vertigo	Rare rotatory vertigo in acute lesions of the PIVC

Continued

■ Table 5-1 CENTRAL VESTIBULAR SYNDROMES—cont'd

Site	Syndrome	Mechanism/Etiology
	Spatial hemineglect (contraversive)	Multisensory horizontal deviation of spatial attention with (right) parietal, temporal, or frontal cortex lesions
	Transient room-tilt illusions	Paroxysmal or transient mismatch of visual and vestibular 3D spatial coordinate maps in vestibular brainstem, parietal, frontal cortex lesions or peripheral vestibular lesions
	Tilt of perceived vertical with body lateropulsion (mostly contraversive)	Vestibular tone imbalance in roll with acute lesions of the PIVC
Thalamus	Thalamic astasia	Dorsolateral vestibular thalamic lesions
	Tilt of perceived vertical (ipsiversive or contraversive) with body lateropulsion	Vestibular tone imbalance in roll
Mesodiencephalic brainstem	OTR (contraversive; ipsiversive if paroxysmal)	Vestibular tone imbalance in roll (integrator: OTR with INC lesions)
	Torsional nystagmus (ipsiversive or contraversive)	Ipsiversive in INC lesions
		Contraversive in riMLF lesions
Mesencephalic brainstem	Skew torsion (contraversive)	Vestibular tone imbalance in roll with MLF lesions
Pontomedullary brainstem	Upbeat nystagmus	Tone imbalance in pitch in bilateral lesions of the central tegmental tract or the brachium conjunctivum
	Tilt of perceived vertical, lateropulsion, OTR	Vestibular tone imbalance in roll with medial and/or superior vestibular nuclei lesions
	Pseudo–"vestibular neuritis"	Lacunar infarction or MS plaque at the root entry zone of the eighth cranial nerve or vestibular cerebellum
	Downbeat nystagmus	Tone imbalance in pitch or asymmetry in the distribution of on-directions of vertical gaze-velocity Purkinje cells
	Transient room-tilt illusion	Acute severe vestibular tone imbalance in roll or pitch
	Paroxysmal room-tilt illusion in MS	Transversally spreading ephaptic axonal activity
	Paroxysmal dysarthria/ataxia in MS	Transversally spreading ephaptic axonal activation
Medulla	Upbeat nystagmus	Tone imbalance in pitch in a paramedian lesion of the cerebellar loop of the central tegmental tract (nucleus prepositus hypoglossi)
Vestibular cerebellum	Downbeat nystagmus	Tone imbalance in pitch or asymmetry in the distribution of on-directions of vertical gaze-velocity Purkinje cells caused by bilateral flocculus lesions (disinhibition)

■ Table 5-1 **CENTRAL VESTIBULAR SYNDROMES—cont'd**

Site	Syndrome	Mechanism/Etiology
	Positional downbeat nystagmus	Disinhibited otolith-canal interaction in nodulus lesions
	Familial episodic ataxia (EA1 with myokymia and EA2 with vertigo)	EA1 = autosomally dominant inherited potassium channelopathy EA2 = autosomally dominant inherited calcium channelopathy
	OTR, perceived tilt of subjective vertical	Tone imbalance with ipsiversive or contraversive tilts caused by lesions of the uvula, flocculus, dentate nucleus, and cerebellar hemispheric lesions
	Epidemic vertigo	Viral infection of cerebellum

INC = interstitial nucleus of Cajal; MS = multiple sclerosis; OTR = ocular tilt reaction; riMLF = right medial longitudinal fasciculus.

(lateral tilt in the frontal plane). In the "normal" position in the roll plane, the subjective visual vertical (SVV) is aligned with the gravitational vertical, and the axes of the eyes and the head are horizontal and directed straight ahead.

Signs and symptoms of a vestibular dysfunction in the roll plane can be derived from the deviations from normal function. A lesion-induced vestibular tone imbalance results in a syndrome consisting of a perceptual tilt (SVV), vertical misalignment of the visual axes (skew deviation), ocular torsion, or a complete ocular tilt reaction (OTR; the triad of head tilt, skew deviation, and ocular torsion).

There is convincing evidence that all of the following signs and symptoms reflect vestibular dysfunction in the (frontal) roll plane:

- OTR
- Skew deviation (skew-torsion sign)
- Spontaneous torsional nystagmus
- Tonic ocular torsion (monocular or binocular), if not caused by infranuclear ocular motor disorders
- Tilt of perceived SVV (with binocular and monocular viewing)
- Body lateropulsion

Ocular motor or postural tilts, as well as maladjustments of SVV, point in the same direction, either clockwise or counterclockwise (as seen from the viewpoint of the examiner). The direction of all tilts is reversed if pathological excitation of unilateral "graviceptive" pathways is the cause of vestibular tone imbalance in roll rather than a lesional input deficit. The combination of static and dynamic signs is not surprising if one considers the functional cooperation of otoliths and vertical SCCs owing to their neuronal convergence within "graviceptive" pathways. These signs and symptoms may be found in combination or as single components at all brainstem levels. Unilateral lesions of vestibulo-cerebellar structures (e.g., uvula, nodulus, and dentate nucleus) can also induce these signs in the roll plane.[10,11] A systematic study of 111 patients with acute unilateral brainstem infarctions showed that pathological tilts of SVV (94%) and ocular torsion (83%) are the most sensitive signs.[12] Skew deviation was found in one-third and a complete OTR in one-fifth of these patients (Table 5-2). Similar frequencies were found in the cerebellar lesions.[10,11]

Current clinical data support the following topographic diagnostic rules based on vestibular signs and symptoms in roll[3,4,10,11,13] (Fig. 5.3):

1. The fundamental pattern of eye-head tilt in roll—either complete OTR or skew torsion without head tilt—indicates a unilateral peripheral deficit of otolith and vertical canal input or a unilateral lesion of "graviceptive" brainstem pathways from the vestibular nuclei (crossing midline at lower pontine level) to the interstitial nucleus of Cajal (INC) in the rostral midbrain and vestibulo-cerebellar structures.
2. Tilts of SVV, resulting from peripheral or central vestibular lesions from the labyrinth to the vestibular cortex, are the most sensitive signs of a vestibular tone imbalance in roll (~90%). In the acute phase, tilts of SVV are more

■ Table 5-2 FREQUENCY OF SUBJECTIVE VISUAL VERTICAL (SVV) TILT, SKEW DEVIATION, OCULAR TORSION, AND OCULAR TILT REACTION (OTR) IN ACUTE UNILATERAL BRAINSTEM AND THALAMIC INFARCTIONS

			Ocular Torsion (%)			
Lesion	Patients (No.)	SVV Tilt (%)	Monocular	Binocular	Skew (%)	OTR (%)
Mesodiencephalic						
Paramedian thalamic	14	64	29*	43*	57	57
Posterolateral thalamic	17	65	13+	20+	0	0
Anterior polar thalamic	4	0	0	0	0	0
Brainstem						
Mesencephalic	16	94	54	38	37.5	25
Pontomesencephalic	12	92	64	18	25	25
Pontine	34	91	47	33	26.5	12
Pontomedullary	13	100	60	20	23	7.7
Medullary (Wallenberg's syndrome)	36	94	27	55	44	33
Total Brainstem	111	94	47	36	31	20

*Additional third nerve palsy.
+Slight torsion of about 2.8 degrees.

pronounced in patients with lesions of the vestibular nuclei (Wallenberg's syndrome) than in patients with vestibular neuritis.[14] The remission of the perceptual deficits over a period of 2 to 4 weeks is very similar in both diseases.

3. All tilt effects—perceptual, ocular motor, and postural—are ipsiversive (ipsilateral eye lowermost) and caused by unilateral peripheral or pontomedullary lesions below the crossing of the "graviceptive" pathways. They indicate involvement of the labyrinth, vestibular nerve, or medial and/or superior vestibular nuclei; the last are mainly supplied by the vertebral artery.
4. All tilt effects in unilateral pontomesencephalic brainstem lesions are contraversive (contralateral eye lowermost) and indicate involvement of the MLF (paramedian arteries arising from basilar artery) or INC (paramedian superior mesencephalic arteries arising from the basilar artery).
5. Brainstem lesions ipsilateral to the vestibular nucleus, near the ascending medial lemniscus (ipsilateral vestibule-thalamic tract, IVTT), also induce isolated ipsilateral deviations of SVV without vertical divergence or ocular torsion.[15]
6. Depending on the damaged region of the cerebellum, unilateral lesions of the vestibulocerebellum induce primarily contraversive (~60%) and more seldom (~25%) ipsiversive deviations.[11] The structure most frequently impaired in cases of contraversive signs is the dentate nucleus.
7. Skew deviation is always combined with ocular torsion (skew-torsion sign). It manifests without head tilt if ascending pontomesencephalic "graviceptive" pathways are affected rostral to the downward branching of the vestibulospinal tract.
8. Unilateral lesions of ascending vestibular pathways rostral to the INC typically manifest with deviations of perceived vertical without concurrent eye-head tilt.

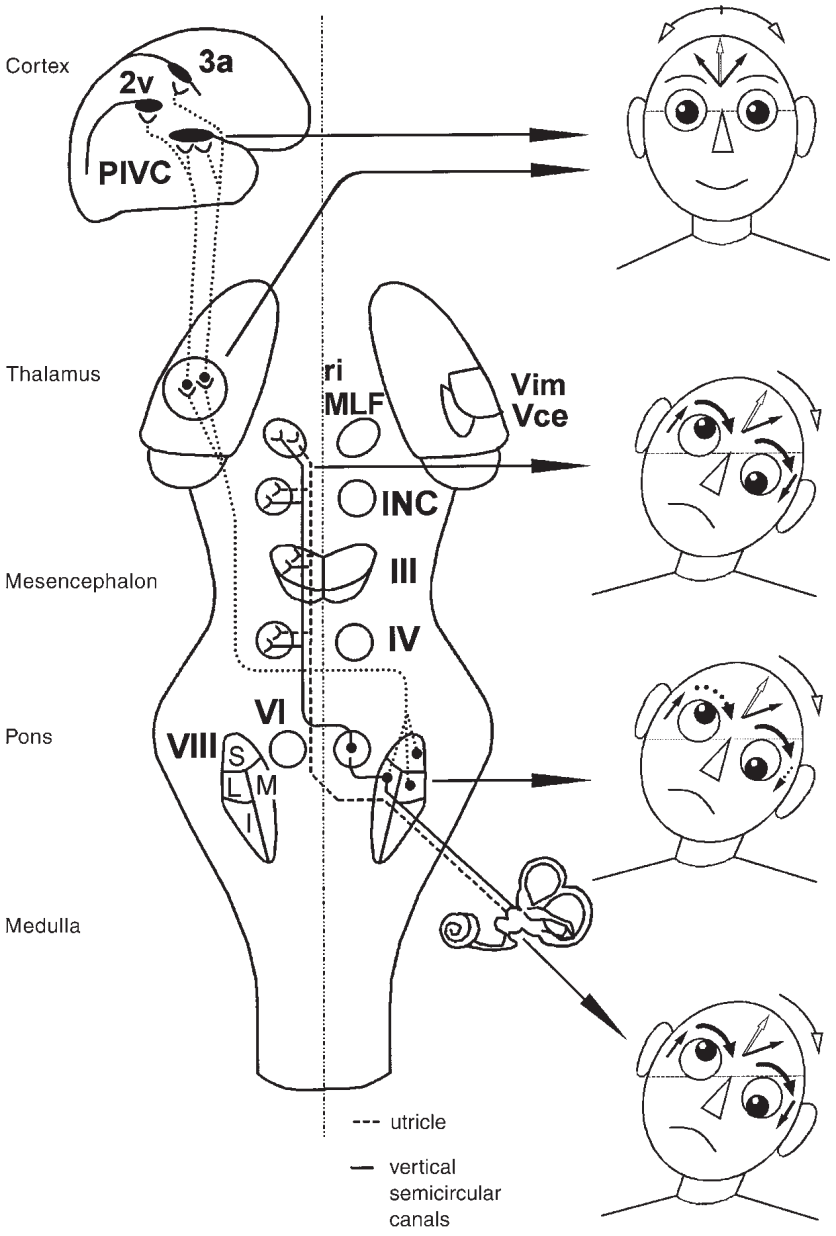

Figure 5.3 Vestibular syndromes in roll plane: graviceptive pathways from otoliths and vertical semicircular canals (SCCs) mediating vestibular function in roll plane. The projections from the otoliths and the vertical semicircular canals to the ocular motor nuclei (trochlear nucleus IV, oculomotor nucleus III, abducens nucleus VI), the supranuclear centers of the interstitial nucleus of Cajal (INC), and the rostral interstitial nucleus of the medial longitudinal fasciculus (riMLF) are shown. They subserve the vestibulo-ocular reflex (VOR) in three planes. The VOR is part of a more complex vestibular reaction that also involves vestibulospinal connections via the medial and lateral vestibulospinal tracts for head and body posture control. Furthermore, connections to the assumed vestibular cortex (areas 2v and 3a and the parietoinsular vestibular cortex [PIVC]) via the vestibular nuclei of the thalamus (Vim, Vce) are depicted. "Graviceptive" vestibular pathways for the roll plane cross at the pontine level. Ocular tilt reaction (OTR) (skew torsion, head tilt, and tilt of perceived vertical, subjective visual vertical [SVV]) is depicted schematically on the *right* in relation to the level of the lesion: ipsiversive OTR with peripheral and pontomedullary lesions; contraversive OTR with pontomesencephalic lesions. In vestibular thalamic lesions, the tilts of SVV may be contraversive or ipsiversive; in vestibular cortex lesions, they are preferably contraversive. OTR is not induced by supratentorial lesions above the level of INC.[3]

9. OTR in unilateral paramedian thalamic infarctions (in ~50%; paramedian thalamic arteries from basilar artery) indicates simultaneous ischemia of the paramedian rostral midbrain, including the INC.
10. Unilateral lesions of the posterolateral thalamus can cause thalamic astasia and moderate ipsiversive or contraversive SVV tilts, thereby indicating involvement of the vestibular thalamic subnuclei (thalamogeniculate arteries). This generally resolves within a matter of days or a few weeks.
11. Acute unilateral lesions of the PIVC and the superior temporal gyrus cause moderate ipsiversive or mostly contraversive SVV tilts lasting several days (temporal branches of the middle cerebral artery or deep perforators).[16,17]
12. Some of the patients with lesions of the PIVC, the superior temporal gyrus, the operculum, and the anterior insula simultaneously show a pusher syndrome.[18] There is a positive correlation between the extent of pushing and the SVV deviation.[19] This occurs more frequently with lesions of the right hemisphere (42%)

than of the left hemisphere (25%) and indicates a close correlation between the control of posture and stance and the vestibular system.

13. An SVV tilt found with *monocular* but not with binocular viewing is typical for a trochlear or oculomotor palsy rather than a supranuclear "graviceptive" brainstem lesion.[20] Complicated ocular motor syndromes are occasionally induced in midbrain lesions by the combination of a central vestibular deficit in the roll plane (caused by the INC lesion) and a concurrent nuclear or fascicular 3rd cranial nerve or 4th cranial nerve palsy ("mixed pattern").
Tilt effects caused by paroxysmal activation of "graviceptive" pathways point in the opposite direction to those caused by lesional inhibition, such as unilateral infarction.[21,22]

Thus, all clinical signs of vestibular dysfunction in roll can be helpful when one is determining not only the level but also the side of the brainstem lesion. If the level of damage is known from the clinical syndrome, the vestibular syndrome indicates the more severely affected side. Conversely, if the side of damage is clear from the clinical syndrome, the direction of OTR, skew deviation, and SVV tilt indicate the level on the brainstem.

Etiology

The two most common causes of tonic OTR are brainstem ischemia (especially Wallenberg's syndrome and unilateral paramedian thalamic plus rostral mesencephalic infarctions) and brainstem tumors[21,23] followed by unilateral thalamic hemorrhages or lower brainstem hemorrhages (cavernous angioma, lymphomas), severe brainstem concussion, MS, or in association with attacks of basilar migraine.

Natural Course and Management

The natural course and management of OTR depend on the etiology. OTR is usually transient; in cases of hemorrhage or infarction, recovery occurs within a few days to weeks. However, it can be permanent, as we observed in a patient with severe brainstem concussion. Following unilateral brainstem infarctions, all features of OTR—postural, ocular motor, and perceptual—disappear naturally and gradually within 4 to 6 weeks or months (repeated measurements made in seven patients over a period of up to 1 to 3 years; Fig. 5.4).[24] We found that repeated measurements of skew deviation, ocular torsion (OT), and tilts of SVV made during a single day showed consistent tilts.[10] Repeated measurements on subsequent days showed a gradual recovery, mostly within 30 days, both for OT and SVV (Fig. 5.5). Some patients, however, maintained a residual OT of a few degrees without a corresponding tilt of SVV for up to 2 years.

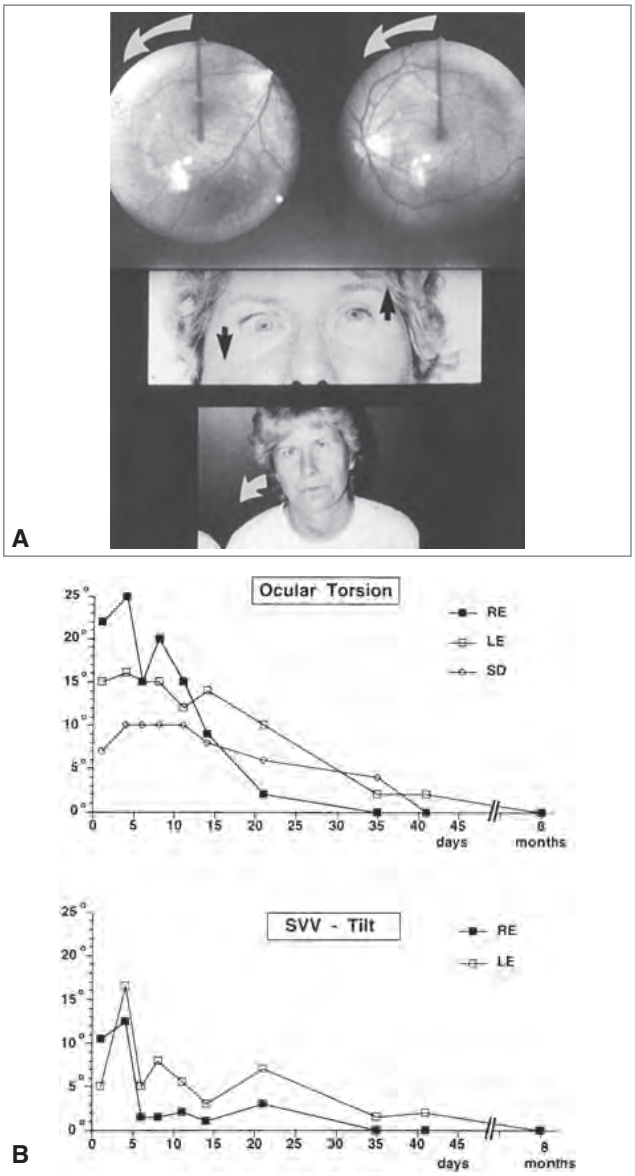

Figure 5.4 **(A)** Patient with a left paramedian thalamic infarction presenting with a complete ocular tilt reaction (OTR) to the right. OTR consisted of contraversive head tilt of 20 degrees (*bottom*); skew deviation of 10 degrees, left eye over right eye; and ocular torsion of 15 to 20 degrees (counterclockwise from the viewpoint of the observer). **(B)** Natural course of ocular torsion, skew deviation (SD), and tilt of subjective visual vertical (SVV, in degrees) shows gradual recovery in 6 weeks. RE = right eye; LE = left eye.[24]

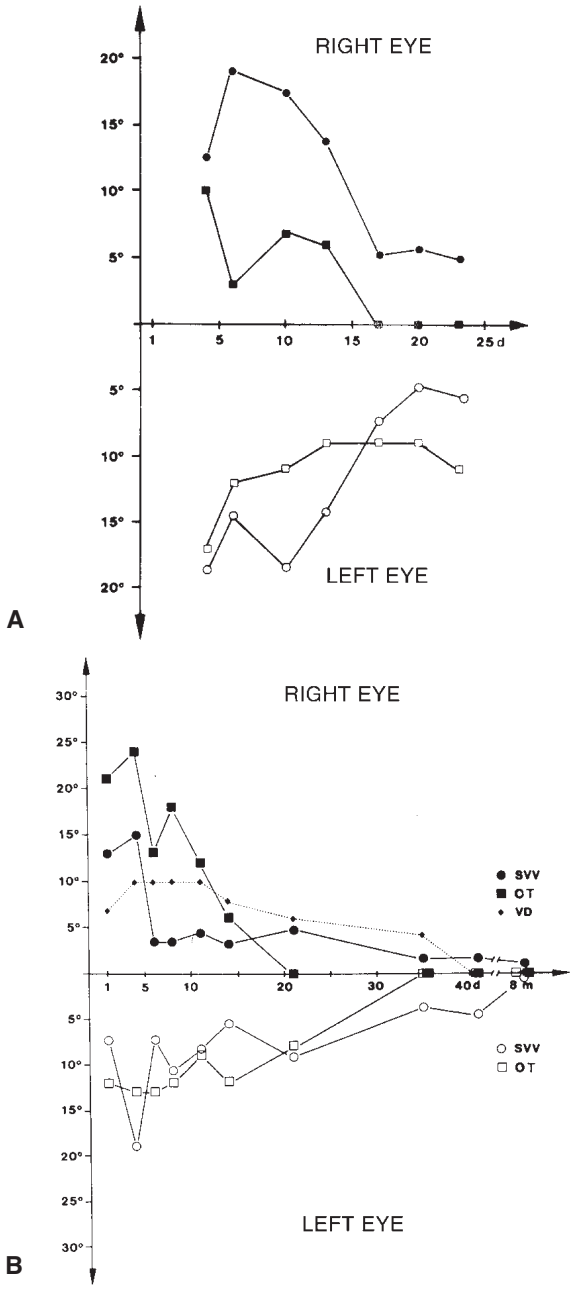

Figure 5.5 Two representative time courses of deviations of subjective visual vertical (SVV) and ocular torsion (OT) (separate for the left and right eyes) in a patient with Wallenberg's syndrome on the left **(A)** and a patient with unilateral lesion of the region of the interstitial nucleus of Cajal (INC) in the rostral midbrain tegmentum **(B)**. Note the dissociated effects in the patient with Wallenberg's syndrome; OT and SVV are deviated most in the ipsilateral left eye. Comparison of individual OT and SVV values in the two patients shows varying dissociations of the net tilt. Both tend to normalize within 4 to 6 weeks, and fluctuations cannot be explained simply by methodological inaccuracy. d = days; m = months; VD = vertical divergence as skew deviation.[9]

Recovery is based on a functionally significant central compensation of a vestibular tone imbalance induced by a unilateral central lesion. The mechanisms underlying central compensation of central lesions may be similar to those of central compensation of peripheral vestibular lesions as the remission of the def icits is very similar.[14] However, although compensatory processes in unilateral peripheral vestibular lesions are mediated via the cortical hemispheres,[25] those in central lesions of the medulla are mediated via the brainstem and cerebellum. [26] Physical therapy may facilitate this central compensation, but this possibility has not yet been proven in a prospective study.

Paroxysmal OTR in MS has been treated effectively with carbamazepine, and baclofen has been reported to be of some therapeutic benefit.

Thalamic and Cortical Astasia Associated with Subjective Visual Vertical Tilts

An association of SVV tilts with f alls is also typical for posterolateral (vestibular subnuclei) thalamic lesions. Thalamic astasia[27] is a condition in which patients without paresis or sensory or cerebellar deficits are unable to maintain an unsupported, upright posture. Postural imbalance with a transient tendency to fall has been reported after therapeutic thalamotomy and thalamic hemorrhage.[28] According to our experience in some 30 patients with acute thalamic infarctions, the posterolateral type may cause contraversive or ipsiversive postural instability with SVV tilts, whereas the paramedian type (if it e xtends into the rostral midbrain) al ways causes contra versive falls.[24] Masdeu and colleagues[29] have described astasia and gait failure with damage of the pontomesencephalic (locomotor) region. Although not discussed, it could also be e xplained in part by a v estibular tone imbalance in roll, especially because skew deviation was described as a feature of the syndrome. Indeed, as seen in an imaging study on vascular thalamic lesions,[30] lesions of the posterolateral thalamus cause an interruption of the vestibular pathways to the temporoparietal v estibular cortex areas of the affected side and also to the contralateral side. Moreover, lesions of the thalamus may cause pusher syndrome with deviations of the body vertical and with or without deviations of the SVV.[18] This condition is not merely a vestibular disorder but reflects a dysfunction of spatial orientation, attention, and postural control—all based on multisensory integration (visual, vestibular, somatosensory).

Cortical infarctions of the middle cerebral artery territory in several studies showed significant, ipsiversive or mostly contraversive, pathological SVV tilts. [16,17,31]

The overlapping area of these infarctions centered on the posterior insula, which, according to functional imaging studies,[32-36] is homologous to the PIVC in monkeys, and the adjacent superior temporal lobe.[37,38] Further areas involved in the perception of verticality are the superior temporal gyrus, the operculum, the anterior insula, and the inferior frontal gyrus.[17] SVV tilts caused by vestibular cortex lesions can also be associated with (a compensatory) body lateropulsion. This finding may explain some cases of the cortical phenomenon "pusher" syndrome,[18] which physical therapists readily recognize.

Torsional Nystagmus

The "graviceptive" input from the otoliths converges with that from the vertical SCCs to subserve static and dynamic vestibular function in roll. This combination of static and dynamic effects[39] is not surprising if one considers how these functions are corroborated. Several studies on OTR, lateropulsion, and SVV were concerned with static effects of vestibular dysfunction in roll.[11,12] These effects persist for days to weeks, during which time they spontaneously subside. In the acute stage of infarction, additional dynamic signs and symptoms occur, which consist of horizontal rotational vertigo and torsional nystagmus.[40,41] Fast phases of rotational nystagmus are contraversive in pontomedullary lesions, whereas the slow phases correspond in direction to the static deviation.

Several distinct and separate lesions (see Fig. 5.2) have been associated with torsional nystagmus: for example, lesions of the vestibular nuclei,[41,42] the lateral medulla,[40,43] in rare cases the MLF (as indicated by an association with internuclear ophthalmoplegia),[44,45] the INC, and the riMLF.[46-48] Fast phases of torsional nystagmus are contraversive in pontomedullary lesions and ipsiversive in paramedian pontine and mesencephalic (INC) lesions (rare exception: contraversive in riMLF lesion).

Jerk-waveform seesaw nystagmus (a torsional nystagmus with elevation of the intorting eye and depression of the extorting eye) is induced by an inactivation of the INC in the rostral midbrain and is also ipsiversive.[48]

The different locations of lesions causing different directions of torsional nystagmus at first appear to be confusing. They can, however, be explained by the tonic torsional shift of eye position along the graviceptive pathways from the vestibular nuclei to the INC. A lesion of the (medial or superior) vestibular nucleus causes an ipsiversive tonic deviation (ipsiversive ocular torsion) with compensatory fast phases of the torsional nystagmus to the contralesional side. In view of the fact that the pathway within the MLF crosses to the contralateral side, an MLF lesion in the pontine and pontomesencephalic brainstem induces a tonic contraversive deviation, and therefore a torsional nystagmus with the fast phases ipsilesional. The same is true for a lesion of the INC. The only exception to these directional rules for tone imbalance along the vestibular graviceptive pathways is the riMLF, a lesion of which causes an (possibly non vestibular) ocular motor tone imbalance in the opposite direction.

Vestibular Disorders in (Sagittal) Pitch Plane

A striking difference between vestibular tone imbalance in the roll and pitch planes is that roll dysfunction is caused by unilateral, and pitch dysfunction by bilateral, lesions of paired pathways in the brainstem or of the cerebellar flocculus.[4] This structural difference probably explains why a vestibular tone imbalance in pitch frequently occurs with various intoxications, metabolic disorders, or degeneration, which is unusual for tone imbalance in yaw or roll, unless as a functional decompensation of an earlier (compensated) tone imbalance. Downbeat nystagmus (DBN) and upbeat nystagmus (UBN) are not merely ocular motor disorders but disorders that also affect orientation and balance. A tone imbalance in the pitch plane manifests as vertical upbeat or downbeat nystagmus, fore-aft head-and-body tilt, and deviation of the subjective straight-ahead toward the direction of the slow phase of the nystagmus.[49]

Despite the numerous clinical studies on UBN and DBN as well as the many hypotheses proposed to explain their pathomechanism, so far the pathophysiology of these disorders has not been clarified.[50-54] There seem to be various forms of DBN responsible for different aspects of the disorder. Several pathomechanisms that cause an instability in the brainstem-cerebellar networks, which normally stabilize vertical gaze, are currently being discussed. For example, an asymmetry

- in the vertical neuronal integrator with a disorder of the saccade generator, which is conspicuous in excentric gaze positions;
- in the central connections of the VOR for vertical eye movements including the otolithic pathways, which explains the frequent dependence on gravity; or
- in the vertical gaze-pursuit system with spontaneous upward drift.

Here the flocculus/parafloccculus seems to play a special role, because its damage leads to a disinhibition of the vestibular pathways of the superior vestibular nucleus to the oculomotor nucleus. This fits with findings from functional imaging studies which have proven that patients

with idiopathic DBN have a hypometabolism or a reduced activity in the flocculus/paraflocculus as well as in the pontomedullary brainstem.[55-57] In contrast, structural MRI found atrophies of the gray matter not in the flocculus/paraflocculus but in the lateral portions of the cerebellar hemispheres (lobule VI) and in the oculomotor vermis.[57]

Clinically, DBN occurs more commonly than UBN and is often permanent (as in idiopathic forms, cerebellar degeneration, or Arnold-Chiari malformation),[58,59] whereas UBN is usually a transient phenomenon. Lesional sites for UBN have been more precisely confirmed by clinical studies (bilateral lesions of the pontomesencephalic junction or the medulla) than those for DBN (bilateral pontomedullary lesions or bilateral flocculus dysfunction). Transitions between UBN and DBN have been frequently described in paramedian pontomedullary lesions. Whereas DBN is more typical for congenital craniocervical malformations (Arnold-Chiari malformation) and cerebellar degeneration, UBN is more typical for MS, bilateral brainstem ischemia (basilar artery thrombosis), or brainstem tumors. UBN and DBN in the primary position of gaze may be the result of various intoxications (without structural lesion).

Downbeat Nystagmus

Box 5-1 summarizes the information given in this chapter about the downbeat nystagmus/vertigo syndrome.

Downbeat nystagmus in the "primary" gaze position, or more particularly on lateral gaze, is often accompanied by oscillopsia and postural instability. This is a clearly defined and, depending on the lesional site, permanent association of symptoms, which often indicates structural lesions of the paramedian craniocervical junction. DBN is present in darkness and with fixation; slow-phase velocity and amplitude increase on lateral gaze or with head extension or head movements in the sagittal (pitch) plane. It may be present only on downward or lateral gaze. Slow-phase velocity is not consistently related to vertical gaze and, contrary to Alexander's law, may even be maximal on upward rather than downward gaze. Nystagmus is a jerk, usually with linear slow phases. It may exhibit changes of exponential velocity in slow phases, both increasing and decreasing.

It has been reported that reversals from DBN to UBN can be provoked by upward gaze deviation,[60] convergence,[61] or transitions from sitting to supine position.[62] DBN and UBN may be the directional counterparts of a vestibular tone imbalance in the pitch plane. The close proximity of the areas causing either UBN or DBN in the medulla agrees with the directional changes between the two. Reversals from UBN to DBN have also been observed.[61]

Oscillopsia should be expected to cause an impairment of postural balance, because retinal image motion is a major cue for body stabilization. However, this kind of "visual ataxia" cannot simply account for the typical postural imbalance, which is a striking feature of the fore-aft body sway and includes a tendency to fall backward. This fore-aft postural instability can be interpreted to be a direction-specific vestibulospinal (or cerebellar) imbalance, because it can be observed when the eyes are closed. We believe that the objective measurable backward tilt represents a vestibulospinal compensation in the direction opposite to the perceived lesional "forward vertigo" that corresponds to downbeat nystagmus.[63] When the eyes are open, a measurable visual stabilization of body sway is preserved, but it does not sufficiently compensate for the visual ataxia. In DBN (more aptly termed "downbeat nystagmus syndrome"), the patient's pathological postural sway with the eyes open depends on the direction of gaze; it increases with increasing amplitude of the nystagmus. Pathophysiologically, it is secondary to a combination of both vestibulospinal ataxia and reduced visual stabilization of posture owing to the nystagmus.

Etiology

Idiopathic cases occur most often (38%), degenerative disorders of the cerebellum in 20% of cases, vascular lesions in 9%, and malformations in 7%. The more seldom causes like toxic drug damage, lesions in MS and paraneoplastic syndromes, vestibular migraine, vitamin B_{12} and thiamine deficiencies, or traumatic and hypoxic injuries occur in decreasing frequency.[58,59]

Nutritional cerebellar syndromes owing to thiamine deficiency, in particular alcoholic cerebellar degeneration (see Box 5-1), not only cause a typical 3-Hz fore-aft oscillation of body sway but also DBN.[59,64-66] Antiepileptic drugs, especially phenytoin and carbamazepine, can produce a reversible DBN with associated cerebellar signs, depending on the dosage of the drugs. Other rare causes are lithium toxicity,[64] felbamate intoxication,[67] toluene abuse,[68] and severe magnesium depletion.[69] Other conditions associated with DBN are MS,[70] familial periodic ataxia, tumors of the posterior fossa, cerebellar degeneration,[71] paraneoplastic cerebellar degeneration,[72] infratentorial vascular diseases such as dolichoectasia of the vertebrobasilar artery,[73] hematomas, cavernomas, syringobulbia,[74] and encephalitis (see Box 5-1).

Management

DBN caused by drugs, magnesium depletion, or vitamin B_{12} deficiency is usually reversible when the intoxication

Box 5-1

DOWNBEAT NYSTAGMUS (VESTIBULAR DOWNBEAT SYNDROME)

Clinical Syndrome

Downbeat nystagmus in the primary position of gaze (no suppression by fixation), increased on lateral gaze or head extension

Associated distressing oscillopsia and postural imbalance with a tendency to fall backward and vertical deviation of straight-ahead

Saccadic downward pursuit

Transitions from downbeat to upbeat nystagmus possible

Incidence/Age/Sex

Incidence depends on etiology

No obvious preference for sex

Rare in children (congenital)

Pathomechanism

Several hypotheses under discussion: Tone imbalance caused by lesions of the pathways mediating (1) signals of the vertical cerebellovestibular neural integrator; (2) central connections of the vertical vestibulo-ocular reflex, including both the vertical semicircular canal and otolith responses (pitch plane); or (3) signals of the vertical smooth pursuit system

Structural or functional lesions involve either:

- The floor of the fourth ventricle between the vestibular nuclei
- The vestibulocerebellar flocculus (intoxication, cerebellar degeneration)

Etiology

Beyond the idiopathic form (38%) the most common causes are cerebellar degeneration, including alcoholic cerebellar degeneration, vascular lesions, cerebellar ectopia, and intoxication (drugs: phenytoin, carbamazepine, lithium)

Other conditions: multiple sclerosis, tumor, hematoma, encephalitis, magnesium depletion, vitamin B_{12} deficiency (see Table 5-1)

Course/Prognosis

- Frequently permanent when caused by structural lesions
- Usually reversible when caused by intoxication or metabolic deficiency

Management

Medical treatment with 4-aminopyridine, gabapentin, baclofen, or clonazepam

Differential Diagnosis

Acquired pendular nystagmus, gaze-evoked nystagmus, upbeat nystagmus, spasmus nutans (infants), vertical congenital nystagmus, ocular bobbing

or metabolic deficiency is reversed. DBN caused by structural lesions in the posterior fossa is usually permanent. It is therapeutically expedient to try to treat the symptoms of persisting DBN with various drugs. In recent years the potassium channel blockers 3,4-diaminopyridine and 4-aminopyridine have been proven to have a positive effect, above all in DBN and also in single cases of UBN.[75-78] For this reason a therapeutic attempt can be made with 5 to 10 mg 4-aminopyridine, 3 times a day (or 10 mg, once or twice a day in the sustained release form) for DBN (Fig. 5.6).

Animal experiments have shown that administration of aminopyridines can increase the resting-state activity and excitability of Purkinje cells[79] and thus improve the reduced inhibitory GABA-mediated influence of the cerebellum on the superior vestibular nuclei. Because epileptic attacks or heart rhythm disorders—especially at higher dosages—can occur in rare cases, an ECG should be performed before and ~60 minutes after the first ingestion of 5 mg 4-aminopyridine to promptly detect any lengthening of the QT interval.[76] This is so far a single, nonstandard therapeutic attempt.

Because UBN generally slowly resolves after an acute occurrence, therapy for its symptoms is often not necessary. In cases of very disturbing oscillopsias due to large amplitudes of UBN and in DBN, one can try 4-aminopyridine (5 to10 mg, three times a day orally),[80] memantine (10 to 20 mg, twice a day),[81,82] or gabapentin (300 to 600 mg twice a day).[82] If both are ineffective, one can try baclofen (5 to 10 mg twice a day orally).[2]

Upbeat Nystagmus (Upbeat Nystagmus Syndrome)

Box 5-2 summarizes the information given in this chapter about the upbeat nystagmus syndrome.

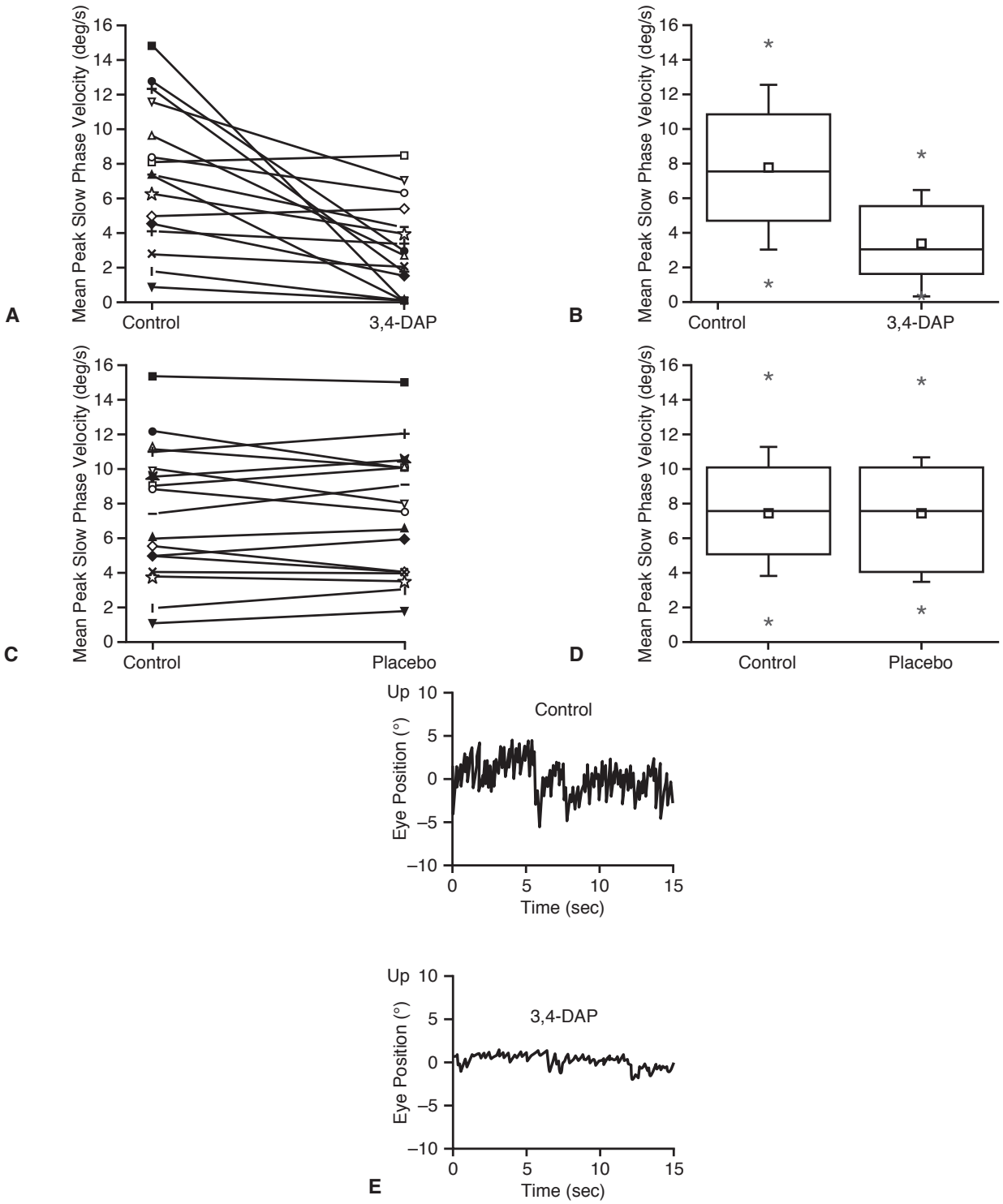

Figure 5.6 Effect of 3,4-diaminopyridine on downbeat nystagmus (DBN): influence of 3,4-diaminopyridine (3,4-DAP) on the mean slow phase velocity of DBN (measured with 2D videooculography). The graphs a-d show the mean slow phase velocity of DBN for each individual patient. **(A)** Controls vs. 3,4-DAP, **(C)** controls vs. placebo. Both graphs **(B)** and **(D)** show a so-called box plot with mean value, median, and 50 percentile as well as standard deviation for controls vs. 3,4-DAP (*B*) and controls vs. placebo (*D*). 3,4-DAP reduced the maximal velocity of the slow phase of DBN from 7.2 to 3.1 deg/sec 30 min after ingestion of 20 mg 3, 4-DAP ($p < 0.001$). **(E)** shows an original recording of vertical eye position before (top) and 30 min after (bottom) ingestion of the medicine. (From Strupp et al, 2003,[75]; with authors' kind permission.)

Box 5-2

UPBEAT NYSTAGMUS (VESTIBULAR UPBEAT SYNDROME)

Clinical Syndrome

Upbeat nystagmus in the primary position of gaze (no suppression by fixation), modulated by static head tilt

Associated distressing oscillopsia, postural imbalance, deviation of straight ahead

Saccadic upward pursuit

Transitions from upbeat to downbeat nystagmus possible

Incidence/Age/Sex

Incidence depends on etiology

No obvious preference for sex

Rare in children (congenital)

Pathomechanism

Several hypotheses under discussion: Tone imbalance caused by lesions of the pathways mediating (1) signals of the vertical cerebello-vestibular neural integrator; (2) central connections of the vertical VOR, including both the vertical semicircular canal and otolith responses (pitch plane); or (3) signals of the vertical smooth pursuit system

Structural or functional paramedian lesions involve either:

- Pontomesencephalic junction (ventral tegmental tract, brachium conjunctivum)
- Medulla (perihypoglossal nuclei)

Etiology

Multiple sclerosis, brainstem tumors, infarction, hematoma, cavernoma, encephalitis, abscess, alcoholic degeneration (Wernicke's encephalopathy), drug intoxication, nicotine (see Table 5-1)

Course/Prognosis

Depending on etiology, gradual improvement by central compensation

Usually reversible when caused by intoxication

Management

Medical treatment with 4-aminopyridine, baclofen (clonazepam)

Physical exercise (eye movements and balance training)

Differential Diagnosis

Acquired pendular nystagmus, gaze-evoked nystagmus, downbeat nystagmus, spasmus nutans (infants), vertical congenital nystagmus, reversed ocular bobbing

UBN in the primary position of gaze with concomitant oscillopsia and postural instability is a pendant of DBN and probably reflects either an imbalance of vertical VOR tone[49] or a downward ocular drift by asymmetrical smooth pursuit commands.[50-54] It has the same causes and involves central eye-head coordination in the pitch plane as mediated by pathways from the vertical SCCs and the otoliths. Because in some cases the manifestations are typically modulated by otolithic input arising from static head tilt, UBN can in a broader sense also be a kind of positional nystagmus. As distinct from DBN, brainstem lesions are often found in patients with UBN. Two separate intra-axial brainstem lesions in the tegmentum of the pontomesencephalic junction and in the medulla near the perihypoglossal nuclei (Fig. 5.7) are likely to be responsible for this syndrome, but there is insufficient evidence to determine whether the cerebellar vermis is involved. In analogy to DBN, UBN can probably indicate bilateral lesions of the pathways mediating (1) signals of the vertical cerebello-vestibular "neural integrator," (2) the central connections of the vertical VOR, or (3) signals of the vertical smooth-pursuit system. UBN arising from pontomesencephalic lesions could result from damage of the superior vestibular nucleus–ventral tegmental tract pathway coursing through the ventral pons and transmitting excitatory upward vestibular signals to the third nerve nucleus.[54] This would lead to an imbalance between downward and upward systems and in a downward slow phase. UBN in lesions affecting the caudal medulla could result from damage in a feedback loop involved in upward gaze-holding.[54]

Etiology

UBN was observed in 26 of 17,900 patients examined at a neurotological clinic in Japan. The incidence rate was

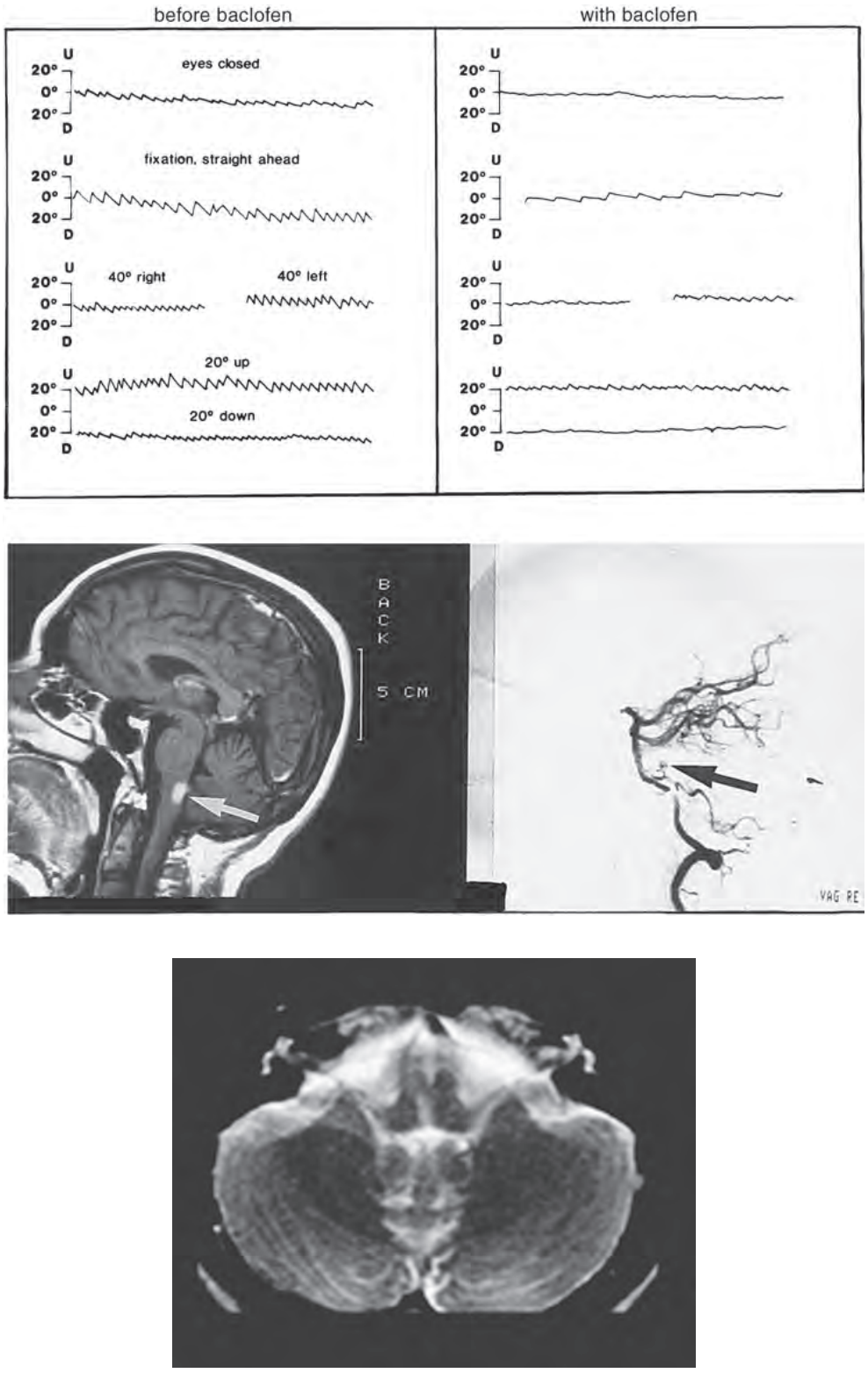

Figure 5.7 Partial suppression of upbeat nystagmus by medical treatment with baclofen. (*Top*) Vertical electronystagmography recordings. (*Bottom*) Magnetic resonance image of a patient with unilateral upbeat nystagmus and a paramedian medullary infarction affecting the area of PMT neurons near the neurons of the perihypoglossal nuclei.

0.145%.[83] The etiology of UBN is in general similar to that of DBN (see Box 5-2). Malformations of the craniocervical junction and cerebellar degeneration seem to be less common than in DBN, whereas brainstem tumors and MS are more common. UBN can be associated with bilateral (paramedian) vascular brainstem lesions, hematoma, cavernoma, MS, encephalitis, abscess, or head injury. It has been repeatedly reported in alcoholic degeneration, especially in Wernicke's encephalopathy[84] and in single cases of Fisher's syndrome,[85] central diabetes insipidus,[86] and Pelizaeus-Merzbacher disease,[87] and has even been associated with middle ear disease.[88] We have seen transient upbeat nystagmus associated with various intoxications, for example, with antiepileptic drugs. UBN can on rare occasions be congenital.[89]

Management

UBN may be associated with severe vertigo, ataxia, and nausea, particularly at first. Affected patients may require vestibular sedatives (e.g., dimenhydrinate or scopolamine) as long as nausea lasts. Depending on the etiology, the natural history of this sign usually shows gradual improvement or disappears, in contrast to DBN, which is frequently permanent. Physical exercise involving fixation, eye movements, and postural balance accelerates central compensation. Medical treatment is possible with baclofen (5 to 10 mg PO daily), which has a beneficial effect on nystagmus amplitude, oscillopsia, and visual acuity in some patients (see Fig. 5.7).[90] Carbamazepine was found to be effective in a single case of upbeat nystagmus caused by MS.[91] In single patients with UBN, Glasauer and colleagues[80] have shown that 4-aminopyridine reduced the peak slow-phase velocity in light from 8.6 to 2.0 deg/sec, but UBN in darkness was not affected. These investigators concluded that 4-aminopyridine reduces the downward drift in UBN by augmenting smooth pursuit commands.

Summary

These studies in UBN and DBN show that a new therapeutic principle has been developed: aminopyridines, as potassium channel blockers that increase the activity and excitability of Purkinje cells, have a beneficial effect on several disorders.

Vestibular Disorders in (Horizontal) Yaw Plane

The clinical signs, both perceptual and motor, of a vestibular tone imbalance in the yaw plane include rotational vertigo, deviation of perceived straight-ahead, lateropulsion of the eyes, past pointing, rotational and lateral body falls, and horizontal nystagmus.

Central vestibular syndromes manifesting purely in the yaw plane occur less frequently than those caused by imbalance in the pitch and roll planes, for two reasons.[4] First, the area of a lesion that can cause a tone imbalance in yaw is comparatively small (root entry zone of the vestibular nerve, medial and superior vestibular nuclei, and the adjacent integration center for horizontal eye movements—the paramedian pontine reticular formation [PPRF]). In contrast, the area of a lesion that can cause vestibular tone imbalance in roll or pitch covers nearly the entire brainstem from the medulla to the rostral midbrain (see Fig. 5.2). The larger extent of the latter area is a result of the greater separation of the vestibular nuclei and the ocular motor integration centers for vertical and torsional eye movements (riMLF and INC). Second, the area of a lesion that can theoretically cause a pure tone imbalance in the yaw plane adjoins and overlaps areas subserving vestibular function in roll and pitch (see Fig. 5.2).There is a multisensory convergence within the parallel neural network of the vestibular nuclei,[92] a lesion of which will cause mixed vestibular syndromes in more than one plane. A study of vestibular nuclei lesion in the monkey demonstrated a combined nystagmus: its horizontal component beat toward the contralateral side after rostral lesions and toward the ipsilateral side after caudal lesions.[93]

Some of the cases described as central variants of vestibular neuritis[94-97] caused by lesions of the medial vestibular subnucleus or the root entry zone of the vestibular nerve were probably not restricted to the yaw plane, because the case descriptions also contain signs and symptoms of VOR tone imbalance in other planes of action. There have been frequent reports that cerebellar infarctions caused by occlusion of the anterior inferior cerebellar artery (which may also supply the rostral vestibular nuclei) also mimic vestibular neuritis.[98-100] The other main cause of confusion with disorders at the entry zone of the eighth cranial nerve is MS.[95]

Vestibular syndromes, when caused by unilateral pontomedullary lesions, commonly result in combined vestibular tone imbalance in more than one plane, such as a combination of torsional and horizontal nystagmus. This tone imbalance may manifest not only in spontaneous nystagmus but also in spontaneous or gaze-evoked ocular deviations. There may be an inappropriate horizontal ocular deviation during attempted vertical saccades (lateropulsion in Wallenberg's syndrome) or an inappropriate torsional deviation during attempted horizontal saccades.

Vestibular Cortex: Locations, Functions, and Disorders

The two major cortical functions of the vestibular system are spatial orientation and self-motion perception. These functions, however, are not specifically vestibular; they also rely on visual and somatosensory input. All three systems—vestibular, visual, and somatosensory—provide redundant information about the position and motion of the body relative to the external space. Although the vestibular cortex function is distributed among several multisensory areas in the parietal and temporal cortices, it is also integrated in a larger network for spatial attention and sensorimotor control of eye and body motion in space.

The term "disorders of higher visual function" is well established in neuro-ophthalmology. It usually correlates circumscribed supratentorial especially cortical lesions with particular dysfunctions of "higher visual perception," recognition, and memory as well as spatial orientation along the ventral and dorsal streams.[101] Analogously one could also define disorders of higher vestibular function.[2] These disorders are characterized by complex perceptual, sensorimotor, and behavioral criteria that exceed the basic perceptions like body motion and motor responses as well as the vestibulo-ocular and vestibulo-spinal reflexes. There are differences and similarities between these higher vestibular/visual disorders. A typical difference is that higher vestibular disorders often involve other sensory modalities, whereas higher visual disorders do not. Similarities are that both manifest with cognitive disturbances of spatial orientation, attention, spatial memory, and navigation. Sometimes, however, it is not possible to differentiate between them. This becomes evident in syndromes like visuo-spatial hemineglect[102,103] or the room tilt illusion with transient episodes of "upside-down vision" induced by a vestibular tonus imbalance.[104-106] Not all disorders of higher vestibular function are caused by lesions of central vestibular structures. Peripheral bilateral vestibular loss, for example, not only causes oscillopsia and postural imbalance due to insufficient vestibulo-ocular and vestibulo-spinal reflexes; it also impairs spatial memory and navigation because of a lack of vestibular input to the hippocampal formation.

Animal studies have identified several distinct and separate areas of the parietal and temporal cortices that receive vestibular afferents, such as area 2v at the tip of the intraparietal sulcus,[107-109] area 3aV (neck, trunk, and vestibular region of area 3a) in the central sulcus,[110] PIVC at the posterior end of the insula and retroinsular regions,[37,38,111] the periarcuate cortex area 6 pa,[112] area 7 in the inferior parietal lobule,[113] and the ventral intraparietal area (VIP) in the fundus of the intraparietal sulcus.[114-117] In view of the strong interconnections between PIVC and the other vestibular cortex areas (mainly 3aV and 2v) as well as the vestibular brainstem nuclei, Guldin and Grüsser[5] postulate that it is the core region within the vestibular cortical system in the monkey. About 50% of the neurons in this region respond to vestibular stimulation in addition to somatosensory, optokinetic, or visual stimulation. This area is involved in the processing not only of vestibular, somatosensory, and visual information that is generated whenever the position of the body changes in relation to the extra-personal space[5] but also of that generated when stationary human subjects perform optokinetic nystagmus.[118,119] Not only do most of these cortical areas receive bilateral vestibular input from the vestibular nuclei, they in turn directly project down to the vestibular nuclei.[5,120,121] Thus, corticofugal feedback may modulate vestibular brainstem function.

Our knowledge about vestibular cortex function in humans is less precise. It is derived mainly from stimulation experiments reported anecdotally in the older literature and from later brain activation studies with positron emission tomography (PET)[30,35,122] or functional magnetic resonance imaging (fMRI)[32-34,36,118] and lesion studies.[17,31] The areas in humans that were activated during caloric or electric vestibular stimulation were located in the posterior insula (first and second long insular gyri) and retroinsular regions (representing PIVC and the posterior adjacent visual temporal sylvian area [VTS] in the monkey), the superior temporal gyrus, operculum, the parts of the inferior parietal lobule representing area 7 in monkey, the depth of the intraparietal sulcus representing monkey area VIP, the postcentral and precentral gyrus, the anterior insula and adjacent inferior frontal gyrus, the anterior cingulate gyrus, the precuneus, and hippocampus most often bilaterally (Fig. 5.8). Simultaneous with these signal increases (activations), signal decreases (deactivations) of areas within the visual and somatosensory systems of both hemispheres were observed.[32,123] Because opposite "activation-deactivation" patterns occurred during visually induced self-motion perception with activations of parietal visual cortex areas and concurrent deactivations of the multisensory (vestibular) cortex,[118,124] a reciprocal inhibitory cortical interaction between the sensory systems was assumed.[124]

Activation of the cortical network during vestibular stimulation is not symmetrical in the two hemispheres. Rather, it depends on three determinants that could be defined in a 2003 study investigating healthy right- and left-handers.[35] The determinants were (1) the subjects'

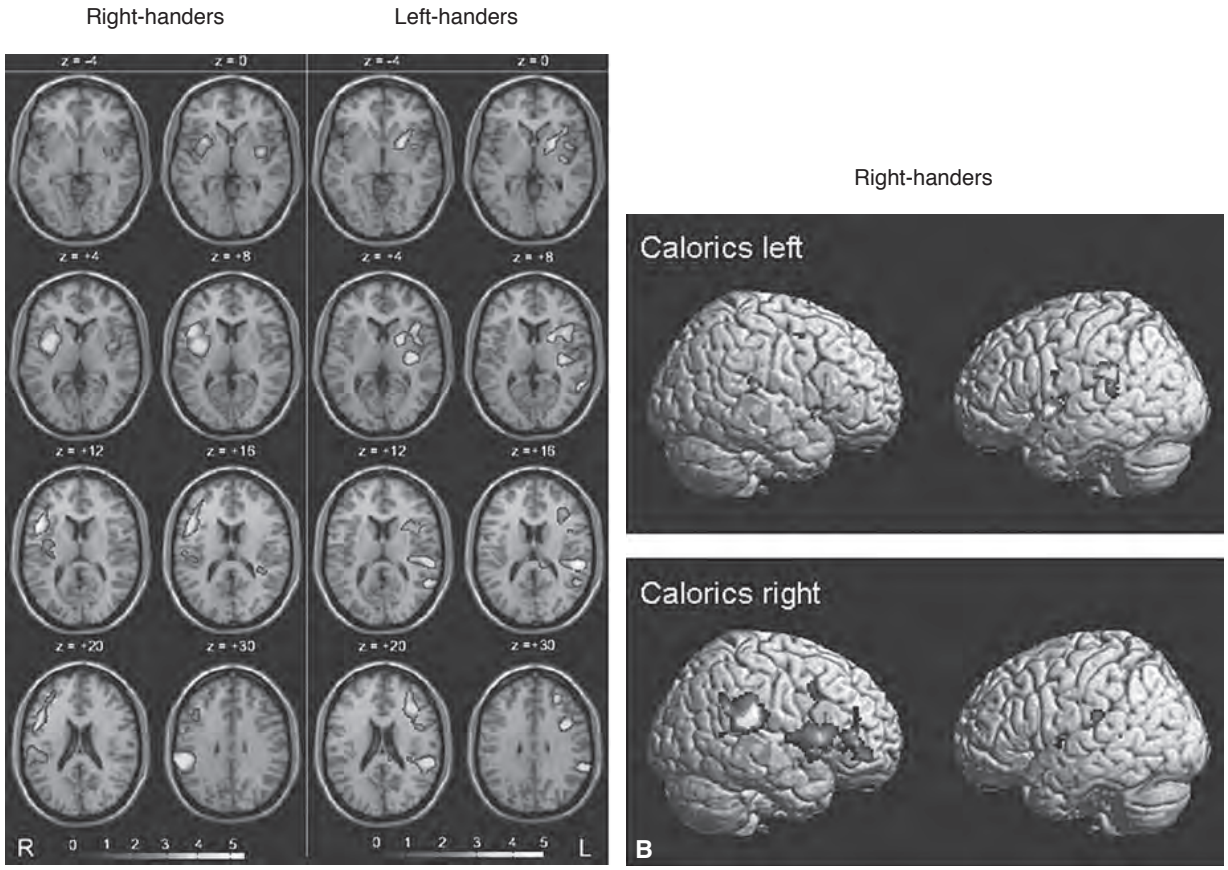

Figure 5.8 **(A)** Areas activated during caloric stimulation (warm water at 44°C) of the right ear in right-handed, and of the left ear in left-handed, healthy volunteers (group analysis ; n = 12; $P < 0.001$; ^{15}O-labeled H_2O bolus, positron emission tomography). Activations were located in the anterior and posterior insula, the superior temporal gyrus, the inferior frontal gyrus, the postcentral gyrus, the inferior parietal lobule, and the anterior cingulum. Note that the activations were more pronounced in right-handers during irrigation of the right ear in the right hemisphere and in left-handers during irrigation of the left ear in the left hemisphere. This finding indicates dominance of the nondominant hemisphere in the processing of vestibular information. **(B)** Lateral views of the surfaces of both hemispheres showing activated areas during caloric stimulation of the right or left ear in right-handers in the superior temporal cortex, temporoparietal junction, insular cortex, and inferior frontal cortex. Compared with the activation pattern during caloric irrigation of the right ear, caloric irrigation of the left ear led to activations that were smaller in both hemispheres and more frequently located within the ipsilateral left hemisphere. These results represent dominance of the ipsilateral vestibular pathways. (Modified from Dieterich et al, 2003,[35] and Bense et al, 2003[125]).

handedness, (2) the side of the stimulated ear, and (3) the direction of the induced vestibular symptoms. Activation was stronger in the nondominant hemisphere, in the hemisphere ipsilateral to the stimulated ear , and in the hemisphere ipsilateral to the f ast phase of v estibular caloric nystagmus.[30,35,126] Furthermore, in right-handed healthy volunteers, who performed allocentric visuospatial judgments (line bisection) with and without galv anic stimulation of the right or left v estibular nerve, the most relevant cortical area for the processing of v estibular information was located in the posterior insula bilaterally, right significantly more than left, including the PIVC. [127] Recently, a meta-analysis of vestibular stimulation studies showed convergent activation in a region near the posterior insula, the right posterior operculum (OP2), embedded in a joint v estibular network that seems to represent the human homologue to the v estibular area PIVC as proposed in nonhuman primates.[128]

Multimodal Sensorimotor Vestibular Cortex Function and Dysfunction

The human vestibular, visual, and somatosensory systems cooperate to determine the internal representation of space and subjective body orientation in unique three-dimensional coordinates, which are either egocentric (body-centered) or exocentric (world-centered). This is not a trivial process; two of the sensory systems are anchored in the head, which moves relative to the trunk. Retinal coordinates—dependent as they are on gaze and head position—and head-fixed labyrinthine coordinates would require continuous updating of the particular eye and head positions to deliver reliable input for adequate ocular motor and motor exploration of space.

Nature seems to have solved this sensorimotor control of a multilink and multiaxis system through multisensory coding of space in either common egocentric or exocentric rather than retinotopic or head-centered coordinates. This encoding has been demonstrated for posterior parietal neurons.[129,130] Spatial information in nonretinal coordinates allows determination of body position relative to visual space, which is a necessary prerequisite for accurate motor response. To obtain such a frame of reference, information coded in coordinates of the peripheral sensory organs (retina, otoliths, SCCs, and proprioceptors such as muscle spindles) must be transformed and integrated.[131] This function is most probably subserved by the temporal and posterior parietal cortex, a lesion of which produces a visuospatial hemineglect.

Karnath and associates[102,132,133] argued that neglect in brain-damaged patients is caused by a disturbance of the central transformation process that converts the sensory input coordinates from the periphery into an egocentric, body-centered coordinate system. The importance of the vestibular input for spatial orientation and the continuous updating of the internal representation of space is evident in the deficient spatial memory in microgravity during spacecraft missions. Large errors are made during prolonged microgravity when subjects are pointing at memorized targets, and it is the lack of knowledge of target position, not limb position, that is causative.[134]

In patients, an inappropriate vestibular input resulting from peripheral or central dysfunction can cause paroxysmal "room-tilt illusions," the result of a mismatch of the two three-dimensional visual and vestibular coordinate maps.[105,106] Furthermore, a plane- and direction-specific tilt of static spatial orientation occurs in disorders of the VOR, such as an eye and body lateropulsion in vestibular nuclei lesions (e.g., Wallenberg's syndrome). Adjustments of subjective straight-ahead also exhibit a lateral shift. Here the tilt of perceived straight-ahead is elicited by the asymmetrical vestibular tone in the brainstem, which reaches the cortex by ascending projections. Vestibular syndromes caused by only cortical lesions have not yet been well defined. Static cortical spatial disorientation may occur as any of the following:

- Paroxysmal room-tilt illusion in parietal or frontal lobe lesions
- Contralateral spatial hemineglect in temporo-insular, inferior parietal, or frontal lobe lesions
- Vertical neglect below the horizontal meridian in bilateral parieto-occipital lesions
- Tilts of perceived vertical (ipsi- or contraversive) and body lateropulsion in unilateral temporo-insular (e.g., PIVC) lesions

Dynamic cortical spatial disorientation with apparent motion or rotational vertigo may occur (1) in vestibular epilepsy with temporoparietal foci and (2) rarely as a transient vertigo in acute lesions of the vestibular cortical network.

Spatial Hemineglect: A Cortical Vestibular Syndrome?

Spatial hemineglect impairs focal attention toward space on the contralesional side. It is most often induced by acute brain damage of the inferior parietal lobule[135] and the temporo-insular region[102] of the right hemisphere[135] and occurs less commonly with acute right or left lesions of the frontal premotor cortex.[136] One case report described a patient who had sequential strokes in both hemispheres. After suffering a right-sided parietal infarct, he had a severe unilateral spatial neglect, which abruptly disappeared after a second, left-sided frontal infarct.[137] Other studies have described single patients with bilateral inferior parietal lobe lesions that manifested in vertical neglect of the lower half-space below the horizontal meridian.[138,139] Mesulam[140] hypothesized that there is a cortical network for directed attention in which the inferior parietal lobule modulates the shift of attention within extrapersonal space, and the dorsolateral frontal area is responsible for generating exploratory motor behavior. In summary, anatomical findings of the imaging studies and the observation of a right hemisphere dominance for processing vestibular input have obvious parallels with anatomical findings in patients suffering from spatial neglect. Damage to the right inferior parietal lobule and temporoparietal junction (TPJ)[135,141,142] as well as the inferior frontal gyrus[136,143] have been observed to correlate with spatial neglect. In addition, more recent studies have found the right superior temporal cortex and the right insula to be critically related to spatial neglect.[102,144-147,

Studies showing that vestibular (caloric) stimulation significantly improved spatial functioning have stressed the important role of the vestibular system in neglect.[148,149] When vestibular stimulation was combined with neck muscle vibration, the horizontal deviation combined linearly, adding or neutralizing the effects observed during application of both types of stimulation.[131] This study also showed that the patients with neglect displaced subjective body orientation ipsilesionally, a behavior that does not result from a disturbed primary perception or disturbed transmission of the vestibular or proprioceptive input from the periphery. Karnath and associates[132,133] argued that the transformation process converting the sensory input coordinates from the periphery into egocentric (body-centered) coordinates is the critical mechanism leading to hemineglect. This process must involve multisensory integration and motor behavior, including eye and hand movements as well as walking trajectory.[150] Spatial hemineglect also includes the back space of the body.[151]

Vestibular Epilepsy

Vestibular epilepsy (vestibular seizures or auras) is a rare cortical vertigo syndrome secondary to focal epileptic discharges in either the temporal lobe or the parietal association cortex[152-154]; multiple areas of both receive bilateral vestibular projections from the ipsilateral thalamus.

If vestibular seizures arise from different areas, the sensorimotor symptomatology may differ as regards apparent rotation or tilt,[155] with or without associated eye, head, and body deviation or epileptic nystagmus. Clinical data on the directions of apparent self-motion or surround-motion are mostly incomplete and imprecise. If the description is exact, as in rotatory seizures ("volvular epilepsy"), then the topographic localization of the underlying pathology is too inexact to permit its allocation to known vestibular areas.

An acute unilateral functional deficit of the vestibular cortex (e.g., in medial cerebral artery infarction) rarely manifests with vertigo,[156-158] unlike lesions in the vestibular area of the brainstem. It is not the functional loss but the focal discharge that causes central vertigo. This has been repeatedly demonstrated by stimulation experiments. Electrical stimulation of the human thalamus during stereotactic neurosurgical procedures induced sensations of movement in space, most frequently described as horizontal or vertical rotation or sensations of falling or rising.[159] These sensations were similar to those induced by stimulation of the vestibular cortex.[152,153]

Vertigo has long been considered a manifestation of epileptic auras.[160,161] Most information on auras (vestibular epilepsy), including case descriptions, comes from older textbooks, for example, those by Bumke and Foerster[162] and Penfield and Jasper,[153] or from review articles.[163,164]

In a later study searching for the human representation of "vestibular cortex," Kahane and associates[165] retrospectively investigated patients with epilepsy who had undergone stereotactic intracerebral electroencephalogram recordings before surgery and looked for patients in whom an illusion of rotation was induced. The investigators stimulated at 44 different loci in the temporal and parietal cortex and found that electrical stimulation of an area in the temporo-peri-Sylvian cortex particularly elicited rotatory sensations. This area included Brodmann areas 40, 21, and 22. Of these, the superior temporal gyrus (STG) and middle temporal gyrus (MTG) preferentially caused illusions of rotation around the subjects' yaw axis, whereas the parietal operculum elicited pitch plane illusions. Kahane and associates[165] thus confirmed earlier findings by Penfield and coworkers,[153,166,167] who had observed sensations of dizziness and rotary bodily movements especially during electrical stimulation of the STG in epileptic patients.

Epileptic nystagmus usually beats contraversive to the seizure focus and may be of vestibular, visual (optokinetic), or cortical ocular motor origin.[168]

Management

Vestibular seizures respond to antiepileptics. First-line drugs are levetiracetam, sodium valproate, carbamazepine, and lamotrigine. If necessary and possible, surgical procedures may be considered.

Paroxysmal Central Vertigo

Nonepileptic paroxysmal vertigo or other vestibular syndromes may result from pathological excitation of various vestibular structures.[169] Most of them occur in MS, but others may be associated with a brainstem abscess (paroxysmal OTR) or an arteriovenous malformation with previous bleeding (repetitive paroxysmal nystagmus and vertigo[42]) or brainstem infarction.

The following manifestations of paroxysmal vestibular syndromes of the brainstem have been described in MS:

- Paroxysmal dysarthria, ataxia, and vertigo[170]
- Paroxysmal OTR[171]
- Paroxysmal room-tilt illusion[172]

Central Vestibular Falls Without Vertigo

There are a few instances of what is probably central vestibular dysfunction. In such cases, patients without paresis or sensory or cerebellar deficits are unable to maintain an unsupported upright stance. They do not, however,

complain of vertigo. Their conditions include thalamic astasia, lateropulsion in Wallenberg's syndrome or in PIVC lesions, and ocular-tilt reaction in pontomedullary or rostral midbrain lesions.

Thalamic Astasia

Postural imbalance with a transient tendency to fall has been noted after therapeutic thalamotomy.[173-175] It has been attributed to muscle hypotonia or neglect and has also been observed after thalamic infarctions[176] and hemorrhages.[28,177] Masdeu and Gorelick[27] described 15 patients with "thalamic astasia," in the absence of motor weakness, sensory loss, and cerebellar signs, as a result of lesions of different causes, all primarily involving superoposterolateral portions of the thalamus but sparing the rubral region. "Typically, when asked to sit up, rather than using the axial muscles, these patients would grasp the side rail of the bed with the unaffected hand or with both hands to pull themselves up."[27] Thalamic astasia is transient and lasts for days or weeks, with the dorsothalamic region being the critical locus. Because posterolateral thalamic infarctions cause tilts of the perceived vertical that are either ipsiversive or contraversive,[24,30] thalamic astasia and tilts of perceived vertical may both reflect a vestibular tone imbalance. Furthermore, what Masdeu and colleagues[29] described as astasia and gait failure with damage of the pontomesencephalic locomotor region involving the right pontine peduncle area may be associated with vestibular dysfunction in roll. Their patient presented with a contraversive skew deviation of 10 degrees.

Thalamic hemiataxia differs from thalamic astasia and rarely occurs in isolation; it is usually associated with hemisensory loss without hemiparesis[178] or hemisensory loss and hemiparesis.[179] The lesions involve the ventral lateral nucleus of the thalamus and the adjacent posterior limb of the internal capsule and the mid- to posterior thalamus containing the dentatorubrothalamic and ascending pathways.[178,179]

Lateropulsion in Wallenberg's Syndrome

Lateropulsion of the eyes and the body is a well-known transient feature of dorsolateral medullary infarction. Affected patients have irresistible, ipsiversive falls but generally no subjective vertigo. Different brainstem lesions from midbrain to medulla cause ipsiversive deviation of the subjective vertical.[10,180,181] Transient OTR and ipsiversive deviations of SVV, which indicate a pathological shift in the internal representation of the gravitational vector, are typically found in Wallenberg's syndrome.[23,181]

We hypothesized that the subjective vertigo is missing in patients with this syndrome (despite a striking tendency to fall sideways) because individual multisensory postural regulation is adjusted to the deviated vertical. Lateropulsion then represents postural compensation of an apparent body tilt contraversive to the lesioned side. Despite the resulting postural imbalance and the conflicting true vertical, the body is continuously adjusted toward what the central nervous system erroneously computes as vertical.[23,181] This hypothesis could explain why patients fall without vertigo or warning signals from the multisensory spatial orientation system. These deficits are centrally compensated during several months via brainstem-cerebellar loops.[26] Lateropulsion without hemiparesis also occurs in cortical lesions. Patients with infarctions of the middle cerebral artery territory are well known to physiotherapists, who call them "pushers." It has been demonstrated that acute lesions of the PIVC cause contraversive tilts of the perceived visual vertical,[31] making it likely that the cortical lateropulsion can also be caused by a vestibular tone imbalance in the roll plane.

Lateropulsion in both dorsolateral medullary lesions and posterior insular lesions spontaneously recovers within days to weeks.[181] The recovery process might be facilitated by physical therapy.

Summary

Central vestibular syndromes are characterized by ocular motor, postural, and perceptual signs. In a simple clinical classification they can be separated according to the three major planes of action of the VOR: yaw, roll, and pitch. A tone imbalance in yaw is characterized by horizontal nystagmus, lateropulsion of the eyes, past-pointing, rotational and lateral body falls, and lateral deviation of the perceived straight-ahead. A tone imbalance in roll is defined by torsional nystagmus, skew deviation, ocular torsion, and tilts of head, body, and the perceived vertical. Finally, a tone imbalance in pitch can be characterized by some forms of upbeat or downbeat nystagmus, fore-aft tilts and falls, and vertical deviation of the perceived straight-ahead. The thus defined syndromes allow for a precise topographic diagnosis as regards their level and side. Most signs and symptoms of central vestibular disorders resolve spontaneously within weeks to months owing to either recovery of the lesion or central compensation and substitution. The predominantly benign course of these syndromes may be facilitated by physical and drug therapy.

Acknowledgment

The authors wish to thank Ms. J. Benson for copyediting the manuscript.

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CHAPTER 6

Postural Abnormalities in Vestibular Disorders

Emily A. Keshner, PT, EdD ■ *Anne K. Galgon, PT, PhD, NCS*

When this chapter was written for the first edition of this book, the vestibular system was considered to play its primary role in the control of posture and balance. In recent years it has been acknowledged that rather than initiate automatic postural reactions, the vestibular system is responsible for governing orientation in space. [1-3] Whereas the somatosensory system provides information about the position and motion of the body with respect to the support surface and the body segments with respect to each other, the vestibular system provides information with respect to gravity and other inertial forces. The central nervous system adapts quickly to the loss of peripheral vestibular inputs from the labyrinths so that it is sometimes difficult to objectively identify symptoms of vestibular deficit. Identification of the role of the vestibular system in posture and orientation has relied on findings of postural and orientation disorders in patients and animals with vestibular abnormalities. [4-8] Most clinical diagnoses are based on subjective complaints, and patient descriptions of a symptom might differ. One might experience a perception of the world spinning about, while another complains of imbalance and falling, yet both could have the same vestibular pathology.[9] Since the process of central nervous compensation proceeds over a lengthy period of time, patients can also have different symptoms when they finally arrive at a clinic, although suf fering a similar deficit. Both clinical and experimental observations have shown that symptoms of vertigo, past pointing, nystagmus, and equilibrium disturbances are the major complaints of patients with partial or total destruction of the vestibular labyrinths.[10] Despite these fairly consistent symptoms, examination of any one patient with postural abnormalities arising from damage to the vestibular system could yield an uncertain diagnosis.[9,10]

The question for the clinician and the clinical investigator is whether any one compensatory strategy would prove most efficient or effective for the population of patients with a vestibular deficit. If one compensatory strategy is best, then a systematic approach to treatment could be followed. But to determine the ef fectiveness of the compensation, we must first determine how to reliably indicate whether the patient suffers from postural dyscontrol, and whether it is vestibular dysfunction that is responsible for the symptoms. Although standard clinical tests of the vestibular system have not changed much, technologies such as virtual reality (VR) and vestibular evoked myogenic potentials (VEMP) allow us to explore vestibular function when combined with other sensory signals (VR) or to specifically identify what part of the labyrinths or vestibular pathways may be affected (VEMP).[11,12]

In this chapter, methods available for testing postural disorders that are associated with vestibular pathology are briefly discussed. Then, postural behaviors that have been quantified and associated with specific vestibular pathologies are described. Finally, the issue of how the postural system compensates for loss or damage to vestibular signals, including the changes that occur with natural aging, are discussed.

Examining the Vestibulospinal System

Advantages and Limitations of Clinical Tests

Although vestibular disorders continue to be diagnosed through measures of the vestibulo-ocular system such as electronystagmography and rotational testing, these cannot fully describe all disorders of the vestibular system. One problem is that tests of eye movement (vestibulo- ocular) integrity and postural (vestibulospinal) function may not be correlated.[9] First, the well-defined loop of the vestibulo-ocular reflex (VOR) does not reveal the integrity of the more complex vestibulospinal pathways, which are intimately involved in the control of posture and balance. Second, tests of the vestibulo-ocular reflex are commonly performed in the plane of the horizontal semicircular canals, whereas vestibulospinal reflexes are dependent on inputs from the vertical semicircular canals and the otoliths.

A clinical test that is able to partially reveal information about vestibulospinal function is the vestibular evoked myogenic potential (VEMP). This is a muscle response generated from acoustical stimulation of one of the otoliths that transduces linear accelerations and decelerations. The cervical VEMP (cVEMP) is a measure of a short-latency response from a tonically contracted ipsilateral sternocleidomastoid (SCM) muscle. The presence, absence, or abnormal amplitude and latency of this muscle response provides diagnostic information about the function of the saccule, the inferior vestibular nerve,[12-15] or the brainstem pathways to the ipsilateral SCM.[15] The ocular VEMP (oVEMP) test provides diagnostic information about the utricle, the superior vestibular nerve, and the associated brainstem pathways to the contralateral inferior oblique ocular muscle (Fig. 6.1). (For more details on otolith tests see Chapters 11 and 12.)

Diagnostic utility of the patterns of oVEMP and cVEMP has been examined for various audiovestibular and neurological disorders, including vestibular labyrinthitis or neuronitis, Ménière's disease, vestibular migraine, benign paroxysmal positional vertigo (BPPV), superior canal dehiscence, Tullio phenomenon, vestibular schwannoma, multiple sclerosis, and spinocerebellar degeneration.[12-16] The relationship between abnormal cVEMP responses and postural dysfunction (i.e., vestibulospinal mechanisms) is still not well studied. Individuals with saccular or inferior vestibular nerve dysfunction and asymmetrical cVEMP

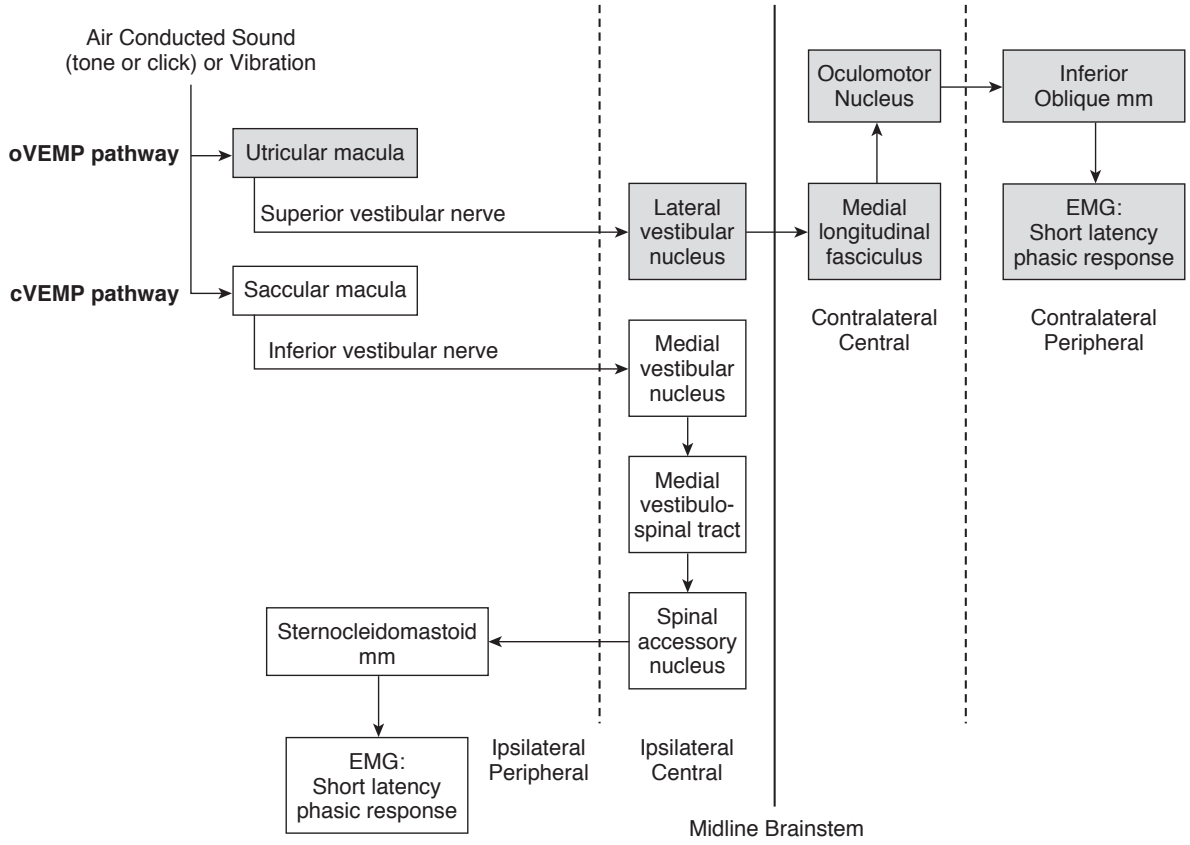

Figure 6.1 Diagram of the cervical (cVEMP in black) and ocular (oVEMP in gray) vestibular evoked myogenic potential pathways from input of an acoustic stimulus to output at the muscle.

responses demonstrate abnormal sensory organization tests as compared with healthy individuals; however , balance performance and self-perception of dizziness was not different from individuals with unilateral caloric weakness and normal cVEMP responses.[17]

Although dynamic posturography is not a direct assessment of peripheral or central vestibular function, it is a useful tool for identifying impairments associated with dysfunction of the vestibulospinal system. [18-20] Dynamic posturography assesses balance rather than specific vestibular function, but response patterns that are specific to individuals with vestibular dysfunction can be elicited with dynamic posturography. Thus, it is a useful adjunct to more traditional methods of testing vestibular function. There are tests of the vestibulospinal system that are more easily and inexpensively available to the clinician than dynamic posturography and that can be manipulated to better reveal vestibulospinal dysfunction (Table 6-1).

■ Table 6-1 **ADVANTAGES AND DISADVANTAGES OF CLINICAL TESTS OF POSTURAL INSTABILITY**

	Advantages	Disadvantages	Test Manipulations (challenge individuals with vestibular compensation)
		Static Tests	
Romberg	Easily performed in clinic Quantify by time till loss of stability	Qualitative Does not test adaptive responses	Narrow base of support Can manipulate sensory stimuli Can add head turns
Stabilometry	Quantitative	Requires a force platform or kinematic or inertial sensors Intersegmental shifts confound results Does not test adaptive responses	Can manipulate sensory stimuli Can add head turns
Vestibular Evoked Myogenic Potential (VEMP)	Specific test of the saccule and inferior vestibular nerve or utricule and superior vestibular nerve.	Requires electromyography of the sternocleidomastoid muscle for the cVEMP and the inferior oblique muscle for the oVEMP	Can apply a tone or click or vibration
		Dynamic Tests	
Stepping Tests	Easily performed in clinic Quantified by the number of steps before loss of stability	Does not test adaptive responses Has not been shown to be reliable	
Tiltboards	Easily performed in clinic Requires adaptation to external forces	Qualitative Amplitude and application of force are not controlled	
Posturography	Quantitative Requires adaptation to external forces	Requires a posture platform	Can manipulate sensory signals
Virtual Reality	Tests the effect of visual information on postural reactions—incorporates perception with postural responses	Requires expensive technology, a knowledge of programming, and some form of kinematic or physiologic measurements	Can manipulate sensory signals (haptic, auditory, visual)

One such manipulation is head movement or head shaking that has been shown to destabilize standing in individuals with vestibular dysfunction.[21] Stabilizing balance is more difficult in humans when the head is moving, because otolith sensory function is less effective at determining orientation.[22] Standing during head shaking becomes more difficult, particularly when vision or somatosensory information is absent or inaccurate,[22] and clinicians can add head turning to postural control tests even when performance appears normal. Several clinical balance and gait observational measures include head turning among the items to test functional balance (see Chapter 21). Clinicians should note, however, that speed of head turning in these clinical tests is not well controlled. Additionally, the loss of stability with right versus left head turns in standing or walking does not correlate with the side of vestibular involvement.[23] However, this manipulation increases the difficulty of a test and may identify functional postural control deficits in older adults and in individuals who may be partially compensated after vestibular hypofunction.

Dynamic Posturography

Automatic Postural Responses

In the 1970s, Nashner reported stereotypical, automatic responses to postural disturbances initiated at the base of support, introducing the measurement of postural reactions on a moving platform as a powerful experimental approach.[24,25] Since that time, the majority of studies of postural kinematics have concentrated on the electromyographic (EMG) responses from muscles in the lower limb, from which most descriptions about restabilizing actions have been drawn.[24-29] Subjects stand on a platform that could be translated in an anterior and posterior direction, or rotated so that the ankles are moved into plantar or dorsiflexion (Fig. 6.2). The expected response to anterior motion of the platform is backwards sway (base of support moved in front of the center of mass), producing a decreased angle at the ankle and a stretch of the ankle muscles on the anterior surface of the body (i.e., tibialis anterior). If the platform moves posteriorly, the subject sways forward (base of support moved behind the center of mass), thereby decreasing the ankle angle and stretching the gastrocnemius and soleus muscles. Rotating the platform into plantar or dorsiflexion would produce equivalent changes at the ankle (see Fig. 6.2), but the center of mass remains in line with the base of support.

Although the monosynaptic stretch reflex does not act functionally to replace the center of mass over the base of support, EMG analysis of the lower limb muscles revealed that the muscles being stretched also responded at

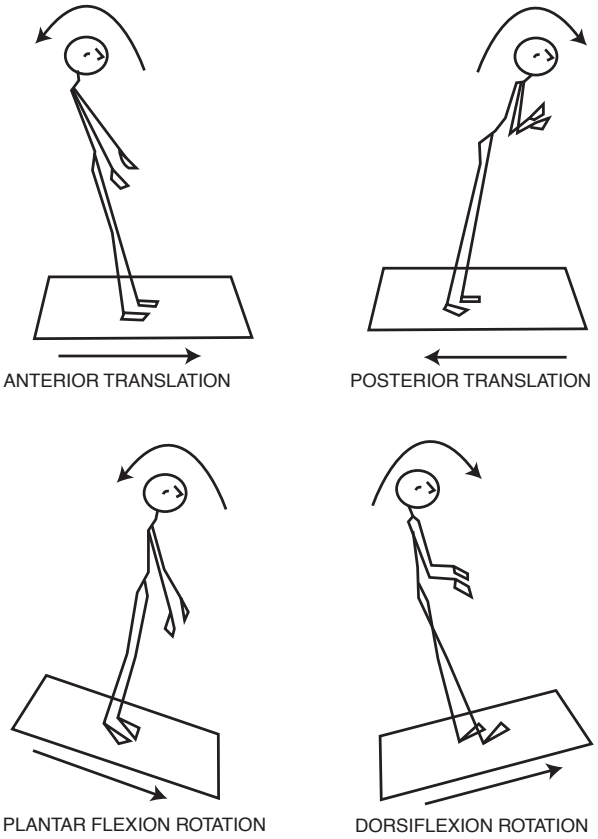

Figure 6.2 Four directions of perturbation on the dynamic posture platform. Note that in anterior translations and plantar flexion rotations, ankle angles increase as the person compensates for the motion of the platform. In posterior translations and dorsiflexion rotations, ankle angles decrease.

latencies longer than the stretch reflex but shorter than voluntary reactions to bring the body back over the base of support. These restabilizing ankle muscle responses (at latencies of 90 to 120 ms) were followed within 10 to 20 ms by the muscle in the upper leg on the same side of the body (i.e., soleus followed by the hamstrings; tibialis anterior followed by the quadriceps). Thus, from these early studies, patterns of muscle activation initiated by ankle proprioceptive inputs, and arising from distal to proximal lower limb muscles, were identified as ascending muscle synergies responsible for restabilization after platform movement.[24,25]

Nashner's original conclusion that the body acts as a rigid, inverted pendulum, reliant primarily on ankle proprioceptive inputs to initiate the restabilizing actions, does not accurately describe the complex, multisegmental actions that occur during postural restabilization.[27,29-32] The "ankle strategy" is most effective when sway is slow and the support surface is firm. When it is impossible to exert adequate torque around the ankle joint, as when the base

of support is narrow or compliant, balance is recovered with flexion at the hip or a "hip strategy."[33] Allum et al[34] concluded that all balance corrections result from one of two basic synergies defined by the timing between the body segments, and that the amplitude of the selected synergy was modulated according to the initial movement velocities at all joint segments. Thus, there is a continuous repertoire of movement strategies for balance rather than two discrete strategies.

With more demanding stimuli (e.g., rapid perturbations and a center of mass moving far beyond the base of support), the ability to take a step has been found to be the relevant criteria for recovery of balance.[35,36] Other studies have also demonstrated that the initiation of the balance reactions is highly dependent both on the ability to predict the occurrence of the perturbation,[37-40] and on the location of the disturbance on the body.[41-44]

Identifying Vestibular Contributions to Automatic Postural Responses

Since the earliest presentation of Nashner 's findings, investigators have been attempting to define the contribution of the vestibular system to the automatic postural responses.[45-49] Studies in which the labyrinthine receptors were directly stimulated by vertically dropping human and animal subjects, thereby producing linear acceleratory stimuli,[46-49] demonstrated that direct labyrinthine stimulation can produce automatic or "triggered" postural reactions in the lower limb. Nashner[26] hypothesized, however, that the vestibular system contributes to the control of lower limb balance reactions only when proprioceptive signals are absent or unreliable. Motion of the dynamic posture platform was made to match the sway at the hip, thereby maintaining a neutral position at the ankle. Proprioceptive feedback from the ankle thereby became unreliable and the subjects would have to rely on vestibular signals. The outcome was that the automatic postural responses were significantly delayed.

If we explore this more carefully, it is probable that the servomechanism did not fully remove the ankle proprioceptor feedback but rather produced distorted or modified signals that altered the automatic postural reactions. Another consideration is that the responses to vestibular inputs during quiet stance are not equivalent to those responses generated during dynamic gait and a loss of balance when there are strong accelerations of the head. To resolve the issue of labyrinthine involvement in the generation of postural reactions to support surface displacements,[50] angular displacement of the ankle was kept equal for both platform translations and rotations, thereby producing equivalent proprioceptive signals from the ankle although the acceleration inputs to the labyrinths were different. When head acceleration and neck and lower limb muscle EMG responses recorded during both types of perturbation did not exhibit the same response patterns, the investigators concluded that labyrinthine signals must be directly involved in the generation of lower limb postural reactions. Differences between the two subject groups were greatest and most consistent during platform rotations,[51] leading these researchers to believe that vestibular loss is best diagnosed using a rotation of the support surface. Further studies using a range of platform translation velocities (5–35 cm/sec) [52] have shown that equivalent head accelerations produced very different hip and knee torques between healthy subjects and patients with bilateral vestibular deficit. This suggests that the magnitude and force pattern of the muscles depends very much on the presence of vestibular inputs from early head movements.

Altering Sensory Cues

Manipulating the visual and somatosensory inputs that are available during dynamic posturography is another method employed to isolate a person' s ability to use vestibular signals. More commonly used protocols to alter somatosensory inputs include a sway referenced platform that matches the normal sway at the ankle during quiet standing, or adding a layer of dense foam to the base of support to make somatosensory inputs less effective.[53] Visual conditions have been controlled by either stabilizing or rotating the visual field in an anterior -posterior plane.[53-55] The most widely used test of altered sensory cues is the sensory organization test or SOT.[56] The SOT systematically measures postural sway in standing under six conditions. This includes condition 1 (eyes open, fixed surface); condition 2 (eyes closed, fixed surface); condition 3 (eyes open, fixed surface, sway referenced visual field); condition 4 (eyes open, sway referenced surface, visual field fixed); condition 5 (eyes closed, sway referenced surface), and condition 6 (eyes open, sway referenced surface, sway reference visual field). The emergence of virtual reality technology has now enhanced the ability of scientists and clinicians to explore the impact of visual perturbations on postural responses (see Table 6-1).[11,57]

Although normal subjects, elderly individuals, and those with vestibular deficit exhibit an increased tendency to fall under conditions of altered sensory input,[53,54, 56, 58, 59] concluding that the cause is a vestibulospinal system unable to compensate for the loss of other sensory signals may be premature. Modification of somatosensory and visual inputs is not necessarily equivalent to a loss of those signals, and the central nervous system may well compensate for distorted or minimized inputs by altering the sensorimotor transformation algorithm. For example, the

system may select a compensatory strategy that relies on enhancing the gains of somatosensory and visual responsiveness to the distorted inputs rather than shifting responsibility for the response onto the vestibulospinal system. Disorientation of labyrinthine-loss patients when visual[54] or somatosensory[60,61] information becomes unreliable can also be explained by the fact that neither signal can adequately specify the orientation of the body in all situations. In natural environments we rely on both visual and vestibular signals to define our orientation in space, and evidence indicates that labyrinthine-deficient individuals become more sensitive to and dependent on visual inputs with vestibular loss.[62-64]

Clinical researchers have attempted to improve the sensitivity of distinguishing between postural control in younger and older adults[64-67] and individuals with vestibular deficits[20,68-70] by adding head shaking to dynamic computer posturography and the sensory or ganization test (SOT). The head shaking SOT (HS-SOT) adds a horizontal head shake to conditions 2 (eyes closed; surface fixed) and 5 (eyes closed; sway referenced). The head shaking component is accomplished by having the individual perform horizontal head turns at a peak velocity of 60 deg/sec for 20 seconds.[20] At least one study,[69] however, noted ceiling effects in condition 2 and floor effects in condition 5 of the HS-SOT in individuals with vestibular deficits, which led to a modification that includes head shaking at additional velocities (120 deg/sec in condition 2 and 15 deg/sec in condition 5).[67,69] Lim et al[68] showed that the HS-SOT conditions 2 and 5 were related to perception of disability through the Dizziness Handicap Inventory up until 6 months after an acute unilateral hypofunction. The SOT without the head shake only correlated in the first 1 week after the acute event. Thus the HS-SOT may be a valuable outcome measure in vestibular rehabilitation for individuals with loss of balance provoked by head movement.[68,70]

In summary, postural responses to support surface displacements have traditionally been tested by: (1) translating a standing subject along the earth's horizontal plane on a moving platform, (2) rotating the foot about the horizontal axis of the ankle into dorsiflexion or plantar flexion, and (3) keeping the platform fixed to the earth horizontal or sway referencing the angle of the platform or visual field to match the angle at the ankle during quiet sway. Experiments using the posture platform have been performed with a wide range of velocities and amplitudes of displacement that alter the transmission of forces from the lower limb to the head and make the comparison of vestibular influences on balance difficult across laboratory settings. Instructions to the subjects and monitoring of past experiences with the paradigm are not regulated.A further restriction on the interpretation of results is that measurement of postural responses should incorporate the mechanics of body sway with the threshold properties and dynamic characteristics of the labyrinthine receptors,[29,52,71-76] because knowing the mechanics of the head and motions of the center of mass is necessary for predicting the role of canal and otolith feedback in restabilization. Despite its limitations, and the multitude of variables that should be controlled, the posture platform continues to be employed as a method for obtaining quantitative measures of postural reactions in clinical populations. Many of the results reported in this chapter have, in fact, depended on the posture platform methodology to examine disturbances in postural control as a result of vestibular dysfunction.

Tests of Quiet Stance

Traditional clinical examinations of vestibulospinal function include tests of self-localization, such as the Romberg test.[77] Initially, Romberg's test of instability was based on a population of patients with proprioceptive loss from tabes dorsalis who were unable to stand with feet together and eyes closed. But the Romberg test is insensitive for detection of chronic unilateral labyrinthine impairment[78] and is highly variable even within a subject.[78,79] Modifying the test by having the patient stand in a tandem heel-to-toe position (sharpened or tandem Romberg) has made the test more sensitive, probably because of the narrowed base of support. Even so, tests of quiet stance fail to measure the adaptive components of the postural response that are essential to dynamic balance during most daily activities.[80]

Tests of quiet stance may indicate the severity of a balance problem because patients with vestibular system damage will demonstrate increased sway and falling when the base of support is constrained (e.g., narrow base of support or compliant surface) during quiet standing. Conclusions about the neural processes contributing to postural imbalance are severely limited. The effect of altered proprioceptive and cutaneous information on low-frequency sway stabilization cannot be determined by tests of quiet standing. Changing velocity of the visual field is a significant parameter controlling body sway during quiet standing,[80] but simple removal of vision does not alter the temporal or spatial organization of the automatic postural reactions.[45] Furthermore, behavioral measures as to how often a subject falls or to which direction he deviates do not convey information about the motor and sensory mechanisms that may be involved in postural control.[81,82] Thus, attempting to assess the integrity of the vestibular system through a test of quiet standing opens the door to

many confounding variables and is far from specific to the vestibulospinal disorders that may produce a postural deficit.[83,84] Despite these limitations, the concept of deviation from the vertical during quiet standing continues to underlie clinical testing of vestibular dysfunction.

There has been renewed interest in quantifying postural sway in clinical examination of individuals with vestibular loss.[85-88] A compliant foam surface has been shown to be very sensitive for identifying vestibular disorders. Varela et al[86] demonstrated increased sway amplitudes between healthy individuals and those with vestibular involvement in standing on compliant surfaces with eyes open and eyes closed. A large-scale survey of balance and vestibular function in the United States used a compliant surface as an indicator of vestibular dysfunction and an increased risk of falling, and found that more than 35% of adults 40 years and older experienced difficulty standing with eyes closed on the compliant surface.[89]

Recent development of clinical applicable testing systems utilizing inertial sensors may help clinicians in both diagnosing impairments[86] and monitoring improvements[85] with vestibular rehabilitation during tests of quiet stance. Inertial sensors attached to pelvis and trunk [86] or sacrum[87] may help capture sway amplitudes during clinical balance and gait tests. Horling et al [85] measured sway standing with eyes closed (Romberg test) with firm surfaces and eyes closed on compliant surfaces in individuals with vestibular loss, extremity proprioceptive loss, and healthy individuals. They found that compared with healthy individuals, those with vestibular loss had increased sway in the eye closed and compliant surface standing condition, and those with proprioceptive loss had increased sway in both the eye closed on firm and compliant surface conditions.

Stabilometry

Stabilometry is a clinical tool that measures anterior-posterior and lateral excursions of the body in subjects standing quietly on a force platform, usually over time.[90-92] During that time, the subject stands quietly on a force plate, and the excursion of the center of gravity is measured across several conditions that can include eyes open, eyes closed, and eyes closed with head extension. Attempts to stress the vestibulospinal system have been incorporated into this system of measurement by altering signals from other sensory pathways. For example, adding a layer of foam rubber (compliant surface) to the base of support to make somatosensory inputs less effective,[53] or placing the subjects within a visually controlled environment to modify visual feedback.[54] This attempt to quantify the classical Romberg test has made the measurement of postural sway during quiet standing more objective, but the mechanisms contributing to the observation of increased sway still cannot be identified. One problem is that changing the position of the body parts (either randomly or through experimenter directive) could shift the center of pressure without affecting the stability of the subject.[90] In general, because the sensory apparatus of the vestibular system is most responsive to changes in acceleration and orientation in space,[93] and because patients with vestibular deficits tend to have normal Romberg signs, tests of quiet standing on a stabilometer are not compelling measures of vestibulospinal function.

Tiltboards

Tilt reactions, or reflexes opposing bodily displacement, traditionally were evoked through a lateral tilt or anterior/posterior tilt of the supporting surface about a horizontal axis.[4-6] On tilting the base of support, the reaction to regain a stable equilibrium occurred by moving the body against the angular momentum and repositioning the center of gravity within the vertical projection of its base of support.[4-8] These reactions have also been elicited in the clinic by simply pushing the patient at the shoulder girdle. Problems with the accuracy of this test are threefold. First, because the tilt reactions are measured by observational techniques, later voluntary responses (greater than 150 ms) rather than automatic postural reactions are being evaluated. Second, the response pattern alters if the force is applied directly to the trunk rather than to the support surface. Third, tilt responses will be organized differently depending on whether your patient is pushed or trips over an obstacle in the environment, whether the application of perturbation is predictable, and whether it is self-induced or elicited.

Stepping Tests

The Unterberger[94] or stepping test of Fukuda[95] examines the ability of patients to turn about a vertical axis when marching or stepping in place. Marked variability in the amount of rotation produced by even the same subject, however, makes these tests unreliable.[9] Patients with severe disruption of the vestibular system may stagger so uncontrollably, that the stepping tests cannot reliably indicate the side of the lesion.[96] A battery of tests developed by Graybiel and Fregly [97] (Ataxia Test Battery) examines subjects standing upright, on one leg, and with feet aligned in tandem position with eyes open or closed, as well as tandem walking in a straight line on the floor or on a narrow rail. This test is useful for patients who have compensated for a labyrinthine deficit because,

when a narrowed base of support is required, even those patients score more poorly than normal subjects on measures of deviation from the straight line or of the number of steps made before falling from the rail.

Virtual Reality Environments

The perception of self-motion and orientation in space is derived from a convergence of vestibular, proprioceptive, and visual signals. To resolve ambiguity between motion of objects in the world and self-motion, we use multisensorial feedback and make perceptual choices about what we believe is happening.[98] Virtual reality (VR) technology (Fig. 6.3) offers the sensory complexities found in the physical world in the controlled environment of the laboratory[99] Individuals become immersed in the training environment so that they feel they are part of the scene [100] and perceive that the world is moving about them.[101] Thus, VR allows us to create a synthetic environment with precise control over a large number of physical variables that influence behavior while recording physiological and kinematic responses.[102]

In a VR environment, healthy subjects have demonstrated large increases in postural sway when the frequency and amplitude of visual information was incongruent with the frequency and amplitude of the somatosensory signals generated by a support surface perturbation.[103] In this case the visual information had no parameters in common with the physical disturbance; thus, it could have been treated as irrelevant to the postural perturbation and ignored. Instead, in both populations the postural responses became more complexly organized by incorporating an additional frequency into the intersegmental response and further increasing the magnitude of the response. Almost all subjects, including those with labyrinthine deficit, shifted from a hip strategy to a partial inverted pendulum when the visual signal varied in frequency, indicating that when the visual signal had nothing in common with the physical disturbance, the response was to stiffen the mechanical system and decrease the degrees of freedom being controlled. Although the additional frequency was present in the responses of the adults with labyrinthine deficiency, their responses did not increase in size suggesting either that, in the absence of labyrinthine inputs, they were unable to produce responses of any greater magnitude,[45] or that modulating the response magnitude was dependent on an ability to calculate the visual-vestibular conflict.

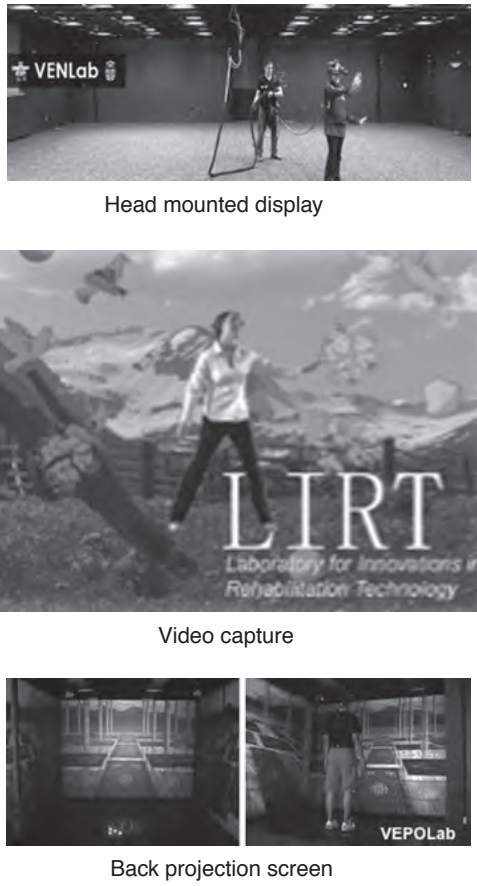

Figure 6.3 Some examples of virtual reality technology in which the performer can freely move while interacting with the visual images. **(A)** The performer can walk about while wearing a head-mounted display as in the VENLab directed by Dr. William Warren at Brown University (http://www.cog.brown.edu/research/ven_lab/). **(B)** Video capture in the Laboratory for Innovations in Rehabilitation Technology directed by Dr. Tamar Weiss at the University of Haifa permits performers to observe themselves interacting with virtual objects (http://staffsw.haifa.ac.il/index.php?option=com_content&view=article&id=1&Itemid=12). **(C)** In the Virtual Environment and Postural Orientation (VEPO) Laboratory directed by Dr. Emily Keshner at Temple University, a dynamic posture platform (NeuroCom International, Inc.) has been placed in front of a 3-wall full field of view back-projection screen to simultaneously perturb the base of support and the visual system of the performer (https://sites.google.com/a/temple.edu/vepo/about-us).

Postural Reactions in Peripheral Vestibular Disorders

A discussion of etiology and diagnostic testing of vestibular system disease is beyond the scope of this chapter, but can be found in other sources.[9,104,105] The focus here will be on those vestibular disturbances that have been found to produce a postural disturbance, and that have been tested for changes in vestibulospinal function. Dysfunction in the vestibulospinal system can be divided into two categories: distortion and deficiency.[106,107] A *deficiency* in the system usually implies that the sensory (i.e., labyrinthine) inputs have been reduced or abolished,

resulting mostly in complaints of unsteadiness and instability. *Distortion* indicates that the signal is present but disturbed, and does not correspond with expectations about the sensory feedback. The result would be inappropriate or false motor responses to the existing situation (e.g., vertigo and ataxia). A summary of postural disturbances is presented in Table 6-2 for the disorders discussed in this chapter.

Deficient Labyrinthine Inputs

Damage along the eighth nerve or within the vestibular labyrinth produces lost or diminished signals from the peripheral vestibular apparatus.[93] Central disturbances originate at the vestibular nuclei or in the central pathways that communicate with the vestibular nuclei. In both cases, patients can experience disequilibrium, imbalance, and ataxia. With unilateral lesions of the peripheral system, the normal symmetry of inputs from the right and left labyrinths become disordered, resulting in a decreased firing rate of the vestibular nuclei on one side. A unilateral lesion affects the system as if the intact side were being stimulated, thus generating an illusion of change in head orientation and movement. The inherent disequilibrium then activates the vestibulospinal system to respond inappropriately, resulting in vertigo, nystagmus, and postural instability.

Another effect of vestibular system stimulation, maintaining tone of the muscles against gravity , appears to be directly correlated with labyrinthine inputs because the

■ Table 6-2 **POSTURAL DISTURBANCES OBSERVED WITH VESTIBULAR DISORDERS**

Peripheral Vestibular Disorder		Clinical or Functional Expression
Deficient Inputs	Need more energy to maintain the upright position	Complaints of fatigue with sustained upright activities
	Instability increases in the presence of inappropriate sensory signals	Complaints of instability in complex sensory environments
	Amplitudes of EMG and torque are inversely related to severity of deficit	Reduced corrective responses when destabilization is detected by head movement
Distorted Inputs	Still able to process vestibular inputs	Postural response normal when vertigo is not active
	Falls increase in the presence of inappropriate sensory signals	Complaints of instability in complex sensory environments
Central Vestibular Disorder	Impaired perception and location of the gravitational vertical	Head and trunk tilt
	Direction-specific ataxia	Lateropulsion and loss of gait trajectory
	Falling tends to occur in the direction of quick phase nystagmus	Lateropulsion results in falls toward the direction of horizontal nystagmus
Aging	Longer response latencies and delayed reaction times	Slow at responding to destabilization
	Diminished sensory acuity and impaired signal detection	Slow at responding to destabilization
	Postural response patterns are temporally disordered	Inappropriate postural response to destabilization

activation of extensor muscles in the extremities of both monkeys[108] and humans[109] with unilateral deficit were enhanced contralateral to the side of the lesion. But postural reactions are more complex than single pathway vestibular reflexes, and cannot be traced and localized as easily as these direct line responses. For example, when both labyrinths are lesioned, an artificial sense of motion does not occur, and neither do the symptoms of nystagmus and vertigo. Yet, equilibrium is still disturbed, suggesting that the balance function of the vestibular system is not a simple response to stimulation of the labyrinthine receptors.

Indicators of Vestibulospinal Deficiency

Unilateral and Bilateral Labyrinthine Deficit

Variability of the responses measured from the many methods of posturography confirms the complexity of control of these disordered postural responses. After repeated attempts to quantify the results of the Romberg test, the most reliable effort seems to be measuring energy of the power spectral densities of the center of force trajectories when maintaining an upright position.[110] Both in this study and in others using force plates to record sway during quiet stance,[10,92] intersubject variability and overlap between normal and clinical populations reduced the strength of the findings. Results suggest, however, that more energy needs be expended to maintain an upright position when visual inputs are removed (eyes closed) from patients with a labyrinthine deficit.

In a series of papers presented by Black and Nashner,[56,106,110] postural sway was recorded through a potentiometer placed at the level of the hips. Patients stood on a platform that could be either earth fixed, or moved proportional to body sway (sway referenced). The visual environment was then manipulated so that patients experienced a visual field that was either: (1) earth fixed, (2) proportional to body sway, or (3) removed by eye closure. Patients with reduced or absent labyrinthine inputs were unstable compared with normal controls only when ankle proprioceptive references were proportional to body sway and visual references were either removed or inappropriate (conditions 2 and 3 above). When the only reliable source of feedback was the vestibular inputs, it was believed that patients with vestibular deficiencies would fall because they were dependent on the somatosensory and visual reference to correctly organize their postural responses. From these studies, Black and colleagues suggested that vestibular deficits could be quantified by systematically altering the sensory information provided by the support surface and the visual surround.[110]

Several problems limit our reliance on these results for clinical diagnosis and measurement. First, patients with unilateral or partial bilateral deficits at times were as unstable as patients with total loss of vestibular function,[81] thus rendering this a poor test of graduated function in the vestibular system. Second, the authors tested only well-compensated patients. As will be discussed later, compensation could occur as a central reorganization in the system. Thus, these experiments may not be testing a vestibular deficit, but rather a compensatory subsystem that responds inadequately to the presented stimuli. Third, patients with postural instability from other, non-vestibular disorders may have test results similar to those of patients with vestibular deficits (Hain and Herdman, personal communication).

Allum and colleagues examined both the latencies and amplitudes of muscle EMG responses on a platform that dorsiflexed the ankle.[27,34,45,50,51,82,109,111] Areas under the EMG bursts in the ankle muscles, soleus, and tibialis anterior, and ankle torque recordings of patients with complete bilateral labyrinthine deficit, were significantly diminished when compared with normal subjects with eyes both open and closed. Using these data, the presence of a linear correlation between EMG amplitudes and the extent of the peripheral vestibular deficit was explored.[112] The population measured included those with intact labyrinths (normal subjects), acute unilateral labyrinthine deficit patients, chronic unilateral labyrinthine deficit patients, and bilateral labyrinthine deficit patients, thus covering a graduated range of labyrinthine function.

A stepwise discriminant analysis technique performed on the data suggested that muscle response amplitudes in the soleus and tibialis anterior muscles, as well as amplitude of torque exerted on the platform, were inversely related to the severity of the labyrinthine deficit. Muscle and torque responses diminished in amplitude as the reception of labyrinthine inputs decreased. Because lower limb EMG activity was still present in the patients with complete loss of labyrinthine inputs, a linear correlation of the amount of EMG activity with extent of peripheral vestibular deficit suggests that lower limb postural reflexes could be triggered by proprioceptive stretch reflexes, but that amplitude modulation is under the control of, or requires the presence of, vestibulospinal signals. EMG activity in the neck muscles was not obviously altered in these patients, implying local control of neck muscle responses. Thus, the effectiveness of the ankle muscle responses to produce a functional forward torque in

patients rotated backwards on a platform was diminished, and these patients tended to fall backwards.

EMG responses of patients with vestibular deficit during horizontal translations of a platform were also examined.[51,61] Although latencies of the postural reactions were produced without significant time delays, segmental organization of the postural response were disordered. Allum et al[112] found that angular velocity measures of rotation of the trunk about the hip were not significantly altered in patients with a bilateral peripheral vestibular deficit, and that a hip strategy was a common component in the postural response to platform rotations. These investigators suggested that whichever movement strategy was selected depended on the initial direction of trunk and head acceleration (which is oppositely directed in platform rotations and translations), and was executed as if preprogrammed from the beginning. Although Horak et al[61] previously stated that hip strategies were not seen in patients with vestibular disorders, new measures of hip torques emphasize the importance of identifying the proper response variables when using dynamic posturography as a diagnostic tool. Runge et al[52] found that the joint torques at the hip and knee were abnormal over a range of velocities during rearward translations for patients with bilateral deficit. Because head accelerations were the same in the patients and healthy subjects, these investigators agreed with others' findings[27,45] that the magnitude and force pattern of the muscles depends on vestibular inputs from early head movements.

We can conclude that patients with partial or total loss of labyrinthine input exhibit diminished amplitudes of EMG response and thus require greater energy expenditure to maintain balance, particularly when another source of stimulation to the system (e.g., visual inputs) has been removed. These patients also exhibit greater sway in sensory conflict situations, and variability between patients is a common clinical occurrence because of the dynamic central compensatory processes. With unilateral deficit, the initial perception of apparent body motion is directed away from the side of the lesion. Postural reactions are usually in a direction opposite to the direction of vertigo, causing a tendency to fall and deviate toward the affected side. Severe postural disturbances occur when these individuals have to rely on vestibular inputs, but the deficit is compensated within 2 weeks after the lesion.[113] Individuals with bilateral deficit exhibit normal postural sway in quiet stance.[61] Removal of vision with eye closure increases this sway by only a small amount.[106] These patients tend to fall backward when their eyes are closed during dorsiflexion tilts of a platform. Their diminished amplitudes of muscle activity presumably result in the reduced restabilizing torques recorded at the ankle, knee, and hip.[114]

Ménière's Syndrome

Ménière's syndrome, or endolymphatic hydrops, is considered to be a vestibular deficiency although it presents as fluctuating vestibular function. Symptoms of acute Ménière's syndrome include hearing loss, tinnitus, and a sensation of fullness or pressure in the ear.[9] Patients with this syndrome exhibit a negative Romberg sign during remission,[115] but symptoms of dizziness and instability can occur for several days following intermittent episodes of vertigo. These episodes appear at irregular intervals for years, and about one-third of the patients eventually develop bilateral involvement.[9]

Objective diagnosis of Ménière's syndrome has been dependent on long-term documentation of the fluctuating hearing loss. Quantitative posturography measures have been able to identify consistent changes in the postural response of these patients, even during periods of remission. The movement pattern of the center of gravity was measured during a stepping test after observing that patients deviated toward the affected side even during the remission period.[115] Stepping was performed in the dark with eyes open and closed, and patients were required to perform at a frequency that was both optimal for normal walking and that elicited a smooth rhythmical pattern (i.e., 1.2 Hz). When eyes were closed, the patients exhibited angular deviations of 30 degrees or more toward the affected side after 8 to 12 seconds of stepping. Time to deviation indicated a degrading central motor program that was initiated by visual inputs, but which required vestibular inputs (in the absence of vision) to be maintained over time.

Measures of sway during quiet standing have included analyses of the pattern of motion, displacement, and power spectrum of the center of gravity. With all of these measures, position of the center of gravity changed in an irregular fashion, and deviated primarily toward the affected side.[116,117] High-frequency components of standing sway were observed during acute phases of Ménière's syndrome, but not during remission periods.[117] Patients with Ménière's syndrome who had not developed vestibular hypofunction, as determined from vestibulo-ocular reflex gains, were also tested under conditions of sensory conflict during quiet standing (see earlier description in section on Indicators of Vestibulospinal Dysfunction).[110] These patients responded very much like well-compensated patients with unilateral or bilateral loss of labyrinthine inputs. The group had nearly normal responses on all trials with the platform fixed to earth horizontal. Responses fell outside

the normal range when either the platform or the visual field was perceptually stabilized, again suggesting dependence on reliable inputs from the vestibular labyrinths during these test conditions.

Indications of Vestibulospinal Distortion

Benign Paroxysmal Positional Vertigo

Patients with BPPV have been examined to study the effects of distorted labyrinthine inputs on posture.[110] The key to this syndrome is that brief episodes of vertigo (usually less than 1 minute) are generated with position change. Paroxysmal positional nystagmus can be observed with rapid changes of position. After a period of several attacks, symptoms can become more prolonged, and include dizziness and nausea lasting for hours or days. [9] Degeneration of the utricular macula releasing otoconia that influence the cupula of the posterior semicircular canal is strongly implicated as the cause of BPPV , and could result from a variety of etiologies (e.g., trauma, infection, ischemia). The intensity of BPPV depends on the velocity of the positional changes, and attacks can be avoided if positions are assumed very slowly.[118]

Because of the positional component of this syndrome, postural changes are easily recorded during quiet stance by having patients alter the position of their head in space. After tilting the head, large amplitudes of anterior-posterior sway and sway ipsilateral to the direction of head tilt were observed.[118] Instability gradually decreased as the vertigo diminished, but with eyes closed, the sway could not be compensated by other inputs, and falling occurred.

Unlike patients with a loss of vestibular inputs, patients with distorted inputs from BPPV reacted normally on a moving platform when forced to rely only on their vestibular inputs. More disturbing to this group of patients were inappropriate (perceptually stabilized) visual circumstances whether the platform was earth fixed or moving proportional to body sway.[81,106,110] Patients with BPPV probably rely primarily on visual information to organize their postural reactions, and have suppressed their response to the potentially unreliable vestibular inputs.

Postural Reactions in Central Vestibular Lesions

One could erroneously assume that function of the vestibular labyrinths is directly representative of the functional integrity of the vestibular system. Although receiving direct inputs from the peripheral labyrinths, the vestibular nuclear complex also receives visual and somatosensory inputs.[93] Convergence of vestibular and somatosensory input onto the vestibulospinal and reticulospinal[119] neurons can take place at the level of the vestibular nuclear complex, at the adjacent reticular formation, and on spinal interneurons[120,121] and motoneurons.[119] However, increasing evidence[122,123] suggests that correct alignment of the head with the trunk and with the gravitoinertial vertical [124] requires that the vestibular system receive ascending somatosensory inputs. To attain an appropriate postural response, a convergence of sensory information from the vestibular, somatosensory, and visual systems is needed to align the body with respect to earth vertical.Thus, with labyrinthine loss, even if the otoliths remain intact, the ability to identify the upright orientation may be impaired.[125] Inputs from either of these modalities are not necessarily redundant because each represents dif ferent parameters and is effective within a particular frequency domain.[126-128] In fact, the frequency of stimulation is important to control with compensated patients because motor output of the visual system and the vestibular system has been found to be frequency dependent. [76,129-132] Thus, it is unlikely that normal postural responses are reflective of the isolated labyrinthine and neck reflexes observed in the decerebrate animal.[133-135] Instead, postural reactions probably emerge from a dynamic coupling of all available sensory signals. Body posture can be oriented to a visual, somatosensory, or vestibular reference frame depending on the task and behavioral goals. It may also be based on an estimated internal representation of body orientation with respect to the environment from memory, thereby incorporating the expected multisensory inputs and flexible postural reactions occurring for each task.[136,137] A convergence of inputs from more than one sensory modality would then be necessary to create this estimated postural orientation. Our studies in a virtual environment[138] demonstrated that converging inputs controlled postural orientation during rotations of the visual scene in pitch and roll. The head and trunk were linked in magnitude and phase, whereas the ankle produced small compensations that were lar gely out of phase with the upper body. We inferred, as in a previous study on a posture platform, that the upper body responded to visual-vestibular signals while the ankle responded to proprioception and changes in ground reaction forces. [30] Buchanan and Horak[139] also reported differential controls of the head and trunk and of the lower limbs when examining segmental organization of postural responses. Thus, the cause of instability in labyrinthine-deficient individuals may be caused by a disorder of head and trunk spatial orientation rather than lower limb instability.[140]

Several clinical findings have been suggested to differentiate between a peripheral and central disturbance in the vestibular system. Gradually increasing disturbances of standing, walking, and falling in the direction of the

quick phase of spontaneous nystagmus have been identified as indications of a central vestibular lesion.[141] Balance disorders as a result of abnormalities of the vestibular nuclear complex have been observed, [142,143] but are poorly documented. The majority of the literature about central vestibular brainstem lesions reports only oculomotor abnormalities, but Brandt et al [143] have attempted to relate well-defined central vertigo syndromes to characteristics of postural imbalance. Briefly, these investigators reported five conditions for which postural imbalance have been consistently reported: downbeat nystagmus vertigo syndrome, ocular tilt reaction, Wallenberg's syndrome, paroxysmal and familial ataxia, and brainstem lesions that mimic labyrinthine dysfunction. One should recognize, however, that structures other than the vestibular system may be damaged and affect balance.

Downbeat nystagmus is specific for a lesion of the paramedian craniocervical junction (30% of cases caused by Arnold-Chiari malformation), inducing a direction-specific vestibulospinal ataxia. Static head tilts modulate the intensity of the nystagmus and the postural sway, suggesting involvement of otolith function. The typical postural imbalance in this condition is an anterior -posterior sway with a tendency to fall backwards, but many of these patients do not complain of vertigo or balance problems. Brandt et al[143] suggest that the backward sway is vestibulospinal compensation to the forward vertigo resulting from the downbeat nystagmus. *Ocular tilt reaction* is actually a triad of responses, including ipsilateral head-trunk tilt, ocular torsion, and ocular deviation. Patients seem to have a readjustment in their perception of the vertical that matches the actual tilt deviation of the eye, head, and trunk. This condition has been observed in patients with brainstem abscess, multiple sclerosis, and acute Wallenberg's syndrome. *Wallenberg's syndrome* is an infarction of the dorsolateral medulla resulting in ipsilateral dysmetria of the extremities, pain and temperature loss, and a lateropulsion of the eyes and head causing the body to deviate toward the side of the lesion and, consequently fall. A scale was developed that grades the severity of this phenomenon and its time course for recovery.[144] Classifications of severity are: Grade I: Moderate head-trunk tilt without imbalance; Grade II: Head-trunk tilt with considerable imbalance, but no falls; Grade III: Head-trunk tilt with imbalance falls with eyes closed; and Grade IV : Head-trunk tilt with imbalance falls with eyes open.

Paroxysmal and familial ataxias share the broad-based, unsteady gait that defines ataxia. Finally , pontomedullary lesions near the vestibular nuclei at the entry of the eighth nerve can mimic a peripheral labyrinthine disorder, and *drop attacks* (a sudden, unpredictable forward falling) can occur with basilar insuf ficiency. Thus, the evidence from clinical reports suggests that a central vestibular dysfunction results in impaired perception and location of the gravitational vertical exhibited throughout the whole body postural system. But with all of these syndromes, other motor structures are af fected as well, and may contribute to the impairment.

Postural Dysfunction with Pathology of Other Sensory-Motor Centers

The vestibular nuclear complex communicates with motor as well as sensory centers.[141] In fact, extensive reciprocal connections between the vestibular nuclei and the*cerebellum*[145] argue for a prominent role of the cerebellum in regulating the output of the vestibulospinal system, and lesions of the cerebellum result in severe postural disturbances. Three kinds of cerebellar ataxia have been identified, suggesting different pathophysiological mechanisms that are dependent on the site of the lesion.[146] A test of the sway stabilizing responses on a posture platform of patients with late cortical atrophy of the*anterior lobe* of the cerebellum revealed that response latencies were within normal limits following dorsiflexion rotations and backward translations on a platform, but amplitudes and durations of response were two to three times greater than normal,[147,148] and habituation to the stimulus was absent.[149] Postural response magnitudes of the patients with anterior lobe damage were scaled correctly when relying on current somatosensory feedback, but the patients were unable to scale their responses when relying on prior experience. Thus, the major effect of anterior lobe cerebellar damage on postural responses may be an impairment of responses based on predictive central set.[148] This may have implication on motor learning and intervention to improve postural performance. Individuals with cerebellar damage may show poor motor learning, show slower recovery of postural control, and require more practice.[150]

A characteristic sway frequency of 3 Hz has been recorded in this population.[151] Intersegmental counterbalancing actions were enhanced in these patients, so that falling was not commonly observed, but they tend to exhibit a stiff-legged gait. Stance ataxia was found to improve with visual feedback, unlike that appearing with vestibulospinal lesions.[152, 153] Thus, in these patients, stabilizing responses occurred, but they lacked the balance between opposing muscle forces and grading of response over time.

The postural system of patients with lesions of the *vestibulocerebellum* (flocculus, nodulus, and uvula) may be so severely impaired that these patients cannot walk.

Ataxia of the head and trunk is observed while sitting, standing, and walking. These patients exhibit unusually large sway in all directions with predominantly low frequencies of less than 1 Hz, and visual stabilization appears to be reduced when the Romberg test with eyes open and closed is compared. These patients tend to fall even when sitting down, which may be a result of diminished intersegmental movement for counterbalancing or to truncal ataxia.[146,152] *Neocerebellum* lesions produce little postural instability or disturbance of stance even with eyes closed. Control of position of the body's center of mass seems to be disturbed since these patients exhibit ataxia during a limb and trunk pursuit task.[146] Reports of head and trunk deviation to the side of the lesion have also appeared.[152]

With *basal ganglia* disorders, such as Parkinson's disease, equilibrium reactions are often delayed or absent.[4] An anticipatory postural response in the soleus muscle normally seen in response to a perturbation of the forearm, is absent or reduced in these patients, [154] although long latency responses to direct stretch of a muscle have been observed to be enhanced in the Parkinson population in both the upper arm[155] and lower limb.[156] Response latencies to sudden platform displacements were found to be within the normal range,[157,158] although the ability to suppress long-latency muscle reactions to a perturbation was impaired in these patients even under supported conditions.[159] When required to scale the magnitude of their responses to amplitude and velocity of backward platform translations, patients with Parkinson's disease produced smaller than normal extensor bursts, larger than normal flexor bursts, and smaller torque responses.[160] On a sinusoidally moving treadmill, patients with Parkinson's disease were able to maintain their balance with eyes open by using their leg flexor muscles whereas healthy subjects activated their extensor muscles. Timing and amplitude of this muscle activity were also impaired. [161] The inability to generate adequate force in the stabilizing muscles appears, therefore, to be hampering successful postural reactions in these patients. Inferring the contribution of the basal ganglia or the impairment of vestibular information during dynamic postural reactions in this patient population is difficult, however, because the motor impairments could be as much an effect of akinesia, rigidity, or aging as of disruptions in the postural control system.

Lesions in the *motor cortex* have also resulted in disturbances to the automatic postural reactions. Patients with spastic paresis rarely exhibit disturbances of posture during quiet standing as in the Romberg test, but reactions to rapid displacements of the support surface indicate deficits in the dynamic postural reactions. [162] Hemiplegic adults demonstrate delayed onsets, a failure to respond, and disparate responses of agonist and antagonist muscles in the paretic lower limb during postural perturbations on a platform.[163] With augmented feedback, such as a warning tone and knowledge of perturbation direction, however, timing of the postural responses improved. [164] When balancing on a seesaw apparatus, patients with spastic hemiparesis minimized the high-frequency, anteroposterior sway on the affected side with a corresponding reduction of the EMG response in tibialis anterior.[165,166] Electrical stimulation of the tibial nerve in patients on the seesaw revealed a delayed and diminished EMG response of tibialis anterior in the affected leg, thereby interfering with the normal compensatory response to displacement of the support surface. Spastic paraparetic patients were observed to produce qualitatively similar results.[165,166]

Children with cerebral palsy were also studied on a posture platform.[167] Their instability seemed to correlate with the clinical diagnosis so that children with spastic hemiplegia exhibited reversals in the expected order of muscular activation, whereas children with ataxia demonstrated normal muscle sequencing yet fell frequently. The timing, direction, and amplitude of their postural reactions were disturbed, particularly when the expected sensory inputs had been altered (see paradigm described in section on Indicators of Vestibulospinal Dysfunction). Thus, postural abnormalities of children with cerebral palsy were due either to muscle incoordination or instability as a result of an inability to deal with sensory conflict.

Results of these clinical studies suggest that the long-latency, polysynaptic postural adjustments can be elicited at the spinal level, but require modulation by supraspinal structures to develop a sufficient response threshold and gain. Possibly there are an inappropriate number of nerve fibers within the damaged motor pathway to excite the motoneuron pool, or the damaged pathway sends a reduced drive to the interneurons at segmental levels that would normally facilitate the polysynaptic reflex response.[162]

Mechanisms for Recovery of Postural Stability

Identification of compensatory mechanisms will improve therapeutic interventions that teach compensation for or adaptation to, destabilizing conditions. These mechanisms are studied through clinical research, but we must be cautious about conclusions drawn about the function of an anatomical site that are based strictly on the absence of motor control in the presence of specific deficits or damage. We must remember that responses generated in the absence of a sensory or motor signal do not reveal the function of that input. Rather, these responses demonstrate how the system operates in the absence of certain inputs.

Sensory Substitution

Vestibular, visual, and somatosensory signals influence the organization of a normal postural response. When any one of these signals is lost or distorted, a central reweighting occurs so that the remaining sensory inputs are used to elicit postural reactions, albeit in some altered fashion (Fig. 6.4). Changes in the postural response organization with loss of labyrinthine inputs have been described in detail in earlier sections of this chapter. Two modifications in particular should be noted. First, in the absence of labyrinthine signals, the normal postural reactions to dorsiflexion of the ankle are elicited, but with significantly diminished amplitudes.[111] Thus, the response does not reach an appropriate gain to maintain stability, and restabilizing torques at the ankle are inadequate to prevent falls.

Second, peripheral vestibular deficit patients tend to hold the neck stiff so that little free head movement occurs. An analysis of the temporal relationship between angular acceleration of the head and trunk in the flexor and extensor directions demonstrates that patients move the head in the same direction as the body (head locked to trunk) associated with absent neck torques, [168] while normal subjects exhibit a counter rotation of the head and body in the sagittal plane. [83,112,123] This finding correlates with clinical observations that vestibular deficit patients increase gain of neck muscles to hold the head stiff in relation to the body. A fast Fourier transform performed on the head and trunk angular acceleration recordings revealed a loss of the normal 2- to 3-Hz peak in the power spectrum of patients with bilateral labyrinthine deficit. [45] This frequency has been cited as the operating frequency for the vestibulocollic reflexes in studies of normal subjects attempting to stabilize the head during vertical and horizontal rotations in the seated position,[76,130] and is typical of natural head movements during locomotion. [169,170] Stiffening of the muscles may, therefore, be one compensatory strategy that actually works against successful restabilization by interfering with the normal balance of movement-dependent torques at the different body segments, and with the reception of stimuli necessary to produce vestibular adaptation.

Somatosensory inputs provide powerful feedback about motion of the limbs and stabilize body sway at the lower frequencies (less than 1 Hz). Studies have shown that sensory input to the hand and arm through contact cues at the fingertip or through a cane can reduce postural

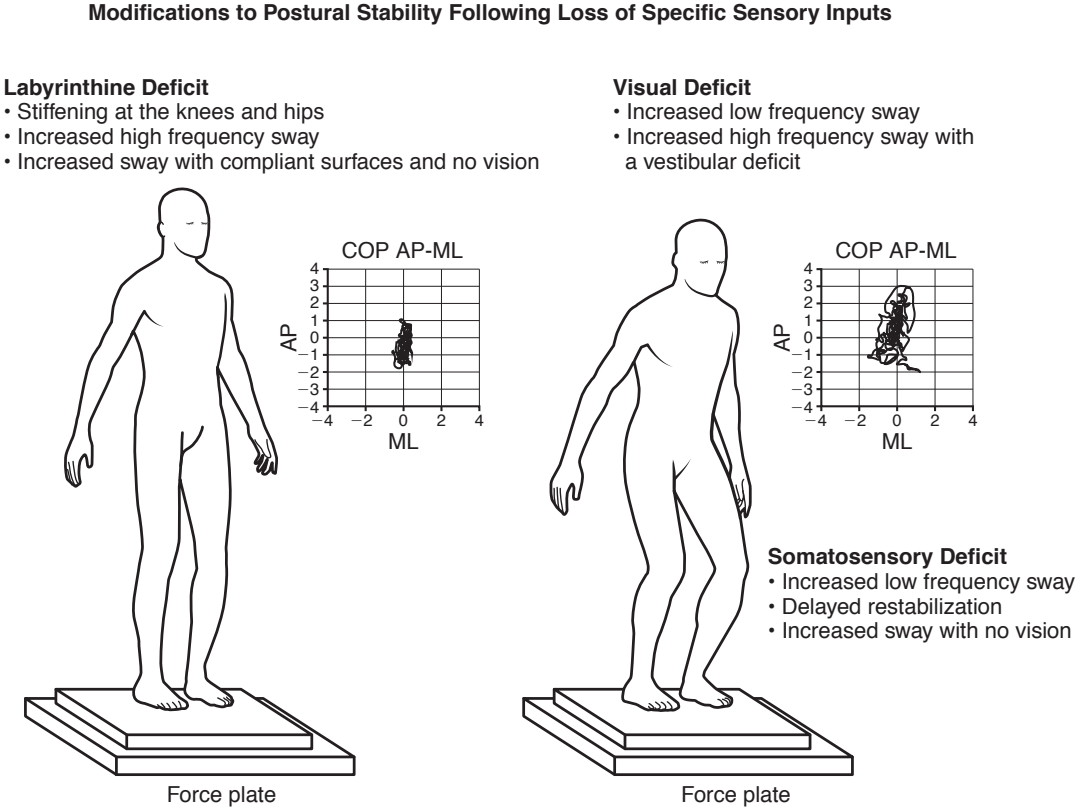

Figure 6.4 Cartoon depicting changes in postural sway patterns and segmental actions following loss of vestibular, somatosensory, or visual signals.

sway in individuals who have no impairments and in patients without a functioning vestibular system, even when contact force levels are inadequate to provide physical support of the body.[159,171] When proprioceptive feedback from the ankle was excluded or suppressed in normal subjects during perturbations on a posture platform, [21,172] a characteristic low-frequency (1 Hz) sway emerged. Postural abnormalities have been observed with impairment of spinal pathways such as occurs with Friedreich's ataxia, a hereditary disorder affecting the spinocerebellar pathways and posterior columns.[173] In the absence of feedback from these pathways to the cerebellum, a significant delay of the restabilizing response of the tibialis anterior muscle following dorsiflexing ankle rotations on a posture platform has been observed. [147] These patients exhibit large lateral sway deviations in the low frequency range (less than 1 Hz) with eyes closed, as do patients with tabes dorsalis.[172] Patients with sensory polyneuropathy of the lower extremities demonstrate ataxia and instability during quiet stance. Falls tend to occur when the eyes are closed, [173] suggesting that visual inputs are necessary along with vestibular inputs in the absence of lower limb proprioceptor signals.

Finally, cervical proprioceptors have been the focus of investigations related to the diagnosis of dizziness and ataxia.[174,175] The neck proprioceptors have intimate connections with the vestibular system and are probably used both as feedforward and feedback to the vestibulocollic reflexes.[176] Cervicogenic dizziness has been a controversial diagnosis, however, because there are no hard signs to identify the neck as the source of the dizziness![175] Karlberg and colleagues[177-179] have engaged in a series of studies to verify ataxia or vertigo of cervical origin. Using posturography in which stance was perturbed by a vibratory stimulus applied toward the calf muscles, they studied patients with recent onset of neck pain and vertigo but normal otoneurological findings. Results demonstrated disturbed postural control in the patients with cervical vertigo that differed from patients with vestibular neuritis. In addition, dorsal neck muscle tenderness and tightness was found in a majority of the patients with cervicogenic dizziness that was diminished following treatment for neck pain.[180] These results suggest that disorders of the neck need to be considered when assessing patients complaining of dizziness, vertigo, and balance disturbances.

Visual signals are used to accurately detect and reduce motion relative to the surround. [181,182] In normal subjects, vision is very influential, but does not appear to be an essential input for the recovery of balance. Many studies have shown that simply removing vision will not produce significant changes in the postural response organization, although greater sway amplitudes may appear.[45,55,58] Instead, visual information is thought to be redundant unless both vestibular and somatosensory inputs are lost.[182] To test the importance of visual inputs in the absence of labyrinthine inputs, sway was measured in subjects standing on a stabilometer placed within a laterally tilting room.[54] At low frequencies of sinusoidal tilt (0.0025 to 0.1 Hz), patients with unilateral and bilateral labyrinthine deficits exhibited sway similar to that of normal subjects. At higher frequencies (0.2 Hz), the patients' sway increased beyond normal limits, indicating that patients with vestibular deficit could rely on visual inputs at lower frequencies, but suffered for the loss of vestibular signals at higher frequencies.[80,183] Labyrinthine deficit patients on a stabilometer were better able to stabilize sway when fixating on a stationary light.[184] Patients became unstable when an optokinetic stimulus was introduced, which indicates that velocity information received through peripheral vision is the cause of the increased sway.

The influence of velocity of the visual field on postural control was further explored in a three-wall virtual environment.[185-187] Visual field motion was found to be very compelling for both healthy young and elder individuals, and produced changes in the postural response organization even when they were standing quietly on a hard surface. This finding justified using a virtual environment to test the effectiveness of a postural training program for individuals with labyrinthine disorders. [188] One woman had a bilateral labyrinthine deficit, and the other suffered from unremitting Ménière's disease that did not respond to treatment. After being trained to stand in the dark on an increasingly unstable sway referenced platform, these individuals were tested on the platform while performing a reaching task in the presence of a rotating visual field. Center of pressure responses were shown to improve significantly both immediately and 2 weeks following the training compared with performance on the task before training.

In summary, patients lacking labyrinthine inputs become more dependent on accurate ankle proprioceptive and visual references to correctly organize their postural responses. Inappropriate or distorted signals along either of these sensory pathways will produce increased sway and falls in these patients. Although the sensory signals often provide congruent information, inputs from any of these modalities are not necessarily redundant because each represents different parameters and is effective within a particular frequency range.[80,126,127] Thus, falls observed in vestibular deficit patients, particularly following a platform perturbation or in the absence of other sensory signals, may be caused by uncontrolled or poorly compensated oscillations of intersegmental structures at particular frequencies of sway.

Compensatory Processes

Compensation for vestibular pathology is a gradual process of functional recovery that is probably of central origin.[189,190] Although adaptation (i.e., long-term changes in gain) of lower limb postural reactions has been observed in patients with labyrinthine deficit,[109,191] these individuals still evidence a great deal of instability . Thus, adaptation to support surface inputs alone is not an effective compensatory process. Numerous structures have been identified as participating in vestibular compensation including the vestibular nuclei, spinal cord, visual system, cerebellum, inferior olive, and more.[190] Thus, focusing specifically on a single site for functional recovery of postural control would be difficult. In fact, studies have shown that in both humans and animals, methods of compensation for vestibular dysfunction are not comparable either across subjects or within a subject for different functions.[190,192] The only consistency seems to be that the goal of postural compensation is to reorganize the neural circuitry so that bilateral stimulation of the vestibular system is kept in balance.

The role of central processing in posture control can be observed in anticipatory adjustments that are preprogrammed and can create the inertial forces necessary to counterbalance an oncoming balance disturbance. [193-195] Subjects asked to co-contract to counteract random sum-of-sines rotations[196] exhibited neural control over a greater bandwidth than when distracted or relaxed, suggesting that feedforward signals could alter the reactive components of the response. Horak and Nashner[33] found that prior experience as well as current feedback information influenced the selection of postural strategies on a translating platform. When subjects had some knowledge of the characteristics of a forthcoming visual displacement, they were able to reduce their postural readjustments even when they did not exert active control over the visual motion.[76] Central control over postural responses can be measured in studies examining predictive processes. For example, Guitton et al[129] assessed the influence of mental set on the relative importance of visual and vestibular cues for head stabilization in humans. Normal subjects and patients with bilateral vestibular deficit were tested on their ability to stabilize their heads voluntarily with visual feedback and in the dark, and while distracted with a mental arithmetic task while being rotated horizontally using a random (white noise) stimulus with a bandwidth of 0 to 1 Hz. Normal subjects stabilized their heads best when voluntarily attempting to keep the head coincident with a stationary visual target. Vestibular deficit patients had comparable response amplitudes with vision present, but much lower amplitudes when vision was removed. The apparent absence of head stabilization when subjects performed mental arithmetic suggested that the short-latency (approximately 50 ms), head-stabilizing reflexes provided little effective head stabilization at these frequencies of rotation.An analysis of response latencies revealed that long-latency or voluntary mechanisms (occurring at greater than 150 ms) were primarily responsible for the observed head stabilization.

Anticipatory presetting of the static and dynamic sensitivity of the postural control system also assists in stabilization of the head at high frequencies. [195,197] Practice or prior experience with a postural task influences EMG output. With practice, decreasing size of the EMG response to a plateau level has commonly been observed during stabilizing reactions,[25,45] suggesting central habituation of these responses at the cortical or spinal levels. Selection of postural strategies on a translating platform is influenced by prior experience as well as current feedback information. [33] When the task is well practiced, subjects are able to combine complex movement strategies and respond quickly under a variety of different posture platform paradigms. Even chronic patients with labyrinthine deficits eventually demonstrate normal sway,[92] indicating that a central regulatory mechanism is compensating for the peripheral dysfunction.

A study of head-stabilizing responses with random frequencies of sinusoidal rotation may have revealed the method by which the CNS is regulating postural compensation.[198] When tested in the dark at high frequencies (up to 4 Hz), compensation for bilateral peripheral labyrinthine deficit manifested as a shift in system mechanics. In the horizontal plane, patients were able to compensate for trunk rotation by increasing stiffness of the head and neck. In the vertical plane, patients maintained a stable head over a broader frequency range than healthy adults, possibly by changing stiffness or increasing the gain of the cervicocollic reflex to compensate for instability at higher frequencies. The inability of patients to stabilize at low frequencies (below 1 Hz) in the sagittal plane suggests that the system requires otolith inputs for quiet standing. The ability to maintain a stable head at more functional higher frequencies, however, suggests that individuals with a labyrinthine deficit have acquired an adaptive strategy that reveals the complex integration of CNS control over the mechanical properties of the system.

Changes in Postural Reactions with Aging

An impaired vestibular system is believed to be involved in the increased instability of the elderly because anatomical studies have revealed a gradually decreasing density of labyrinthine hair cell receptors beginning at age 30, and a steeper decline in the number of vestibular receptor ganglion cells beginning around 55 to 60 years of age. [199-201] Although caloric measures of the peripheral vestibular

system have demonstrated declining function with age,[200] these changes are not present in the central vestibular neurons. The gradual loss of labyrinthine acuity with age prompts viewing the elderly population as a model for compensation to vestibular dysfunction, but in the elderly, sensory loss occurs as a slow process along several feedback pathways, not just the vestibular pathways.[199-204] Thus, their compensatory approach to postural instability may not be the same as in those patients who have experienced an acute but sustained loss of a single input (see Table 6-2).

Age-related trends in the vestibulo-ocular and optokinetic reflexes have been shown to correlate well with anatomical changes found in the peripheral vestibular system.[202,203] Declining gains and increased time delays have been seen in the optokinetic response (OKR), and decreased VOR gains with larger phase leads appeared at frequencies below 1.5 Hz. This would suggest that elderly subjects have an increased reliance on the visual tracking reflexes, and that increased visuomotor processing time could contribute to feedback delays and poor performance.[202,204] Thus, elderly individuals will have more difficulty detecting sensory signals indicating a loss of stability, and when they do, longer response latencies may well interfere with their ability to produce timely stabilizing reactions. Older individuals who showed decline in OKN and VOR function also showed poorer performance on clinical measures of balance and gait as compared with those with less decline of these functions.[203]

If conflicting sensory information is not suppressed, but instead can influence and modify the weighting of all other sensory inputs, then older individuals may be particularly disturbed in the presence of conflicting visual and somatosensory information. Postural responses of older subjects were poorly organized during combined support surface perturbations and mental distraction tasks,[205,206] and the predictive components of posture were absent.[207] Results from a number of studies[208-210] suggested that when two simultaneous tasks were required (e.g., postural stabilization and manipulation), postural adjustments were delayed in the elderly. In an elderly subject with disruption or deterioration of the mechanisms controlling, any distraction from postural control may be dangerous. In fact, when maintenance of an upright position and stabilizing gaze were both required, an increased incidence of falling appeared in the elderly.[211-213]

Falls among the elderly are a major public health concern because they are the leading cause of injury-related death and of nonfatal injury in the United States.[214,215] The dynamic process of maintaining an upright posture is compromised in the elderly as evidenced by this increased incidence of falling. Lengthened response latencies, increased static sway, and muscle weakness have been cited as contributing to falls in the elderly,[216,217] but none of these have been identified as causal factors. Attentional demands and disorganized postural strategies are more global parameters that have been targeted as potentially generative causes of falls.[114,208,218-220] Increased instability on a moving platform[221,222] and during locomotion[223-225] has also been attributed to an increasingly rigid trunk with aging. It has been observed that older adults stiffen their trunk to decrease the controlled degrees of freedom in an effort to make themselves more stable,[219,226] yet they still produce reduced head stabilization in space.[227,228]

A longitudinal study of elderly fallers[229] has found significant changes in the mean frequency of postural sway in the medial-lateral direction. A low-frequency component was identified in this plane suggesting a slow postural drift during quiet standing.[230] On a posture platform, the stabilizing muscle synergies, found to appear in a temporally consistent fashion in young healthy subjects, exhibit a disorganized order of onset in the elderly.[231] Latencies of EMG responses and of reaction times are increased in the elderly population[218,231,232] as is quiet sway.[233,234] Although both sway induced by platform perturbation and quiet sway measures demonstrated significant aging-related decreases in stability,[235] the differences between young and elderly were more pronounced for the perturbed sway data. Some of the quiet sway measures, however, were more successful in distinguishing elderly fallers from nonfallers.[235] A study of elderly individuals on a rotating posture platform[114] has explored whether delayed latencies of lower limb muscle responses are responsible for the failure to produce torque outputs necessary to compensate for unexpected falling. Results indicate that a disordered temporal relationship between tibialis anterior and soleus muscles, which are concurrently activated in younger individuals,[45] resulted in decreased stabilizing ankle torques. Weakness of the tibialis anterior muscle has been described in the elderly,[217] and could be a major contributor to this diminished torque response.

Increased stiffening between the head and trunk has also been reported in elderly subjects when attempting to keep the head stationary in space when the trunk was moving[226,227,236] and during whole body postural control.[42] Elderly subjects exhibited larger joint torques and greater trunk motion than young adults when attempting to stabilize on a translating platform and when standing on a narrow beam,[42,227,237,238] which could be a result of a stiffer mechanical system. If the elderly locked their head to their trunk to decrease the controlled degrees of freedom, these greater torques would be transmitted to the head. These data imply that elderly subjects rely on active trunk mechanics to coordinate head and trunk motion but the stiffer

trunk results in less damping of the ascending forces and, therefore, poorer stabilization. Thus, impaired balance in the elderly may be produced by altered response synergies that are generated by delayed vestibulospinal and proprioispinal reflex responses and a postural strategy to increase stiffness of the aging musculoskeletal system.

Summary

We can draw the following conclusions about mechanisms that contribute to postural stability from the existing data. First, central neural processes influence stability in the form of automatic, long-latency reactions, voluntary movements, and changes in both the passive mechanical properties (e.g., stiffness) and active force outputs (e.g., joint torques) of the system. Second, the presence or absence of specific sensory inputs (e.g., vestibular or proprioceptive) alters the magnitude or temporal onset of the muscle response pattern, whereas distortion of sensory inputs seems to rearrange the directional organization of the muscle response patterns. Third, learning, attention, and predictive processes influence the performance of postural reactions, as does the motor activity in which the individual is currently engaged when the postural behavior is required. Finally, a particular compensatory strategy adopted by a patient may interfere with, rather than assist, postural stability.

Clinicians and researchers need to identify the preplanned and automatic components of a postural response to determine how best to influence the postural response organization. Clinicians will need to have knowledge of the strengths and limitations in tests of postural control and strategies to challenge the postural system (changing visual, somatosensory cues or altering the complexity of sensory information) with compensation to identify functional-related balance problems. Recognizing the multiple factors that contribute to the outcome of a postural response should assist clinicians in determining the approach and effectiveness of their intervention strategies for retraining and restoration of postural function.

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CHAPTER 7

Disability in Vestibular Disorders

Natalia A. Ricci, PhD, PT ■ *Helen S. Cohen, EdD, OTR, FAOTA*

Vestibular disorders are characterized by vertigo, disequilibrium, oscillopsia, and autonomic signs. These problems can contribute to reduced independence in activities of daily living and health-related quality of life (HRQoL).[1-3] Most vestibular disorders are relatively benign, but they can still cause significant problems in the ability to function.[1] Vestibular disorders are also burdensome to society as a drain on the economy ,[4] because of lost work hours, decreased participation in society outside the home, and increased health-care costs. Diagnostic testing, alone, does not provide information about the need for intervention. The patient's overall health, functional status, and personal goals are important in determining the need for care and the type of intervention.[5]

"Dizziness" and vertigo are among the most common complaints in medical practice, affecting approximately 20% to 30% of people in the general population. [6] These symptoms are relatively more common in the older population. From the National Health Interview Survey, USA, among 37 million elderly persons, 7 million persons (19.6%) reported a problem with dizziness or balance in the preceding 12 months. [7] On a German survey (n = 1003), dizziness/vertigo had a prevalence of 22.9% within the last 12 months and an incidence (first episode) of 3.1%. For vestibular vertigo, the prevalence was 4.8% and the incidence was 1.4%.[8]

Despite the statistics cited above, the significant impact of vestibular disorders on functional performance is still not widely recognized. For example, the International Society of Physical and Rehabilitation Medicine sponsored a workshop to identify the health conditions that are most associated with disability and are amenable to rehabilitation interventions.[9] Participants, who were physicians and other health-care providers who specialize in rehabilitation, were generally unaware of vestibular disorders. In total, participants identified 103 rehabilitation-relevant health conditions, some of which were related to vestibular diseases, such as hearing loss and falls. Participants failed to identify disorders of vestibular function or other diseases of the inner ear as significant and treatable problems. This chapter will elucidate the concept of disability, the interface between disability and vestibular disorders, and the assessment tools used to evaluate functional status.

Concepts of Disability

Confusion in disability terminology impairs communication among investigators, clinicians, and patients. [1] The National Institutes of Health (NIH) used the term "functional limitations" to refer to limitations in functioning in the immediate environment for basic self-care skills and other essential activities of daily living. [10] The NIH used the term "disability" to describe how the individual interacts with the social environment, and used the term "societal limitations" to describe how barriers defined by laws or social policy restrict functional performance.

One of the first widely used models, the Nagi disability framework, used basic concepts of active pathology ,

impairment, functional limitation, and disability.[11,12] In this framework, disability was defined as "limitation in performance of socially defined roles and tasks within a sociocultural and physical environment,"[11] similar to the NIH definition of disability.

The International Classification of Impairments, Disabilities, and Handicaps (ICIDH)[13] was an attempt to develop a standard terminology for the consequences of disease.[5] The ICIDH classified three central concepts related to disease and health conditions: impairment, disability, and handicap. In this model, disability was defined as "any restriction or lack of ability to perform an activity in the manner or within the range considered normal for a human being."[13] In other words, it defined disability with reference to individual functioning. Handicap was defined as "a disadvantage that limits or prevents the fulfillment of a role that is normal (social and cultural factors) for that individual." The ICIDH used impairment, defined as abnormalities of body structure or system function, as the cause of disability and handicap.

In the Disablement Process model,[14] disability was defined as a process of the impacts of chronic and acute conditions on individual function and in society. That model combined the concepts of disability and handicap, possibly leading to confusion.

The medical model of disablement is based on the theory that disability is a consequence of body structure or functional decline, a personal problem. By contrast, the social model of disablement is based on the theory that disability is a result of the lack of opportunities in a person's physical and social environment; thus, the problem is caused by society.[5,15,16] Both points of view are valid. In real life, health conditions and social/environmental factors affect many of the same people. The process of disablement is probably caused by the complex relationship between biological and social factors.[16]

The biopsychosocial model of disability is currently the dominant perspective.[12,17] Therefore, the ICIDH was revised and replaced with the International Classification of Functioning, Disability and Health (ICF).[18,19] The ICF complements the International Classification of Diseases, Injuries, and Causes of Death (ICD-10), to give a more complete understanding of individual people and population health status.[5,16,20] The classification covers any disturbance in terms of functional states following the domains: body functions, body structures, activities, and participation.[5,15,18] The loss or abnormality of a body part or function is impairment. Difficulties in the ability to execute activities are limitations, and problems in involvement in life situations are participation restrictions. These functional states can be influenced positively or negatively by contextual factors, including environmental or personal. In this taxonomy, disability combines impairments, limitations, and restrictions as the negative effect between a health condition and contextual factors (environmental and personal factors).[17,19] The ICF has been adopted by the World Report on Disability as the conceptual framework for understanding functioning and disability.[19,21]

Using the ICF concepts, reduced vestibular function, as indicated by tests described elsewhere in this book, is impairment. For example, a vestibular impairment caused by damage to the vestibular nerve might cause a reduced vestibulo-ocular reflex gain. Many people with vestibular disorders have limitations in performing activities of daily living.[8] Natural or man-made environmental factors can trigger vestibular symptoms, such as poor lighting, heights, open space, sound pollution, or rough terrain.[4,22] Those environmental interactions can cause anxiety, depression, and social isolation because of restrictions in participation outside the home.[8]

Assessments of Disability

Measurements of health status (i.e., function/disability and quality of life) are crucial components of patient care, the use of which help the clinician to design rehabilitation programs, contribute to policy goals, and monitor the effectiveness of health care.[23] Many disability assessments have been developed, some more useful than others. Most of them are self-assessments, in which the patient rates him- or herself. Detailed questions and detailed rating scales may help the patient to describe performance more accurately than open-ended questions. Because disability is subjective, however, outcomes from self-rating scales have limitations that may reflect the patient's beliefs more than the patient's actual status.[23] Therefore, self-rating may not correlate with the impressions of the patient's significant others, that is, the people closest to the patient. Illiteracy and use of questions that do not apply to that patient's individual situation can confound assessment and result in unreliable answers.[24]

Some strategies can be used to address some weaknesses of the self-assessments. Patients can under- or overestimate their performance skills, so some performance tests or direct observation can be useful.[24] For example, to evaluate difficulty walking, the clinician may ask the patient about it, use some objective tests in which the patient may perform as well as possible to impress the clinician, or just observe the patient walking into or out of the office when the patient is unlikely to be aware of being observed. The patient's significant others may be intimately involved with the diagnostic and rehabilitation process.[25] Therefore, evaluating their perceptions of the patient's ability to function is important. Families and

friends may over- or underestimate patients' abilities. Sometimes those perceptions are not health related but reflect some long-established patterns in some people's interpersonal relationships.[26] Patients may perceive themselves as more independent than their significant others may perceive them, especially for personal care skills, as suggested by scores on the Vestibular Disorders Activities of Daily Living scale (VADL).[26] Similarly, on the Vertigo Symptom Scale (VSS), spouses tended to overestimate vertigo severity compared with patients.[25] So, the challenge is not to choose one perspective over another, but to combine them.[27] These additional kinds of assessments can increase costs and time during evaluations. Some behaviors may not be observed because they are unique to the patient's living environment or are too personal, or because of the subjective nature of perception of quality of life.

The points of view of different professionals affect their assessments of disability. For example, physicians are most concerned with symptoms and signs; physical therapists and occupational therapists are more concerned with the way people perform daily life activities; counselors and psychologists may be more concerned with the emotional and social impacts of the health condition. All of them are concerned about HRQoL. Once the clinician knows what to measure, he or she should identify relevant measurement instruments.[28] Fortunately, the available self-perceived measures are low cost, easy to use, and require minimal equipment. Some scales are more time consuming than others, and some scales place difference emphases on self-care activities or psychosocial interactions. Therefore, the clinician should select the level of detail desired for screening, the specificity of the scale for different constructs, and the comprehensiveness of the scale. For non–English-speaking patients, the availability of the scale in the patient's native language may also affect the choice of a scale.

Some other factors also affect the selection of a scale. The interests and needs of the individual patient, or target population, the difficulty understanding the items and questions, the severity of the problem, and the time needed to use the measure are factors to consider. The clinician should know if the scale is sensitive to change, so that treatment results can be quantified, and should know about the psychometric properties of the scale, such as the validity and reliability. A good scale has high reliability, sensitivity, specificity, and validity.

Furthermore, the clinician must choose between a generic scale and a disease-specific scale. Generic health status measures are broadly applicable across diverse diseases, disease severity, health status, health interventions, and demographic subgroups. They summarize concepts that apply to different health conditions and populations. Disease-specific assessments are designed for special patient populations with the goal of measuring clinical changes after implementing a plan of care.[29] We discuss the most widely used generic and specific assessments for patients with vestibular disorders.

Generic Disability Assessments

Three generic assessments have been used with patients who have vestibular disorders. The Short-Form-36 Health Survey (SF-36)[30] has been widely used to evaluate HRQoL. The Functional Independence Scale (FIM)[31] is a better choice for a more detailed functional activity level. The ICF[18] has been used for a comprehensive evaluation of disability.

The SF-36 was designed as a generic indicator of health status (functioning and well-being) in any patient group and in population surveys.[23] This 36-item scale has been used worldwide, with several translations, with good validity and reliability.[30] It is divided into two categories with four domains per category: physical functioning, role-physical, bodily pain, general health, vitality, social functioning, role-emotional, and mental health. The first four domains are in physical health; the other four domains are in mental health. The assessment uses a 5-point scale. For the final score, some norm-based scoring algorithms must be used. The final score ranges from 0 to 100, with 100 indicating the best HRQoL. One study showed that, after vestibular rehabilitation, the SF-36 was less responsive to change (Physical domain: 6.67% and Mental domain: 4.35%) than the Dizziness Handicap Inventory (DHI), a specific-measure (11.94%).[32] Another study revealed that patients with chronic dizziness had scores 38% to 87% of normative values for the 8 domains, with the lowest scores on role-physical and role-emotional domains.[33] It is an important assessment and reveals poor HRQoL for patients with vestibular disorders, but it has some weaknesses. It does not seem suitable to describe changes. Some items are not related to symptoms of vestibular disorders, such as the domain for bodily pain. The scale is cumbersome, and the scoring method can be time-consuming in daily practice compared with disease-specific measures.

The 18-item FIM is a well-normed scale, developed by the Uniform Data System for Medical Rehabilitation to measure physical and cognitive disability in terms of level of care required. It is applicable to patients of all ages and with all diagnoses, and it is especially useful to planning and quantifying rehabilitation outcomes.[31] It is not a comprehensive scale, because it covers only basic activities of daily living and not complex, instrumental activities such as shopping or driving. The assessment includes

physical and cognitive domains covering self-care, sphincter control, mobility, locomotion, communication, and cognition.[23] The FIM uses a 7-point scale. The total score is calculated by summing all the items. The total score can range from 18 to 126; higher scores indicate greater independence. The ratings indicate actual performance in daily life rather than the patient's potential for performance, and scores may be based on observation. Using the FIM can be time-consuming, however, and staff must be trained to administer it. Some items can be performed easily by patients with vestibular disorders, such as eating and sphincter control, which may be the reason why elderly patients with vestibular disorders presented high FIM total scores (118.5 points) in one study.[34]

The ICF is a comprehensive assessment with 1,434 categories.[4,35,36] The ICF uses a 5-point scale, and an item can also be scored 8 (not specified) or 9 (not applicable). Evaluating all ICF categories is impossible, so two alternative strategies have been developed. ICF categories can be linked to related items in disease-specific questionnaires, [3] which begs the question of whether the ICF is needed and if the diagnosis-specific questionnaires are more useful. Also, core sets have been developed that include only categories that are typical and essential to describe a specific health condition.[9,20,35] Several core sets have been developed for health conditions that cause common disabilities. A vestibular core set was published recently.[37]

Two studies examined patients with Ménière' s disease using the ICF.[38,39] Most of the effects listed by the patients came from impairments (70%), following by activity limitations (39%), participation restrictions (47%), environmental factors (16%), and personal contextual factors (28%). In the category of sensory functions, impairments in hearing, symptoms of vertigo, tinnitus, and imbalance were the most frequent. In the category of activity limitations, patients reported problems with communication, difficulty walking in the dark, and driving in specific circumstances (long roads, unfamiliar place, and dark). In the category of participation, patients were restricted for work and for performance of recreational and leisure activities. In the category of environmental contextual factors, patients reported changes in diet to manage their condition. Patients also described problems with uncertainty of life because of the unpredictable course of their Ménière's disorder.

Müeller et al[4] developed an international standard for the description of functioning and disability in patients with vertigo and dizziness based on the ICF. They found 471 concepts in the patients' interviews that were linked to 142 different ICF-categories (40 body functions categories, 62 activity and participation categories, and 40 environmental factors). Four percent of concepts could not be linked to specific ICF categories, most of them related to personal strategies of coping. Alghwiri et al[3] described and compared the ICF to the concepts covered by eight clinical self-report measures used in vestibular rehabilitation. The questionnaires provided 164 items with a total of 312 meaningful concepts. The concepts were linked to 51 different ICF categories: 19 categories related to "body functions," 30 categories to "activities and participation," and 2 categories related to "environmental factors." Forty-two concepts could not be linked to any of the ICF components, including "moving the head into different directions," "moving in or out of the bathtub or shower," and "avoiding certain activities, positions." These studies highlight the importance of the relationship between the individual's ability to function and the environment. Although the ICF has a huge range of categories, some important concepts for understanding disability in people with vestibular disorders are not included in the ICF classification system (e.g., transferring into and out of the bathtub or shower).[3] Some of those concepts are included in one disease-specific assessment, the VADL.[40,41] Those concepts should be added to a future revision of the ICF.

Generic measures have a place in rehabilitation for comparing outcomes in disparate groups of patients and across facilities and even nations. Unfortunately , these measures may not address problems of concern for particular health conditions. Also, they are not sensitive enough to vestibular disorders to capture the impact of vertigo and disequilibrium on the patient's everyday life.[42] Assessments that are specific to a particular health problem are more likely to be relevant in everyday practice and for therapeutic treatment planning.

Disability Assessments that Are Specific to Vestibular Disorders

A wide variety of disability-specific measures are available, but many of them are not comparable because they assess different aspects of disability.[20] Specific self-rated scales have been developed to evaluate patients with vestibular disorders such as the DHI,[43] Vertigo Handicap Questionnaire (VHQ),[44] VSS,[45] Activities-specific Balance Confidence scale (ABC scale), [46] Dizzy Factor Inventory (DFI),[47] UCLA- Dizziness Questionnaire (UCLA-DQ),[48] Vertigo, Dizziness, Imbalance Questionnaire (VDI),[49] VADL,[40,41] Vestibular Rehabilitation Benefit Questionnaire (VRBQ),[50,51] and Vestibular Activities and Participation (VAP).[52] The questionnaires have different scopes; therefore, they include several different constructs or specific items.[3,4,51]

The 25-item DHI is the oldest and most widely used self-assessment for patients with vestibular disorders. The

DHI evaluates the physical factors and the functional and emotional consequences of vestibular disease.[43] The questionnaire is divided into physical (seven items), emotional (nine items), and functional (nine items) subscales. It uses a 3-point scale: "yes" (4 points), "sometimes" (2 points) and "no" (0 points). The total score is obtained by summing the item scores; the maximum possible score is 100. Higher scores indicate more severe handicap. It has high internal consistency ($\alpha = 0.89$) and reliability ($r = 0.97$).[43] It has been translated into several languages including some cultural variations (Argentinian Spanish,[53] Italian,[54] French,[55] Arabian,[56] Japanese,[57] Hebrew,[58] Brazilian Portuguese,[59] Chinese,[60] Spanish,[61] Swedish,[62] and German[63]).The three response choices—yes, sometimes, or no—make the DHI quick and easy to use. Interpretation may seem easy but the meaning of items rated "sometimes" may be unclear. Also, it does not provide a broad view of the patient, and it is unable to detect small changes. Despite the DHI's responsiveness after vestibular rehabilitation, the scale is not useful for treatment planning, because it does not indicate which tasks or skills should be addressed during rehabilitation.[23] In particular, although the DHI items are relevant to symptom context,[42] the omission of important self-care skills is a significant weakness.

The 22-item VHQ was created to describe common social, psychological, and behavioral context experienced by patients with vestibular impairment. It is divided into 4 domains: restriction of activities, social anxieties, fear of vertigo, and severity of vertigo attacks. It uses a 5-point scale, 0 (no handicap) to 4 (maximum handicap).The final score is calculated based on the "percentage handicap," in which the item average score is multiplied by 25 giving a maximum score of 100%. The VHQ has high internal consistency for the total score ($\alpha = 0.93$) and test-retest reliability showed no significant change.[44] The questionnaire has been translated into German.[64] This scale provides a good understanding of the patient's anxieties and behavioral restrictions that lead to handicap, so it can be helpful for psychologists and other professionals who work with coping strategies. It is not so useful for physical therapists and occupational therapists, who are concerned with performance of daily life activities.

The 27-item VSS evaluates symptoms related to vestibular disorders. It is divided into four subscales: somatization, autonomic symptom, acute attack of vertigo, and vertigo of short duration.[45] It is scored on a 6-point scale from 0 (never) to 5 (very often, more than once a week). The subscale total score is calculated by summing item scores and dividing them by the number of items; higher scores indicate more frequent symptoms. It has good internal consistency and reliability, but less is known about its responsiveness. The scale has been translated into four languages (German,[65] Afrikaans,[66] Turkish,[67] and Spanish[68]). This questionnaire is about symptoms; it is not intended to measure other constructs. The VSS may be useful when assessing patients who have difficulty describing their symptoms.

The 16-item ABC scale is frequently used to evaluate patients with vestibular disorders, but it is the only scale that was not developed for this purpose. It was developed to assess self-perceived balance confidence in performing daily activities in community-dwelling seniors.[46] It is scored on an 11-point scale, from 0 ("no confidence") to 100% ("complete confidence") indicating how confident the patient perceives him- or herself to be when performing activities that involve balance. The total score is the mean of all items. When used with patients with vestibular disorders, the ABC scale has a negative moderate correlation with the DHI ($r = -0.635$), high internal consistency ($\alpha = 0.95$), and good reliability for individual items (ICC = 0.67 to 0.92).[71] It is widely used and has been translated into several languages (German,[69] Canadian French,[70] Turkish,[71] Chinese,[72] and Swedish[73]). It has been used for a variety of health conditions including stroke, Parkinson's disease, and lower limb amputation as well as vestibular disorders and disequilibrium of aging. The ABC scale includes some important activities such as walking up and down stairs, but it does not address skills that may be affected by vertigo such as rolling over in bed. Thus, the ABC scale, alone, may be insufficient for many patients. It is, however, an easy tool to use and may give some patients some insight into their balance problems.[24]

The 44-item DFI was developed for use as a preliminary screening or as part of a comprehensive assessment battery.[47] The inventory was based on the Multidimensional Pain Inventory and the DHI but with new items added based on the author's clinical experience. It uses a 5-point scale to assess 3 domains (symptom factors, responses of significant other, and activity level). Further testing for reproducibility, validity (discriminant and convergent), and responsiveness analysis should be performed.[42] The domain of "responses of significant other" is a strength of the scale. The omission of self-care tasks and the length of time to complete the scale are weaknesses.

The 5-item UCLA-DQ uses a 5-point qualitative scale for patients to characterize their vertigo based on frequency, intensity, impact on daily activities, impact on quality of life, and fear of dizziness. The original paper that described this questionnaire did not provide data about its psychometric properties.[42] Therefore, the clinician should use caution when interpreting the results and comparing results from different samples and patients in

clinical practice. It has been translated into Spanish[61] and Swedish.[74] The items about daily activities and quality of life are generic, putting unrelated concepts together such as "driving" and "taking care of yourself." The questionnaire has limited utility, because it does not address any items in detail,[24] but it may be used as a quick screening before a comprehensive assessment.

The 36-item VDI uses a 6-point scale to assess 2 domains including 14 questions about symptoms and 22 questions about HRQoL. Each domain is given a total score based on the sum of the items. [42] The scales have good internal consistency (VDI symptoms: 0.86; VDI HRQoL: 0.92) and reliability (VDI symptoms: 0.81;VDI HRQoL: 0.87),[49] but responsiveness was modest. The VDI has low to moderate correlation with the SF-36 physical and mental components. No English version is available because it was developed in Spain but aTurkish version[75] is available.

The 28-item VADL evaluates the self-perceived impact of vestibular impairment on daily life activities. It is divided into three domains: functional (12 items), ambulation (9 items), and instrumental (7 items). It is scored on a 10-point scale from 1 (Independent) to 10 (Too difficult and no longer performs that activity) with an additional rating of "Not Applicable." Subscores and the total score can be based on either the mean or the median of the item scores. The VADL has high internal consistency for the total score and for the subscales, and good test-retest reliability over 2 hours.[40] It has been translated into Brazilian-Portuguese,[76] Italian,[77] and Korean.[78] A Spanish version is available from the author . The VADL focuses on daily activities and does not address symptoms or emotional components.[24] Strengths of the VADL are the detailed list of self-care skills in the functional domain and the use of a detailed scale that provides a specific rating of the patient's level of performance. The 10-point scale may be difficult for some patients to select a level of function.[24] Clinical experience suggests that the detailed scale provides the basis for a useful structured interview during the clinical visit.

The 22-item VRBQ assesses the subjective sensation of vertigo and its consequences in 2 domains, symptoms (11 items) and quality of life (1 1 items), using a 7-point scale. To obtain the final subscores and the total score, some special calculations must be made.The score calculation is an attempt to quantify the difference between the patient's current state and a state that is normal for the individual, but is not easy to apply in daily practice. The questionnaire has good internal consistency for the total score (0.73), strong reliability (ICC = 0.92), and a moderate responsiveness.[50,51] Although the attempt to improve the value of the subscores and total scores is admirable, the VRBQ does not provide new insights into the problem of disablement, because all the items were derived from preexisting scales such as the DHI, VSS, VHQ, DFI, UCLA-DQ, and VADL. The omission of self-care activities is a significant weakness.

The 34-item VAP is scored on a 5-point scale. It examines the activities and participation of people with vestibular disorders. Its relationship to the ICF is a strength. The total score is obtained by calculating the mean of the item scale values after excluding the "NotApplicable" items. The VAP total score had excellent test-retest reliability (ICC = 0.95) and poor to excellent agreement per item (κ 0.41–0.80).[52] The scale provides a variety of activities that involves walking and instrumental tasks, but the omission of important personal self-care tasks is a significant weakness.

Many of the scales evaluate frequency and intensity of symptoms, movements that trigger symptoms, and emotional problems, such as the DHI, VRBQ, and DFI. To assess performance of daily life activities, the VADL and VAP are the best options. A rapid screening can be performed using the UCLA-DQ, but a more comprehensive evaluation of personal factors can be assessing by the DFI. No scales assess the impact of the physical environment on vestibular disorders. The ideal assessment would include domains for symptoms, self-care activities, mobility skills, instrumental activities, emotional impact of symptoms, and the impact of environmental factors.

Disability and Vestibular Disorders

Many studies have examined disability in patients with vestibular disorders. We review those studies in this section. Note that some studies used the normed instruments described above, but others did not.

In a study with community-dwelling elderly , the prevalence of subjective complaints of dizziness was 45%; the movements that elicited this symptom were "standing from a lying position" (50%), followed by "turning the head" (48%) and "standing from a sitting position" (38%).[79] All these movements are performed several times a day and are components of various activities of daily living. People with vestibular disorders often reduce their levels of activity and avoid such movements as they attempt to avoid trig - gering the symptoms. That habit, however, may cause secondary problems such as stiff neck, headache, and muscle weakness,[22,80] which then decreases their functional status, in a downward spiral.

Aratani et al[81] reported that 42% of elderly people with vestibular disorders complained of having dif ficulty performing 7 to 15 activities. The most difficult task was "walking close to home," following by "having

a bath/shower." Activity in outdoor environments, such as walking close to home, required greater attention, head and gaze fixation, head and body orientation, and body stability. Those functions can be impaired by vestibular disorders. Having a shower can be difficult for people with vertigo and balance disorders for several reasons. Balance is challenged because the floor is slippery and because individuals shift their weight to bend over to wash their hair or lower extremities. Vision is challenged when people bathe without their eyeglasses, when they have water or shampoo in their eyes, and when the bath or shower stall does not have an overhead light. Also, vestibular disorders cause decreased gain of the vestibulo-ocular reflex, resulting in blurred vision.

People with vestibular disorders sometimes have difficulty performing more complex tasks such as shopping and community-level skills, driving, some work-related tasks, and participating in some leisure activities[22,80,82,83]. Cohen et al[82] compared healthy subjects and subjects with vestibular disorders using the Driving Habits Questionnaire with some additional questions. Patients reported reduced driving skills compared with healthy subjects, particularly in challenging driving situations (alone, in the rain, at night, during rush hour, on the highway, changing lanes, in ramped garages, and pulling into and out of parking spaces), but driving habits and crashes did not differ. Visual information is very important to spatial orientation for people with vestibular impairments. They may become more disoriented than healthy people in an environment with reduced visual stimuli or with visual conflict (such as cars passing). Also, when driving, these patients tend to avoid making head movements, or they move their heads slowly, to avoid eliciting vertigo.

Gutiérrez-Márquez et al.[83] reported that of 139 patients with vestibular disorders who had paid jobs, 51% had problems performing their jobs. The median number of days off work was 7 for patients with unilateral weakness, 5 for patients with bilateral vestibular weakness, and 12 for patients with central problems. Yardley et al[80] found that 12.3% of patients who work took days of f work because of vertigo, and 25% had difficulty carrying out a job satisfactorily. For people with Ménière's disease, the unpredictability of the disorder and the hearing loss cause many patients to leave their jobs and either stop working or find different kinds of work.[84]

Most people with vestibular disorders avoid leisure activities or reduce their participation, especially activities related to exercises and body movements. [22] Even a simple, sedentary hobby such as crocheting may be avoided by patients, because the position of the head can trigger symptoms. Thus, even basic daily tasks, such as taking a shower, and more vigorous activities, such as physical exercises, cause anxiety and avoidance behavior. Being inactive and limiting movements and interaction with the environment may limit vestibular compensation and may lead to a cycle of escalating anxiety and disability[80] The anticipation of problems is especially anxiety-provoking for patients with uncontrollable and unpredictable attacks of vertigo, leading them to restrict their participation even more.[4,22,43]

The influence of contextual factors has not yet been well explored in people with vestibular disorders.[4] Some environmental and seasonal factors, such as temperature variations and air pollution, have been shown to be correlated with vertigo events in benign paroxysmal positional vertigo (BPPV).[85,86] The theory, which sounds far-fetched, is that environmental factors may interfere with metabolism of endolymph, causing free-floating particles. That theory is not supported by strong evidence.

Stronger evidence for the role of contextual factors comes from work by Ganança et al, [87] who found that 53.1% of elderly people with vestibular disorders had fallen inside their homes. The bathroom was the most common place to fall (38.1%). The most common tasks in which they were engaged before the fall were walking (53.1%), ascending/descending stairs (10.9%), changing position (9.4%), and taking a shower (6.3%). Those tasks could be performed safely and without falls with some minor changes in the environment, such as using a cane while walking and installing grab bars near the toilet and in the shower stall. Clinicians should consider the important role of modifying contextual factors during treatment planning.

The social environment in which people live is also an important aspect of the environmental context. [18] For people with vestibular disorders, the limits to their independence in self-care activities and mobility skills, fear of falling, avoidance of some tasks, and decreased participation in activities outside the home leads to restricted social networks and consequent decreased sense of well-being and quality of life. [22] Wiltink et al [88] found that 28.3% of people with vertigo reported symptoms of at least one anxiety disorder (generalized anxiety , social phobia, or panic) compared with only 5.1% of the control group. Having supportive family and friends might be a related contextual factor because those people might assist the person with vertigo.[4] Conversely, the lack of supportive significant others, or the presence of people who are unsupportive because they do not understand the impact of vestibular disorders on function performance, can be a barrier to participation and developing a sense of self-efficacy and well-being.

Health professionals may have different perceptions of patients' functional limitations, and these dif fering

perceptions of disablement could affect treatment planning. For example, when physicians were asked about the impact of dizziness on their patients' quality of life, physicians perceived less impact than patients. [48] A patient may continue to perceive him- or herself as disabled because of vertigo, despite the professional' s objective observation to the contrary. To ameliorate a sense of decreased self-efficacy, that patient will continue to seek medical care, if not with the original professional then with others.[41] This type of behavior has the potential to increase health-care costs significantly.

Some vestibular disorders are associated with specific functional limitations and patterns of disability. For example, most people with BPPV (84%) and Ménière's disease (85%) complain significantly more about vertigo than people with unilateral peripheral vestibular weakness (39%). Unsteadiness, however, was significantly more common in patients with unilateral peripheral weakness (83%), than in patients with Ménière's disease (53%) and BPPV (22%).[89]

In BPPV, the episodes are often provoked by everyday activities on which patients perform pitch rotations of the head, such as looking upward, bending forward, and rolling over toward the affected side.[90,91] Those movements are often performed during routine activities such as trying to reach for an object, making the bed, putting on socks or shoes, washing the hair at home or at the beauty salon, being reclined in a dentist's chair, changing a light bulb, or shaving.[91]

Ménière's disease is a special case, because many patients have two different states of functioning, either normal or incapacitated. In a survey of functional limitations in Ménière's disease, patients who considered their attacks to be mild (17%) were able to continue working or participating in an activity during an attack, but patients who considered the attacks to be moderate (56%) were forced to stop their activity and lie down.The remaining 27% had trouble with the attacks even while lying down. [92] Cohen et al[84] found that during Ménière's attacks patients have difficulty performing activities that require good hearing, such as using the telephone and social communication, but also activities that involve balance control. Vertigo and balance problems mainly influence the physical dimension of functioning, whereas hearing loss influences the psychosocial dimension.[93] To modify Helen Keller's famous dictum, vertigo and balance disorders cut you off from doing things, but hearing loss cuts you off from people.

Patients with bilateral vestibular hypofunction have difficulty maintaining balance and are at risk for falls when the support surface is unstable, when postural control is challenged, and when ambient light is limited.[94] In a study comparing patients with bilateral and unilateral peripheral vestibular weakness, balance impairments and the Timed Up and Go test together explained 78% of the variance in DHI scores of the patients with bilateral vestibular hypofunction, but they explained only 13% of the variance in DHI scores of unilateral weakness patients. Thus, balance impairments are probably more closely related to functional limitations in patients with bilateral vestibular weakness or loss. [95] Their well-known symptoms of oscillopsia and reduced dynamic visual acuity may lead to particular difficulty driving, walking in crowds, and participating in sports. With practice, many of those deficits may be reduced or ameliorated.

Central vertigo is generally associated with severe disequilibrium, oscillopsia, vertigo, and additional neurological signs.[6] In a study that compared patients with peripheral and central vestibular disorders on the VAP and the DHI, patients with central involvement had significantly worse scores than did patients with only peripheral involvement.[52]

Summary

We have pointed out some differences by diagnosis, but having a vestibular disorder, in general, is more significant for functional limitations than the etiology of the specific disorder.[41] Patients, in general, perform tasks more slowly than normal. They limit their activities within and outside the home: they restrict driving to local roads and avoid highways, and they may need time off from work or be less efficient or effective at work. Many patients also limit their social activities as a result of visual vertigo elicited indoors in large spaces such as some houses of worship, some community centers or auditoriums, and in crowds or unstructured, outdoor spaces because of visual vertigo and disequilibrium.

Using a wide range of strategies discussed elsewhere in this book, the functional impairments and limitations in activities and participation that lead to disability in vestibular disorders can be prevented or ameliorated. The contextual factors, especially the physical environment, can sometimes be altered or adapted to improve function. In vestibular rehabilitation, the clinician's goal for every patient is always to reduce symptoms, increase safety and independence, and improve the quality of life.The use of valid and reliable standardized assessments can help to achieve those goals.

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CHAPTER 8

Vestibular Compensation—Recovery after Unilateral Vestibular Loss

Ian S. Curthoys, PhD ■ *G. Michael Halmagyi, MD*

Sudden, complete unilateral loss of vestibular function in normal, healthy individuals results in a dramatic series of symptoms—strong sensations of turning (vertigo), nausea, rapid eye movements (nystagmus), oscillopsia (the illusion that the visual world is moving as they move their head), falling to the affected side, gait ataxia, postural instability, distortions in the perception of body orientation and movement, and inadequate compensatory responses to head movement. We will use the term unilateral vestibular deafferentation (UVD) syndrome to refer to these symptoms. Such patients are greatly distressed. Over the first few days most of the symptoms decline and the patient's distress correspondingly diminishes. For most patients, these symptoms have disappeared within a few weeks, and the patients return to their normal lifestyle and are happy with their recovery. The term used to describe that general recovery is vestibular compensation, and superficially it seems that there is a full recovery and vestibular function has returned. Indeed, in a few patients this is exactly what does happen: some patients with vestibular neuritis experience all these symptoms, but as their vestibular neuritis diminishes, their peripheral vestibular function, as shown by new specific, objective tests, is completely restored. However, in most patients there is little or no restoration of vestibular function, and when their peripheral vestibular function is tested by these new tests of canal and otolith function, the permanent loss is clearly shown. Nevertheless, most patients with permanent UVD recover and are happy—they are "well-compensated." They do not experience vertigo, nausea, oscillopsia, or postural unsteadiness (see Box 8-1).

That the objective tests show the permanent vestibular loss implies that well-compensated patients must have developed some means of overcoming the loss of this major sensory system. The big question is how? What are these patients doing to help themselves so much? This puzzle is sharpened by the fact that some patients, after apparently identical vestibular loss, continue to complain about their UVD syndrome for years after the event, and are so unhappy they repeatedly visit their clinician to attempt to overcome their poor compensation. It is our contention that the well-compensated patients have learned behaviors to minimize the effects of their loss—for example, to generate saccades or blinks during head movement—whereas the poorly compenated patients have not learned these strategies.

The vestibular system has a major role in the control of many responses and so the symptoms of the UVD syndrome are complex and varied, and many factors can affect recovery. Simply listing the symptoms and their change over time does not help in understanding the UVD syndrome and its recovery. Vestibular compensation is not a

Box 8-1

ABBREVIATION GLOSSARY

ABR — auditory brainstem response

BVD — bilateral vestibular deafferentation

contralesional — on the opposite side to the affected ear (i.e., the healthy side)

COR — cervical ocular reflex

cVEMP p13 — the initial positivity at 13 msec of the cervical vestibular evoked myogenic potential

GABA — gamma-amino-butyric acid

ipsilesional — on the same side as the affected ear

LVN — lateral vestibular nucleus

MVN — medial vestibular nucleus

OCR — ocular counterrolling

OTR — ocular tilt reaction

oVEMP n10 — the initial negativity at 10 msec of the ocular vestibular evoked myogenic potential

pitch — rotation around the interaural axis of the head

roll — rotation around the naso-occipital axis of the head

SVH — subjective visual horizontal

SVV — subjective visual vertical

UVD — unilateral vestibular deafferentation

VEMP — vestibular evoked myogenic potential

VOR — vestibulo-ocular reflex

yaw — rotation around the vertical axis of the head

"unitary" inexorable recovery of all the functions affected, as the term "vestibular compensation" implies. Different vestibular-controlled responses recover at different rates, and some of these responses do not recover at all.

Why does UVD cause these symptoms? Why aren't these symptoms permanent? How can there be any recovery? The answers lie in the neural basis of the UVD syndrome, the changes in the pattern of neural activity over time, mainly in the vestibular nuclei of the brainstem, but also in the changed behaviors that patients develop to overcome vestibular loss. Loss of one labyrinth causes changes in neural activity in the two vestibular nuclei on either side of the midline of the brainstem. During vestibular compensation the altered pattern of neural activity returns to something approaching normal, and as it does so, some of the symptoms of the UVD decrease.

Three facts must be taken into account:

1. Neurons in the first vestibular relay nucleus in the brain, the medial vestibular nucleus in the brainstem, receive input from the peripheral vestibular receptors in the inner ear and relay output to many destinations. Most importantly, the outputs are relayed to the neural structure controlling eye movements and posture, and also to thalamic structures probably responsible for the sensation of body position and movement.
2. Neurons in the paired vestibular nuclei on each side of the brainstem communicate across the midline with each other. This so-called commissural interaction plays an important role in normal vestibular function and in vestibular compensation.
3. Most importantly, the activity of neurons in the vestibular nuclei is not determined solely by input from the peripheral vestibular system. These neurons also receive input from many other neural structures, and any of those inputs can modulate the activity of vestibular nucleus neurons and so influence the behavioral responses (and perception) during UVD and its recovery. For example, visual input, spinal input, reticular input, and cerebellar input all project to neurons in the vestibular nucleus and can modify the activity of vestibular neurons.

The multi-input character of neurons in the vestibular nuclei is important for understanding why so many seemingly unrelated influences can affect the UVD syndrome and its recovery (Fig. 8.1). The transmission of vestibular activity can be affected by the cerebellum and the reticular formation. For example, axons from cerebellar neurons project directly onto neurons in the vestibular nucleus, allowing a very direct, fast, and powerful control of transmission from the periphery to the neurons controlling vestibular responses. That cerebellar modulation can range from silencing the transmission of vestibular information to enhancing the gain of the transmission, and the evidence is that cerebellar input can have profound effects on vestibular compensation.

In this chapter we discuss

- the causes of vestibular loss
- the symptoms following UVD
- the recovery after UVD
- the relevant neural mechanisms

We review the behavioral, clinical, and experimental evidence concerning neural transmission of vestibular information, how it is modified by unilateral loss, and what

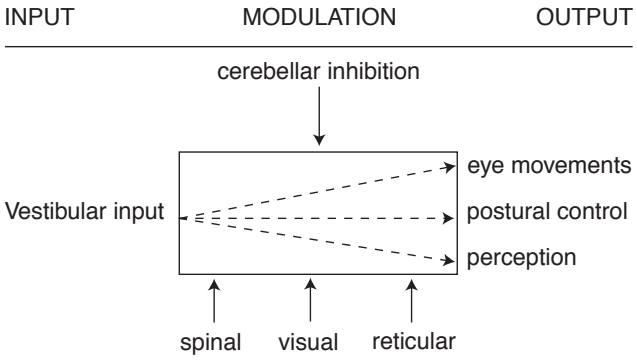

Figure 8.1 A simple general schematic illustration to represent the responses that can be controlled by vestibular input and how transmission of the vestibular input to govern those responses can be modulated by various sources: visual, spinal, reticular, and cerebellar. This scheme should not be taken too literally—for example, there is evidence that visual input differentially affects eye movements and posture.

happens over time as recovery takes place. For other reviews of vestibular compensation, see references.[1-21]

Many species exhibit responses after UVD that recover quickly over time in a way that parallels human recovery, although the exact speed of recovery varies considerably from one species to another. For example, monkeys, cats, rats, guinea pigs, and humans show ocular nystagmus after UVD. However, in some of these species (rats and guinea pigs), this spontaneous nystagmus disappears within about one day, whereas in other species (such as humans) the disappearance takes longer. Given this similarity across species, the reasonable assumption is made that similar neural processes underlie the recovery in these different species, and that presumption is the basis for using results from experimental studies of vestibular compensation on animals to understand human vestibular compensation.

One conundrum is that although most patients recover and lose most of their acute symptoms relatively quickly, in some patients the recovery is incomplete; they continue to experience symptoms and are distressed, even years after the UVD. The term used to describe such patients is "poorly compensated." In other individuals who have recovered and appear to be fully functional ("well-compensated" patients), there can be a near total relapse—called decompensation—under particular conditions such as severe stress.

Causes of Vestibular Loss

Patients with UVD are a very heterogeneous group, and that heterogeneity is a major reason for the hugely variable results of vestibular compensation. This group of patients is heterogeneous because of differences in the cause, the extent of the loss, and the preexisting level of vestibular function. Animal studies show the very different courses of compensation according to the different causes of the UVD.[22]

There are many possible causes of unilateral loss of peripheral vestibular function. Among them are disease, trauma, surgery (e.g., for removal of vestibular schwannoma), and therapeutic intratympanic injection of gentamicin for the treatment of peripheral vestibular disorders. The reason for carrying out the UVD can be a determinant of the severity of the symptoms and the rate of recovery. In the very unusual case in which a normal healthy person suddenly loses all peripheral vestibular input from one ear, the person experiences the very dramatic symptoms described above. At the other extreme, someone who has had a slowly growing vestibular schwannoma on one vestibular nerve—and then had that schwannoma and the entire vestibular nerve removed at surgery—may exhibit almost no symptoms after the UVD. Most probably, this is because there has been a progressive vestibular loss over some time, and the patient has been progressively compensating as the schwannoma grew. So, by the time the UVD surgery takes place, the patient has no effective vestibular function in the affected ear. Therefore, there is no UVD syndrome and thus no vestibular compensation takes place after surgery. Without knowing the preoperative level of vestibular function and having an understanding of the mechanism of vestibular compensation, these totally opposite UVD syndromes would be inexplicable.

The anatomy of the peripheral vestibular nerve is also important for understanding the symptoms experienced by patients, their recovery, and the variability between patients. Although most vestibular afferent neurons course in the vestibular divisions of the eighth cranial nerve, some vestibular neurons course in the cochlear division of the eighth nerve. That means that if a patient undergoes a procedure, such as surgical vestibular neurectomy, after which there is still remaining hearing, then it is possible that some residual vestibular neurons may still be projecting from the affected labyrinth to the brainstem and that this residual vestibular input may trigger postoperative vestibular symptoms.

Many procedures are used for therapeutic intervention in peripheral vestibular function. For example, neuroma removal usually involves entire removal of the eighth cranial nerve, whereas vestibular neurectomy involves surgical sectioning of one division of the nerve (e.g., the superior vestibular nerve) rather than the entire nerve. The various

vestibular sensory regions project their information to the vestibular nucleus in different bundles of fibers within the vestibular nerve. That knowledge is also important for understanding recovery processes, because if just one division of the vestibular nerve is surgically sectioned (e.g., the superior vestibular nerve), then there is still substantial vestibular input projecting to the vestibular nuclei via the inferior vestibular nerve.

One procedure, now very widely used to treat unilateral peripheral vestibular dysfunction because of its relative safety, is intratympanic injection of a solution of the ototoxic antibiotic gentamicin—injected via a fine needle inserted through the eardrum into the middle ear (see Carey [2004] for a review).[23]

At low doses, gentamicin is taken up into the inner ear through the round window of the cochlea and fairly selectively attacks the cilia of vestibular receptors, disabling their response to natural vestibular stimuli, but with little effect on cochlear receptors, so hearing is minimally affected. Gentamicin attacks vestibular receptors in a progressive fashion; Type I receptors at the striola of the otoliths and the crest of the crista are most vulnerable.[24] Physiological evidence shows these receptors and the afferent neurons terminating on them convey information primarily about *changes* in vestibular stimulation—the onset of an angular or linear acceleration—and if they are lost, then one would expect that information about changes in vestibular stimulation would be affected early. Recent evidence shows that there are behavioral changes that correspond to that progressive loss.[25] During the course of the gentamicin treatment, the cell bodies of the receptors may be preserved and the receptor cilia on these vestibular receptor cells may regrow following the procedure, so that vestibular function (and possibly vestibular symptoms) may eventually reappear following gentamicin treatment.[26] In such patients, what may appear as vestibular compensation is in fact the return of peripheral vestibular function. A similar situation applies in some cases of vestibular neuritis when the affected peripheral neurons recover from temporary loss and start to function again. These are not examples of vestibular compensation but of *restoration* of peripheral vestibular function. Until recently, it was not possible to demonstrate this restoration of function conclusively, but with new selective tests of canal and otolith function, this vestibular restoration can be clearly identified. The conclusion: one needs to be careful in using data from patients with vestibular neuritis to understand vestibular compensation.

The intratympanic gentamicin injection procedure is now so successful that surgical UVD procedures are, with the exception of schwannoma surgery, becoming less common. However, in patients after gentamicin, the final level of residual vestibular function is unknown unless post-procedural testing is undertaken. After the same intratympanic gentamicin injection procedure, some patients may lose almost all vestibular function, whereas others may lose much less.

One of the themes of this chapter is to emphasize the importance of having baseline data about the level of vestibular function *before* any therapeutic procedure. This should be complemented by data about the level of vestibular function *after* the procedure. These data are especially important for the supposedly healthy ear. After the UVD procedure, it will be the neural output from this remaining labyrinth that will be of prime importance for the person's stability. Is the "healthy" ear really healthy? Getting such data is a very modest investment of time in relation to the possible outcome of a patient who compensates poorly because the supposedly healthy ear was in fact dysfunctional before the UVD. Such patients may spend many years in therapy and rehabilitation in an attempt to overcome their poor vestibular compensation, which was caused by an inappropriate procedure.

To measure precisely the pattern and resolution of the UVD syndrome in humans, it is best to study the same patients before and after surgical deafferentation of one intact labyrinth. Although such patients and the facilities for studying them are rare, some long-term quantitative data on the precise sensory and motor consequences of UVD of such human patients is available.[27-29]

The intense disequilibrium after UVD has both sensory and motor components, which can be categorized into static or dynamic symptoms. Static symptoms are present continuously, even when the person is totally stationary. Dynamic symptoms occur during movement. We consider each category in turn.

Static Symptoms

Spontaneous Nystagmus

Immediately after UVD, there is a spontaneous, mainly horizontal, conjugate ocular nystagmus. Both eyes show rapid eye movements, so that both eyes appear to be beating *away from the affected (lesioned) side*. Recordings of eye position show that the eye movements consist of slow eye deviations (called slow phases) toward the affected side, followed by rapid eye movements (called quick phases) away from the affected side (Fig. 8.2). The slow phases are not apparent to an observer and can only be detected by recording the eye movements. To an observer, both eyes appear to be beating away from the affected ear. This spontaneous nystagmus can be reduced or entirely

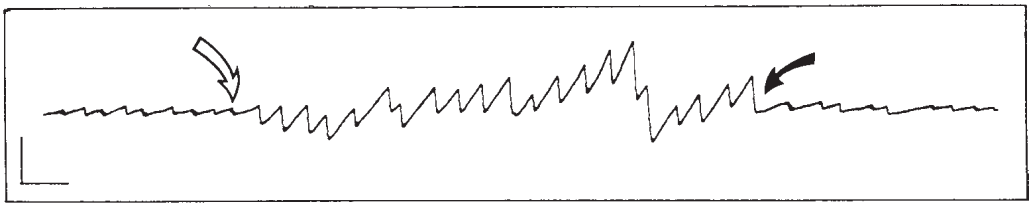

Figure 8.2 Peripheral vestibular nystagmus. Oculographic recording showing a left-beating, primary-position nystagmus that is obvious only when visual fixation is removed (open arrow), and is quickly suppressed again when visual fixation is permitted (filled arrow). Peripheral vestibular nystagmus can be detected clinically by viewing the fundus of one eye while occluding the other. The patient had a right vestibular neurectomy the previous day. Upward deflections indicate rightward eye movements; downward deflections indicate leftward eye movements. Bars: 10 deg and 1 sec.

suppressed by visual fixation, and such suppression by vision is an important way of identifying that the nystagmus is a result of peripheral vestibular dysfunction. Nystagmus caused by central dysfunction is not reduced or abolished by vision. To detect spontaneous nystagmus, one needs to have a way of viewing eye movements in darkness or without any visual fixation stimuli present. Two ways of doing this are Frenzel glasses or video recordings of eye movements in total darkness while the eye is illuminated by (invisible) infrared light. In the week after the occurrence of the UVD, the strength of the spontaneous nystagmus in darkness (the slow phase eye velocity) declines, but for some patients there is always a very small spontaneous nystagmus present in darkness as a permanent legacy of their UVD.

In spontaneous nystagmus, the vertical eye movement components are small or absent, presumably because the two vertical semicircular canals in the labyrinth cause essentially opposite vertical eye movements: anterior canal activation causes upward movement, and posterior canal activation causes downward movement. So, the simultaneous loss of both these sensory organs in the one labyrinth will result in the two vertical components cancelling each other.

Why Does Spontaneous Nystagmus Occur?

In animals, the UVD causes a total loss of vestibular afferent input to the vestibular nucleus on the affected side (the *ipsilesional* vestibular nucleus), so there is a great reduction in neural activity of neurons in that ipsilesional nucleus. Simultaneously, there is an increase in the resting activity of neurons in the vestibular nucleus on the healthy side (the *contralesional* vestibular nucleus). So, the UVD generates an imbalance in neural activity analogous to that produced by a real head rotation toward the intact ear. In both cases, the UVD and the rotation, the responses and the sensations are similar—the person perceives him- or herself as *rotating*, there is nystagmus, and there are corrective postural responses. In UVD patients, over time that neural imbalance decreases, and as it does, the static symptoms diminish.

Ocular Tilt Reaction

After UVD, in addition to spontaneous nystagmus, there is a static otolith component that is called the ocular tilt reaction.[30] Just as an angular rotation of the head causes an imbalance in neural activity between the two vestibular nuclei, so a maintained head tilt also causes an imbalance. Also, just as the canal-induced post-UVD symptoms can be seen as compensatory for a head angular acceleration, so the post-UVD otolithic responses (skew deviation, ocular torsion, and head tilt) can be seen as compensatory for a roll head tilt. It seems that these static otolithic symptoms are also caused by the neural imbalance between the vestibular nuclei.

The static otolithic symptoms consist of

1. *skew deviation* of the two eyes. The ipsilesional eye is positioned lower in its orbit relative to the (vertical) position of the contralesional eye in its orbit. This can result in double vision (diplopia), but the extent of the skew deviation is rarely large and usually resolves rapidly.[30-32]
2. *conjugate ocular torsion*. Both eyes roll in the orbit (around the line of sight) and adopt a maintained rolled eye position with the upper pole rolled toward the lesioned ear. This maintained ocular torsional position can be up to 15 degrees of ocular torsion relative to the pre-operative torsional position (Fig. 8.3).[31,33,34]
3. *roll head tilt* toward the lesioned side. When tested without visual input, the patient has a small roll head tilt toward the lesioned ear.

The mechanism by which the unilateral otolithic loss causes this ocular tilt reaction is speculative. It is most likely similar to the mechanism of the spontaneous nystagmus that

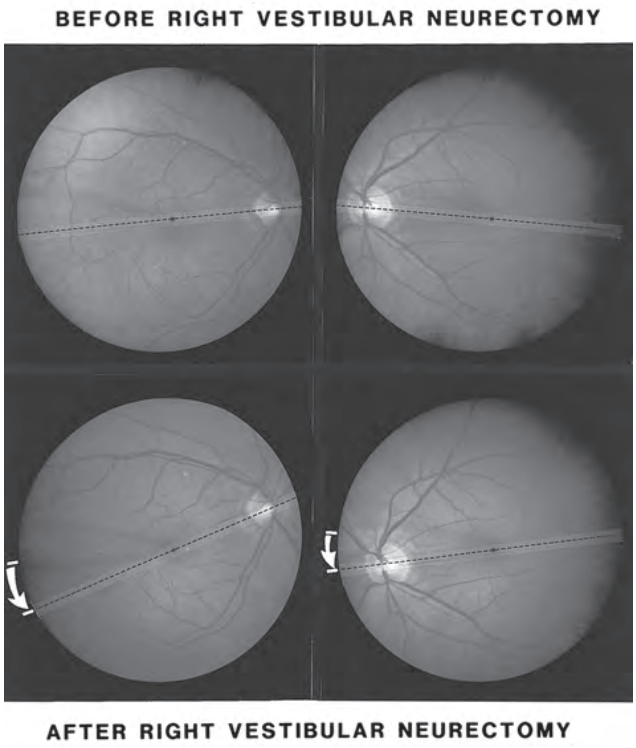

Figure 8.3 Fundus photographs of the left and right eyes of a patient before (*top*) and 1 week after (*bottom*) right vestibular neurectomy. After unilateral vestibular deafferentation (uVD), there is tonic torsion of the 12 o'clock meridian of each eye toward the patient's right side. The change in torsion angle measures 17 degrees in the right eye and 15 degrees in the left eye. When the patient was asked to set a luminous bar to the perceived visual horizontal in an otherwise darkened room, he set the bar tilted toward his right side by 14.2 degrees when viewing with the right eye and by 15.1 degrees when viewing with the left eye. (From Curthoys et al, 1991.[29])

occurs after UVD: it probably reflects an imbalance in average neural activity in the two lateral vestibular nuclei (LVN) because of decreased resting activity in otolithic secondary vestibular neurons in the ipsilesional vestibular nucleus, resulting from the loss of input from primary otolithic neurons.[35]

The aberrant ocular torsion position resolves slowly over time, but some residual ocular torsion seems to be a permanent legacy of a UVD.[29] How can it be measured? Examination of the fundus of the eye of such patients shows that the UVD causes the torsional position of the eye to change: the upper pole of both eyes has rolled to the lesioned side.[29] However, when the torsion is small, it is difficult to detect by fundus observation. A more sensitive way is to use the change in perception of the subjective visual horizontal or vertical, which closely corresponds to the change in ocular torsional position.[29,36] That measure of visual perception can be achieved by asking subjects in an otherwise darkened room to set a rotatable visible line so that it is where they perceive the visual horizontal to be. Patients with unilateral vestibular loss set this line so that it is too "low" on their affected side.[29,32,37,38] Comparison of the ocular torsional position and the subjective visual horizontal (SVH) shows that patients rotate this visual line so that its image lies close to their (torted) retinal meridian. This test, using settings to the SVH or subjective visual vertical (SVV), is called the bias test. The clinical significance of these findings is that careful standardized measurement of the subjective visual horizontal (or vertical) of patients, seated with head erect in an otherwise totally darkened room using a dim light bar, gives valuable diagnostic information about vestibular (mainly otolithic) function (see also Chapter 12).[29,38] A significant tilting of the subjective visual horizontal (greater than 3 degrees from the true horizontal) indicates vestibular, probably mainly otolithic, hypofunction on the side to which the patient tilts the bar.

We have measured the torsion and the perception of SVH in patients before and after unilateral vestibular neurectomy and demonstrated that the size of the change in the perception of the SVH caused by the UVD corresponded almost exactly to the change in ocular torsional position caused by the UVD, and that torsion position and perception recovered in parallel over time (see Chapter 12).[38-40]

Posture

After UVD, there is an offset in posture toward the affected side, which can be demonstrated by a variety of simple tasks that indicate that patients with UVD tend to lean or fall toward the affected side, especially in the early period after the UVD.[41] This position "offset" is called lateropulsion, and it decreases and disappears within about a month.[42]

Dynamic Symptoms

Semicircular Canal Responses

The dynamic VOR response is tested by accelerations of the subject—either angular or linear. For practical reasons, low-frequency (less than 1 Hz), low-acceleration, sinusoidal horizontal rotation has been used extensively in the past to test dynamic vestibular function.[43-45] Unfortunately, the results of such tests are indefinite, because at low frequencies the oculomotor response to the acceleration can be controlled by a variety of different sensory and motor systems apart from the vestibular system,[28,46] as shown by the fact that patients without vestibular function can generate responses to these stimuli.

Because of the failure to exclude extraneous sources of oculomotor control, there may *appear* to be recovery

of dynamic vestibular function. However, when careful measures with specific vestibular tests are made, the clear answer is that there is little or no such recovery of dynamic purely vestibular function. It can appear that recovery takes place, possibly as patients learn new modes of responding to the sinusoidal rotational test stimulus. These very low sinusoidal frequencies are not physiological; most natural head movements are high-acceleration, high-frequency stimuli. The head accelerations during walking or running, which post-UVD patients complain about, have high acceleration (2000 to 3000 deg/sec/sec) and high frequencies (5 to 12 Hz).

In order to measure dynamic vestibular function *specifically*, it is necessary to use high accelerations. One such test is the head impulse test (HIT), which uses brief, passive, unpredictable, angular accelerations in the natural range (2000 to 3000 deg/sec/sec). A simple version of this test can be conducted anywhere. This test consists of the clinician facing the patient and holding the patient's head at arm's length. Then, the clinician turns the patient's head in an abrupt unpredictable horizontal head rotation of about 20 degrees in about half a second, while the patient is asked to stare at the tip of the clinician's nose and not blink. The head is turned to be pointing at a target location—the movement is "turn and stop"—with no rebound. The term used to describe this kind of head rotation is a "head impulse." Although it is very brief, this abrupt head rotation has a peak angular velocity of around 200 to 300 deg/sec and a peak angular acceleration of 2500 to 4000 deg/sec/sec (or higher). In normal subjects, this head rotation results in a short-latency (about 10 ms), smooth, compensatory eye movement that compensates for the head turn so that the subject's gaze remains fixed on the clinician's nose irrespective of whether the rotation is to the left or right (Fig. 8.4). This means that, in normal subjects, VOR gain (the ratio of eye velocity to head velocity) is close to 1.0. For a patient after a unilateral loss, the result is very different. If the patient's head is rotated to the affected side, the UVD causes a marked reduction in VOR gain for these ipsilesional head rotations, whereas the VOR gain for head rotations to the healthy side (contralesional rotations) is only modestly reduced (Fig. 8.5).

The inadequate eye movement response during the ipsilesional head rotation means that, during the head rotation, the patient's eyes move with the head off the target, and so accumulate a position error which is usually corrected by a saccade at the end of the head movement, which takes the patient's gaze back to the target, the clinician's nose. It is that corrective saccade (called an "overt" saccade) that the clinician can (with a little training and practice) detect, and which is the tell-tale sign of an inadequate VOR.[28,47]

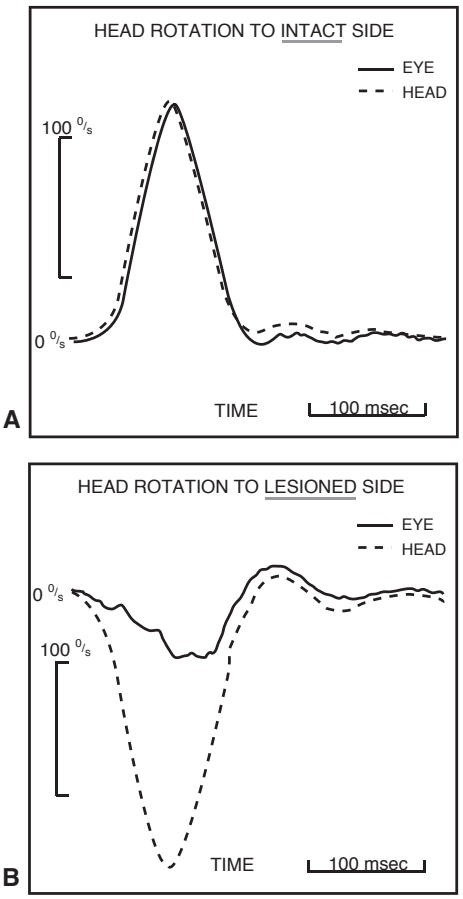

Figure 8.4 Time series of the eye velocity and head velocity response during a single horizontal passive head impulse from a patient 3 years after unilateral vestibular neurectomy. Head velocity is shown in dashed lines, eye velocity in continuous lines. **(A)** For rotation to the intact side, eye velocity mirrors head velocity. **(B)** In contrast, for rotations to the lesioned side, eye velocity is systematically smaller than head velocity from the onset of head rotation. (From Halmagyi et al, 1990.[28])

Testing over time shows that the ipsilesional VOR gain during the first 100 msec, before other sources of oculomotor control can affect the eye movement response, remains (apparently permanently) at around 25% of normal, whereas the contralesional VOR gain remains at around 80% of normal values.[2,28,48,49-52] A similar asymmetry of the VOR is also found in animals.[50-53]

After that first report of the overt catch-up saccade in 1988, some clinicians complained that in some patients with a known vestibular loss, it was not possible to detect this saccade at the end of the head turn toward the affected ear. When we measured the responses of UVD patients carefully with scleral search coils, we discovered that some UVD patients can manage to generate the corrective saccade actually *during* the head turn.[54] These saccades

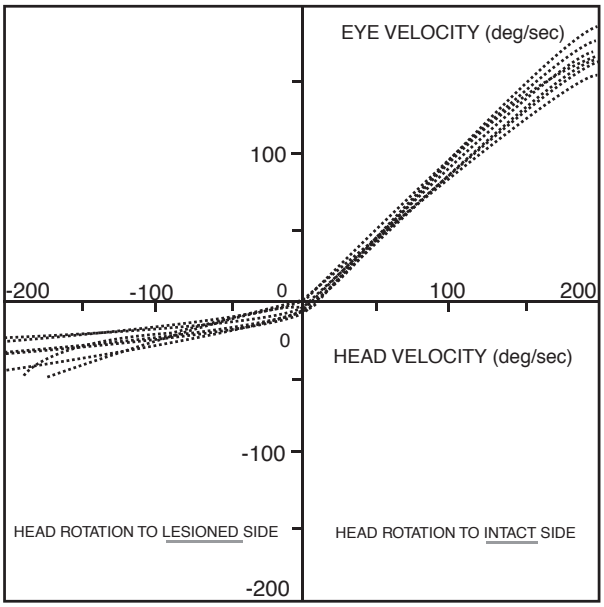

Figure 8.5 Plots showing the relationship between horizontal eye velocity, as a function of the corresponding horizontal head velocity during a head impulse for 20 horizontal head impulses in a patient who had undergone a left vestibular neurectomy 3 years previously. For head turns to the affected side, the eye velocity is very small and does not match the head velocity as it does for head turns to the intact side. This results in a profound VOR deficit in response to head impulses directed toward the affected side. In contrast, the VOR in response to head impulses directed toward the intact side is close to normal. (From Halmagyi et al, 1990.[28])

during the head turn are not detectable by the naked eye and so would not be observed by the clinical observation. We called these hidden saccades "covert" saccades and their existence means that it is necessary to actually measure the eye movement response during the head turn, rather than relying on the unreliable subjective clinical observation (Fig. 8.6). It is still the case that if an overt saccade is detected clinically, then the patient does have a vestibular loss. However, if no saccade is detected by clinical observation, no conclusion can be drawn—the patient may be normal or may have a unilateral vestibular loss and be generating a covert saccade to conceal that loss. Only objective measurement of the eye movement can resolve the latter question, and this was made possible by our development of a small, lightweight, high-speed, head-mounted video camera on minimal slip goggles. When we measured the eye movement during head turn, this camera system was fast enough (250 Hz sampling rate) to detect those small covert saccades and remove any ambiguity. We called this new system vHIT, and it has been in very successful use in clinics now for 3 years.

Covert saccades are a very successful way of dealing with the challenge facing a patient with a unilateral vestibular loss during a head movement, and it may be that those patients who recover well are using these covert saccades to protect their visual stability—that is a research question at present.[54] Why is a covert saccade

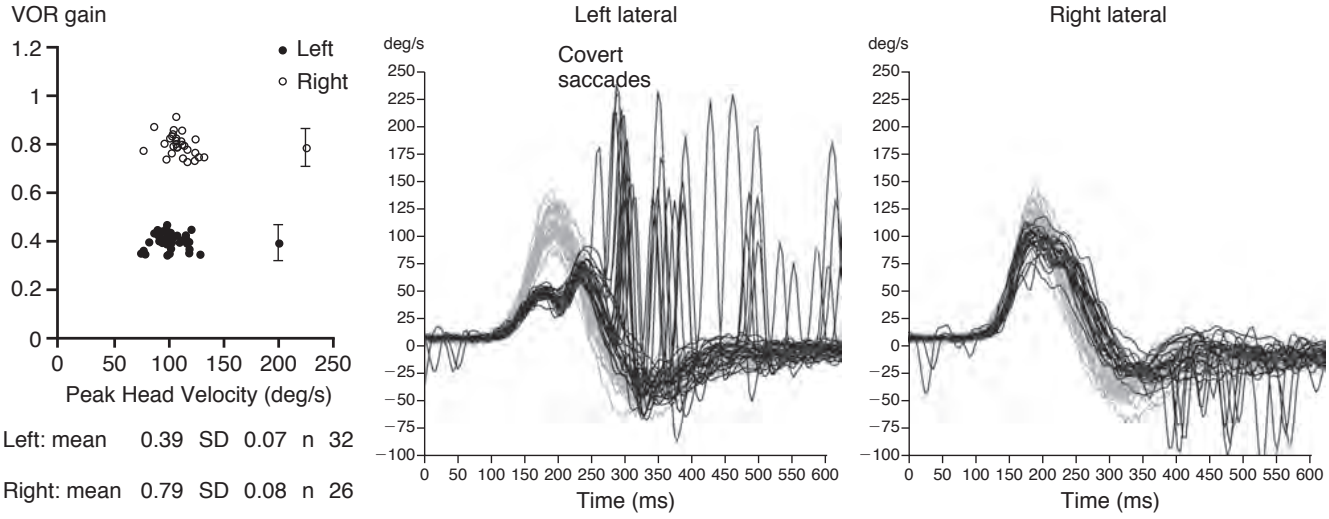

Figure 8.6 Time series of the head velocity stimulus and the eye velocity response during head turns to the left (affected) side and to the right in a patient with left superior vestibular neuritis. Many records have been superimposed, and for convenience of comparison, the eye velocity is inverted to show its relationship to head velocity, and the records for leftward rotations are inverted to allow comparison with the records for rightwards head rotation. For head turns to the affected side, the VOR gain is low (about 0.4) and there are many saccades occurring during the head movement. These would be very difficult to detect by the naked eye, and they are referred to as covert saccades. For head turns to the healthy (right) side, the eye velocity matches head velocity closely, and VOR gain is about 0.8.

successful? In such a UVD patient, if there is no covert saccade, the image is dragged across the retina at about the same velocity as the velocity of the head turn, and so causes a smeared, blurred visual image that lasts about as long as the head turn—usually 200 msec or so. It is this smear that is held to be so aversive for UVD patients, who complain about the blur and bounce of their visual experience during head movements. The perceptual effects of smear can be reduced or eliminated by a saccade. There are two reasons for this reduction: (1) the very high eye velocity of a saccade, which lasts only 20 msec or so, will reduce the visibility of the smear; and (2) visual perception is suppressed by central neural processes just before and during a saccade by a phenomenon known as saccadic suppression.[55-57] The combination of these two factors is an effective means of greatly reducing or eliminating the perceptual effect of the retinal smear. So, the brief ballistic saccade during the head turn regains the target and is probably as effective as the slow, compensatory, eye velocity correction from vestibular input for gaze maintenance (Fig. 8.7). Of course, a blink will also achieve the same result. Below we consider the role of covert saccades in vestibular compensation.

After UVD, there is a small symmetrical loss of VOR gain for pitch head rotations for both pitch nose down and pitch nose up. There is a larger loss for roll rotations.[58-61] When the roll head movement is toward the lesioned side, there is a large reduction in VOR gain; when the movement is toward the healthy ear, the drop in gain is not as great. In fact, when the rapid head rotation is aligned to be in the plane of one pair of the vertical canals (at about 45 degrees to the main pitch and roll planes) the UVD effect becomes clear, and the asymmetric VOR can be detected, because each vertical canal forms a synergistic pair with its opposing partner.[62] Impulses in these planes are called LARPs (left anterior-right posterior) and RALPs (right anterior and left posterior).

Dynamic Otolithic Responses

After UVD, there are deficits in otolithic dynamic responses. During dynamic otolithic stimulation as the head is rolled at constant velocity toward the lesioned ear, the

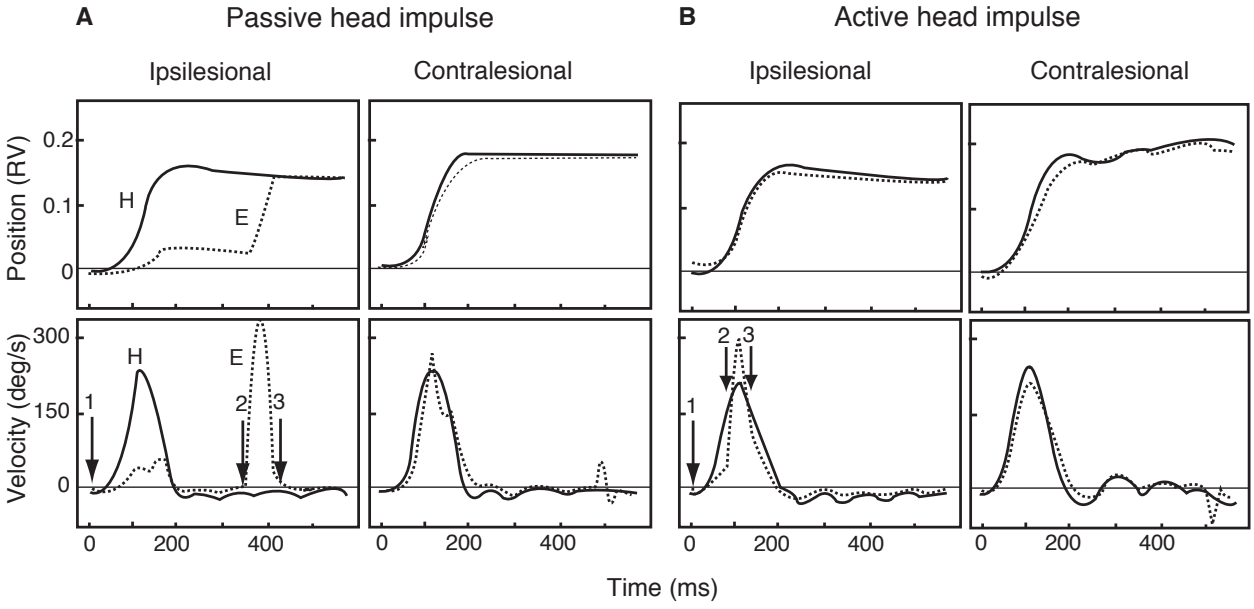

Figure 8.7 Time series of eye and head velocity during typical passive **(A)** and active **(B)** head impulses for both ipsilesional and contralesional head rotations. The eye velocity response has been inverted to allow close comparison to the head velocity. The start of the head rotation is indicated by the arrow (1), the start of the compensatory saccadic eye movement by (2), and the end of that compensatory saccade by (3). During passive ipsilesional head rotations, a large gaze error develops during the head rotation, and that error persists until the saccade occurs after the end of the head rotation and acts to return the patient's eyes to the target. With a comparable active head rotation **(B)** to the ipsilesional side, this compensatory saccade occurs *during* the head rotation so that the gaze error is small, and no corrective saccade is needed after the end of the head rotation. Close inspection of this low-noise, search-coil record shows the presence of this small but very effective saccade (a covert saccade) during the active head rotation. (From Black et al, 2005.[63])

magnitude of the linear acceleration stimulation of the otolithic receptors causes a small compensatory ocular torsion response—a roll of both eyes around the line of sight—called ocular counterrolling (OCR) in the direction opposite to the head roll tilt. OCR is one of the few accepted measures of otolith function.[64,65] Schmid-Priscoveanu et al tested patients before and after UVD and found at testing shortly after UVD, there was an asymmetry in their OCR response with smaller OCR for roll-tilts of the head toward the affected side. This OCR asymmetry recovers fairly quickly[66]—it is not like the permanent asymmetry of the angular VOR.

Impulses of linear acceleration (so-called head "heaves")—in which the patient's head is given a brief unpredictable lateral head movement toward or away from the affected ear[67,68]—also reveal an asymmetrical oculomotor response after UVD.[69,70] This lateral or linear head movement causes a horizontal eye rotation in normal subjects that appears to be caused by the otoliths because angular head rotations are minimal: after UVD, there is an asymmetry with a smaller horizontal eye velocity for linear accelerations directed to the operated ear (as with canals, ipsilesional stimulation reveals the smaller response) than for linear accelerations directed to the intact ear. However, this otolithic response asymmetry also disappears fairly quickly.[71]

Otolithic Responses to Sound and Vibration

Testing dynamic otolith function, and measuring any change in otolith function after UVD, has posed a major challenge because the usual way of delivering the linear acceleration stimulus has been to move the whole head (or body) of the patients using a sled or centrifuge. These devices are large, very expensive, dangerous, and totally impractical in a clinic. However, there is another way of generating linear accelerations: bone conducted vibration (BCV), which is composed of a series of linear accelerations of the mastoids.[72] Physiological evidence from guinea pigs shows that these linear accelerations, although brief, are a very effective means of activating one class of otolith neurons—otolith irregular neurons.[73] The evidence is that 500-Hz BCV is selective for otolith irregular neurons; few canal neurons are excited at that frequency. The sensitivity of these neurons is extraordinarily high; some neurons show clear activation at stimulus levels that are at or below ABR threshold.

Similarly, ACS can also activate these same otolith irregular afferent neurons, presumably by bending the cilia by a mechanism that is still not understood. This selective response of otolithic afferents to ACS and BCV allows one to use ACS and BCV to index dynamic otolith function in the clinic.[74-76] At lower frequencies (e.g., 100 Hz), canal neurons can also be activated by BCV (unpublished observation). This physiological evidence underpins the use of moderate intensity BCV and ACS as new ways of testing dynamic otolith function in the clinic.[77] Delivery of 500-Hz BCV to the midline of the forehead at the hairline—a location called Fz—has been shown to cause about equal stimulation of both ears simultaneously and so is a simple clinically effective way of simultaneous bilateral dynamic otolith stimulation.[78]

There are very extensive neural projections from the otoliths to many muscle groups throughout the body, and thus otolith activation results in myogenic potentials in these muscle groups. The two main potentials that have been measured for clinical tests of otolith function are the cVEMP and oVEMP, although many others have been reported. In response to 500-Hz Fz BCV, patients with UVD show a marked asymmetry of these VEMPs with little recovery over time.[72,79-81]

The Cervical Vestibular Evoked Myogenic Potential—the cVEMP

Sound-evoked saccular neurons project to and synapse on neurons in the ipsilateral vestibular nuclei, and inhibitory neurons in the vestibular nuclei project to ipsilateral spinal motoneurons and inhibit them.[82,83] So, if the SCM is tensed, and BCV is delivered, there is a short latency (about 13 msec to peak) positive (inhibitory) response recorded by surface EMG electrodes over the tensed SCM. The stimulus can be either a light tap to Fz with a tendon hammer or a brief (6 msec) burst of modest vibration (about the intensity delivered by a body massager or an electric toothbrush) delivered by a Bruel and Kjaer minishaker 4810.[79]

Thus, a short 500-Hz tone burst results in the cervical vestibular evoked myogenic potential—the cVEMP—consisting of an initial positive potential at about 13 msec latency followed by a negative potential at about 23 msec.[84-86] The cVEMP is mediated by an uncrossed inhibitory sacculo-collic response. In healthy subjects, the 500-Hz Fz BCV results in symmetrical cVEMPs, and in patients with complete UVD, there is reduced or absent cVEMP p13 on the ipsilesional SCM in response to Fz BCV or ACS stimulation of the affected ear (Fig. 8.8). There is little or no recovery over time; this diagnostic indicator is clear even in long-term UVD patients.

The Ocular Vestibular Evoked Myogenic Potential—the oVEMP

Similarly, in response to the same 500-Hz Fz BCV stimulus, the myogenic potential recorded beneath the eyes as the subject looks up (i.e., elevates the eyes to tense the inferior oblique muscles) is called the ocular vestibular evoked myogenic potential (oVEMP).[86-91] This is a

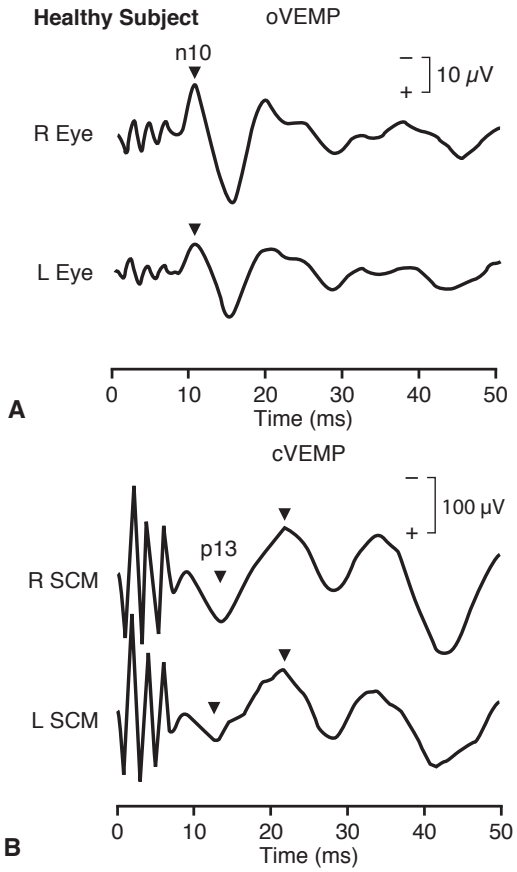

Figure 8.8 Examples of the averaged vestibular evoked myogenic potentials (VEMPs) in response to repeated 7-msec bursts of 500-Hz bone-conducted vibration delivered to the forehead at the junction of the midline and the hairline (Fz) in a healthy subject. **(A)** Electrodes beneath the eyes record a small negative (excitatory) myogenic potential beneath both eyes at a latency of about 10 msec as the subject looks up, and in a healthy subject, these n10 potentials are about equal beneath both eyes. This is the oVEMP n10. Unilateral utricular loss leads to a reduced or absent n10 beneath the eye opposite to the affected side. **(B)** Electrodes over the tensed SCM muscles record a small positive (inhibitory) myogenic potential at a latency of about 13 msec (p13) after the onset of the BCV. This is the cVEMP p13, and in a healthy subject, the p13–n23 potentials over both SCMs are about equal in amplitude. Unilateral saccular loss leads to a reduced or absent p13–n23 over the SCM on the affected side. (From Manzari et al, 2011.[92])

stimulus-locked ocular VEMP with the small initial negative (excitatory) potential at about 10 msec (called the oVEMP n10), which has since been shown to reflect predominantly utricular activity.[89,90] In healthy subjects, in response to 500-Hz Fz BCV, the oVEMP is of equal amplitude beneath both eyes (see Fig. 8.8). However, in patients after UVD, the oVEMP n10 is reduced or absent beneath the eye opposite the affected ear, because the oVEMP n10 is mediated by a crossed utriculo-ocular pathway. The oVEMP is a crossed, excitatory, ascending, utriculo-ocular response.[72,89,90] Unilateral vestibular loss results in reduced or absent oVEMP n10 responses beneath the eye opposite to the affected ear, and there is little or no recovery over time.[72]

These new VEMP tests allow measurement of dynamic utricular and saccular function, and when used together with vHIT, allow testing of all 10 vestibular sensory regions.[77] In this way, it is possible to identify the exact level of patient vestibular function, even during acute vertigo after UVD, and to have objective measurement of the changes (if any) of vestibular function over time.

Sensory Components

After UVD, patients exhibit two different false spatial sensations. Both illusions occur while the patient is stationary, and they usually resolve quickly (within a few days). One illusion is of an angular rotation in yaw, and the other is an illusion of roll-tilt toward the side of the UVD. In darkness or with eyes closed, the patient judges him- or herself to be rotating around the long axis of the body, toward the side of the UVD (i.e., ipsilesional rotation). However, if patients' eyes are open, then they perceive that they are stationary and that the world is rotating around them in the opposite direction (i.e., toward their intact side). This false sensation of rotation is called vertigo. Vertigo is powerful, but complex, and can be difficult for patients to describe.

Measures of the perception of angular and linear acceleration after UVD show poorer perception for accelerations directed to the lesioned ear. UVD patients underestimate the magnitude of both angular and linear accelerations directed to the ipsilesional side.[27,93] The linear acceleration perception has been studied using roll-tilt stimuli, and it was found that UVD patients underestimated roll-tilts to their affected ear, compared with these same patients' accurate performance before UVD.[27] Others have confirmed asymmetries of roll-tilt perception.[37,39,40] This perceptual deficit reduces with time, but the recovery is never total.

RECOVERY

Recovery of Static Symptoms

The static oculomotor and postural components resolve progressively over the first month or so. In contrast, detailed measures show that the dynamic symptoms do not change very much over time. These symptoms may appear to recover, as the patient learns new behaviors to conceal the loss, but careful testing that prevents patients from using these behaviors shows permanent deficit.

Recovery of Dynamic Responses

Recovery of Dynamic VOR—Slow Phases

Early studies of dynamic VOR recovery suggested that there was considerable recovery of the dynamic VOR after UVD.[94] But for practical reasons, those studies were restricted to measuring the slow-phase eye velocity response to low-frequency, low-acceleration sine-wave rotations (with accelerations of the order of tens of deg/sec/sec). When studies with natural values of head accelerations (of the order of thousands of deg/sec/sec) were used, it became clear that the recovery of dynamic VOR for ipsilesional head rotations was small or nonexistent. In patients tested before and after UVD, it was found that, for ipsilesional rotations, there was a large drop in VOR gain immediately after the surgical loss, and over the next year there was very little recovery in gain. The mean velocity gain at 1 week for 11 patients was 0.25 +/– 0.21 (sd). The mean velocity gain at 1 year after UVD was 0.27 +/– 1.14 (sd).[28,47] These patients are very unusual, because tests before the UVD showed that their horizontal canal function was normal. They underwent the surgical UVD to treat intractable benign paroxysmal positioning vertigo, which affected their posterior canal.

Is there any recovery of VOR gain at all? Yes, there appears to be some improvement in VOR gain for ipsilesional rotations in the low-frequency, low-acceleration range.[95,96] However, it is not known how functionally effective this small gain recovery is. As shown above, for higher (natural) head accelerations as encountered during most natural activities such as driving or sport, the evidence is that there is very little or no recovery of VOR gain. Cervical afferent input is one of the many other potential inputs that could modify and substitute for absent vestibular function (see Fig. 8.1). Potentiation of cervical sensory input may be relevant for improving responses to low-frequency head movements, especially for active head movements. There is now evidence from monkey behavioral studies that shows that cervicoocular responses are potentiated after UVD and BVD and that potentiation may assist in recovery of oculomotor responses to low-frequency head movements.[95-97] Recently, there has been evidence of neural mechanisms substituting for vestibular input at low accelerations.[96] In primates, it has been found that extravestibular inputs (neck proprioceptive and efference copy information) substitute for vestibular inputs after vestibular loss and act to stabilize gaze. It is possible that at low head velocities, especially if actively generated, these substitution mechanisms improve performance. It is clear from the permanent loss of ipsilesional VOR that these mechanisms are not adequate at high (natural) passive head accelerations, because such tests of long-term, well-compensated patients show the permanent loss. However, these mechanisms probably account for a portion of the subjective improvements. The evidence from Halmagyi et al[98] shows that neck input seems to have little or no role in generating slow-phase eye velocity responses to unpredictable passive, angular high-acceleration head rotations. It may be important for the generation of saccadic corrections.

The paradox is that despite this very inadequate recovery of the VOR to natural dynamic stimuli, many post-UVD patients return to a normal lifestyle and are just not troubled by the UVD and the VOR loss that our precision measures show that they have. They have "compensated" but evidently not by synaptic changes in the vestibular nuclei for natural head accelerations, because their measured eye velocity for unpredictable passive ipsilesional head rotations remains poor. Why then are they not troubled by their inadequate VOR? Two possibilities, which are not necessarily mutually exclusive, are (1) that the modest recovery of function for low-frequency stimuli is enough, or (2) that they are using other strategies, such as different patterns of eye-head coordination, to overcome their loss. Our preliminary evidence suggests the latter, and that saccades may be the key. In the area of vestibular compensation, the main focus has been on the compensatory slow-phase eye velocity without adequate recognition of the fact that changes in saccadic pattern are a very effective way of overcoming unilateral vestibular loss as suggested in 1989 by Alain Berthoz, who coined the term "saccadic substitution."[99] We suggest the focus for future studies of plasticity in the compensation of oculomotor responses in human patients after UVD should be on saccades, rather than on slow-phase eye velocity.[55]

Permanent deficit of dynamic VOR function after UVD shows that vestibular rehabilitation of UVD patients should be aimed at developing new behaviors that substitute for the dynamic vestibular loss—for example, substituting saccades for the inadequate slow compensatory eye velocity, rather than trying to restore slow-phase eye velocity that cannot be restored.

These different patterns of saccadic responses after UVD lead to the hypothesis that the different groups of patients using these different saccadic strategies may show different patterns of vestibular compensation.[55] During testing at the stage of the acute attack, some VN patients already show changes in their pattern of saccades from overt to covert, but it remains to be seen whether these "early coverters" in fact compensate better.

Summary

In Box 8-2, we list the permanent legacies of unilateral vestibular loss, and in Table 8-1 we summarize the UVD symptoms and the general extent of their recovery. Most symptoms are at their maximum within the first day after the UVD.

Box 8-2

PERMANENT LEGACIES OF UVD

1. Spontaneous nystagmus directed away from the lesioned ear.
2. Ocular torsional position with both eyes rolled to the lesioned side.
3. Inadequate perception of roll-tilt toward the lesioned side.
4. Inadequate perception of horizontal angular acceleration directed to the lesioned side.
5. Inadequate horizontal VOR for high accelerations directed to the affected ear.

Clinical Evidence Concerning Factors Affecting the UVD Syndrome and Its Recovery

Vestibular Restoration

In some patients, the peripheral vestibular function may be totally restored.[92] A recent example comes from a patient diagnosed with acute superior vestibular neuritis. The patient's vestibular function was measured at the height of an acute attack of vertigo caused by unilateral superior vestibular neuritis, and then again 3 months later, without any special medication or rehabilitation in the interim. The results showed that at attack during an ipsilesional head turn, there was a major drop in VOR gain for passive, unpredictable, high-acceleration horizontal head rotations, whereas the VOR gain for head turns to the healthy side was only slightly reduced. However, at testing 3 months

■ Table 8-1 **UVD SYNDROME: SYMPTOMS AND EXTENT OF RECOVERY**

UVD Syndrome	Description/Type	Extent of Recovery
Oculomotor symptoms:		
Spontaneous nystagmus	Rhythmic eye movements with quick phases directed away from the affected ear and toward the intact ear Visible when all visual fixation has been removed	Decreased at 1 week Largely but incompletely recovered at 1 year
Ocular torsion	Maintained roll position of both eyes so the upper poles of both eyes roll toward the lesioned side Visual perception is affected in a corresponding fashion	Largely recovered within 6 months (but not totally—some torsion still present 12 months after UVD)
Skew deviation	Maintained disconjugate vertical eye position with the ipsilesional eye being lower in the orbit than the contralesional eye	Resolves within a few days
Caloric nystagmus	Absent nystagmus to caloric stimulation of the horizontal canal of the affected ear	Permanent loss
Horizontal VOR to passive low angular accelerations	Reduced VOR gain for low-frequency, low-angular-acceleration stimulation in an ipsilesional direction	Steady improvement over months and virtually resolved within 1 year
Horizontal VOR to high (natural) angular accelerations (head impulses)	Substantially reduced VOR gain (25% of normal values) for high-frequency, high-angular-acceleration stimulation (natural values) in an ipsilesional direction	Little change over 1 year and apparently a permanent loss

Continued

■ Table 8-1 UVD SYNDROME: SYMPTOMS AND EXTENT OF RECOVERY—cont'd

UVD Syndrome	Description/Type	Extent of Recovery
	Reduced VOR gain (80% of normal values) for high-frequency, high-angular-acceleration stimulation (natural values) in a contralesional direction	Little change over 1 year and apparently a permanent loss
	Net result—asymmetrical VOR	Little change over 1 year and apparently a permanent VOR asymmetry
Pitch VOR to passive high angular accelerations	Reduced VOR gain for both pitch nose-down and pitch nose-up accelerations	Little change over 1 year—apparently permanent
Roll VOR to passive high angular accelerations	Substantially reduced VOR gain for high angular acceleration to stimulation in an ipsilesional direction	Little change over 1 year
	Reduced VOR gain for high angular accelerations in a contralesional direction	Little change over 1 year
Canal plane head impulses (left anterior–right posterior or right anterior–left posterior)	Substantially reduced VOR gain (25% of normal values) for high-frequency, high-angular-acceleration stimulation (natural values) in an ipsilesional direction Reduced VOR gain (80% of normal values) for high-frequency, high-angular-acceleration stimulation (natural values) in a contralesional direction Net result—asymmetrical VOR	Little change over 1 year
Ocular counter-rolling	Decreased ocular torsion during head rotations toward the ipsilesional ear	Large asymmetry initially and decreased asymmetry over time
Horizontal eye movements to impulses of interaural linear acceleration (head heaves)	Smaller eye rotations for linear accelerations directed toward the affected ear	Resolution unknown
Vestibulospinal symptoms:		
Roll head tilt	Roll head tilt toward the affected side present when visual fixation has been removed	Resolves within weeks but a small roll head tilt may be a permanent legacy (especially in animals after UVD)
Postural disequilibrium at rest	Poor postural stability when vision is removed	Reduces but incompletely resolved within a year
Dynamic postural disequilibrium	Poor postural stability when given dynamic challenges	Reduces over 1 year
Cervical vestibular evoked myogenic potential (cVEMP)	Reduced or absent short-latency inhibition of ipsilesional sternocleidomastoid EMG response to intense air-conducted clicks or bone-conducted vibration	Permanent loss

■ Table 8-1 **UVD SYNDROME: SYMPTOMS AND EXTENT OF RECOVERY—cont'd**

UVD Syndrome	Description/Type	Extent of Recovery
Ocular vestibular evoked myogenic potential (oVEMP)	Reduced or absent excitation of the contralesional EMG of the inferior oblique to intense air-conducted sound or bone-conducted vibration	Permanent loss
Perceptual symptoms:		
Lateropulsion	Sensation of falling or being pushed toward the affected side	Improved by 1 week; resolved within 1 year
Vertigo	In darkness: illusion that the patient is rotating in yaw toward the affected side *OR* In light: illusion that the world is rotating in yaw toward the healthy side	Resolved within a few days
Roll-tilt illusion	In darkness, the illusion is that the person is roll-tilted toward the affected side	Completely resolved within a few days
Yaw angular acceleration perception	Underestimation of the magnitude of yaw angular accelerations toward the affected ear	Resolution unknown
Linear acceleration perception	Underestimation of the magnitude of linear accelerations toward the affected ear	Largely but incompletely resolved at 1 year

later, the VOR for these same ipsilesional rotations had fully returned. Similarly, the test of utricular function (the oVEMP n10) at the acute attack shows an absent oVEMP n10 contralateral to the lesion, indicating that the utricular function on the affected side was not present at the acute phase, but 3 months later, utricular function had fully returned. (In this patient, the saccular function was not affected by the neuritis—it was present at the acute phase and also at recovery, probably because the neuritis was confined to the superior division of the vestibular nerve). Without the results of the tests of peripheral vestibular function, these responses would be classed as an excellent example of vestibular compensation. However, the objective measures of peripheral vestibular function show the improved performance is a result of actual restoration of peripheral vestibular sensory function (Fig. 8.9).

The processes of compensation are unlikely to help patients with short recurrent bouts of paroxysmal vestibular dysfunction as occurs in Ménière's disease, because the process of compensation takes some time (days or weeks) to be implemented, whereas in patients with fluctuating vestibulopathies such as Ménière's disease, the attacks of vertigo are brief compared with the time required for compensation. In humans, vestibular compensation takes 3 to 5 days to get under way and a month or more to achieve a functionally useful level.

Poor Compensation

Most patients recover well after UVD and have an apparently normal lifestyle with good quality of life. But some patients do not. The reason for the poor recovery is puzzling. In many cases, they present with similar symptoms to the well-compensated patients: their UVD procedure is similar, but they do not recover as well as others. "Recovery" has a large subjective component, and it is probable that some of these patients had expected a much better outcome. Detailed comparison of the vestibular performance of such patients has so far not been able to identify any clear differences post-UVD between well- and poorly compensated patients (see Box 8-2).

Patients with poor compensation exhibit a syndrome called chronic vestibular insufficiency, which consists of sensations of disequilibrium, gait ataxia (especially with restricted vision on unstable surfaces), and oscillopsia. The ataxia is particularly evident when vision is disrupted and

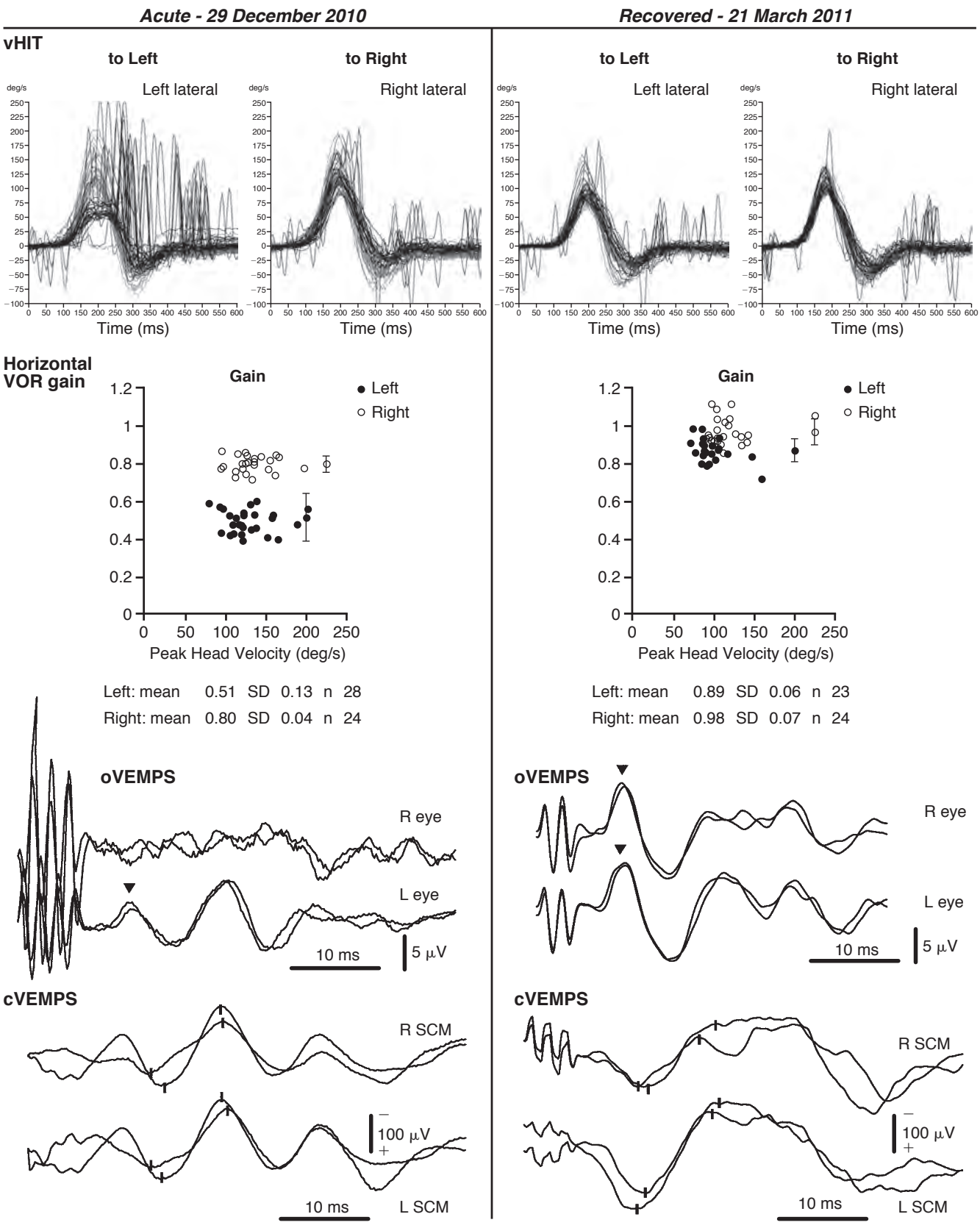
Acute - 29 December 2010
Recovered - 21 March 2011
vHIT
to Left
to Right
Left lateral
Right lateral
deg/s
Time (ms)
Horizontal VOR gain
Gain
Left
Right
Peak Head Velocity (deg/s)
Left: mean 0.51 SD 0.13 n 28
Right: mean 0.80 SD 0.04 n 24
Left: mean 0.89 SD 0.06 n 23
Right: mean 0.98 SD 0.07 n 24
oVEMPS
R eye
L eye
10 ms
5 μV
cVEMPS
R SCM
L SCM
100 μV

Figure 8.9 Tests of peripheral vestibular function on the same patient on two occasions,[92] separated by about 3 months. On the first occasion the patient was tested during an acute attack of vertigo, which was diagnosed as superior vestibular neuritis. The tests of horizontal canal function showed that, for head turns to the left (the affected side), there was a marked reduction in slow-phase VOR gain (about 0.5) and many covert saccades, indicating a major loss of function of the horizontal canal. The VOR for head turns to the intact side was minimally affected (gain of 0.8). At testing 3 months later, the VOR gain for left head turns had returned to normal values, and there were no covert saccades. The test of utricular function showed that initially the oVEMP n10 was absent beneath the right eye (opposite the affected left ear), but the oVEMP also returned to normal and symmetrical values at testing 3 months after the attack. The marked recovery of function shown by tests of peripheral vestibular function justifies the conclusion that there has been a complete restoration of normal vestibular function within 3 months, without special medication or rehabilitation. (From Manzari et al, 2011.[92])

proprioception is challenged (e.g., by asking the patient to stand with eyes closed on a soft, thick, foam rubber mat). Oscillopsia refers to the sensation of apparent movement of a physically stationary object during head movement (e.g., the world appears to bounce during jogging, or appears to slide or smear during a rapid head movement during driving). Oscillopsia is evident when the person's head is moved abruptly (high accelerations) either passively or actively—during running or looking from side-to-side while crossing a road or during a sudden head turn while driving.

Why should this poor compensation occur? In some cases, it seems that events (such as postoperative complications) within the first few days after the operation may be important for determining the success or otherwise of the eventual recovery. There is some evidence that the early stages of compensation—the initiation of vestibular compensation—are vulnerable and may be interrupted by other events. In other words, there may be a sensitive period for the establishment of vestibular compensation.[100-101] Certainly there is neural and behavioral evidence from animal studies that underpins this distinction between the initiation and maintenance of vestibular compensation (Vibert et al).[32] The UVD procedure itself may inadvertently generate the conditions for poor compensation—if the UVD procedure leaves some vestibular fibers intact and operational, the neural signals from these fibers may interfere with the vestibular compensation process.

Some poorly compensated patients may have had inadequate vestibular function or even central (e.g., cerebellar) deficits before the UVD procedure.[102] Thus, the UVD procedure is potentially dangerous, and that is why careful pre-procedural testing is needed to ensure there is an adequate level of function of the remaining labyrinth and an absence of central deficits before the UVD procedure is undertaken. Patients may suffer postural disequilibrium and gait ataxia virtually for the rest of their lives if the UVD procedure is carried out inappropriately. Rehabilitation procedures are of potential value to such poorly compensated patients, but prevention is preferable to rehabilitation.

In human patients, it is unwise to contemplate a UVD when the level of vestibular function in the supposedly healthy ear is not adequate. For example, in Ménière's disease it is frequently the case that both ears are affected by the disease, but one ear is more affected than the other. If a UVD procedure is carried out on the more affected ear in such patients, the patient will be relying on a partly dysfunctional ear to provide full vestibular input.[103] It is not too surprising that in such cases the eventual result will be poor compensation or chronic vestibular insufficiency. The sole remaining labyrinth will provide all the vestibular input for oculomotor, postural, and perceptual control for that person for the rest of his or her life. Is the level of function in that remaining, supposedly healthy labyrinth adequate to carry that burden? With the new tests of peripheral vestibular function (vHIT, oVEMPs, and cVEMPs), all peripheral vestibular sense organs can be evaluated.[77]

Decompensation

An apparently related phenomenon is decompensation. Decompensation refers to situations in which vestibular compensation is nullified and the patient relapses partially or completely, and the UVD syndrome returns—the spontaneous nystagmus, the sensations of vertigo, and the postural disequilibrium reappear. Some situations, such as highly stressful ones, may trigger decompensation,[104-106] but even changing the vestibular environment may also do so[106]: Reber et al reported that when well-compensated rats were exposed to a brief interval of microgravity during parabolic flight, they decompensated and showed spontaneous nystagmus and other signs on removal of the gravity stimulus. That decompensation disappeared within seconds of a return to 1 *g*. This suggests that changing the

otolithic stimulation of the intact ear was sufficient to elicit this brief decompensation. These data and other data suggest that otolithic input from the remaining ear plays an important role in compensation.[12,107]

Psychological Factors

Of course, psychological factors play a major role in compensation, but it is difficult to determine their precise role.[108] Are psychological factors causal in determining the outcome of UVD, or are they caused by the procedure itself? Bowman[109] measured the personality characteristics of well- and poorly compensated patients and found that poorly compensated patients showed higher scores on tests of somatic awareness (attention to bodily sensations) than control subjects. However, it is not clear whether this result occurred because their poor compensation had encouraged them to focus attention on themselves or whether this personality style had been present before the UVD procedure and caused their poor compensation. It is not surprising that people with a predisposition to hypochondriasis would find the symptoms after UVD distressing. Patients who have had a UVD for treatment of a life-threatening neuroma may be expected to respond more positively compared with patients who have chosen to have a UVD to alleviate a much less threatening condition. (See Chapters 18 and 19 for more discussion of psychological factors.)

Medication

At the start of this chapter, we showed the variety of influences on transmission of neural information in the vestibular nucleus. Not only do vestibular neurons themselves use a variety of different neurotransmitters, but they receive input from many other brain regions that can modulate the activity of these neurons and the transmission and processing of vestibular information. Because of the multi-input character of these neurons, a host of different medications can influence the UVD syndrome and its recovery. Whether any one neurotransmitter is the "key" in such a complex system may well be a rather useless question. For reviews of medications see references.[3,110-113]

Evidence from Animal Studies

Despite the similar time course of vestibular compensation in humans and animals, the data from animal studies of vestibular compensation do not transfer directly to human studies. In animal studies, the UVD is carried out on normal, healthy, young animals and is usually surgical and usually complete. There is no attempt in most animal studies to preserve hearing. The analogous human patients would constitute a tiny minority of patients undergoing a UVD—most patients tend to be middle aged and have had a chronic progressive disease of one labyrinth that has been ongoing for months or years, the other labyrinth may be affected, and the surgeon endeavors to preserve hearing. One clear result is that different symptoms—oculomotor and postural—recover at different rates.[114-115] Blocking of neurogenesis in the vestibular nuclei of cats after UVD acts to impair recovery of their postural balance, but it has no effect on the recovery of spontaneous nystagmus.[115] This differential rate of recovery is clearly shown in guinea pig compensation, in which the spontaneous nystagmus recovers quickly, but the roll head tilt (longitudinal twist) is present many months after the UVD (Fig. 8.10).

Again referring to our simple schematic diagram of information transmission in the vestibular system (see Fig. 8.1), one can understand how it is that many different sensory manipulations can influence UVD and the rate of recovery. Deprivation of all visual input after UVD impedes the compensation of dynamic VOR responses to low acceleration stimulation[96] and the static component of roll head tilt,[116,117] but seems to have little effect on the reduction of spontaneous nystagmus after UVD.[94] Visual inputs do augment the diminished motor responses to linear acceleration[118] and the deficient righting reflexes[119] that occur after UVD. Visual motion deprivation delays recovery of locomotor equilibrium.[120-123]

Similarly, stimulation of proprioceptive receptors appears to facilitate the recovery of dynamic postural equilibrium,[124] and deprivation of proprioceptive input appears to retard the recovery of postural equilibrium. Cervical proprioceptive input could be important in static compensation, because head restraint retards resolution of head tilt and spontaneous nystagmus, at least in guinea pigs.[125] Somatosensory proprioceptive deprivation appears to retard static compensation,[118] whereas somatosensory proprioceptive stimulation appears to facilitate the restoration of dynamic postural equilibrium.[124] Acute spinal lesions can produce a temporary decompensation of static postural symptoms.[104,105]

Only scant data exist on the effects of vestibular stimulation or vestibular deprivation on static or dynamic compensation. In frogs, otolithic stimulation hastens, whereas otolithic deprivation delays, static compensation of roll-tilt of the head.[126] In cats, low-frequency combined visual-vestibular stimulation helps reverse the deficit in VOR gain, which occurs in response to low-frequency stimulation

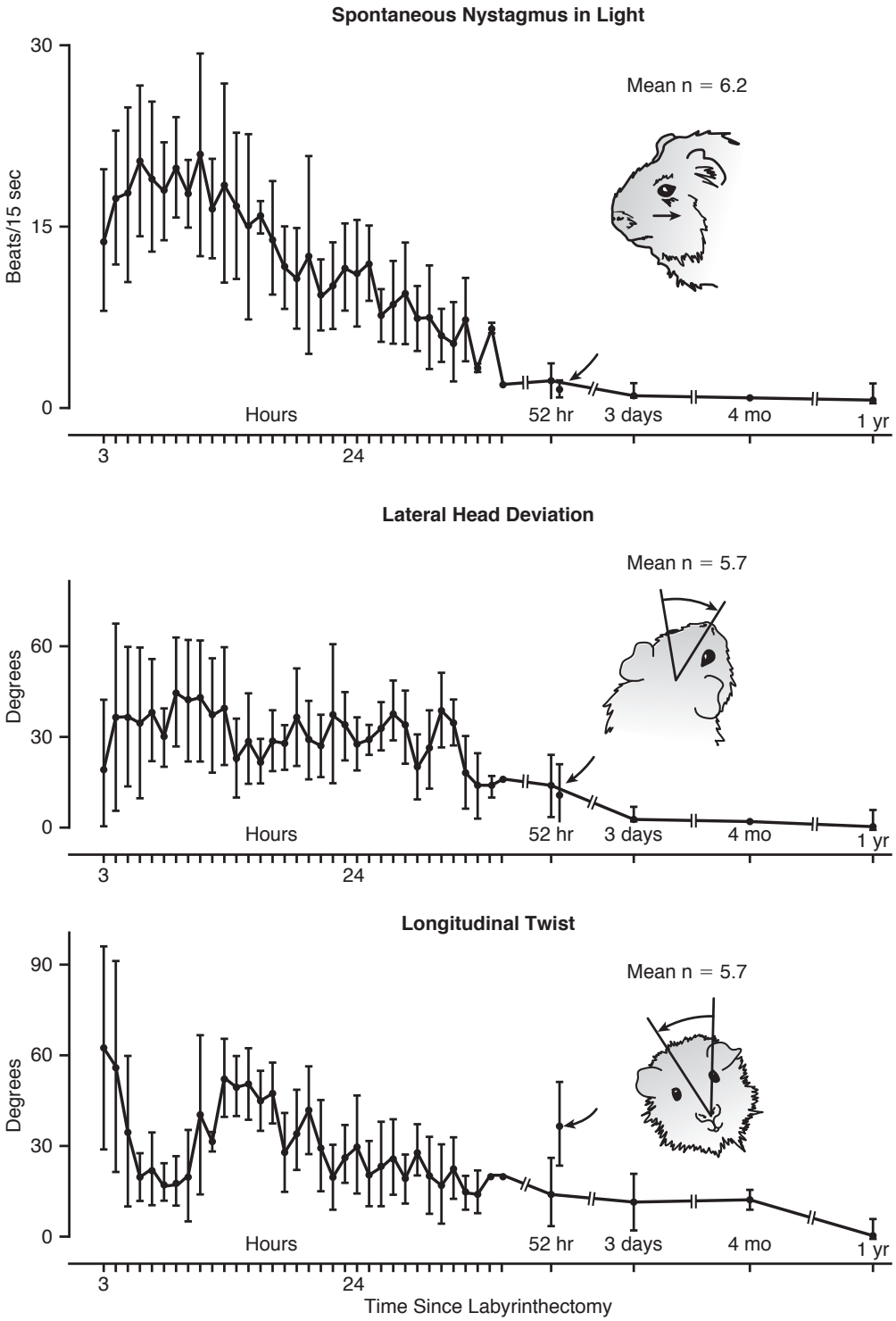

Figure 8.10 Different symptoms of UVD recover at different rates as shown by averaged measures of nystagmus and posture of guinea pigs over time after UVD. The nystagmus decays very quickly—within about 52 hours. However, the roll-tilt of the head shows a completely different recovery profile, and at testing 4 months after UVD, there was still a small but significant head roll-tilt present.

following UVD,[127] but has no effect on the asymmetry of the VOR at high frequencies.

VOR Plasticity

There is a long history of studies of vestibulo-ocular responses that show that the gain of the VOR is modifiable in normal healthy subjects. Gonshor and Jones [128,129] required subjects to wear reversing spectacles during the course of their usual daily activities. To attain a stable retinal image in this situation, the VOR gain must decrease and even reverse. VOR testing in darkness at regular intervals during this procedure showed that the VOR gain did decrease, although not by the full amount required, and there was no reversal. (Unfortunately, the VOR testing in this study used predictable low-frequency sinusoidal rotations rather than pure tests of VOR function using natural angular accelerations, so we cannot be sure that the measured VOR gain increase was purely vestibular.) This result triggered an explosion of interest in VOR plasticity. It appeared that mechanisms responsible for this VOR plasticity may be used by UVD patients to assist their recovery; however, more recent evidence indicates this does not happen.

There is no doubt that the gain of the VOR can be modified with quite remarkable speed in normal healthy subjects—even within a few minutes. However, closer consideration shows that such evidence is not really relevant for the situation confronting the UVD patient in vestibular compensation. Some of the procedures that have been developed for the study of VOR plasticity do assist patients to improve their normal everyday functioning, but we suggest these improvements are not a result of any change in the intrinsic vestibular component of the VOR. If VOR was as modifiable as the VOR plasticity literature suggests, then diagnostic tests of vestibular function would be useless and even unnecessary. Patients would learn to overcome their inadequate vestibular input and generate compensatory responses even when they had total unilateral peripheral vestibular loss.

It is certainly the case that, during vestibular compensation, patients do learn a variety of behavioral strategies to minimize the effect of their vestibular deficit, but all indications are that any plasticity of the purely vestibular dynamic slow-phase eye velocity response is very modest at natural head angular accelerations. For UVD patients, we think that most of the "plasticity" takes place because of contributions of other sensory systems and cognitive control: the patient learns new behaviors to conceal the vestibular deficit. This achieves the sought-after goal of returning the patient to a normal lifestyle.

The peripheral vestibular function of UVD patients differs substantially from that in normal healthy subjects: the entire peripheral vestibular input from one labyrinth has been removed or severely compromised, thereby depriving the brainstem of both the resting discharge of these neurons and its modulation by head movement. Possibly, the challenge of such a loss for processes of vestibular plasticity is too large: the UVD itself disrupts central vestibular processing of vestibular information. In particular, a group of neurons concerned with perseverating the vestibular neural response (the velocity storage integrator) is compromised or disabled, and that may disrupt the neural substrate needed for vestibular modification.

Rehabilitation

Vestibular Compensation and Rehabilitation

Many years ago, Cawthorne and Cooksey suggested a number of exercises to assist in the rehabilitation of patients with vestibular disorders.[130,131] Those exercises are similar to those in use today. Doubts about the efficacy of such exercises have been largely dispelled.[132-135] If there is no change in purely vestibular function, how can these exercises benefit patients? How can patients improve? As we have shown, substitution of other responses can effectively conceal the vestibular deficit and so protect the patient from receiving smeared retinal images during head movements. Such substitution is possible when there is active control of the response by the patient, and we suggest that the Cawthorne-Cooksey exercises and other such exercises are acting to teach patients how to substitute these other responses to conceal and thus overcome their sensory deficit. The results of others[136] agree with our own results.[63] The active VOR gain is enhanced during active voluntary head movements, and during active head impulses, patients can learn to preprogram a small eye movement response (a small corrective saccade) during the ipsilesional head movement that can effectively hide their inadequate VOR during that ipsilesional head rotation.[63] (See also Chapters 9 and 22.)

It is our contention that the development of new eye-head coordination strategies should be facilitated by training with active head turns. If one asks the same patient to maintain gaze in a comparable fashion to the passive head impulse test described above while they *actively* turn their own head abruptly to left or right, most patients learn to preprogram a small saccade to correct for inadequate VOR and to insert this saccade *during* the head movement (see Fig. 8.7).[63] This is an important observation, because it shows that patients are learning to substitute this (covert)

saccade for the deficient vestibular slow phase. Also, it shows that this (covert) saccade acts to minimize the effect of the UVD on the patients' permanent dynamic VOR deficit. Another way of concealing a VOR inadequacy is by a blink, which, by completely removing the retinal image, very effectively prevents smear of the retinal image. Blinks are common during head movements in normal healthy subjects and in UVD patients. Ironically, most tests of VOR (including the head impulse test) prevent patients from using blinks. They require patients to keep their eyes wide open during testing. In other words, the testing procedure itself requires patients to suppress one of the strategies (blinks) that they may have learned to assist their vestibular recovery.

Patients appear to learn new patterns of eye-head coordination. However, the initial preference of many patients is to restrict head movements totally—to "lock" the head on body. This decreases the opportunity for the patients to learn any new pattern of eye-head coordination. In our view, patients should be encouraged to execute eye-head refixations—for example, by the spouse giving them passive, unpredictable horizontal head impulses for a few minutes per day.

We suggest that the process of vestibular rehabilitation should be thought of as an opportunity for other non-vestibular sensory inputs and cognitive-behavioral strategies to increase their role in controlling the patient's equilibrium.

Neural Evidence Concerning Recovery after UVD

Overview

This is a huge area, and in this chapter we refer only to the major features. For reviews of neural processing of vestibular sensory input, see references.[137-141]

There is one simple principle that helps us to understand the neural basis of vestibular responses and vestibular compensation, and that is "balance." That term is often used to denote the function of the whole vestibular system, but it also coincidentally applies to the neural mechanism of vestibular operation. When a person holds the head still, the neural activity in the pair of vestibular nuclei (one on each side of the brainstem) is approximately equal—balanced. If there is a horizontal (yaw) angular acceleration in one direction, for example, to the person's left, then it causes an imbalance in neural activity between the vestibular nuclei. Many neurons in the ipsilateral (left) vestibular nucleus increase their rate of firing of action potentials, while many neurons in the right vestibular nucleus concurrently decrease their firing. The result is an imbalance in neural activation between the two nuclei. Changes in perception, and also in corrective responses (vestibulo-ocular and vestibulo-spinal), are generated by that neural imbalance. If the acceleration is prolonged, there is nystagmus.

Another way of generating such an imbalance is by silencing the input from one labyrinth in a patient at rest, as occurs in a UVD. In both cases, the responses and the sensations are similar: the person perceives him- or herself as rotating, there is a nystagmus, and corrective postural responses occur. Most real-life angular and linear accelerations are usually fairly short, and at the end of the acceleration the system returns to its balanced state. But after UVD, the imbalance at rest—without any imposed angular or linear acceleration stimulus—persists for hours or days. Recordings of neural activity have shown that the imbalance at rest is progressively reduced within the first few days; the very active cells in the contralesional vestibular nucleus decrease their activity, and silenced neurons in the ipsilesional vestibular nucleus start to fire again. As that imbalance is reduced, so the behavioral and perceptual symptoms decline.[142]

Why should rebalancing occur? What processes act to remove the imbalance and equalize firing in the two nuclei? In the guinea pig after a UVD, this happens in the space of about one day, and by 52 hours after UVD, the average activity on both sides is about equal.[142] The spontaneous nystagmus disappears, although the dynamic deficits remain. There is no substantial change of peripheral vestibular afferents of the remaining ear.[143] As the balanced neural activity returns, the static behavioral symptoms disappear. Concurrently, other sources of control of neural transmission through the vestibular nuclei (e.g., cerebellar input) are changing. The following examines this process in more detail (Fig. 8.11). This and the following summarize material from references.[141,144-150]

The physiological characteristics of primary afferent neurons on the remaining (intact) side do not appear to change after loss of the contralateral labyrinth, which had been considered as a possible mechanism subserving vestibular compensation.[151] However, it does appear that after such a contralateral loss there is a higher proportion of irregular afferents on the intact side: 34% post-lesion versus 22% pre-lesion, and the proportion of regular neurons similarly decreases.[151]

Within the medial vestibular nucleus, there are two types of neurons responding to horizontal (yaw) rotation. Both neuron types discharge spontaneously when the head is stationary. During yaw angular head accelerations in an ipsilateral direction, Type I neurons in the ipsilateral vestibular nucleus increase their firing, and these neurons decrease their firing for yaw head accelerations in the contralateral direction. These neurons receive direct input

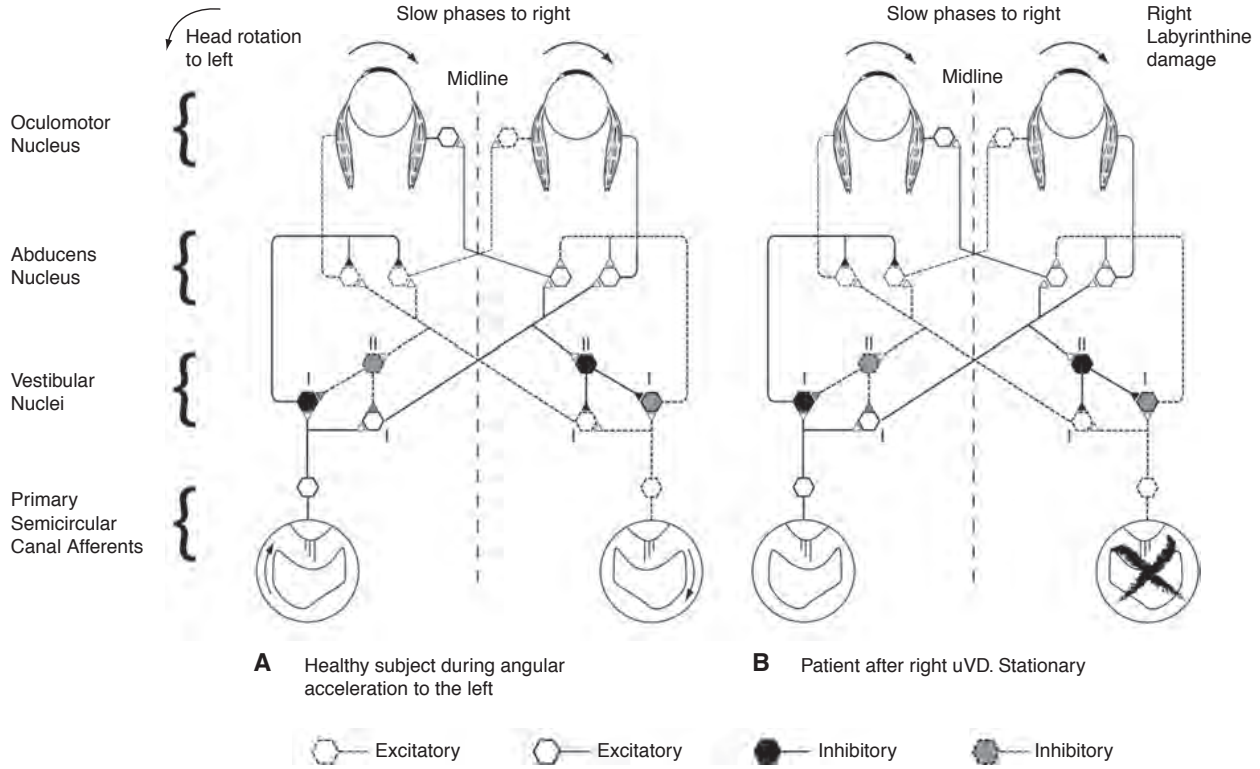

Figure 8.11 To show the neural basis of the nystagmus after UVD and how a UVD patient at rest **(B)** has vestibular activity closely similar to a person undergoing a rotation in the direction of the intact ear **(A)** (so head rotation left is equivalent to right UVD). The left panel shows the responses in some of the identified neural connections of the vestibulo-ocular pathways during a leftward yaw head acceleration. Excitatory neurons are shown as open hexagons; inhibitory neurons as closed hexagons. Activated neurons are shown as thick darker traces; neurons disfacilitated (or inhibited) are shown as light, dashed traces. The sequence is as follows: during ipsilateral acceleration, the primary neurons from the left semicircular canal are activated (continuous lines) while those from the right are inhibited (dashed lines). The excitatory input projects to the ipsilateral (left) vestibular nucleus and activates two types of neurons; first, excitatory Type I neurons, which project to contralateral abducens nucleus, excite those neurons, and so generate the slow compensatory eye movement response; and second, inhibitory Type I neurons in the left vestibular nucleus, which project to the ipsilateral abducens and so act to silence the ipsilateral abducens neurons during the slow phase. As a result of this perfectly complementary excitation and inhibition, there is a smooth, conjugate, slow-phase eye-movement response to the acceleration stimulus. Note that this response of the Type I neurons is *further* enhanced because the excitatory Type I neurons synapse on (inhibitory) Type II neurons in the contralateral vestibular nucleus, silencing their activity even further (even less than the disfacilitation received from the right horizontal semicircular canal). It is stressed that this is a very basic figure showing just one of the many neural circuits controlling vestibulo-ocular responses, some of which do not have the commissural connections shown here.

from the ipsilateral peripheral vestibular labyrinth. Other neurons within that same ipsilateral v estibular nucleus (Type II neurons) sho w precisely the opposite pattern of response: they increase their firing for contralaterally directed head accelerations and decrease it for ipsilateral head accelerations. These Type II neurons respond in such a mirror-image fashion because they are driven (indirectly) by cells from the other labyrinth on the other side of the head. These indirect connections cross the midline and are called commissural connections, and the y play a major role in vestibular compensation.[152] Type II neurons are inhibitory neurons and project their inhibition onto Type I neurons. Type I and Type II neurons work synergistically. During an acceleration, Type I neurons increase their firing, because they are receiving activation from the periphery and also *simultaneously* receiving reduced inhibition

from Type II neurons. This synergistic mechanism means there is enhanced sensitivity of Type I neurons during acceleration compared with the sensitivity of peripheral vestibular neurons. It is a "push-pull" system, and it functions to generate a fast, sensitive response.

Immediately after UVD, there are changes in the activity of both types of neurons on the lesioned side while the animal is absolutely stationary. The resting discharge rate of ipsilesional Type I neurons is substantially decreased, and the resting discharge of ipsilesional Type II neurons is substantially increased.[1] These Type II neurons thus exert even more inhibition than usual on the ipsilesional Type I neurons, presumably driving them to silence and so completely disinhibiting the Type I neurons on the intact side.

When the input from one labyrinth is removed by a UVD, Type I neurons in the ipsilesional nucleus are silenced, resulting in an imbalance in neural activity between the paired vestibular nuclei. Their silence relieves Type I neurons in the opposite ("contralesional") vestibular nucleus of inhibition, and so they can fire at a higher rate. As the Type I neurons on the lesioned side progressively start to fire again, the neurons in the intact side will receive progressively more inhibition, further enhancing the restoration of balanced neural activity. So, the decreased activity of ipsilesional Type I neurons reflects the loss of peripheral excitatory drive *and* the increased inhibition from the contralateral labyrinth mediated by the inhibitory Type II neurons.

How do these neural changes relate to the UVD syndrome and its recovery? The static motor symptoms and the perceptual illusions of vertigo and roll-tilt are likely caused by the imbalance in average neural activity between cells in the vestibular nuclei on each side of the brainstem. The deficits in dynamic responses are probably a result of the decreased dynamic sensitivity of vestibular nucleus neurons as documented by Markham et al (1977).[145]

It should be noted that, in general, the restoration of static equilibrium (i.e., static compensation) is remarkably robust: very little appears to hasten or hinder it. That robustness is in contrast to the restoration of dynamic equilibrium—dynamic compensation—which appears to depend at least in part on intact visual, vestibular, and proprioceptive sensory inputs. Dynamic compensation is usually incomplete. Tighilet et al (2007)[115] showed the clear dissociation between the recovery of spontaneous nystagmus as opposed to the recovery of postural compensation.

In the hours, days, and weeks that follow, there is a remarkable change in the activity of vestibular nucleus neurons. The discharge rates of both Type I and Type II neurons on the lesioned side return much closer to normal, even though the cells in the ipsilesional nucleus are no longer receiving any neural input from its labyrinth. Are these changes caused only by a neural process such as adaptation, or is there an "error signal" that triggers those changes? Does the removal of peripheral vestibular input trigger synaptic or membrane change that acts to restore the balance between the two nuclei? There is evidence for changes both at the synaptic and cell membrane level during compensation.[153-155]

How could the neural balance be restored? Adaptation is one possibility: that the firing rate of the more active contralesional Type I neurons reduces over time so these neurons exert less inhibition onto neurons on the ipsilesional side. Another is that the neurotransmitter receptors in the ipsilesional Type I neurons become less sensitive to the inhibitory transmitter released by the overactive inhibitory Type II neurons. Most interest has focused on GABA.[156-164] A reduction in the efficacy of the neurotransmitter receptor for GABA would allow for greater activity by ipsilesional Type I neurons. In this way, ipsilesional Type I neurons would start to return to normal levels of resting discharge, because they would not be affected by as much inhibition as before. This idea is referred to as "down regulation of GABA sensitivity" being responsible for the restoration of balanced activity in the two vestibular nuclei.

The UVD generates a cascade of neural changes—for example, astrocytes and microglia rapidly increase in the vestibular nucleus.[165,166] Over time, there are significant anatomical changes in the vestibular nuclei,[167-170] although it seems that the behavioral effects of compensation occur before sprouting.[171] Evidence from Campos-Torres et al (2005)[172] and Lacour et al (2009) [173] indicates that the effects of silencing peripheral neurons by chemical means (tetrodotoxin) as opposed to silencing by surgical sectioning of the nerve, are not identical. Tetrodotoxin does not cause degeneration, and Campos-Torres et al (2005) [172] found it causes no glial changes in the vestibular nuclei, as opposed to the dramatic increase in glia following sectioning or damage to the vestibular nerve. Such results suggest that the "error signal" may well be a chemical factor released by the injured neurons.

Recent studies have recorded the characteristics of neurons in slices of brainstem tissue taken from animals that have compensated for various durations after UVD. These studies record neurons either extracellularly or intracellularly from such brainstem slices. In this situation, it is not possible to identify Type I and Type II neurons, because there is no peripheral vestibular input. The cells are divided arbitrarily according to their membrane characteristics into Type A and Type B neurons, which have very different action potential profiles. Type A

neurons are tonically firing neurons; Type B neurons tend to be phasic. Researchers have reported that, after UVD, the membrane characteristics of Type B neurons appear to change.[174,175] These previously phasic neurons appear to take on much more tonic characteristics. There have been other reports of very long-lasting changes in intrinsic membrane characteristics after prolonged inhibition.[176,177] It is likely that membrane changes are responsible in part for the changes in global neural activity to UVD we have described above.

Angular versus Linear Acceleration

Otolithic neurons project to the lateral and descending vestibular nuclei but also send a branch to the cerebellum. Recordings from LVN neurons show that most neurons show an increase in firing for such ipsilateral roll-tilt stimuli.[35] Following a UVD, there is a decrease in the proportion of ipsilesional LVN neurons and a decrease in their average resting activity.[178,179] So, a similar imbalance occurs to natural linear accelerations as described above for natural angular accelerations. Again, that imbalance is mimicked by a removal of all the peripheral vestibular neural input from one side.

Cerebellum

The cerebellum plays an important role in vestibular compensation,[180] which has not been fully recognized because so much focus has been on the vestibular nucleus. Data on the effects of lesions of the cerebellum or its connections on vestibular compensation are contradictory. Whereas some cerebellar lesion studies show a marked delay in the resolution of spontaneous nystagmus, others show no effect.[180-182] Although bilateral occipital lobectomy has no effect on the resolution of spontaneous nystagmus, it does impede the recovery of the VOR to low-acceleration stimulation.[94] Lesions of the brainstem or transcerebellar vestibular commissures do not impede the vestibulospinal symptoms of static compensation, at least in mammals.[183,184] This suggests that input from the contralesional (intact) vestibular nucleus is not essential for static compensation. Kitahara et al have shown that neurons in the flocculus of the cerebellum play an important role in the early stages of vestibular compensation for static symptoms.[185,186] They have proposed that neurons in the flocculus may inhibit the hyperactive Type I neurons in the contralesional vestibular nucleus, and so act to reduce the overactive Type I neurons, which would in turn act to relieve inhibition on the ipsilesional Type I neurons.[187-189] Human patients with cerebellar lesions show slow compensation.[190]

Neural Network Models of Vestibular Function and Compensation

These physiological changes have been incorporated into neural network models of the VOR that model the possible mechanisms of the UVD syndrome and compensation.[191-195] Cartwright's physiologically realistic neural network model was trained on guinea pig eye-movement responses before and after UVD and showed that changes in activity in various stages through the neural network determine the responses.[192-195] The model pointed to neurons on the intact side as being especially important, because their gain changes most.

Summary

The vestibular system is a very fundamental sensory system whose activity influences many motor systems as well as generating perceptual experiences. So, disruption of vestibular function has far-reaching consequences. After UVD, some symptoms recover, but some do not. Over time, patients resume their lifestyle mainly, we think, because of this partial "patchwork" recovery and because they learn a variety of new behaviors to allow normal function. However, when the appropriate tests are carried out, probing purely vestibular function and preventing some of the other "strategies" that the patient may have learned, the permanent loss of many vestibular functions is very clear.

Taking such patients into a clinic and trying to assess the mechanism of their recovery, based on a few classical measures of oculomotor performance using unnaturally low values of head acceleration and restricting the patient's use of other strategies (e.g., excluding blinks during HIT testing), may not reveal the subtle but effective compensatory strategies that patients actually use in real life. It was only by using high-speed, high-resolution measures of eye movements that we detected the very small, but very effective, covert saccades during the passive high-acceleration head movements, and it is these covert saccades that we now think may hold one of the keys to understanding compensation of oculomotor symptoms. Saccades are the new frontier in vestibular compensation, and the major question now is what triggers them.

Substitution of other responses can effectively conceal the vestibular deficit and so protect the patient from receiving smeared retinal images during head movements. The permanent deficit of dynamic VOR function to natural accelerations after UVD shows that vestibular rehabilitation of UVD patients should be aimed at developing new saccadic behavior that substitutes for the dynamic vestibular loss, rather than trying to restore something that cannot be restored.

The Final Question—Are They Happy?

Although vestibular compensation appears to be a simple recovery of function, there is overwhelming evidence that there are many different processes taking place during vestibular compensation. It is also clear that the various processes recover at different rates, or in some cases, do not recover at all, while new behaviors are being learned to substitute for the lost vestibular function. Many patients do learn these new behaviors, and have a satisfactory recovery—they compensate. But some patients, for reasons that we do not yet understand, do not appear to learn the successful behaviors and are unhappy. The challenge is to identify how these poorly compensated patients can be taught to improve their lifestyle. With our colleagues, we are seeking to teach such patients strategies that will help them.

Acknowledgments

The revision of this chapter is supported by theAustralian National Health and Medical Research Council (632746) and the Garnett Passe and Rodney Williams Memorial Foundation. We thank Ann Burgess for her meticulous proofreading of this manuscript.

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CHAPTER 9

Compensatory Strategies for Vestibulo-ocular Hypofunction

Michael C. Schubert, PT, PhD

Normal Vestibulo-ocular Reflex

Normal activities of daily life (such as running) can be associated with head velocities of up to 550 degrees per second (deg/sec), head accelerations of up to 6,000 de g/sec^2, and frequency content of head motion from 1 to 20 Hz. [1,2] Only the vestibular system can detect head motion over this range of velocities, accelerations, and frequencies. Additionally, the latency of the vestibulo-ocular reflex (VOR) has been reported to be as short as 5 to 7 msec.[3,4] In contrast, ocular following mechanisms, such as smooth pursuit, generate slower eye velocities (~60 deg/sec), have relatively long latencies (up to 100 msec), [5,6] and fail at frequencies exceeding 1 Hz.[7] For subjects with vestibular loss that has not been compensated adequately, head movements can significantly limit participation in activities of daily life.

Abnormal Vestibulo-ocular Reflex

People who sustain vestibular damage typically experience vertigo, disequilibrium, head motion–induced oscillopsia, spontaneous nystagmus, and postural instability.[8-11] In particular, when a subject with loss of vestibular function makes rapid head rotations in the direction of the damaged labyrinth, the eyes do not remain fixated on a visual target, reflecting a deficient VOR. As a result, visual acuity during head rotation is degraded.[12,13] As a form of compensation, the brain can generate surrogate eye rotations that compensate for the VOR.[14,15]

Compensatory Strategies

Individuals with vestibular loss use different compensatory strategies to improve their ability to see clearly during a head rotation.[16] Compensatory mechanisms include substitution or modification of a saccade, increased gain of the cervico-ocular reflex (COR), the use of a centrally preprogrammed eye movement, and, perhaps, enhancement of the smooth pursuit system.

The substitution of a saccade in the direction of the deficient VOR (compensatory saccades [CS]), also known as covert saccade because of their occurrence during the head rotation and thus not visible to the clinician's eye during ipsilesional whole-body and head-only rotations, has been identified in persons with loss of vestibular function.[14,15] Recently, it has been reported that these saccades are unique in that they occur with reduced latencies of 50 to 150 msec from the onset of a head rotation (Fig. 9.1),[15,17] occur during both predictable and unpredictable head movements,[14,17] and have been hypothesized to be of vestibular origin.[14]

It appears that CS also function to improve gaze stability associated with ipsilesional head rotations. The influence of the PPS (compensatory saccade) in reducing gaze instability was initially reported in 1988. Segal and Katsarkas[18] studied the VOR using unpredictable whole-body rotations in three persons with unilateral vestibular hypofunction (UVH) as a result of an eighth cranial neurectomy for removal of a vestibular schwannoma. The investigators

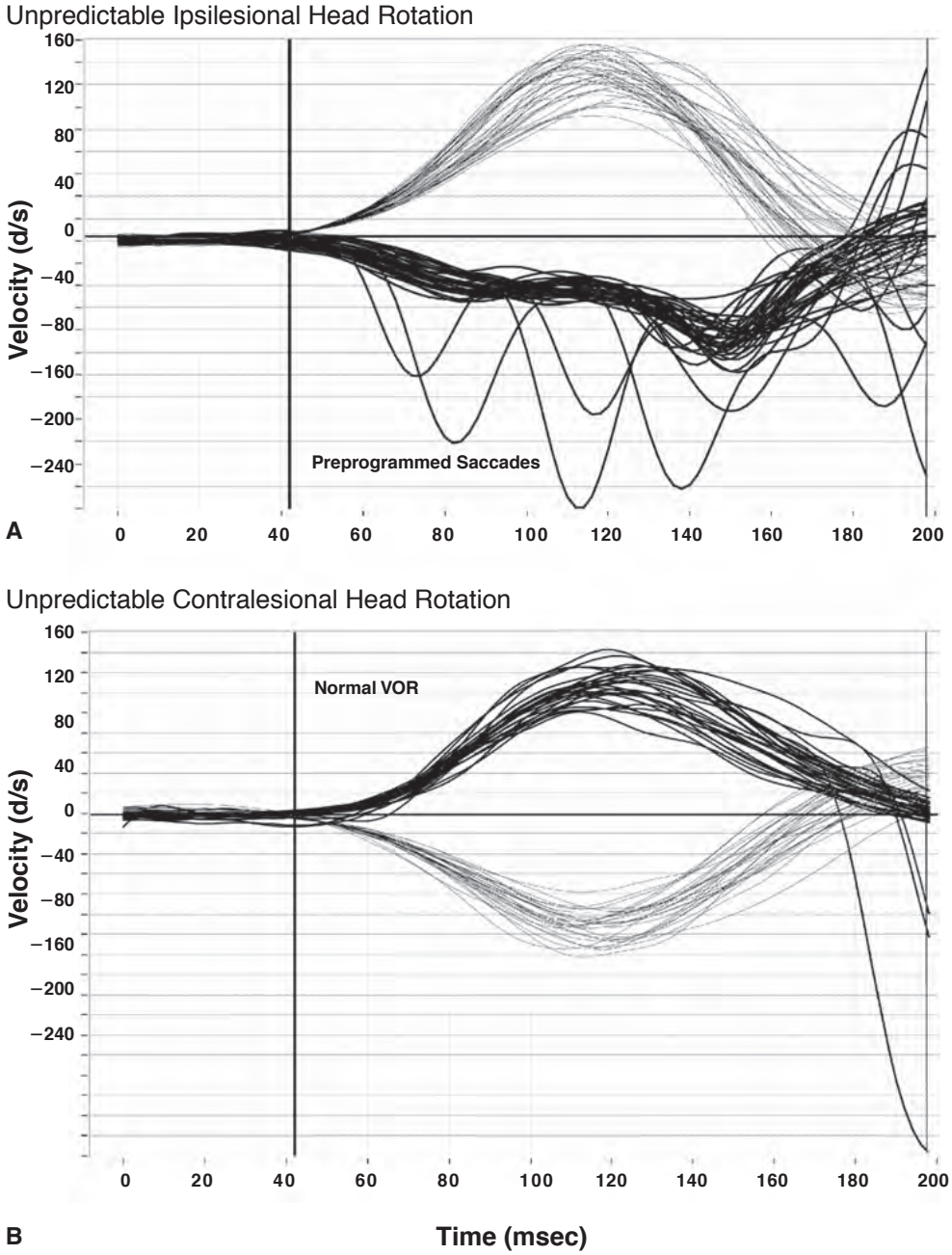

Figure 9.1 Compensatory (preprogrammed) saccades vs. normal vestibular-ocular reflex (VOR) in a person with unilateral vestibular hypofunction (UVH). *Dark traces r*eflect head velocity, and *lighter traces* eye velocity. The *vertical line* marks the onset of the head rotation. **(A)** The slow eye velocity is deficient in relation to the head velocity (low VOR gain), and compensatory saccades (CS) are recruited. Note that the direction of the CS is the same as that of the slow velocity. **(B)** The slow eye velocity is similar to the head velocity for a normal VOR gain.

described a type of saccade that corrected for 28% to 59% of the slow-component error associated with ipsilesional head thrusts. Later, Bloomberg and colleagues[19] reported a similar synergistic relationship between slow-component vestibular and saccadic eye movements for the purpose of improving gaze stability. More recently, we have shown that the gaze position error was reduced when compensatory saccades were recruited as part of the gaze-stabilizing strategy.[20]

The otolith-mediated translational VOR (tVOR) has also been shown to benefit from the symbiotic relationship between VOR and saccades. Shelhamer and associates[21] adaptively increased the gain of the tVOR in human

subjects (with normal vestibular function) by exposing them to lateral sinusoidal translations and target motion for 20 minutes. These researchers showed that saccades occurred in the direction of the slow eye movements and accounted for 32% of the tVOR after adaptation. On the basis of these previous studies, saccades not only have been shown to occur in the direction of the vestibular slow component but also appear to require little time for their recruitment and utility in reducing gaze instability.

Saccadic Modifications

A variety of behavioral modifications to the saccadic oculomotor system have been reported for patients with vestibular hypofunction when the experimental paradigms have involved a head rotation (a condition that normally requires a VOR). Three are described here.

Kasai and Zee[16] reported that when patients with bilateral vestibular deficits were asked to shift their gaze to a target with the eyes and head, they were found to generate an initial saccade to the target of decreased amplitude (undershoot). The researchers hypothesized that the brain intentionally undershoots the target, anticipating the inability of the VOR to keep the eyes still during the head rotation. Through the use of a saccade of insufficient amplitude, the eyes then "drift" to the target with the head motion.

Several studies have established that during an ipsilesional unpredictable yaw head rotation away from a centrally positioned visual target, a saccade is generated in the opposite direction to the head rotation, back toward the target.[14,18-19,21-24] Most of the investigators agree that these types of compensatory saccades:

- Are recruited for the purpose of assisting gaze.
- Improve gaze stability, although they do not completely match the velocity of head motion.
- Are inversely correlated with VOR gain.[14,18,19,24,25]
- May occur more than once during a single head rotation.[14,21,23-25]

The studies also demonstrate a great deal of variability in recruitment of CS. Some patients appear to use CS only for high-acceleration or large-amplitude head motions.[14,16,19,23-26]

It has also been shown that the latency to generate a CS during an ipsilesional unpredictable yaw head rotation away from a centrally located visual target is much shorter than visually guided saccade latencies (70 to 130 msec versus 200 msec, respectively).[14]

Cervico-ocular Reflex

The COR parallels the VOR and is thought to contribute a slow-component eye rotation in the direction opposite to head rotation in the place of the deficient vestibular system. The difference, however, is that the eye rotation from the COR is generated from receptors in the joints and ligaments of the upper cervical vertebrae.[27] For eye movements to be discerned as being generated from the cervical spine and not the vestibular system, the head must remain still. The COR is quantified from the ratio between eye and trunk velocities (absolute value of peak eye velocity ÷ peak trunk velocity). As a compensatory mechanism for vestibular loss, the COR should move the eyes in the direction of the moving trunk. When the trunk is rotated to the left under a still head, the relative head position (static) is the same as if the head had rotated to the right. A leftward trunk rotation (relative rightward static head position) should therefore elicit a leftward eye motion. Although the COR is well described in animal models, its presence in humans is controversial.

In 1906, Barany first demonstrated eye motion in rascally rabbits when the trunk was rotated beneath a still head.[28] In later studies, various researchers demonstrated that sectioning the dorsal roots from C1 to C4 or injecting anesthetic into the cervical joints of rabbits produced nystagmus.[29,30] Recording from cat abducens motor neurons, Hikosaka and Maeda[27] showed in 1973 that stimulating the second or third cervical dorsal roots caused facilitation of the ipsilateral abducens motor neurons and increased firing rate from the contralateral vestibular nucleus. Additionally, contralateral abducens motor neurons were inhibited. Stimulation below the fourth cervical joints did not generate the same facilitation and inhibition responses. These researchers demonstrated synapses between second and third cervical vertebral joints and abducens oculomotor neurons and vestibular nuclei. As a form of control, the second or third cervical joints were anesthetized with lidocaine injection, after which the nystagmus could not be elicited.

A later study by Gdowski and McCrea,[31] in the squirrel monkey, yielded further evidence of the influence of neck proprioceptive information on the vestibular nuclei. The investigators compared firing rates (sensitivity) recorded from secondary afferents of the horizontal semicircular canals while the primate's trunk was rotated beneath a still head (passive neck rotation) with those during whole-body rotation (each rotation parameter was measured at 0.5 Hz, 40 deg/sec and at 2.3 Hz, 20 deg/sec). They reported the gain of the COR was similar regardless of frequency of the test (0.4 ± 0.04 for 0.5 Hz and 0.33 ± 0.05 for 2.3 Hz). This study showed that neurons in the vestibular nuclei are sensitive to neck rotation in primates.

Similar COR gain values were recently reported in rhesus monkey.[32] Yakushin et al[32] studied COR and VOR gain recovery across frequencies of motion ranging

from 0.02 to 6 Hz in monkeys with all six semicircular canals plugged. Before surgery, the measured COR gains were negligible and only observed at those frequencies below 1 Hz. After surgery, the COR gains were significantly increased (~0.15) when measured at frequencies below 3 Hz. Over the next 3 months, the COR gains increased to ~0.4. When the COR and VOR were summed, the compensated response gains were ~0.4 to 0.8.

Recording from rhesus monkey neurons in the medial and lateral vestibular nuclei after bilateral vestibular labyrinthectomy, Sadeghi et al reported nearly 50% of the vestibular neurons responded to passive stimulation of the neck. This correlated with an increase in the COR gain response (~0.25 gain).[33]

The identification of a COR as a means of gaze stabilization in humans is controversial. Part of the controversy is because of the methods used to study the COR, and in part to the great variability in reported function of the COR. See Chapter 31 for a review of cervicogenic dizziness.

Subjects with Vestibular Hypofunction

The majority of studies that have investigated the role of the COR as a compensatory strategy have done so in patients with bilateral vestibular hypofunction (BVH).[16,22,34-37] Kasai and Zee[16] first reported that COR gain varied from 0.27 to 0.5 in three subjects with complete loss of vestibular function. Others have reported similar COR gain values.[22,34-37] Of particular interest was a case study involving a subject in whom complete loss of vestibular function was diagnosed from caloric irrigation.[34] The investigators reported that the COR gain decreased from 0.51 to 0.17 as the central vestibular system demonstrated recovery of the VOR gain (both VOR and COR measured at 0.2 Hz, 40 deg/sec). This finding implies that the COR was a compensatory mechanism transiently useful until the gain from the VOR recovered.

Evidence for the existence of a COR in a person with UVH was recently reported in an 81-year-old woman.[38] The authors described a COR gain of 0.1 ± 0.04 during 0.3-Hz trunk rotation. In this patient, the COR gain was increased to 0.32 ± 0.13 after 5 weeks of gaze stabilization exercises (Fig. 9.2). Oddly, the direction of the COR was not compensatory. This evidence suggests that the COR can be potentiated, although its functional relevance is still uncertain.

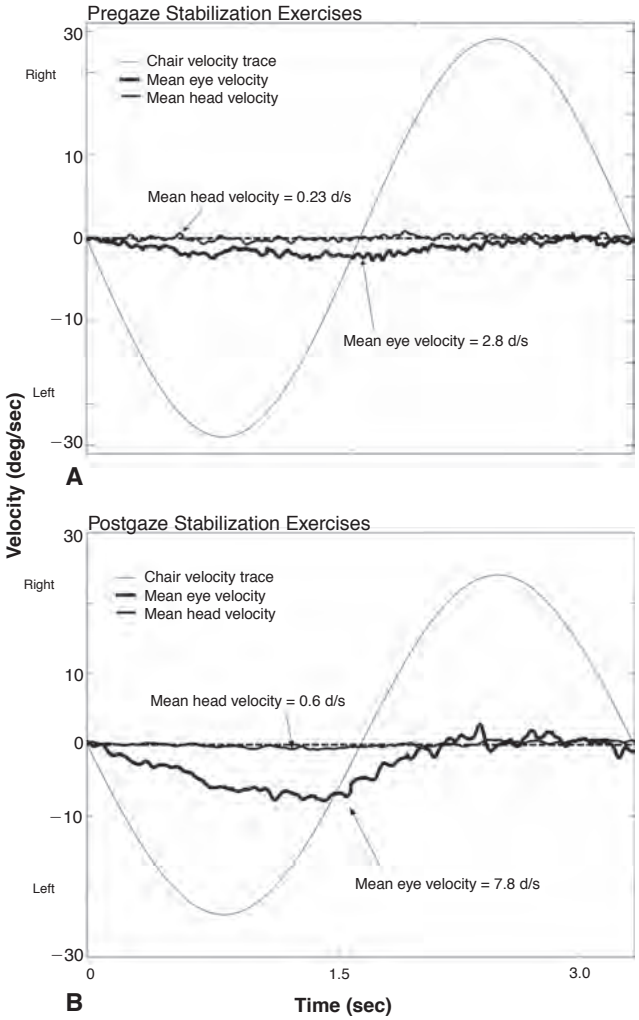

Figure 9.2 Cervico-ocular reflex (COR) in a patient with unilateral vestibular hypofunction (UVH). The chair, eye, and head velocities are plotted. The subject's head was fixed in space to prevent head motion. **(A)** The COR gain (eye velocity/chair velocity) was at 0.1 before the patient started a gaze stabilization exercise program. **(B)** After the program, the COR gain had increased to 0.32. Note the very low head velocity, demonstrating that this eye velocity is not from the vestibular system.

Effects of Prediction

When individuals with vestibular hypofunction can predict head movement, gaze stability improves. This process has been shown in two ways. First, VOR gain (eye velocity ÷ head velocity) is larger during predictable head movements toward the defect than during unpredictable head movements toward the defect in individuals with vestibular hypofunction.[15,17,39-43] Second, visual acuity during head motion (dynamic visual acuity [DVA]) is better during predictable head rotations than during unpredictable head rotations.[12,44] Enhancement of VOR gain (gaze stability) and DVA with predictable head movement is believed to be caused by mechanisms such as central preprogramming and efference copy of the motor command.

We have found evidence that individuals with vestibular hypofunction can generate a high-velocity slow

phase (HVSP) for ipsilesional predictable head rotations (Fig. 9.3*A*).[15] These unique eye rotations occur with velocities greater than the velocity limit imposed by inhibitory cutoff (at 100 deg/sec). Additionally, we have evidence that the HVSPs occasionally occur before the onset of the head rotation and, therefore, are not a vestibular-generated eye rotation (Fig. 9.3*B*).

Recently, new evidence has been reported suggesting the brain can generate a very short-latency eye rotation.[45] In guinea pig with bilateral loss of vestibular function (streptomycin injection) allowed to make active head rotations, the mean anticipatory slow-phase eye motion latency was 0.1 +/ 2.5 msec. This latency is much shorter than the latency observed in either a healthy guinea pig (~7 msec) or one with a unilateral vestibular loss (~ ≥15 msec).

Recording from neurons in the medial and lateral vestibular nuclei in the rhesus monkey after bilateral vestibular labyrinthectomy, Sadeghi et al confirmed that neuronal sensitivity to active head rotation (compared with passive head rotation) was increased, verifying the role that efference copy has as a substitute for absent vestibular information.[33]

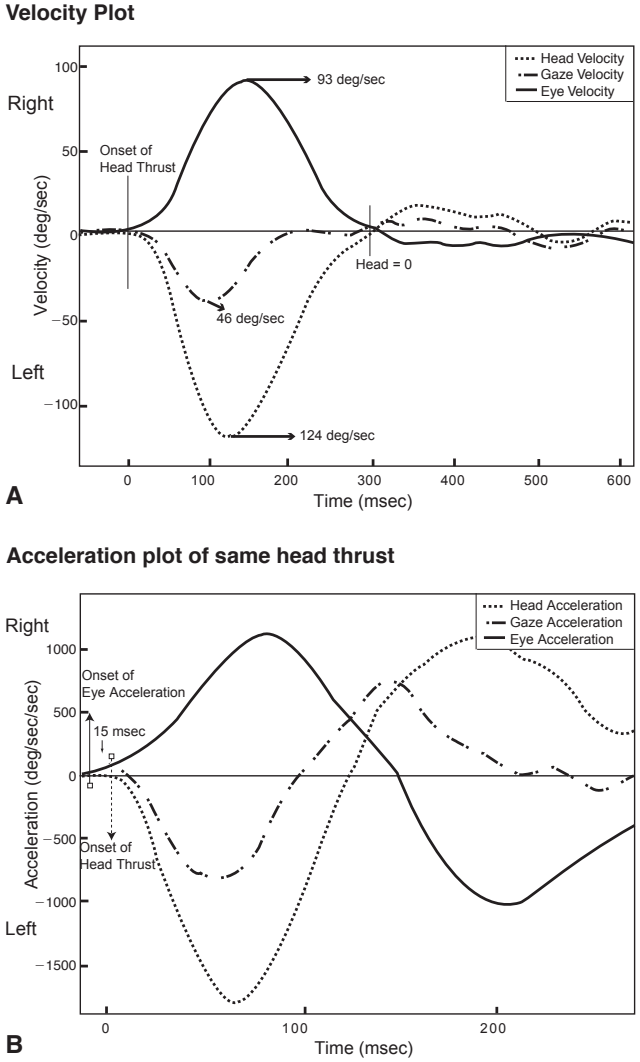

Figure 9.3 Velocity and acceleration plots of a high-velocity slow phase during a predictable head thrust in a subject with bilateral vestibular hypofunction (BVH). **(A)** The head is thrust to the left at 124 deg/sec, above the level of inhibitory cutoff, yet the subject generates a slow eye velocity of 93 deg/sec, for a gain of 0.74. **(B)** The acceleration plot reveals that the slow eye velocity precedes the head rotation, demonstrating that this type of eye response is not generated from the vestibular system.

Enhanced Smooth Pursuit

Smooth pursuit eye movements are used to track a moving visual target, typically without head rotation. Smooth pursuit gain is the ratio of eye velocity to target velocity (eye velocity ÷ target velocity). Like the vestibular system, smooth pursuit is highly modifiable.[46,47] Generally, smooth pursuit functions best at frequencies within 1 to 2 Hz[7] and at velocities within 60 deg/sec. However, for motivated individuals, smooth pursuit has been demonstrated to function well at velocities of 90 deg/sec.[48]

Subjects with vestibular hypofunction may be able to use smooth pursuit as a means of substitution for the deficient VOR, although limited data are available to support this possibility. In a study by Bockisch and colleagues,[49] subjects with BVH were reported to have smooth pursuit gains that were, on average, 9% greater than those in healthy controls. The researchers also documented that these patient subjects were able to generate higher smooth pursuit velocities (peak velocity 40 deg/sec). They concluded that smooth pursuit may be a useful compensatory mechanism for a deficient VOR. More research regarding smooth pursuit and VOR interaction is needed to discern whether smooth pursuit may be a useful oculomotor mechanism to adapt for people with vestibular hypofunction.

Optokinetic Reflex

The optokinetic reflex (OKR) is generated from central oculomotor pathways and involves a combined saccade and smooth pursuit eye rotation. It is a normal response that typically occurs when an individual follows a moving object that takes up at least 60% of his or her visual field. There is recent evidence that the OKR might be modifiable and able to contribute to gaze stability during head rotation. Tilted mice lack otoconia because of a genetic mutation. Interestingly, these mice have higher OKR gain compared with control mice. The authors presumed this is caused by an adaptation strategy between the otolith and visual systems to improve gaze stability.[50]

Summary

Evidence strongly supports the notion that people with vestibular hypofunction use different strategies for gaze stabilization. In time, we hope that these data will reveal methods that enable rehabilitation to be customized to ward the unique adaptive strategies and capacities of each patient.

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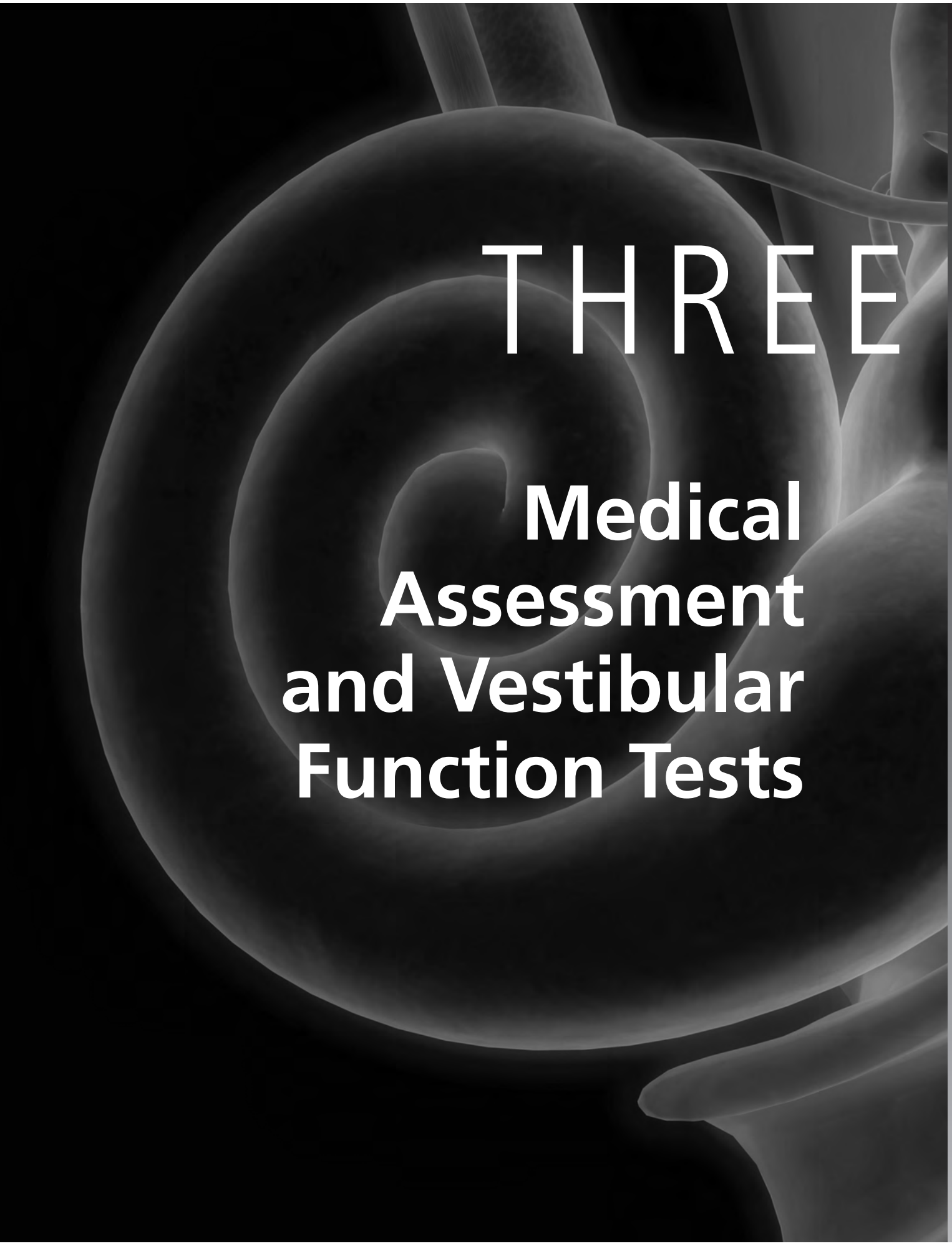

THREE

Medical Assessment and Vestibular Function Tests

CHAPTER 10

History and Clinical Examination

Ronald J. Tusa, MD, PhD

Management of the dizzy patient depends on history, bedside clinical examination, and laboratory testing. This chapter covers the first two portions of this evaluation. An accurate history is needed to determine the onset of the problem, description of the symptoms, and, most important, how the symptoms affect the individual's lifestyle. This last element is crucial to obtain because some individuals may have bedside clinical and laboratory evidence of chronic vestibular loss on one side but may be primarily affected by some other cause of dizziness, such as migraine or anxiety. The bedside clinical examination can be used to distinguish peripheral from central vestibular problems, the extent of loss, and how acute the problem may be. Laboratory testing (see Chapter 11) confirms the provisional diagnosis that was based on history and clinical findings, quantifies the degree of loss, provides evidence of central compensation, and shows evidence of an aphysiological component.

History

The history is by far the most important part of the evaluation. Unfortunately, taking a good history from the start can be extremely tedious, because the patient's complaints are often vague and also can be complicated by anxiety-provoked symptoms. For this reason, I divide the history into those elements that help with the diagnosis and those that lead to goals for management, including physical therapy.

Elements that Help with the Diagnosis

The tempo, symptoms, and circumstances of the patient's primary complaints are the three key items in the history (Table 10-1).

Tempo

The objective of establishing tempo is to determine whether the patient has an acute attack of dizziness (within 3 days or less), chronic dizziness (more than 3 days), or spells of dizziness. To do so, the clinician must be sure the patient describes the *first* onset of the symptoms. Did it happen suddenly, or did it develop very slowly? Was it provoked by anything, or did it occur spontaneously? Did the patient have a cold or some other illness around that time? If the patient suffers from spells, the clinician should try to determine the average duration of the spells in seconds, minutes, or hours. It is important to have the patient describe in detail the first spell, the most severe spell, or the last spell that he or she can clearly recall.

Symptoms

What the patient means by "dizziness" should be expanded on. *Dizziness* is an imprecise term used to describe a variety of symptoms, each of which has a different pathophysiological mechanism and significance (Table 10-2). If the patient cannot describe the symptoms, the clinician should ask whether the symptom causes problems primarily in the head or with balance. It is important to determine if the

■ Table 10-1 KEY ITEMS IN THE HISTORY OF THE DIZZY PATIENT

Disorder	Tempo	Symptoms	Circumstances
Vestibular neuritis	Acute dizziness	Vertigo, disequilibrium, nausea and vomiting, oscillopsia	Spontaneous, exacerbated by head movements
Labyrinthitis	Acute dizziness	Vertigo, disequilibrium, nausea and vomiting, oscillopsia, hearing loss and tinnitus	Spontaneous, exacerbated by head movements
Wallenberg's infarct	Acute dizziness	Vertigo, disequilibrium, nausea and vomiting, tilt, lateropulsion, ataxia, crossed sensory loss, oscillopsia	Spontaneous, exacerbated by head movements
Bilateral vestibular deficit or >7 days from a unilateral vestibular defect	Chronic dizziness	Dizziness, disequilibrium, occasionally oscillopsia	Induced by head movements, walking. Exacerbated when walking in the dark or on uneven surfaces
Mal de débarquement	Chronic dizziness	Rocking or swaying as if on a boat	Spontaneous while lying or sitting Rarely occurs while in motion
Oscillopsia	Chronic dizziness	Subjective illusion of visual motion	Spontaneous with eyes open
Anxiety/depression	Chronic dizziness	Lightheadedness, floating, or rocking	Induced by eye movements with head still
Benign paroxysmal positional vertigo	Spells: seconds	Vertigo, lightheadedness, nausea	Positional: lying down, sitting up or turning over in bed, bending forward
Orthostatic hypotension	Spells: seconds	Lightheadedness	Positional: standing up
Transient ischemic attacks	Spells: minutes	Vertigo, lightheadedness, disequilibrium	Spontaneous
Migraine	Spells: minutes	Vertigo, dizziness, motion sickness	Usually movement-induced
Panic attack	Spells: minutes	Dizziness, nausea, diaphoresis, fear, palpitations, paresthesias	Spontaneous or situation
Motion sickness	Spells: hours	Nausea, diaphoresis, dizziness	Movement induced, usually visuovestibular mismatch
Ménière's disease	Spells: hours	Vertigo, disequilibrium, ear fullness from hearing loss and tinnitus	Spontaneous, exacerbated by head movements

■ Table 10-2 SYMPTOMS OF DIZZINESS

Symptoms	Mechanism
Disequilibrium: imbalance or unsteadiness while standing or walking	Loss of vestibulospinal, proprioception, visual, and/or motor function, joint pain or instability, and psychological factors
Lightheadedness or presyncope	Decreased blood flow to the brain
Sense of rocking or swaying as if on a ship (mal de débarquement)	Vestibular system adapts to continuous, passive motion and must readapt once environment is stable Anxiety
Motion sickness	Visuovestibular mismatch
Nausea and vomiting	Stimulation of medulla
Oscillopsia: illusion of visual motion	Spontaneous: acquired nystagmus Head-induced: severe, bilateral loss of the vestibulo-ocular reflex
Floating, swimming, rocking, and spinning inside of head (psychologically induced)	Anxiety, depression, and somatoform disorders
Vertical diplopia	Skew eye deviation
Vertigo: rotation, linear movement, or tilt	Imbalance of tonic neural activity to vestibular cerebral cortex

symptoms have changed since the onset of the problem. If the patient has spells, have him or her describe in detail the initial spell and the last severe spell.

Disequilibrium

Disequilibrium is imbalance or unsteadiness while standing or walking. It is caused by a variety of factors, including diminished or double vision, loss of vestibular function, defects in proprioception from peripheral neuropathy or spinal cord lesions, defects in motor function from central nervous or peripheral nervous system abnormalities, joint pain, and psychological factors.

Lightheadedness

Lightheadedness, or presyncope, is usually related to momentarily decreased blood flow to the brain. Patients with anxiety or depression also commonly use the symptom of lightheadedness to describe their dizziness.

Sense of Rocking or Swaying as if on a Ship

Patients can find the sensation of *rocking or swaying*, as if on a ship, very disturbing. It frequently occurs for a few days after a prolonged sea or air voyage and persists for a few days. Sometimes, it persists for months to years and is then referred to as *mal de débarquement*.[1] The reason for prolonged symptoms in these cases is unknown. No consistent abnormalities are found on magnetic resonance imaging (MRI) or electronystagmography (ENG) testing.[1,2] When symptoms are prolonged, patients are extremely bothered to the point that their lifestyle is disrupted. Such patients are very difficult to manage. They usually feel better when in motion, and I encourage such patients to engage in physical activity. Small doses of anti-anxiety medication are sometimes helpful (see Chapters 18 and 29 for further discussion).

Motion Sickness

Motion sickness consists of episodic dizziness, tiredness, pallor, diaphoresis, salivation, nausea, and, occasionally, vomiting induced by passive locomotion (e.g., riding in a car) or motion in the visual surround while standing still (e.g., viewing a rotating optokinetic stimulus). Motion sickness is believed to be caused by a sensory mismatch between visual and vestibular cues.[2] Patients with migraine disorder are particularly prone to motion sickness, especially during childhood. Twenty-six percent to 60% of patients with migraine have a history of severe motion sickness, compared with 8% to 24% of people in the

normal population[3,4] (see Chapter 15 for further discussion). The cause for this relationship is not clear Symptoms of increased motion sensitivity are often reproduced when the patient is exposed to a moving full-field visual target.

Nausea and Vomiting

Nausea with or without vomiting is a result of stimulation of the solitary and vagus centers in the medulla. In peripheral vestibular lesions, these symptoms are usually mild to moderate and in proportion to the degree of vertigo: in benign paroxysmal positional vertigo (BPPV), nausea is usually mild and vomiting is rare; in labyrinthitis and vestibular neuritis, nausea is moderate and vomiting may occur during rapid head movement. Severity of symptoms varies in central lesions according to the site of lesion. For pontine strokes (e.g., anterior inferior cerebellar artery [AICA] syndromes), the degree of nausea and vomiting is similar to that in peripheral vestibular defects. For dorsal medulla strokes (e.g., posterior inferior cerebellar artery [PICA] syndromes), nausea and vomiting are extreme and out of proportion to the level of vertigo.[5] For all other central vestibular structures (cerebellar, fourth ventricle floor, interstitial nucleus of Cajal, thalamus, and vestibular cortical lesions), nausea and vomiting are usually mild or absent.

Oscillopsia

Oscillopsia is the subjective illusion of visual motion. It differs from vertigo, in that oscillopsia occurs only with the eyes open, whereas vertigo occurs with the eyes open or closed. Patients occasionally interpret oscillopsia as "dizziness." There are two types of oscillopsia: (1) Spontaneous oscillopsia is caused by acquired nystagmus and is a result of apparent motion of the visual scene caused by movement of the retina (*retinal slip*); (2) Head movement–induced oscillopsia occurs in patients with severe, bilateral loss of the vestibular-ocular reflex (VOR), which is frequently experienced after ototoxicity caused by aminoglycosides. This form of oscillopsia occurs only during head movements and is caused by the lack of the gaze-stabilizing features of the VOR.

Floating, Swimming, and Spinning inside the Head (Psychological Symptoms)

Sensations of floating, swimming, or swimming "inside the head" are frequently the symptoms of anxiety (panic attacks, agoraphobia, obsessive-compulsive disorder), somatoform disorders (including conversion), or depression (see Chapter 18 for further discussion).

Vertical Diplopia

Vertical diplopia is double vision in which the two images line up vertically. The diplopia is not present if either eye is covered. Vertical diplopia is commonly a result of a skew eye deviation from peripheral or central otolith dysfunction.

Vertigo

Vertigo is the illusion of movement of the self or the environment caused by sudden imbalance of tonic neural activity in the vestibulocortical pathway (labyrinth–eighth nerve–vestibular nucleus–vestibular thalamus–vestibular cortex). It is due either to normal head movements (physiological), lesions that cause loss of function (ablation) of vestibular pathways on one side (e.g., vestibular neuritis), or mechanical problems of the inner ear (e.g., BPPV).The direction of vertigo depends on the structures involved. Rotational vertigo in the horizontal plane is caused by horizontal semicircular canal (SCC) dysfunction, which commonly occurs from labyrinthine (e.g., labyrinthitis or Ménière's) or eighth nerve dysfunction (vestibular neuritis). Rotational vertigo in the torsional plane (clockwise or counterclockwise direction) is caused by anterior and posterior SCC dysfunction on one side from a central lesion in the dorsal medulla. Tilt—lateral translation or lateropulsion—is caused by utricle dysfunction, which can be caused by lesions in the labyrinth or eighth nerve, but more commonly occurs from central defects. Lesions in central vestibular pathways may cause nystagmus, skew eye deviation, and lateropulsion but rarely cause rotational vertigo.

Circumstance

The clinician must determine the circumstances in which the patient's dizziness occurs. Dizziness may be provoked only by certain movements, such as standing up after lying down for at least 10 minutes (orthostatic hypotension) or vertical or oblique head movements (lying down, turning over in bed, or sitting up, BPPV). If eye movements without head movement cause dizziness, and there is no eye movement disorder (such as ocular misalignment or an internuclear ophthalmoparesis), the symptom is not likely to be caused by a vestibular or neurological problem. When dizziness occurs without provocation (spontaneous), and it is vestibular in origin, it is commonly exacerbated by head movements.

International Classification of Vestibular Symptoms

In an attempt to achieve an international consensus of vestibular disorders, a committee of the Barany Society met and published their first document. Their first paper classified vestibular symptoms.[6] Table 10-3 shows a modification of their diagrams based on the concepts we use at Emory. The main symptoms include dizziness, vertigo,

■ Table 10-3 CLASSIFICATION OF VESTIBULAR SYMPTOMS*

Classification	Spontaneous vs Triggered	How Triggered	Symptom
Dizziness	Spontaneous		Spinning or non-spinning
	Triggered	Positional	Spinning or non-spinning
	Triggered	Head-motion	Spinning or non-spinning
	Triggered	Visual-induced	Spinning or non-spinning
	Triggered	Sound-induced	Spinning or non-spinning
	Triggered	Valsalva-induced	Spinning or non-spinning
	Triggered	Orthostatic	Spinning or non-spinning
	Triggered	Other triggered	Spinning or non-spinning
Vestibulo-visual	Spontaneous		External vertigo
	Spontaneous		Visual tilt
	Spontaneous or triggered	Head still vs head moving	Oscillopsia
	Triggered	Head moved	Visual lag
	Triggered	Head moved	Movement-induced blur
Postural symptoms	Triggered	Upright	Unsteadiness
	Triggered	Upright	Directional pulsion
	Triggered	Upright	Balance-related near fall
	Triggered	Upright	Balance-related fall

*(Adapted from Bisdorff et al, 2009.[6])

vestibular-visual symptoms, and postural symptoms. Each of these may occur spontaneously or may be triggered. Readers interested in the details of the classification from that publication should refer to the original article.

Other Helpful Elements in the History

How the Dizziness Affects the Patient's Life

One of the most useful questions to ask in order to determine appropriate management is "How does the dizziness affect your life?" Three patients with an incomplete peripheral vestibular loss from vestibular neuritis on one side may give three different responses. The first patient may state that he is not affected at all by the dizziness, but he just wants to be reassured that it is nothing seriously wrong; this response would not require extensive evaluation and management. A second patient may state that she has no unsteadiness while walking but can no longer play golf or tennis because of her balance; this patient may require only a high-level physical therapy exercise program. A third patient may state that he is completely devastated by the dizziness and will not leave the house, drive, or participate in any social activities. This patient requires extensive counseling and physical therapy by the physician and physical therapist. He may also need medication and psychological counseling to better cope with the symptom.

Medications

The clinician must make sure to obtain a complete list of all of a patient's prescription and over-the-counter medications. A number of drugs can cause dizziness, some of which also are used to treat dizziness (see Chapter 14). Some over-the-counter medications, such as diphenhydramine and meclizine, also cause dizziness.

What the Patient Believes Is Causing the Dizziness

Another overlooked question that should be asked of a patient with dizziness is "What do you think is causing your dizziness?" Sometimes the patient has a specific concern that may not be addressed routinely by the health care provider. Unless this concern is addressed, the patient may leave the clinic unsatisfied with the visit.

Elements that Lead to Goals for Management, Including Physical Therapy

Subjective Complaints

Two components of the history and examination are helpful in developing treatment goals. The first is obtaining a list of the patient's subjective complaints. One method of doing so is to present a written list of symptoms that the patient can simply check off (see Item 1 of the Patient Questionnaire in Appendix A at the back of the book). The second is to quantify the intensity of specific symptoms, for instance, by using a *Visual Analogue Scale* (VAS). An example would be the head movement VAS shown in Figure 10.1. It consists of a 10-cm line anchored with words on both ends. The person rates the symptom intensity while sitting quietly and then while actually performing a task. The results are expressed as the difference between the baseline measure and the measure after performing the task. In the example (see Fig. 10.1), the results are the intensity of dizziness, as measured on a 10-cm line, while sitting quietly and then after 1 minute of horizontal head movements at a 1-Hz frequency. Separate papers containing the line are used for each measure.

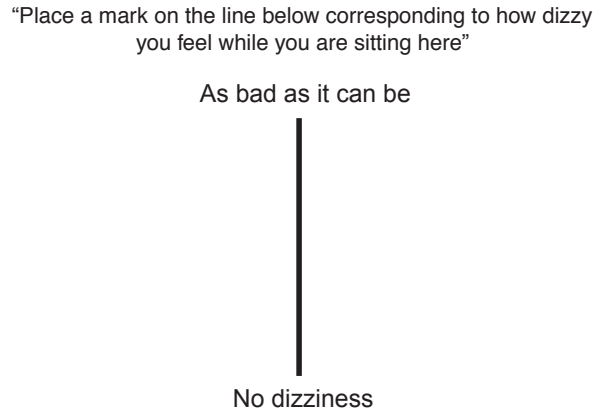

Figure 10.1 Head movement Visual Analogue Scale (VAS). The patient is instructed to place a mark on the line corresponding to how dizzy he or she feels while sitting and then while performing a task. For this scale, test-retest reliability is $r = 0.59$, based on a separate sample of patients with unilateral and bilateral vestibular loss (n = 25).

Impact on Functional Activities

The effect of dizziness on the patient's functional activities can be determined from Section A of the Multidimensional Dizziness Inventory found on the last page of the Patient Questionnaire (see Appendix A at the end of the book).

Perceived Disability

The perceived disability caused by a patient's dizziness can be determined from the Disability Scale, shown in Box 10-1.[7] In this scale, the patient picks the best statement out of six that best fits how he or she feels. The clinician must remember that this is *perceived* disability or handicap. The scale has been validated for degree of perceived disability in patients with unilateral vestibular loss (UVL) and bilateral visual loss (BVL). A score of 4 or higher is correlated with poor outcome of vestibular rehabilitation; that is, the dizziness is unlikely to change with rehabilitation. This scale has high test-retest reliability ($r = 0.97$).[8]

Box 10-1

DISABILITY SCALE*

For the following, please pick the *one* statement that best describes how you feel:

_____ Negligible symptoms (0)

_____ Bothersome symptoms (1)

_____ Performs usual work duties but symptoms interfere with outside activities (2)

_____ Symptoms disrupt performance of both usual work duties and outside activities (3)

_____ Currently on medical leave or had to change jobs because of symptoms (4)

_____ Unable to work for over one year or established permanent disability with compensation payments (5)

*Numbers in parentheses are individual scores for scale. A score of 4 or higher is correlated with poor outcome from vestibular rehabilitation; that is, that patient's condition is unlikely to change with rehabilitation. (Adapted from Shepard et al, 1990.[7])

Fall History

In the fall history, the clinician should obtain a description of any falls (where, when, lighting, what was the patient thinking about), the frequency of falls, the last occurrence, and whether any injuries occurred with the falls. It is also important to obtain information about "near falls"—those events in which patients would have fallen if they had not caught themselves by gripping furniture or some other object or if someone else had not caught them.

Confidence in Balance

The patient's confidence in balance can be obtained with the Activities-Specific Balance Confidence Scale (ABC) developed by Powell and Myers.[9] This scale can be self-administered or done through interview. Each item is rated 0% to 100%. The lower the score, the greater the patient's fear of falling. There is a moderately strong negative correlations (r = -0.64) between the Dizziness Handicap Inventory and the ABC in patients with complaints of dizziness.[10]

Interference with Daily Activities

Interference with daily activities includes basic activities of daily living (ADLs) such as dressing, bathing, getting on and off a toilet, preparing a simple meal, and performing light housekeeping; whether driving is restricted or altered; and whether the patient's employment is affected by dizziness.

Problems that May Interfere with Recovery

Several features of a patient's status may interfere with recovery from dizziness: medications, a list of which can be obtained from item 10 of the Patient Questionnaire (see Appendix A at the end of the book); anxiety or depression, the presence of which can be obtained from item 12B of the Appendix (Positive Affect, Negative Affect scale [PANAS]); the patient's support system at home or work; and whether the patient would have secondary gains from being ill.

Physical Examination

Table 10-4 lists the portions of the physical examination that should be performed on every patient with dizziness to facilitate diagnosis. Visual fixation reduces or suppresses horizontal and vertical nystagmus generated by peripheral vestibular defects. Therefore, some portions of the examination are optimally done with either Frenzel lenses or a video infrared camera to block fixation (Table 10-5).

Spontaneous Nystagmus

Peripheral Vestibular Disorders

Selective lesions in the peripheral vestibular pathways result in spontaneous nystagmus caused by the unopposed higher spontaneous neural activity in the intact vestibular pathways (Fig. 10.2). For example, vestibular neuritis on one side results in peripheral vestibular nystagmus because of the unopposed activity of the lateral and anterior SCC activity on the intact side.[11] The lateral and anterior SCCs project to the ocular motor nuclei via the medial vestibular nucleus. Peripheral vestibular nystagmus after acute loss of vestibular function on one side is a *static defect*, because it occurs even with the head still. Static defects from

■ Table 10-4 **PHYSICAL FINDINGS FOR DIAGNOSIS OF DIZZINESS**

Physical Finding	Pathology
Spontaneous nystagmus present	Acute unilateral vestibular loss, or brainstem/cerebellum abnormality
Skew eye deviation (vertical eye misalignment)	Disruption of peripheral or central utricle pathway
Decreased vestibulo-ocular reflex	Chronic vestibular hypofunction
Eye movements and vertigo elicited during maneuvers	Usually, inner ear debris from benign paroxysmal positional vertigo Rarely, central positional vertigo or nystagmus, perilymphatic fistula, hypermobile stapes, Ménière's disease, superior semicircular canal dehiscence
Visual tracking impaired	Brainstem abnormality
Imbalance while standing or walking	Peripheral vestibular or central problem

■ Table 10-5 WHEN TO USE FRENZEL LENSES OR VIDEO INFRARED (IR) CAMERA DURING TESTING TO BLOCK FIXATION

Part of Physical Examination	Use Frenzel Lenses or Video IR?
Look for spontaneous nystagmus	Yes
Assess vestibulo-ocular reflex (VOR):	
Dynamic visual acuity	No
Head impulse test	No
Head-shaking nystagmus test	Yes
Look for skew eye deviation	No
Examine visual tracking and saccades	No
Look for eye movements elicited during maneuvers	Yes
Examine stance and gait	No

peripheral vestibular loss resolve spontaneously in 1 to 2 weeks, without any intervention. Simultaneous bilateral vestibular loss does not cause spontaneous n ystagmus, because there is no asymmetry between the two sides.

Three features of spontaneous n ystagmus can be used to separate peripheral (inner ear or eighth cranial nerve) vestibular disorders from central vestibular disorders (Table 10-6). First, nystagmus that is caused by peripheral disturbances can be decreased with fixation, but nystagmus resulting from brainstem and cerebellum

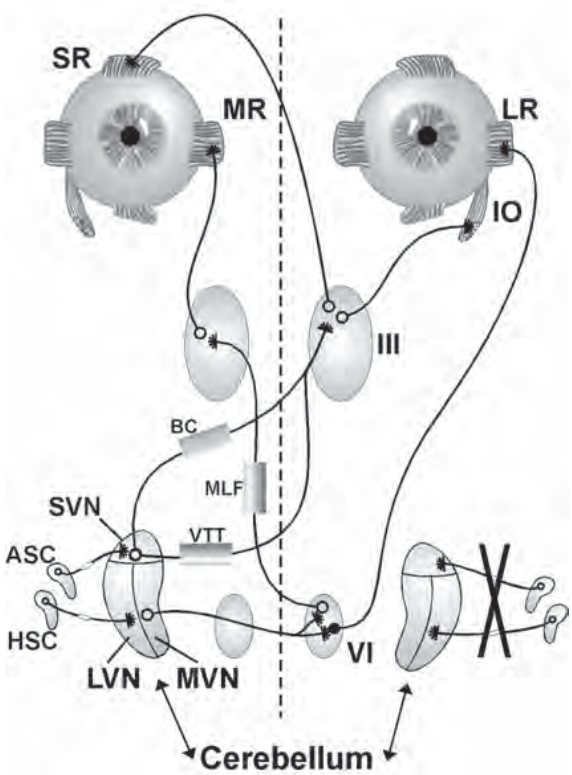

Figure 10.2 Peripheral vestibular loss on the left side from vestibular neuritis. This disorder disrupts the portion of the vestibular nerve from the horizontal semicircular canal (HSC) and anterior semicircular canal (ASC) (X). The ASC and HSC project to the superior vestibular nucleus (SVN) and the medial vestibular nucleus (MVN). The SVN in turn projects to the oculomotor nucleus (III) via the brachium conjunctivum (BC) and the ventral tegmental track (VTT). The MVN projects to the abducens nucleus (VO) and the III nucleus via the medial longitudinal fasciculus (MLF). The VI nucleus projects to the lateral rectus muscle (LR). The III nucleus projects to the superior rectus (SR), medial rectus (MR), and inferior oblique (IO) muscles. (Adapted from Tusa, 1998.[12])

■ Table 10-6 FEATURES THAT DISTINGUISH PERIPHERAL FROM CENTRAL VESTIBULAR NYSTAGMUS

Feature	Peripheral Vestibular Nystagmus	Central Vestibular Nystagmus
Effect of fixation	Nystagmus decreases	Nystagmus either does not change or it increases
Direction of gaze	Usually mixed plane (horizontal and torsional)	Usually single-plane horizontal (torsional or vertical)
Effect of gaze	Nystagmus increases with gaze toward direction of quick phase	Nystagmus either does not change or reverses direction

lesions usually cannot. The clinician can easily test this feature by examining one eye with an ophthalmoscope while having the patient fixate on a target with the other eye.[13] Then the clinician covers the fixating eye with a hand to determine whether the nystagmus increases. Other ways to check for the effect of fixation on spontaneous nystagmus are listed in Table 10-7. Second, the direction of jerk nystagmus in peripheral causes is primarily horizontal, and torsional to a small degree. The slow phases of the horizontal and the torsional component move the eyes toward the involved ear (Fig. 10.3). One must remember that nystagmus is defined by the direction of the quick phases, so a left vestibular neuritis causes right-beating and right torsional nystagmus. Third, nystagmus from peripheral vestibular disorders varies in amplitude and velocity according to eye position in the orbit (gaze dependency). On the first day of a peripheral vestibular disorder, the nystagmus is found when the eyes are gazing center, or toward or away from the side of the lesion (third-degree nystagmus), but the nystagmus is most brisk during gaze away from the side of the lesion. Within a few days, nystagmus may be found only during gaze center and gaze away from the side of the lesion (second-degree nystagmus). Within 1 week, nystagmus may be present only during gaze away from the side of the lesion (first-degree nystagmus).

Central Vestibular Disorders

Figure 10.4 illustrates the structures and pathways that mediate central vestibular nystagmus, and Table 10-8 describes the neural mechanisms.

■ Table 10-7 **USEFUL METHODS OF OBSERVING NYSTAGMUS WITH FIXATION BLOCKED**

Tool	Technique
Ophthalmoscope	View optic nerve of one eye while covering the other eye
Ganzfeld	Have patient stare at blank wall or blank sheet of paper
Video infrared camera	View eyes with infrared camera inserted in blackout goggles
Frenzel lenses	View eyes while patient is wearing ≥20-diopter lenses inserted into face mask

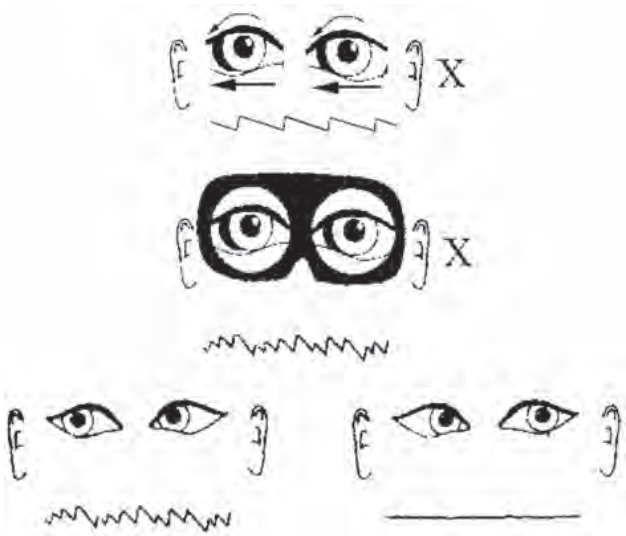

Figure 10.3 Peripheral vestibular nystagmus. This figure depicts disruption of the left superior division of the eighth nerve from vestibular neuritis (X). (*Top*) Mild right-beating and right torsional nystagmus when fixation is present. (*Middle*) Vigorous nystagmus when fixation is suppressed in a subject wearing Frenzel goggles. Nystagmus is labeled according to the direction of the quick phases. Eye movement trace is shown below the face. By convention, eye position to the right is up, and eye position to the left is down. (*Bottom*) The effect of horizontal eye position on nystagmus. The intensity of nystagmus increases when the patient looks in the direction of the quick phases. (Adapted from Brandt, 1991.[14])

Skew Eye Deviation

Skew eye deviation is a vertical misalignment of the eyes caused by a peripheral or central otolith defect. It is part of the ocular tilt response. Each otolith innervates four eye muscles via a three-neuron arc. The central connections of the utricle on one side are shown in Figure 10.5.[15] The projection to the ocular motor nuclei causes the vertical eye deviation and torsion during head tilt. The projections in the lateral and medial vestibular spinal tracts mediate the head tilt during the ocular tilt reflex. Acute loss of function of the utricle on one side from eighth nerve section or vestibular neuritis causes a pathological ocular tilt response because of the unopposed excitation of the intact utricle.[16]

Figure 10.6 depicts the findings in a left-sided lesion (leftward pathological ocular tilt response [OTR]). Excitation of the right superior rectus and oblique muscles causes elevation and intorsion of that eye, and excitation of the left inferior rectus and oblique muscles causes depression and extorsion of that eye. This combination causes a skew eye deviation. Excitation of the neck muscles innervated by the intact vestibulospinal tracts causes a left head tilt. Leftward pathological OTR can be caused

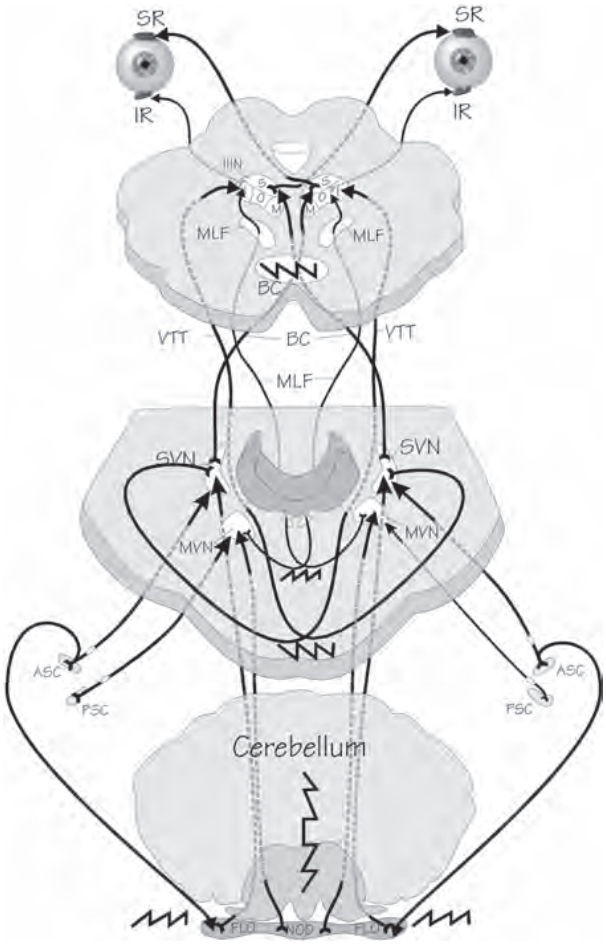

Figure 10.4 Central vestibular pathways and lesion location for central vestibular nystagmus. Downbeat nystagmus occurs whenever the tone within the central pathways from the anterior semicircular canals (ASCs) is relatively higher than the tone within the posterior semicircular canals (PSCs). This can occur from lesions of the cerebellar flocculus (FLO) on both sides caused by disinhibition of the superior vestibular nucleus (SVN). It can also occur from bilateral lesions of the medial longitudinal fasciculus (MLF), which carries input from the PSC to the third nerve (III) nuclei. The III nucleus includes the superior rectus nucleus (S), inferior rectus nucleus (I), medial rectus nucleus (M), and inferior oblique nucleus (O). Upbeat nystagmus occurs whenever the tone within the central pathways from the PSC is relatively higher than the tone within the ASC. This can occur from lesions of the ventral tegmental tract (VTT) or the brachium conjunctivum (BC), both of which carry input from the ASC to the third nerve nuclei. Torsional nystagmus occurs when both ASC and PSC central nuclei are lesioned on one side, as in dorsolateral medullary lesions (Wallenberg's syndrome). Periodic alternating nystagmus is characterized by velocity-constant jerk nystagmus directed to the right for 1 to 2 minutes and then nystagmus to the left for 1 to 2 minutes, with a nystagmus-free interval in between. It occurs whenever the MVN is disinhibited from the cerebellar nodulus (NOD). Seesaw nystagmus occurs from unilateral inactivation of the interstitial nucleus of Cajal. (Adapted from Tusa, 1998.[12])

■ Table 10-8 VESTIBULAR NYSTAGMUS CAUSED BY CENTRAL LESIONS

Nystagmus	Pathology	Possible Mechanism
Torsional nystagmus	Dorsolateral medulla lesion	Decreased tonic neural activity to the INC from anterior and posterior SCC on one side
Downbeat nystagmus	Lesion of cerebellar flocculus or floor of fourth ventricle	Decreased tonic neural activity to the INC from posterior SCC on both sides
Upbeat nystagmus	Lesion of brachium conjunctivum or dorsal upper medulla	Decreased tonic neural activity to INC from central anterior SCC on both sides
Seesaw nystagmus	Unilateral lesion of INC	Unilateral inactivation of INC on one side
Periodic alternating nystagmus	Cerebellar nodulus lesions	Unstable (high gain) neural activity in the MVN
Latent nystagmus	Loss of cortical binocular visual input to the NOT usually from congenital esotropia	Decreased tonic neural activity to MVN from the NOT on one side when one eye is covered. (NOT provides all the visual input into the MVN)

INC = interstitial nucleus of Cajal; MVN = medial vestibular nucleus; NOT = nucleus optic tract; SCC = semicircular canal.

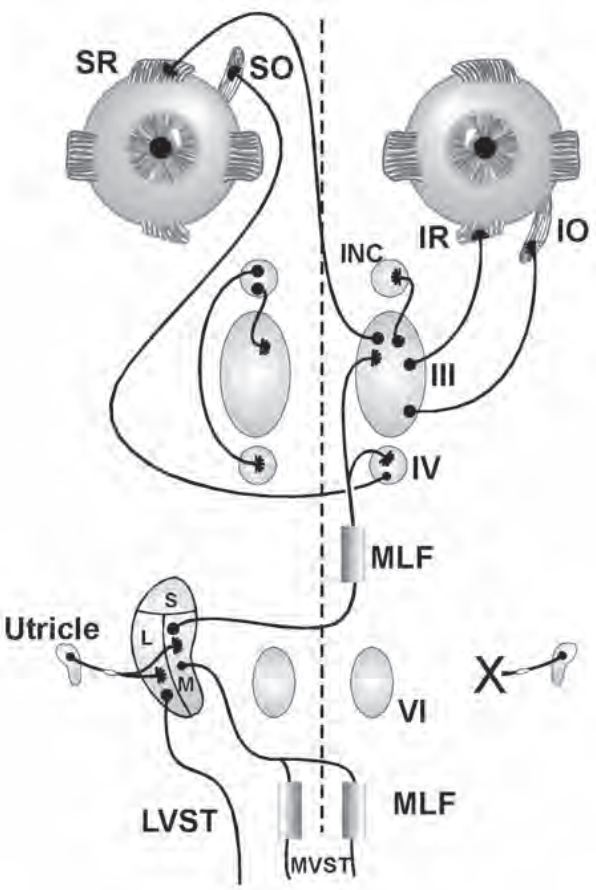

Figure 10.5 Otolith pathway from the left utricle. This figure depicts disruption of the left utricular division of the eighth nerve from vestibular neuritis. The utricle projects to the lateral (L) and medial (M) divisions of the vestibular nucleus. These portions of the vestibular nucleus project to the medial vestibular spinal tract (MVST) and lateral vestibular spinal tract (LVST). In addition, the medial division of the vestibular nucleus projects to the trochlear (IV) and oculomotor (III) nuclei via the medial longitudinal fasciculus (MLF). The IV projects to the superior oblique eye muscle (SO), and the III projects to the superior rectus (SR), inferior rectus (IR), and inferior oblique (IO) eye muscles. INC = interstitial nucleus of Cajal; VI = abducens nucleus; S = superior vestibular nucleus. (Adapted from Tusa, 1998.[12])

by destructive lesions along the pathway extending from the utricle on the left side, such as in the left utricle (labyrinthitis), left vestibular nerve (vestibular neuritis), left vestibular nucleus (Wallenberg's syndrome), right medial longitudinal fasciculus (left internuclear ophthalmoparesis [INO]), right rostral interstitial nucleus of the medial longitudinal fasciculus, and right interstitial nucleus of Cajal.

Vestibular-ocular Reflex

When vestibular function is lost on one side, two types of abnormalities emerge within the VOR. The first abnormality

Figure 10.6 Pathological ocular tilt response from left-sided peripheral vestibular defect. This defect causes the head to tilt to the left, the eyes to have a static torsional component to the left, and a skew eye deviation resulting in a right hypertropia. In the light during bedside examination, the only finding that may be readily appreciated is the skew eye deviation.

is a *static imbalance* caused by a difference in the tonic discharge rate between the vestibular nuclei on the two sides of the brainstem. The tonic resting firing rate of most or all of the primary afferents on the lesioned side are reduced.[17] Resting firing rates of type I nuclei neurons on the lesioned side are acutely absent. In comparison, the resting firing rates of the neurons on the normal side double because of inactivation of the inhibitory commissural path. This imbalance results in a spontaneous nystagmus. Even though the primary afferents may not recover, the relative resting firing rates are readjusted centrally within several days after onset (loss of spontaneous nystagmus). About a week after vestibular neuritis occurs, spontaneous nystagmus can be completely suppressed in the light. On average, though, the relative resting firing rates are less than in control animals. The second abnormality is the loss of *dynamic sensitivity* during head rotation because of the loss of half of the push-pull combination. This results in a decreased gain (eye velocity/head velocity) of the VOR.

The following three bedside tests examine the changes in the dynamic sensitivity of the VOR (Table 10-9). The results of all three VOR bedside tests are useful for patients with unilateral or bilateral loss of vestibular function (such as vestibular neuritis or ototoxicity). Results of the dynamic visual acuity and head-impulse tests are strongly positive in patients with bilateral vestibular loss, but the head-shaking nystagmus test result is not positive in this group.

Head-Impulse Test

Head-impulse test is sometimes referred to as head thrust test. The clinician asks the patient to fixate on a target and then grasps the patient's head to perform passive horizontal and vertical head impulses, observing the eyes during the impulses. After a head impulse, the observation of a

■ Table 10-9 **BEDSIDE TESTS OF THE VESTIBULAR-OCULAR REFLEX (VOR)**

Test	Procedure	Result
Vestibular dynamic visual acuity (DVA)	Static, distant visual acuity is determined with the head still Dynamic visual acuity is then determined while the patient's head is oscillated manually at 2 Hz	A dynamic visual acuity of 3 or more lines above static visual acuity indicates a vestibular defect
Head impulse	The patient fixates a distant visual target, and eye position is observed immediately after a small impulse of the head to the left and right	A refixation saccade after the head impulse indicates decreased VOR If the head impulse elicits a refixation saccade when the patient is fixating a target at near, the test should be repeated with the patient looking at a distant target in order to clearly have a positive result, especially in older patients
Head-shaking nystagmus	Clinician pitches the patient's head down 30 degrees and oscillates the head horizontally 20 times	Elicitation of jerk nystagmus during this procedure indicates a vestibular imbalance

refixation saccade indicates decreased VOR.[18] It is important that the patients are tested while they are wearing their usual glasses, because the VOR is calibrated for visual inputs through those glasses. Table 10-10 shows the sensitivity and specificity of the head-impulse test with respect to the caloric test for UVL. For complete UVL caused by nerve section, the sensitivity and specificity are 100%. For incomplete UVL from a variety of causes, the median sensitivity is 39%, and the specificity is 95%. Sensitivity of testing can be improved by (1) pitching the head down 30 degrees to place the horizontal SCC in the plane of movement and (2) making the head impulse unpredictable.[19] Table 10-11 shows how the head impulse test results vary with the severity of UVL.[20] For example, for moderate paresis of the caloric test (50% to 75% weakness), 19 patients had a negative head impulse test (90% of the total tested) and 2 patients had a positive test (10% of the total tested).

Head-Shaking Nystagmus Test

While wearing Frenzel or IR goggles, the patient is asked to close the eyes, pitch the head down 30 degrees, and then rapidly oscillate the head 20 times horizontally. After the head oscillation, the patient opens the eyes, which the clinician observes for nystagmus. The presence of nystagmus immediately after this procedure indicates a vestibular imbalance.[21] This sign may persist indefinitely after a peripheral or central unilateral vestibular lesion. Table 10-12 shows the sensitivity and specificity of the head-shaking nystagmus test with respect to the caloric test for UVL. For a variety of types of UVL, the overall sensitivity is 46%, and the specificity is 75%. Table 10-13 shows the frequency of a positive head-shaking nystagmus test with respect to the severity of UVL (canal paresis).[22]

Clinical Vestibular Dynamic Visual Acuity Test

The clinical vestibular dynamic visual acuity test compares visual acuity with the head still to visual acuity with the head moving. Visual acuity with the head still is measured first using a visual acuity chart. The patient is then asked to read the smallest possible line on the chart while the examiner manually oscillates the patient's head horizontally at 2 Hz so the face moves 1 or 2 inches in either direction—above the frequency at which pursuit eye movements can track the target. If the VOR is normal, the patient's eyes will move smoothly in the opposite direction of the head movement so that ocular fixation is always maintained. The patient should be able to read either the same line as when the head was still (initial static visual acuity) or the next line above it, which has larger letters. If the patient can read only lines more than 3 lines above the initial static visual acuity line, he or she likely has a vestibular defect (Fig. 10.7). After vestibular adaptation exercises, dynamic visual acuity improves, possibly because of the development of preprogrammed or anticipatory eye movements.[23,24]

■ Table 10-10 BEDSIDE HEAD-IMPULSE TEST COMPARED WITH CALORIC TEST FOR UNILATERAL VESTIBULAR LOSS (UVL)*

Type or Cause of UVL	Specificity (%)*	Specificity (%)*	No. of Patients	Study
Complete UVL caused by nerve section	100	100	20	Halmagyi and Curthoys (1988)[18]
Complete UVL caused by nerve section	100	100	12	Foster et al (1994)[25]
Various	39	97	112	Harvey and Wood (1996)[26]
Various	35	95	105	Harvey et al (1997)[27]
Various	34	100	150	Beynon et al (1998)[20]
Various	45**	91	265	Perez and Rama-Lopez (2003)[28]
Various	71	82	176	Schubert et al (2004)[19]

*Caloric test used as the standard.
**The authors state more than 50% canal paresis is needed for the head impulse test to be positive.

■ Table 10-11 HEAD-IMPULSE TEST (HT) AS A FUNCTION OF CANAL PARESIS

Canal Paresis	No. of Patients	Negative HT Result	Positive HT Result
Normal (0–25)	76	76 (100%)	0 (0%)
Mild paresis (25–50)	23	23 (100%)	0 (0%)
Moderate paresis (50–75)	21	19 (90%)	2 (10%)
Severe paresis (75–100)	30	7 (23%)	23 (77%)

■ Table 10-12 HEAD-SHAKING NYSTAGMUS TEST COMPARED WITH CALORIC TEST*

Sensitivity (%)*	Specificity (%)*	No. of Patients	Study
40	60	108	Wei et al (1989)[29]
95	62	85	Takahashi et al (1990)[30]
27	85	116	Jacobson et al (1990)[31]
44	65	105	Burgio et al (1991)[32]
42	85	197	Goebel and Garcia (1992)[33]
35	92	105	Harvey et al (1997)[27]
38	79	290	Asawavichianginda et al (1997)[22]
Average: 46	*Average:* 75		

*Caloric test used as the standard.

■ Table 10-13 HEAD-SHAKING NYSTAGMUS TEST (HSN) AS A FUNCTION OF CANAL PARESIS

Canal Paresis	Positive HSN Result
Normal (0–20)	22%
Mild paresis (21–25)	24%
Moderate paresis (25–50)	28%
Severe paresis (>50)	62%

(Adapted from Asawavichianginda et al, 1997.[22])

■ Table 10-14 MANEUVER-INDUCED VERTIGO AND EYE MOVEMENTS FROM MECHANICAL PROBLEMS IN THE INNER EAR

Positive Result of	Disorder(s)
Position testing	Benign paroxysmal positional vertigo Central positional nystagmus Central positional vertigo Dehiscence of superior semicircular canal Perilymphatic fistula
Pressure testing, tragus movement, or Tullio phenomenon	Dehiscence of superior semicircular canal Ménière's disease Perilymphatic fistula

Maneuver-Induced Vertigo and Eye Movements

If there is a mechanical problem (e.g., BPPV), nystagmus can be elicited by certain maneuvers. Therefore, in addition to looking for spontaneous nystagmus, the clinician should perform certain maneuvers that may evoke nystagmus (Table 10-14).

Position Testing

The *Hallpike-Dix* test result is positive in patients with BPPV (Fig. 10.8). Nystagmus from BPPV should begin within 30 seconds and last less than 30 seconds. If nystagmus persists while the patient is in this position and is not present when the patient is sitting, it is likely caused by a central disorder (central positional vertigo). The only exception to this statement is BPPV caused by cupulolithiasis. In this condition, otoconia are attached to the cupula of the SCC, so the Hallpike-Dix test will result in persistent nystagmus and vertigo (see Chapter 20 for further

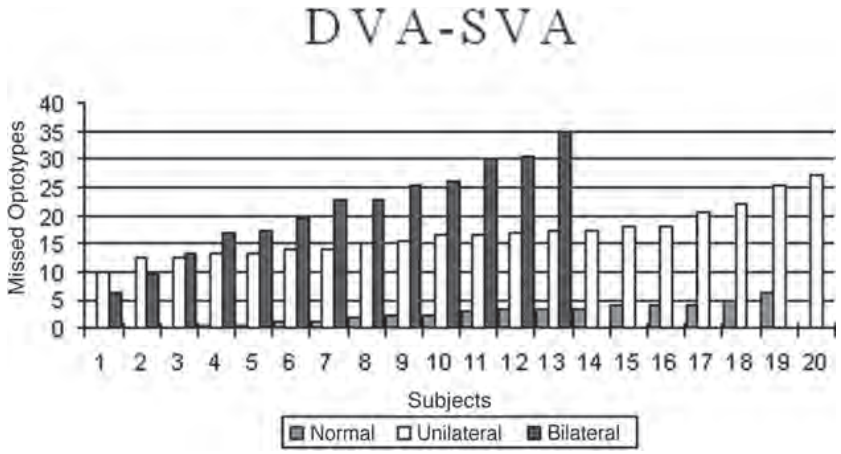

Figure 10.7 Dynamic visual acuity (DVA) scores in patients with vestibular hypofunction and normal controls. On the ordinate is the number of letters (optotypes) missed on a standardized visual acuity (SVA) chart called the ETDRS chart (used in the Early Treatment Diabetic Retinopathy Study). This chart has five letters on each line. On the abscissa are the rankings of individual subjects that had the best visual acuity (to the left) to the most impaired (to the right). *Gray bars* represent controls or subjects seen in the clinic for dizziness who did not have a vestibular defect according to bithermal water caloric testing. *White bars* represent subjects with unilateral vestibular loss (25% or greater asymmetry). *Black bars* represent subjects with bilateral vestibular loss according to caloric testing (<20 degrees of peak slow-phase velocity on four irrigations) and rotary chair test (gain < 0.1).

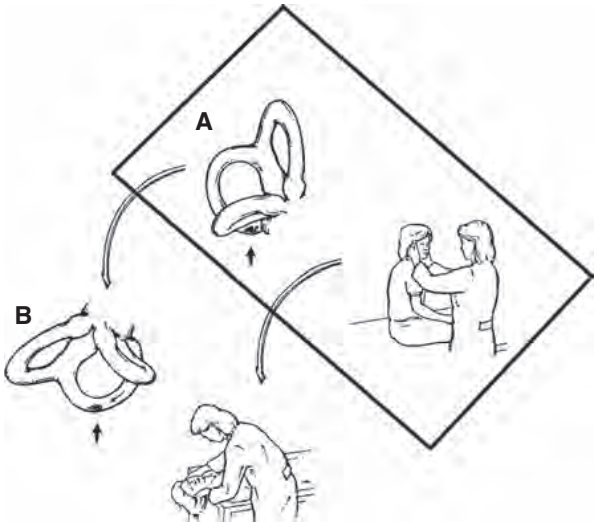

Figure 10.8 Hallpike-Dix test for benign paroxysmal positional vertigo (BPPV). **(A)** In this test, the patient sits on the examination table and the examiner turns the head 45 degrees horizontally. **(B)** The examiner then quickly brings the head and trunk straight back "en bloc" so that the head is hanging over the edge of the examination table by 20 degrees. Nystagmus is sought, and the patient is asked whether he or she has vertigo. Although not shown in the figure, the examiner then brings the patient up slowly to a sitting position with the head still turned 45 degrees, and looks for nystagmus again. The test is repeated with the head turned 45 degrees in the other direction. This figure also shows movement of debris in the right posterior semicircular canal (*black arrows*) during the test. In this example, the patient would have nystagmus and vertigo when the test is performed on the right side, but not when the test is performed on the left side.

details). Positional nystagmus may also be seen in patients with a variety of other mechanical problems of the inner ear. These other disorders are central positional vertigo, central positional nystagmus without vertigo, and perilymphatic fistula (a hole between the endolymph and perilymph or between the perilymph and middle ear).

Pressure Testing, Tullio Phenomenon, and Tragus Movement

Nystagmus or drift of the eyes should also be assessed after positive and negative pressure directed to the external auditory canal (Hennebert's sign), Valsalva, or loud noise (Tullio phenomenon). A positive response is found in patients with perilymphatic fistula, hypermobile stapes, and, occasionally, Ménière's disease or hydrops.

Visual Tracking

Smooth Pursuit Eye Movements and Cancellation of the Vestibulo-ocular Reflex Both smooth pursuit eye movements and VOR cancellation are slow tracking movements that maintain images of small moving targets on the fovea. During smooth pursuit eye movements, the head is kept still. During VOR cancellation, the head is moving synchronously with the target. This movement is referred to as *VOR cancellation*, because the VOR must be suppressed during the head movement; otherwise, the image of the target could not be maintained on the fovea. The patient is asked to track a small target that is moving slowly (20 degrees per second [deg/sec]) both horizontally and vertically, with the head still (smooth pursuit). VOR cancellation can be measured by having the patient fixate on a small target that moves synchronously with the head movement. The easiest way to do this is for the clinician to grasp the patient's head with both hands and gently move it back and forth at 1 Hz. The clinician moves his or her own head synchronously with the patient's head and asks the patient to follow the clinician's nose.

A unilateral peripheral vestibular lesion does not impair either smooth pursuit or VOR cancellation unless the spontaneous nystagmus from the lesion is so high that it prevents the eye tracking systems from functioning normally. In contrast, a lesion in the parieto-occipital frontal cortex, frontal cortex, pontine nuclei, cerebellar vermis, or cerebellar flocculus does cause deficits in smooth pursuit and VOR cancellation for targets moving toward the side of the lesion. During smooth pursuit for target motion toward the side of the lesion, there are catch-up saccades because of decreased pursuit gain (gain = slow-phase eye velocity ÷ target velocity); this process is sometimes referred to as *saccadic pursuit*. During VOR cancellation toward the side of the lesion, a horizontal jerk nystagmus occurs. An example of saccadic pursuit in a right-sided lesion is shown in Figure 10.9. In patients with cerebellar degeneration or other bilateral disorders, smooth pursuit and VOR cancellation are impaired in both directions.

Saccadic Eye Movements

Saccadic eye movements are very rapid changes in eye position. These eye movements can be tested by having the patient follow a target that rapidly changes position. Having the patient fixate on the clinician's nose assesses steady fixation. Having the patient look between the clinician's nose and a finger held approximately 20 degrees eccentrically assesses voluntary saccades; this evaluation is repeated several times to the left, to the right, up, and down. During this test, the clinician should determine whether the saccades have normal amplitude and velocity. For saccades to eccentric targets, the amplitude should be normal or not more than 10% hypometric, and never hypermetric for saccades back to center, the amplitude should be normal or no more than 10% hypometric or

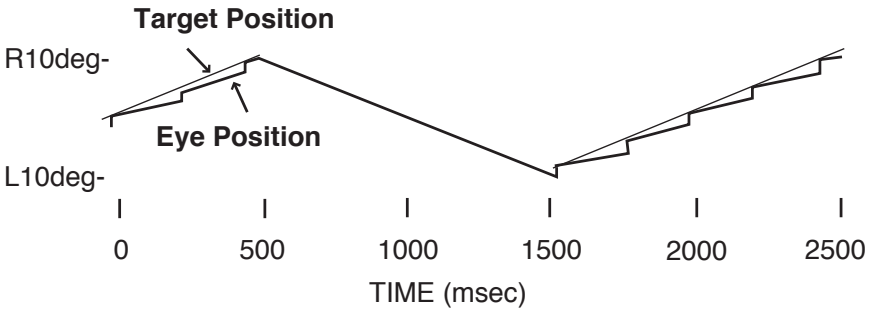

Figure 10.9 Impaired smooth pursuit to the right. The *light line* represents target trajectory. The *dark line* represents eye position. R10deg and L10deg represent eye positions (right and left 10 degrees, respectively) within the orbit.

hypermetric. Saccade velocity should be brisk and equal in the two eyes. The patient should also be told to follow an optokinetic drum or tape to assess quick phases of nystagmus.

Peripheral vestibular defects do not impair saccades. In contrast, unilateral lesions of the cerebellum or its afferent and efferent connections can cause hypermetria in one direction and hypometria in the other direction. A unilateral lesion involving the cerebellar vermis from a superior cerebellar artery infarction results in contralateral hypermetria and ipsilateral hypometria. Infarction of the lateral medulla (Wallenberg's syndrome) also results in ipsilateral hypermetria and contralateral hypometria, presumably because of deafferentation of the fastigial nucleus from infarction of the inferior cerebellar peduncle. The definition of saccade parameters are listed in Table 10-15.

Cerebellar vermal lesions result in hypometric saccades to the right and to the left. Slow saccades can be caused by a number of different disorders, including a decrease in the number of burst cells in the paramedian pontine reticular formation (PPRF) and rostral interstitial nucleus of the medial longitudinal fasciculus RiMLF. Generally, lesions involving the midbrain cause vertical saccade slowing, whereas lesions involving the pons cause horizontal saccade slowing. Slow saccades can also be caused by internuclear ophthalmoparesis (INO). An INO occurs when the pathway between the sixth and third nerve nuclei is disrupted. In an INO, saccades in the eye moving toward the nose (adducting) are slower or limited compared with those in the eye moving away from the nose (abducting). Slow saccades can also occur because of problems in the neuromuscular junction (e.g., myasthenia gravis, Miller-Fisher syndrome). Finally, slow saccades may occur because of eye muscle problems (thyroid eye disease, progressive external ophthalmoplegia).

Stance and Gait

The Romberg test, "Sharpened" Romberg (heel-to-toe tandem stance) test, Fukuda's Stepping Test (FST), and retropulsion test should be performed to evaluate stance and gait and tandem gait.

In the Romberg tests, the patient is asked to stand with feet slightly apart and arms folded across the chest with eyes open for 30 seconds and then with eyes closed for 30 seconds. A *positive* Romberg test result is one in which the patient is stable with eyes open but loses balance with eyes closed. A positive Romberg result

■ Table 10-15 **SACCADIC EYE MOVEMENT DEFICITS**

Saccade Parameter	Definition	Lesion Location
Latency	Time from target step to beginning of saccade	Increased latency seen primarily with lesions in cerebral cortex (visual attention defects) or brainstem (defects in initiation)
Velocity	Peak speed of saccade	Decreased velocity seen primarily from lesions in the pons (burst cells)
Accuracy	How close amplitude of saccade matches amplitude of target step	Decreased or increased amplitude primarily determined by cerebellar vermis and pathways to brainstem

occurs in patients with severe proprioceptive defects from a peripheral neuropathy and may be found in patients with acute vestibular defects. The Romberg test is also useful in identifying a functional component, suggested when the patient rocks backward on the heels yet, remarkably, does not fall or have an exaggerated sway during the test. In the Sharpened Romberg test, the patient is asked to stand with feet in heel-to-toe position, first with eyes open and then with eyes closed. A *positive* Sharpened Romberg test result is found in patients with the same disorders that cause a positive Romberg result, in those with chronic vestibular defects, and in some normal individuals older than 65 years.

For the Fukuda Stepping Test, the subject steps in place for 50 steps with arms extended and eyes closed.[34] Progressive turning toward one side of more than 30 to 45 degrees is abnormal. An abnormal Fukuda Stepping Test can occur from an asymmetric vestibulospinal reflex tone from peripheral or central vestibular tone asymmetry or loss. It can also occur from musculo-skeletal asymmetries including leg-length discrepancy, sciatica, muscle imbalance, or asymmetric joint problems from the hip down. Based on 126 consecutive patients with unilateral vestibular loss, abnormal deviation toward the side of the lesion (45 deg or more deviation) occurred in 50.0% of cases and toward the intact side in 24.6% of cases. [35] In a study that measured FST as a function of degree of caloric weakness on ENG, FST was only sensitive in detecting vestibular weakness when the weakness was severe on ENG (greater than 75% weakness). Furthermore, the FST abnormality decreased with time following the vestibular defect.[36]

In the retropulsion test, the patient stands with the feet slightly spread apart and is instructed to just take one step backward if pulled backward at the hips by a mild force. The result is positive if the patient must take three or more steps backward or falls backward like a log. The retropulsion test has a positive result in patients with basal ganglia disorders (progressive supranuclear palsy, Parkinson's disease) and disorders that disrupt frontal lobe–basal ganglia projections (normal-pressure hydrocephalus, leukoareosis).

Gait and tandem gait should be examined for cerebellar ataxia, decreased head-on-body movements during turns (vestibular hypofunction), and shuffling gait in Parkinson's disease. Other features of gait help identify a functional component, including knee buckling without fall, small-amplitude steps, uneconomical posture and movement, excessive slowness in gait, and fluctuations in levels of impairment.[37,38]

Summary

An accurate history is needed to determine the onset of the problem, description of the symptoms, and, most important, how the symptoms affect the individual's lifestyle. It is the primary piece of information that determines how the patient should be managed. The bedside exam is very helpful in determining if the patient has vestibular hypofunction on one or both sides, BPPV, a central defect in the posterior fossa, or psychogenic features.

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CHAPTER 11

Vestibular Function Tests

Michael C. Schubert, PT, PhD

The vestibular system senses motion of the head for the purposes of maintaining stability of images on the fovea of the retina and stability of postural control during that head motion. In normal function, the vestibular receptors provide an elegant and accurate representation of the motion of the head in three dimensions. This information is then carried along central vestibular pathways to control the reflexes and perceptions that are mediated by the vestibular system. Disorders of vestibular function result in abnormalities in these reflexes and lead to sensations that reflect abnormal information.

The output of the vestibular system can grossly be considered a motor response reflected in an ocular or postural behavior. The ocular signature of the vestibular system is typically measured from the vestibulo-ocular reflex (VOR), and the postural behavior is typically measured using force plate platform equipment. In this chapter, we discuss the common testing techniques of the vestibular system, categorized by ocular or posturographic behavior. Further delineations are made that incorporate testing the behavioral outcome of the vestibular function.

Electronystagmography (ENG)

In the 1800s, it was realized that a voltage of the eye existed and varied with eye rotation.[1] Because the cornea is positively charged with respect to the retina, a dipole (i.e., battery) is formed (Fig. 11.1). This corneo-retinal potential (CRP) has a baseline (eyes stationary, positioned in neutral) potential difference of 1 millivolt and provided the first technique useful to record eye motion, called electro-oculography (EOG).[2,3] By the early 1900s, the CRP had become the standard method to record eye motion.[4] Using horizontally and vertically positioned surface electrodes, the CRP can be detected as the polarity of the eye rotates toward or away from those electrodes. Because a torsional eye rotation (being purely clockwise or counterclockwise) does not cause a vertical or horizontal deflection of the globe within the orbit, the electrodes cannot detect a relative change in the CRP. Thus, torsion eye rotations cannot be monitored with EOG.

In addition to a method for recording eye rotation, ENG also refers to a battery of tests. As a test battery, the ENG includes measures of oculomotor function (including gaze stability, smooth pursuit, and saccades), positional and positioning testing, and the caloric exam. With the development of video-based recording of eye movements, the test battery is often referred to as VNG, video nystagmography.

Oculomotor Function

Various oculomotor systems and mechanisms can contribute to visual fixation of a target on the fovea of the retina, in addition to the vestibular system. These non-vestibular mechanisms include the gaze stability (or gaze holding), smooth pursuit, and saccadic oculomotor systems. Influences such as target position, target velocity, and head velocity are the stimulus variables the brain uses

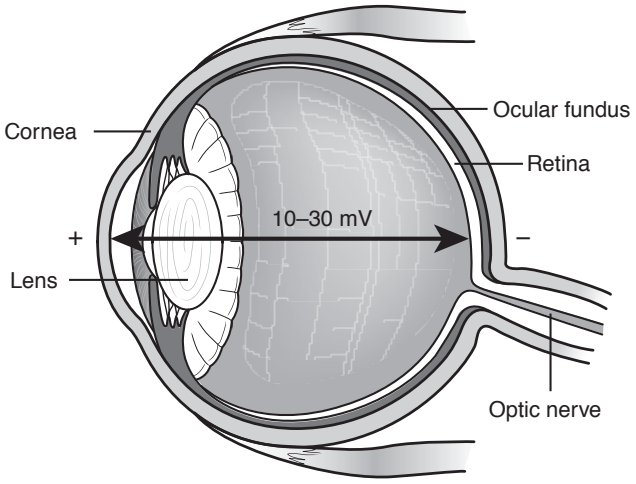


Figure 11.1 Corneo-retinal potential (CRP). The cornea of the eye is positive and the retina is negative. As the eyes move horizontally or vertically toward surface electrodes positioned both horizontally and vertically, the CRP can be detected. The electrodes are recording the relative change in CRP, not potentials from ocular muscles. Torsional (roll) eye rotations are not recorded, because the CRP does not change relative to the electrode position.

to determine which oculomotor system is recruited for gaze stability. The smooth pursuit, saccade, and vestibulo-ocular motor systems each have ranges at which they function most effectively. Each system also has limitations at which point it becomes ineffective. There is overlap among the effective ranges such that multiple mechanisms can combine for the purpose of stabilizing gaze. The purpose of investigating the oculomotor systems as part of the ENG test battery is to identify pathology within the central oculomotor and vestibular systems. Smooth pursuit and saccade testing are used to identify pathology within the brainstem and cerebellar regions. The inability to maintain gaze stability can be caused by either peripheral vestibular or central oculomotor pathology.

Gaze Stability

Gaze stability is the ability to hold the eyes stationary and is tested in primary gaze and after the eyes are moved 30 degrees up, down, right, and left. Gaze stability testing is useful to identify problems with the peripheral vestibular or central oculomotor pathways. Subjects should be able to maintain gaze without any eye motion. Abnormal gaze stability occurs when an eye rotation moves the eye off the target (i.e., jerk nystagmus, ocular flutter). Acute lesions of the peripheral vestibular system (i.e., vestibular neuritis) cause a spontaneous nystagmus that disrupts gaze stability. Various pathologies of the central nervous system also can disrupt gaze stability (e.g., cerebellar stroke).

Smooth Pursuit

The smooth pursuit system is used to stabilize a moving target on the fovea of the retina during low velocities and frequencies of head motion or target motion. Pursuit is generated from multiple cortical regions in addition to the brainstem and cerebellum. As a result, tests of pursuit are not as useful for localizing central pathology. Lesions of the pursuit system may present with unilateral or bilateral pursuit deficits. In cases of unilateral pursuit deficits, the deficits are most apparent with pursuit toward the side of lesion. Although cerebral cortical structures may be involved, the lesions typically involve the brainstem pathways.[5]

The smooth pursuit oculomotor system can be quantified by comparing target velocity with eye velocity (gain = eye velocity/target velocity) while subjects follow a moving target (Fig. 11.2). Abnormality of gain and asymmetry between pursuit to the left and right (or up/down) are the measures of interest. Most studies agree that the smooth pursuit system is ineffective at maintaining fixation of a moving target when the velocity or frequency of the target exceeds 60 deg/sec or 1 Hz, respectively.[6-9] However, two studies reveal that the smooth[10] pursuit system can assist gaze stability at target velocities from 75 to 90 deg/sec.[10,11] When analyzing smooth pursuit, the age of the patient must be considered, because smooth pursuit performance is dependent on age. Other measures of pursuit that may be measured are latency (~125 msec) and acceleration (range: 40 to 200 deg/sec/sec).[10,12]

Saccade Oculomotor System

Saccades are rapid, conjugate eye rotations that quickly move the eyes to place the fovea on the target of interest. Saccades do not therefore maintain gaze stability during head movement, in contrast with the pursuit system, but instead quickly reposition the eyes. They are generated from neurons within the brainstem, basal ganglia, frontal eye fields, and cerebellum (and of course require healthy cranial nerve and extraocular muscle function). Saccades can be useful to locate site of pathology (i.e., internuclear ophthalmoplegia) within the central nervous system. Abnormalities in saccade testing do not occur as a result of peripheral vestibular pathology.

Saccades are tested by asking subjects to move their eyes to a newly repositioned target or between two stationary targets. Often, the saccade testing is done using multiple target amplitudes (Fig. 11.3). Horizontal saccades generally occur from excitatory burst neurons within the parapontine reticular formation of the pons.[13] Vertical and torsional saccades typically occur from excitatory burst neurons within the rostral interstitial nucleus of the medial longitudinal fasciculus of the midbrain.[14] Other central regions are able to generate horizontal and vertical/torsional

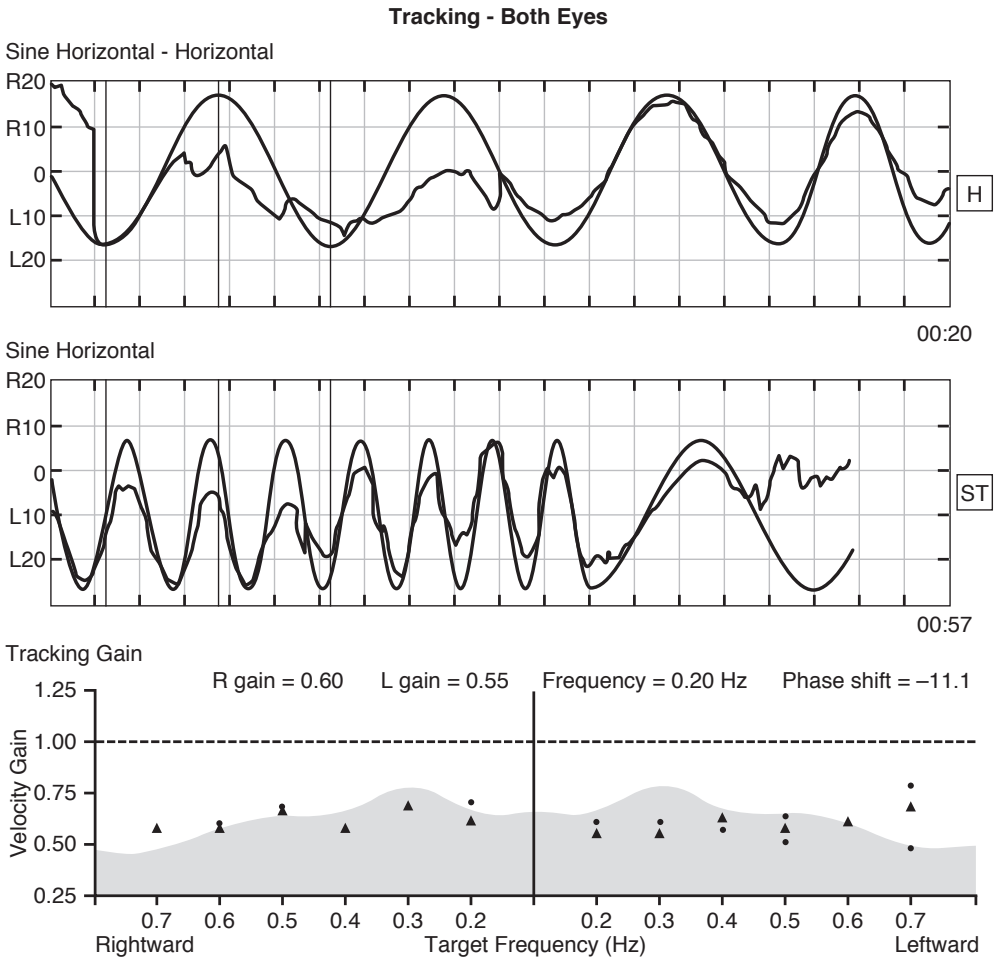


Figure 11.2 Raw trace and analysis plot for smooth pursuit testing. The top two plots display the target moving in a horizontal sinusoid (left to right) at progressively larger frequencies. In the bottom analysis plot, the shaded area represents abnormal performance. The pursuit gain is plotted (y-axis) and is abnormally low for the majority of the testing frequencies (x-axis).

saccades.[5] Important variables in saccade testing are the latency, velocity, and accuracy of the eye rotation. Acceleration and amplitude are also considered. The latency to initiate a visually guided saccade is ~200 msec.[15] Saccadic eye velocities range from 250 to 600 deg/sec. The velocity of the saccades is proportional to the amplitude the eye must move to fixate the target. This relationship between saccade amplitude and velocity is called the main sequence and is useful to confirm healthy function of the saccadic system. Saccades are also typically identified by their exceptionally rapid accelerations that range from 12,000 to 40,000 deg/sec/sec.[15,16]

For more in-depth reading on testing of the human oculomotor system, the reader is encouraged to see Jacobsen and Shepard 2008.[17]

Positional and Positioning Tests

Positional testing refers to placing the patient into various head or whole body positions to reveal those positions that cause nystagmus.[18,19] Different laboratories use different protocols. Typically, the patient's body is placed into a minimum of four different positions (i.e., supine, side lying, head right, and head left) to investigate nystagmus induced by specific head positions relative to gravity. Positional testing is considered abnormal if the slow component eye velocity (SCEV) is greater than 5 deg/sec in any position; less than 6 deg/sec and persistent in 1/2 of the tested positions; or less than 6 deg/sec and transient in all positions. The positioning test is the Dix-Hallpike. Unlike the positional tests, in which the patient is brought slowly into the various positions,

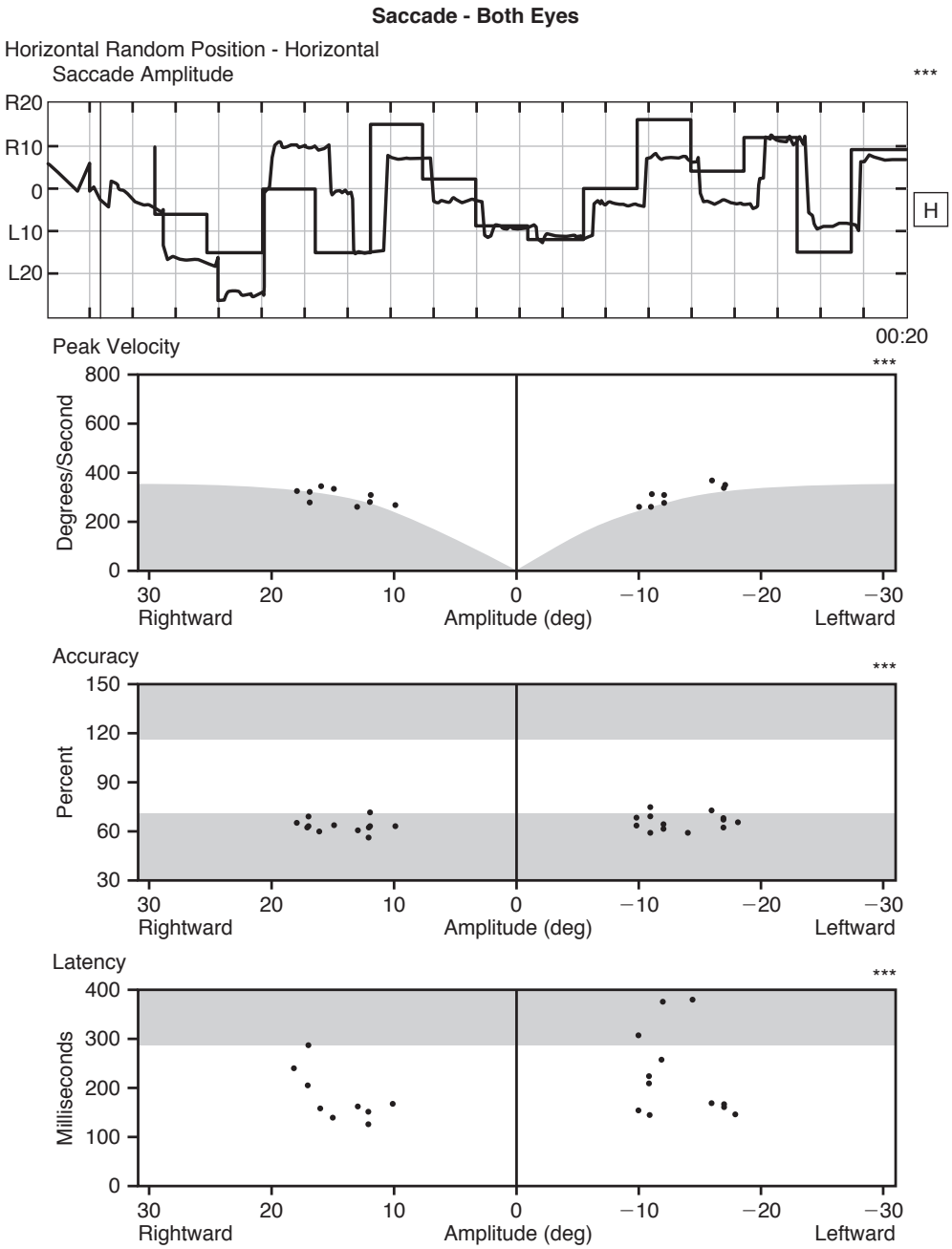


Figure 11.3 Raw saccade traces and summary analysis for velocity, accuracy, and latency. The top plot is for the horizontal channel and shows a random saccade target amplitude (clean trace) and the subject's eyes (jagged trace) following the target position. The velocity and latency of the saccades are normal, and the accuracy is hypometric.

the Dix-Hallpike is performed more rapidly to determine if the *change* in head position will elicit nystagmus. With the exception of the Dix-Hallpike test, positional testing is not considered diagnostic without other components of the ENG and/or clinical exam.

Semicircular Canal Function

The physiological assessment of semicircular canal function requires mechanical stimulation of the cupula and functioning peripheral v estibular afferents. When the cupula moves, nystagmus should be generated (Fig. 11.4).

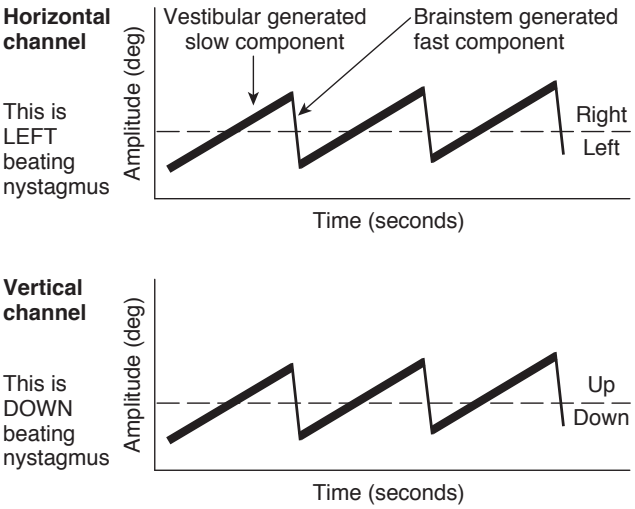


Figure 11.4 Vestibular nystagmus. Nystagmus is named using the fast component. Eye motion is most commonly described with the patient as the reference (i.e., their right/left). The graphing convention denotes the upper ½ plot as a right/left rotation, and the lower ½ plot is up/down rotation—depending on the channel.

The nystagmus is often displayed as part of the vestibular function testing and enables the clinician to discern whether the patterns of eye movements are abnormal.

Caloric Exam

The caloric test is the "gold standard" for identifying peripheral unilateral vestibular hypofunction (UVH).[20] By introducing a cold or warm stimulus into the external auditory canal, a temperature gradient is created within the temporal bone. The change in temperature is the greatest for the lateral aspect of the temporal bone and the least for the medial aspect. In the presence of gravity, this temperature gradient results in the convective flow of endolymph that deflects the cupula of the horizontal semicircular canal and generates nystagmus (Fig. 11.5). For this reason, the position of the head during a caloric examination is important. The caloric stimulation also causes some direct hair cell stimulation and changes in pressure across the middle ear, which will also cause cupular deflection, contributing to the resulting nystagmus.[21-23] The caloric test is particularly useful for determining the side of a deficit because each labyrinth is stimulated separately. The resulting nystagmus enables unique data that are analyzed by two means:

1. The peak SCEV from irrigations of the right ear are compared with the peak SCEV caused by irrigations from the left ear. These values are used in Jongkees formula to determine the relative symmetry in the response to stimulation of the horizontal semicircular canals (SCCs) (Fig. 11.6).[24]

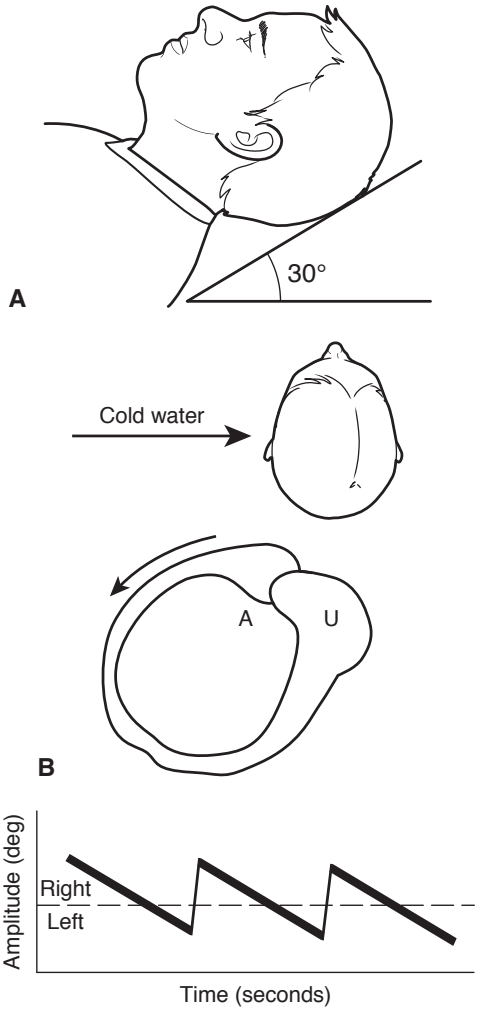


Figure 11.5 Mechanism of the caloric response. The head must be positioned so the endolymph of the horizontal SCC can move in the direction of the gravitational vector **(A)**. In this example, water cooled to 30 deg Celsius is flushed into the left external auditory canal ear (and separately the right ear) for about 1 minute. This creates a temperature gradient in the middle ear that causes endolymph to move within the left horizontal semicircular canal **(B)**. As a result, the cupula is deflected and right-beating nystagmus is generated **(C)**. Direct hair cell stimulation and changes in pressure across the middle ear also cause cupular deflection, contributing to the resulting nystagmus.

$$\text{Unilateral weakness (or relative asymmetry)} = \frac{(LC + LW) - (RC + RW)}{(LC + LW + RC + RW)} \times 100$$

The values are generated from irrigations that are either 7 degrees Celsius above or below body temperature (LC – left cool; LW – left warm; RC – right cool; RW – right warm). Cold irrigations inhibit the vestibular afferents of the stimulated horizontal SCC and create an

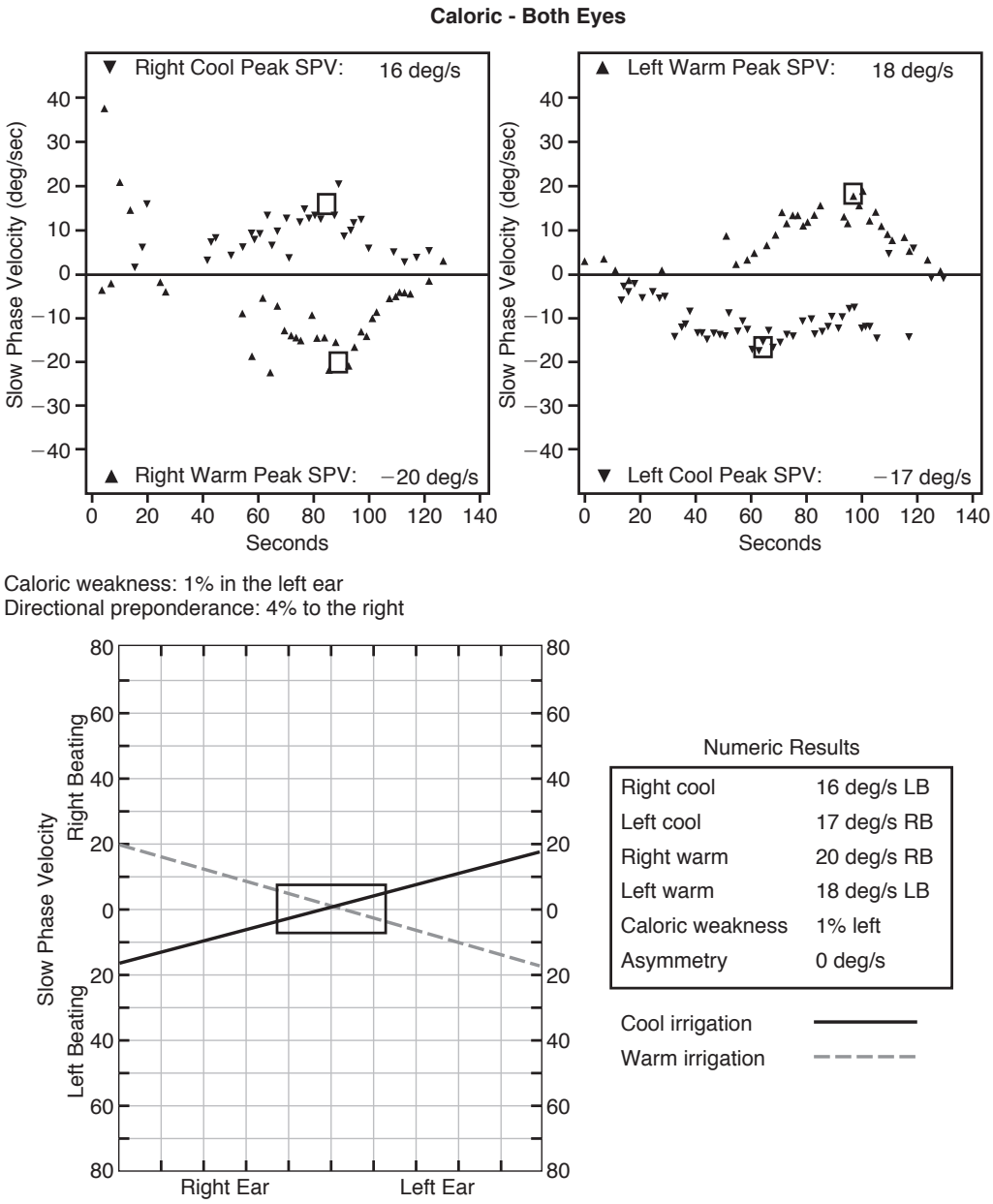


Figure 11.6 Summary of ENG analysis. Top plot shows individual peak slow-phase velocity for warm and cool irrigations chosen by an examiner. The peak SPV values are in the upper right corner of each plot. The bottom plot is a different rendering of the analysis, known as a butterfly plot. Values are summarized in the legend. The junction of the irrigation plots (thin and thicker lines) should be within the center-positioned rectangle representing normal variability. This is a normal ENG exam.

inhibitory nystagmus pattern on the irrigated side. Warm irrigations excite the vestibular afferents of the stimulated horizontal SCC and create an excitatory nystagmus pattern on the irrigated side. Therefore, you would expect a right cool irrigation to generate a left-beating nystagmus and a right warm irrigation to generate a right-beating nystagmus (Fig. 11.7). This can be remembered using the mnemonic COWS: cold opposite warm same—referring to the temperature of the irrigation and the expected direction of the fast component of the nystagmus.

2. The preference for the nystagmus to beat toward one direction or another is also of interest. Directional preponderance (DP) is a measure of the difference in the magnitude of nystagmus

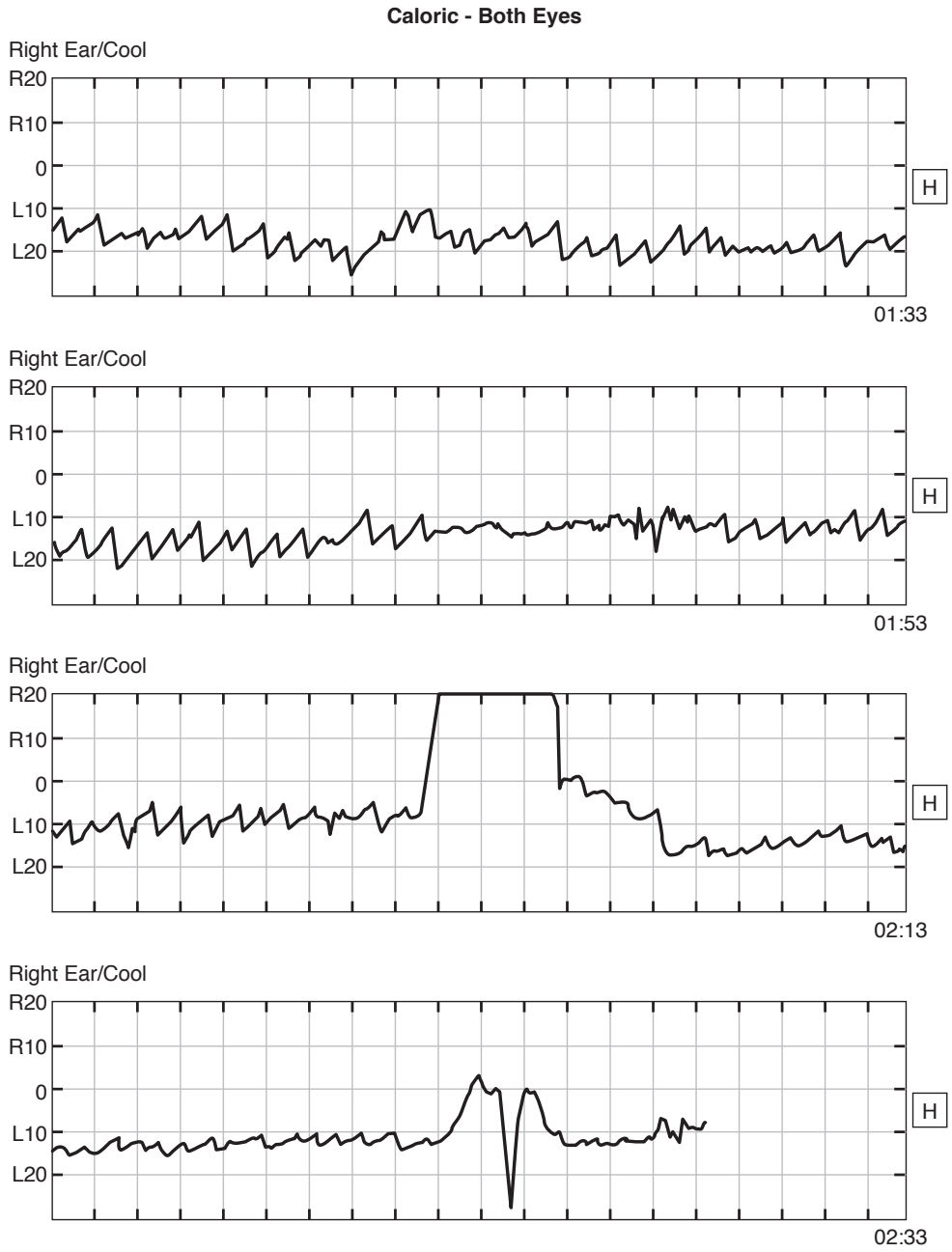


Figure 11.7 Raw tracing of continual nystagmus following cool irrigations for right ear. Vertical graph lines represent time in 1-sec epochs. Horizontal graph lines represent 10 deg eye movements. This is a normal caloric response for this irrigation.

for irrigations that cause a right beat as compared with the irrigations that cause a left beat. The formula compares the peak SCEV for irrigations that cause right-beating nystagmus (RW and LC) with the peak SCEV for irrigations that cause left-beating nystagmus (LW and RC):

$$\frac{DP = (RW + LC) - (LW + RC)}{(RW + LC + LW + RC)} \times 100$$

For both unilateral weakness and directional preponderance, most vestibular laboratories consider values greater than 25% different as being significantly abnormal. The caloric test is limited in its capacity to diagnose pathology , however, because only the horizontal SCCs and the afferents from the superior vestibular nerve are stimulated. The vertical semicircular canals are not assessed. Additionally, that stimulation corresponds to a frequenc y (0.025 Hz) that is

much lower than the natural frequencies of head movement (1 to 20 Hz).[25,26] A summary of the critical aspects of the caloric exam is provided in Box 11-1.

Occasionally, the warm and cool water (or air) irrigations will suggest a 100% loss of vestibular function in one or both ears with no nystagmus having been generated. In this case and others (e.g., the technician suspects a poor stimulation caused by limited temperature exposure), ice water is irrigated into the suspect external auditory canal, which provides a greater temperature gradient and normally causes a robust endolymph flow and inhibition of the horizontal semicircular canal afferents. In the case when no nystagmus is generated by the normal warm and cool water irrigations, the ice water stimulus may generate nystagmus. To confirm that the observed nystagmus is caused by horizontal SCC stimulation, the patient is placed in the prone position, which, because of the reversal of the canal relative to the gravitational vector, will reverse the flow of the endolymph, reverse the cupular deflection, and similarly reverse the direction of the nystagmus. If the nystagmus does not reverse or if no nystagmus is present, then the hypofunction from the horizontal SCC is considered complete (Fig. 11.8).

Rotational Chair Testing

The rotational chair test is the "gold standard" for identifying bilateral vestibular hypofunction (BVH) and the extent of central nervous system compensation to vestibular hypofunction.[20] The rotary chair test provides a physiological stimulus, because rotating the patient causes endolymph flow (with relative excitation and inhibition) from both horizontal SCCs. This form of test is a closer approximation of natural head velocities as compared with the caloric test. The rotational chair test is indicated when bilateral vestibular hypofunction is suspected, when testing vestibular function in children, when the caloric test cannot be performed because of tympanic perforations or anatomical asymmetries of the ear, when the caloric exam is abnormal for function in each horizontal SCC, whenever serial testing is desired, and when trying to determine the extent of compensation.

In individuals with normal vestibular function, nystagmus should be generated during rotational chair testing. In individuals with vestibular hypofunction, nystagmus will be generated, but the magnitude of the SCEV will vary with the extent of the lesion. Data from the rotational chair test are collected by two means: step velocity and sinusoid paradigms (Fig. 11.9). In the step velocity paradigm, the chair is stationary and accelerates to a predetermined peak velocity, continues to rotate at that velocity, and is then stopped suddenly. The test is commonly performed using peak velocities of 60 and 240 deg/sec in each direction. The higher velocity steps can be used to lateralize the hypofunction caused by inhibitory cutoff. For the sinusoid tests, the chair oscillates in yaw (left/right) at varying frequencies (e.g., 0.01, 0.02, 0.04 etc. . . . up to 1.28 Hz), typically with peak velocities that remain constant at 50 or 60 deg/sec.

The typical parameters that are measured from the rotational chair test include Gain, Phase or Time Constant, and Asymmetry. The VOR gain is expressed as the ratio of eye velocity to head velocity (eye velocity/head velocity). Under ideal conditions, and when the eyes are not converged, the VOR gain is –1, implying a compensatory eye velocity equal to the head velocity but in the opposite direction. The gain of the VOR during rotational chair testing is typically much lower than –1, for a variety of reasons (equipment, alertness) (Fig. 11.10). Each vestibular testing laboratory should determine their normal values for VOR gain. Generally, the VOR is considered if there is a gain asymmetry, defined as a greater than 26% difference in the response to rotations to the right and left.

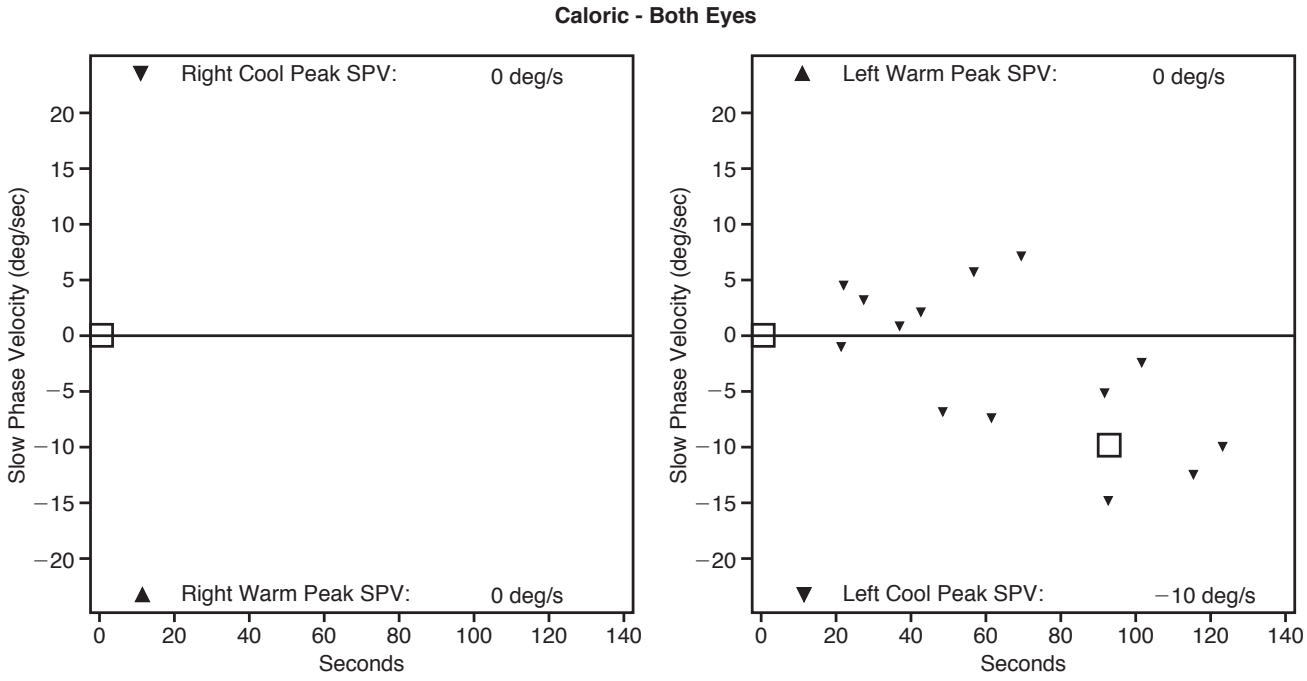
VOR phase and VOR time constant are measures of the transduction process and velocity storage system. Phase is measured with the sinusoid testing paradigm, and the time constant is measured with the step velocity testing paradigm. Both measures are useful variables for assessing the integrity of the vestibular system. Phase is a measure of the timing relationship between the eye and

Box 11-1

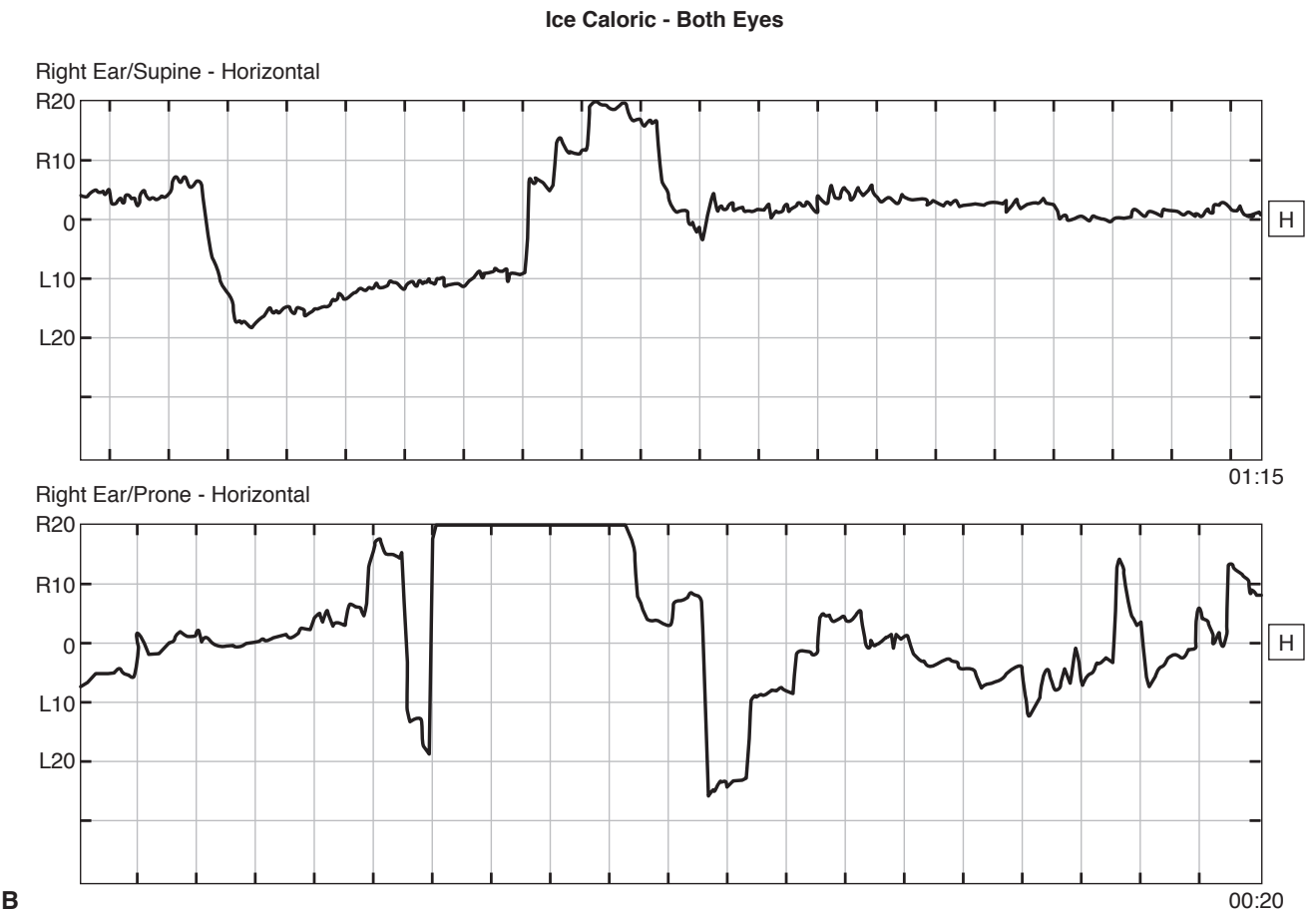
SUMMARY OF THE CALORIC EXAM

- Nystagmus occurs primarily because of endolymphatic flow caused by convection current and eighth nerve excitation/inhibition.
- Typical water irrigations involve temperatures 7°C above and 7°C below body temperature.
- Cold Opposite Warm Same (COWS).
- Most labs consider UW/RVR and DP to be abnormal when ≥26%.
- Only examines the horizontal semicircular canal and related afferents.
- Ice water irrigation commonly applied *only* when an absent response occurs or the technician suspects a poor irrigation.
- Water irrigations more robust, air is useful in the case of perforated TM.

UW – unilateral weakness; RVR – reduced vestibular response; DP – directional preponderance; TM – tympanic membrane



A



B

Figure 11.8 Abnormal ENG and ice water caloric examination. **(A)** Irrigations into the right ear (cool and warm) generate no nystagmus. In contrast, note the symbols present for the left cool and warm water irrigations. **(B)** Ice water irrigation into the right ear causes no nystagmus—regardless of supine or prone positioning.

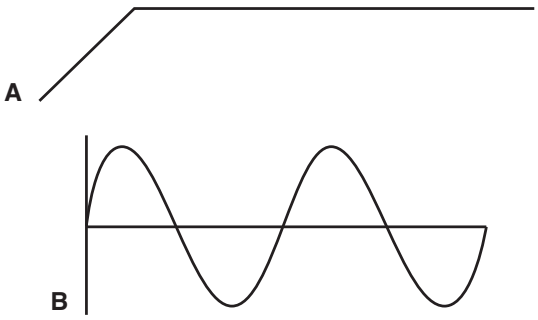


Figure 11.9 Profiles of rotational chair stimuli. **(A)** Trapezoid or step velocity profile. The chair accelerates to a constant velocity, typically 60 and 240 deg/sec. **(B)** Sinusoid profile. The chair oscillates from left to right with peak velocity near 60 deg/sec although the amplitude changes to generate unique test frequencies (typically 0.01, 0.02, 0.04, 0.08, 0.16, 0.23, and 0.64 Hz). Many laboratories use both test profiles because of their unique benefits.

head velocity. Ideally, the eye and head velocities should arrive at equivalent points (i.e., maximum velocity or zero crossings) at the same time. Because the eye and head velocities are in opposite directions, these equivalent points would be 180 deg out of phase. By convention, this is described as zero phase shift (Fig. 11.11). If the eye and head velocities do not arrive at equivalent points at the same time, there is said to be a phase shift, which is labeled as the eye velocity either leading or lagging the head velocity.

In the step velocity test, the cupula will deflect as the chair is accelerated to a preset velocity and the endolymph in the horizontal SCC moves relative to the head. The deflected cupula will return to its rest position (and cease afferent firing) after ~6 sec because there is no further acceleration.[27] However, the neural activity in the medial vestibular nucleus continues to store the velocity signal, which perseverates the nystagmus, which will gradually decay in an exponential fashion. The rate of decay of the nystagmus is measured and called the time constant of the vestibular afferents. The time constant is defined as the time needed for the peak SCEV to reduce to 37% of its peak velocity, and is considered normal between 10 and 25 seconds. The time constant is determined from the slower velocity step rotations (60 deg/sec). Because the time constant and phase are measures of the same processes, the time constant can be determined mathematically using phase data from the 0.01-Hz sinusoid rotation.

During head rotations, the endolymph within the semicircular canals is initially stationary relative to the earth, and thus moves relative to the head (and bony semicircular canal). This relative difference between the stationary endolymph and the rotating head causes endolymph flow and cupular deflection. During rotational chair testing, afferents on the side to which the chair is rotating will be excited, but those on the opposite side will be inhibited. Typically, the nystagmus generated is recorded during ~60 sec of rotation and for ~60 sec upon stopping the chair When the chair stops, the endolymph continues to move

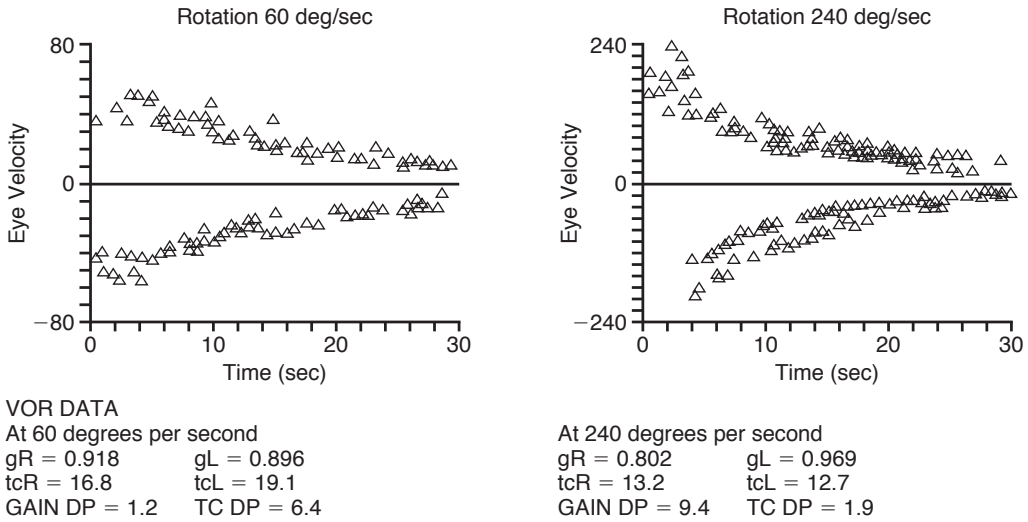


Figure 11.10 VOR gain and time constant during 60 and 240 deg/sec rotations (step profile). Note the magnitude of the VOR gain and the duration of the time constant. The plots appear to normally reduce in velocity over time.

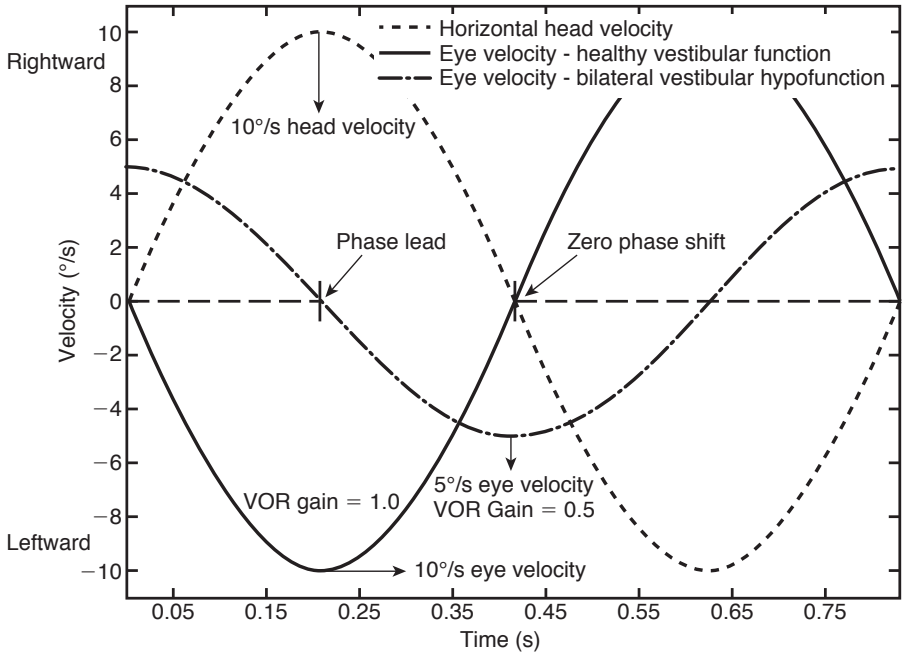


Figure 11.11 Simulated eye rotation during low-frequency sinusoidal head rotation. Positive numbers along ordinate indicate rightward velocity rotation; negative numbers indicate leftward velocity rotation. Arrow line styles match simulated eye velocities. For individuals with healthy vestibular function, as the head rotates to the right at 10 deg/sec, the eyes move to the left at 10 deg/sec, and the eye and head velocity reach zero at the same time (gain = 1, zero phase shift). For those with bilateral reduced vestibular function, eye velocity may be one half or less with respect to head velocity (5 deg/sec in this example, gain = 0.5), and the eyes cross zero velocity in advance of the head crossing zero velocity (eye position leads the head position—phase lead). VOR gain = eye velocity/head velocity. (Reprinted from Schubert and Minor. Vestibulo-ocular physiology underlying vestibular hypofunction. *Phys Ther*. 2004;84(4):373-385, with permission from the American Physical Therapy Association. This material is copyrighted, and any further reproduction or distribution requires written permission from APTA.)

relative to the stationary head, causing the cupulae to deflect in the opposite directions, which causes the excitation-inhibition pattern within the afferents of the horizontal SCCs to reverse. For example, a chair rotation to the right will excite the right horizontal SCC afferents by causing an ampullopetal cupular deflection, and inhibit the left horizontal SCC afferents by causing an ampullofugal cupular deflection. The resultant nystagmus should be right beating. Gradually, the nystagmus will stop, at which point the examiner stops the rotating chair. As soon as the chair stops its rightward rotation, the endolymph within the left horizontal SCC moves such that the cupula is deflected in an ampullopetal direction. Thus, the left horizontal SCC afferents are excited, but the right horizontal SCC afferents are inhibited because of the ampullofugal cupular deflection. The resultant nystagmus should be left beating. This, too, will gradually decline and is used in determining the time constant of the VOR.

In peripheral vestibular pathology, there is a decrease in the time constant and an abnormal phase lead (at low testing frequencies), which never recover. In central pathologies (i.e., cerebellar), the time constant can be abnormally long and there can be an abnormal phase lag. Although the time constant and phase measures do not recover, the VOR gain to low velocity rotations (i.e., 60 deg/sec) does recover. In fact, the rotational chair can be a useful measure of dynamic compensation when the VOR gains for the 60 deg/sec step velocity rotations are normal or when the asymmetry from the sinusoid paradigm tests demonstrate an asymmetry less than 25%. In functional recovery from a chronic unilateral vestibular hypofunction, the VOR gain during 60 deg/sec rotations should be symmetrical. In the case of the faster 240 deg/sec step rotations, VOR gain will still be abnormal for ipsilesional rotations.

The extent of pathology from rotation chair data is determined by comparing VOR gain and phase (or time

constant) measures with established normal values and comparing the gain values for rotations to the right and to the left. Rotary chair testing is limited in its diagnostic ability, because only the horizontal SCCs are assessed to determine extent of pathology. A summary of the critical aspects of the rotational chair exam is provided in Box 11-2.

Otolith Function

Recent advances in vestibular diagnostic testing have extended the region of identifiable pathology to include the otolith organs.[28-30] The vestibular-evoked myogenic potential (VEMP) tests have become an important component of the vestibular function test battery. Two types of VEMP studies exist, cervical and ocular. Each test examines for the presence and absence of the response, abnormally low threshold (i.e., 70 to 80 dB nHL), and/or abnormally large amplitude wave form when the stimulus is applied.

The cervical VEMP (cVEMP) involves recording from electrodes placed over the sternocleidomastoid muscle, which must be contracted during the test. When clicks are introduced to the ipsilateral ear, a disynaptic inhibitory pathway originating from the saccular afferents travels via the medial vestibulospinal tract to ipsilateral sternocleidomastoid (SCM) motoneurons. A transient inhibition of the *ipsilateral* contracted SCM therefore occurs, which is recorded from the electrodes. In healthy vestibular function, an initial inhibitory potential (occurring at a latency of 13 msec after the click) is followed by an excitatory potential (occurring at a latency of 21 msec after the click), with symmetric amplitude and threshold waveforms. For patients with vestibular hypofunction, the cVEMPs may be absent on the side of the lesion. The cVEMP is considered a test of the saccule and the inferior vestibular nerve afferents. The saccule has been implicated as the site of afferent stimulation during cVEMP testing, because saccular afferents provide ipsilateral inhibitory disynaptic input to the SCM muscle,[31] are responsive to click noise,[32-34] and are positioned close to the footplate of the stapes. Therefore, the saccular afferents are subject to mechanical stimulation.[29,32]

The ocular VEMP (oVEMP) is a newer test that places surface electrodes on the superior cheek region, positioned over the inferior oblique (IO) muscle. During the test, subjects must be looking up to bring the IO muscle closer to the electrodes. When clicks to the ear are introduced, or bone vibration to the skull is applied, the *contralateral* IO muscle is excited and EMG is recorded. In individuals with normal vestibular function, the latency of the first waveform is excitatory at 10 msec followed by an inhibitory waveform with a 16-msec latency. The oVEMP is considered a test of the utricle and the superior vestibular nerve afferents. The utricle has been implicated during oVEMP testing based on patients with superior nerve vestibular neuritis (confirmed abnormal caloric and head impulse (thrust) test), who also had absent or reduced oVEMPs.[35] VEMP testing has become particularly useful in assisting the diagnoses of superior canal dehiscence syndrome and Ménière's disease. For further reading on VEMP testing, please see Chapter 12.

Box 11-2

SUMMARY OF THE ROTATIONAL CHAIR EXAM

- Gain at all test velocities should be symmetrical in normal function.
- The higher velocity (>100 deg/sec) step rotations better identify pathologically low VOR gains.
- Lower velocity step rotations are used to determine the time constant and degree of compensation.
- Increased phase lead (sinusoid test) indicates reduced time constant (UVH or BVH).
- Time constant should be >10 sec.
- Only examines the horizontal semicircular canal and related afferents.
- Abnormal high gain and/or long time constant indicate cerebellar pathology (lost inhibition).
- Low-velocity step rotations should be symmetrical when central compensation has occurred.

VOR – vestibulo-ocular reflex; UVH – unilateral vestibular hypofunction; BVH – bilateral vestibular hypofunction

Visual Perception Tests

The subjective visual vertical (SVV) and subjective visual horizontal (SVH) tests are quantified behavioral tests that determine the individual's perception of vertical or horizontal. The behavioral response is presumed to assess otolith function and the central pathways mediating gravitational afference. The visual perception tests cannot be used to uniquely detect saccular or utricular pathology. With the SVV test, patients are asked to align a dimly lit luminous bar (in an otherwise darkened room) with what they perceive as being vertical. With the SVH test, patients are asked to align a similar bar with what they perceive as being horizontal. Subjects with normal vestibular function align the bar within 2.5 deg of true vertical or horizontal, whereas patients with vestibular pathology (central or

peripheral) align the bar greater than 2.5 de g off-vertical (or horizontal).[36,37] The SVV/SVH tests are not able to distinguish central from peripheral pathology. Dieterich and Brandt observed abnormal tilt in 94% of patients with acute unilateral brainstem lesions,[38] and Min et al observed greater than 90% of subjects with acute vestibular neuritis have ipsilateral deviation of SVV.[39] In brainstem pathology, the direction of tilt is usually on the same side of the lesion (ipsiversive), as long as the pathology is caudal to the upper pons. In pathology of the cerebellum (or lesions above the midbrain), the SVV tilt is usually contraversive (opposite the direction of lesion).[40] Whether the SVV test or the SVH test can detect chronic UVH is the subject of debate.[41-43]

Posturography

Recording the postural responses of someone with dizziness has important functional rele vance. Computerized dynamic posturography (CDP) uses force plate technology in measuring center of pressure (COP) under v arious challenging conditions. COP represents the location of all the floor reaction forces from the portions of the body making contact with the force plate. CDP has its main value in objectifying postural responses, documenting change, and identifying malingering.[44] CDP, however, has no diagnostic capability.

In normal function, the maintenance of equilibrium occurs with contribution from the long tracts of the corticospinal and the shorter tracts of the corticobulbar systems (collectively called the pyramidal system), as well as tracts originating from the brainstem nuclei and the basal ganglia (the extrapyramidal systems). The pyramidal system is primarily responsible for controlling f ine, isolated, and multipurpose volitional movement of the limbs, head, and neck. The tracts originate in the cortex and run the length of the spinal cord. The extrapyramidal system is responsible for controlling large, yet cruder motion patterns that are primarily reflexive and postural. These include the vestibulospinal and reticulospinal tracts.

The CDP exam typically includes both a sensory and motor component. The sensory component involves a battery of postural control tests used to assess the patient' s ability to integrate vision, vestibular, and somatosensory inputs to maintain balance. This is typically done by asking subjects to stand for a period of time under six unique conditions (Fig. 11.12). The subject is asked to stand as still as possible, while the area and velocity of the patient's COP is recorded (Fig. 11.13). Abnormal patterns are recognized based on results from the 6-item sensory e xam (Table 11-1).

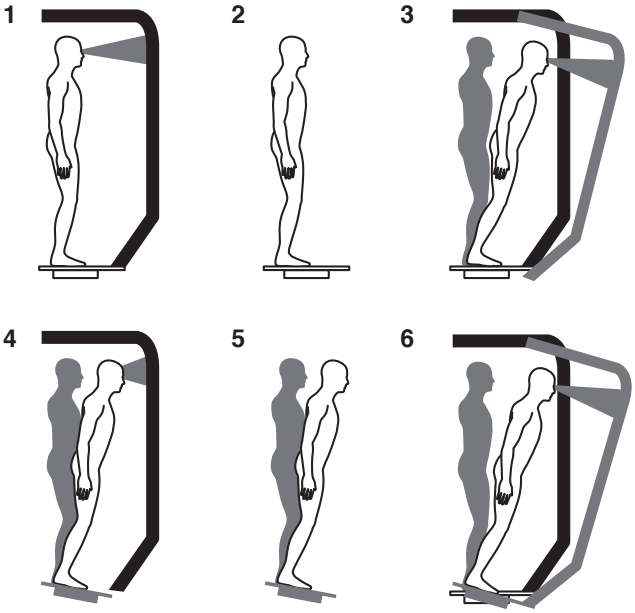


Figure 11.12 The six condition sensory organization test. Condition 1 does not alter any of the three primary sensory contributors for balance. Condition 2 measures center of pressure with eyes closed. Condition 3 distorts visual feedback. Condition 4 distorts proprioceptive feedback. Condition 5 distorts proprioceptive feedback and eliminates vision. Condition 6 distorts visual and proprioceptive feedback, forcing the subject to rely on vestibular input.

The motor component of the CDP commonly includes the motor control test (MCT) and the postural e voked responses test. The MCT assesses postural responses to sudden forward and backward translations of the support surface. The amplitude of the translation varies depending on the test condition. During each translation, the latency to the onset of movements to recover the COP, the distribution of weight through the le gs, and the amplitude of the responses are recorded (Fig. 11.14). It is expected that as the translational perturbation increases, so do the responses. When abnormal, this test indicates possible lesions within the musculoskeletal or long tracts of the neurological system.

Another part of the MCT measures the subject's ability to adapt to a repeated stimulus. In this test, the platform is tilted either up or down at the same amplitude. Normally, when the platform tilts up, muscle spindles from the gastrocnemius muscle are stretched, triggering the monosynaptic stretch reflex to the spinal cord and back.This leads to contraction of the gastrocnemius, which is actually a destabilizing response given the tilt of the platform. In time, a subject should adapt to the stimulus and use less force (smaller amplitude) to maintain balance (Fig. 11.15).

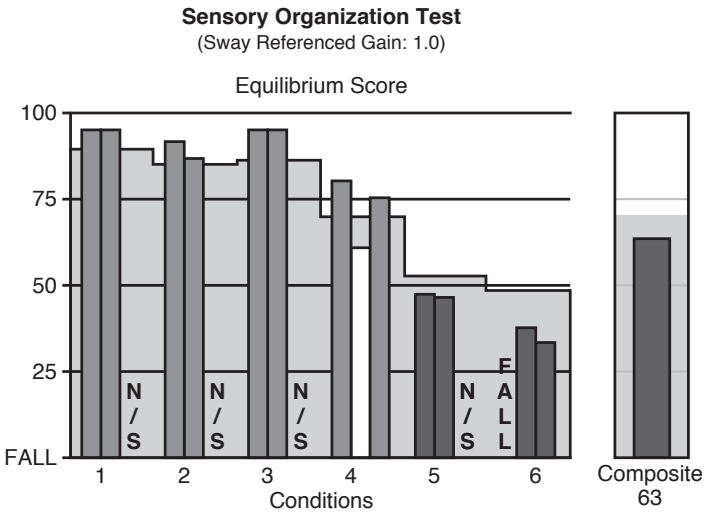


Figure 11.13 Sensory organization test report from the Equitest. Shaded regions represent performance outside normal range. Bars outside the shaded region denote less sway; bars within the shaded area denote more sway and abnormal performance. The patient was not able to maintain balance during the more challenging fifth and sixth conditions. Image courtesy of Natus Medical Incorporated.

■ Table 11-1 **SUMMARY OF ABNORMAL POSTUROGRAPHY PATTERNS**

Abnormal Pattern	Abnormal Test Condition	Interpretation
Visual vestibular dysfunction	4, 5, 6	Suggests difficulty using visual and vestibular input or vestibular input for stance
Visual preference	3, 6	Suggests abnormal reliance of vision for stance
Visual preference/vestibular dysfunction	3, 5, 6	Suggests abnormal reliance on vision and difficulty using vestibular input alone
Somatosensory dysfunction	2, 3, 5, 6	Suggests difficulty using foot support surface and vestibular input
Vestibular dysfunction	5, 6	Suggests difficulty using vestibular input for stance

Surface electromyography from the medial gastrocnemius and tibialis anterior muscles can gi ve important information about the short, medium, and long-loop latency responses e xpected during such tilts. The short-latency (~40 msec) response represents the monosynaptic stretch reflex from the gastrocnemius to the spinal cord and back. The medium-latency (~90 msec) response represents the reflex loop including the gastrocnemius muscle and multiple spinal segments as well as the brainstem. The long-latency (~120 msec) response involves the cortex and leads to contraction of the tibialis anterior muscle as the body attempts to right itself (Fig. 11.16). F or a more thorough read on measuring postural control, please see Chapter 6, Postural Abnormalities in Vestibular Disorders.

Summary

Laboratory measures of v estibular function testing continue to evolve. Within the last 10 years, kno wledge and testing of the otolith end organs has broadened our ability to diagnose lesions within the peripheral v estibular system. The rapid rate at which accelerometers and video cameras continue to become cheaper and faster improves our ability to measure function within the balance system. Even now, video goggle equipment has enabled clinicians to objectify the head impulse test. Re gardless of the imminent advances in vestibular function testing, the primary impetus for its use will be our suspicion of pathology generated from our clinical skills.

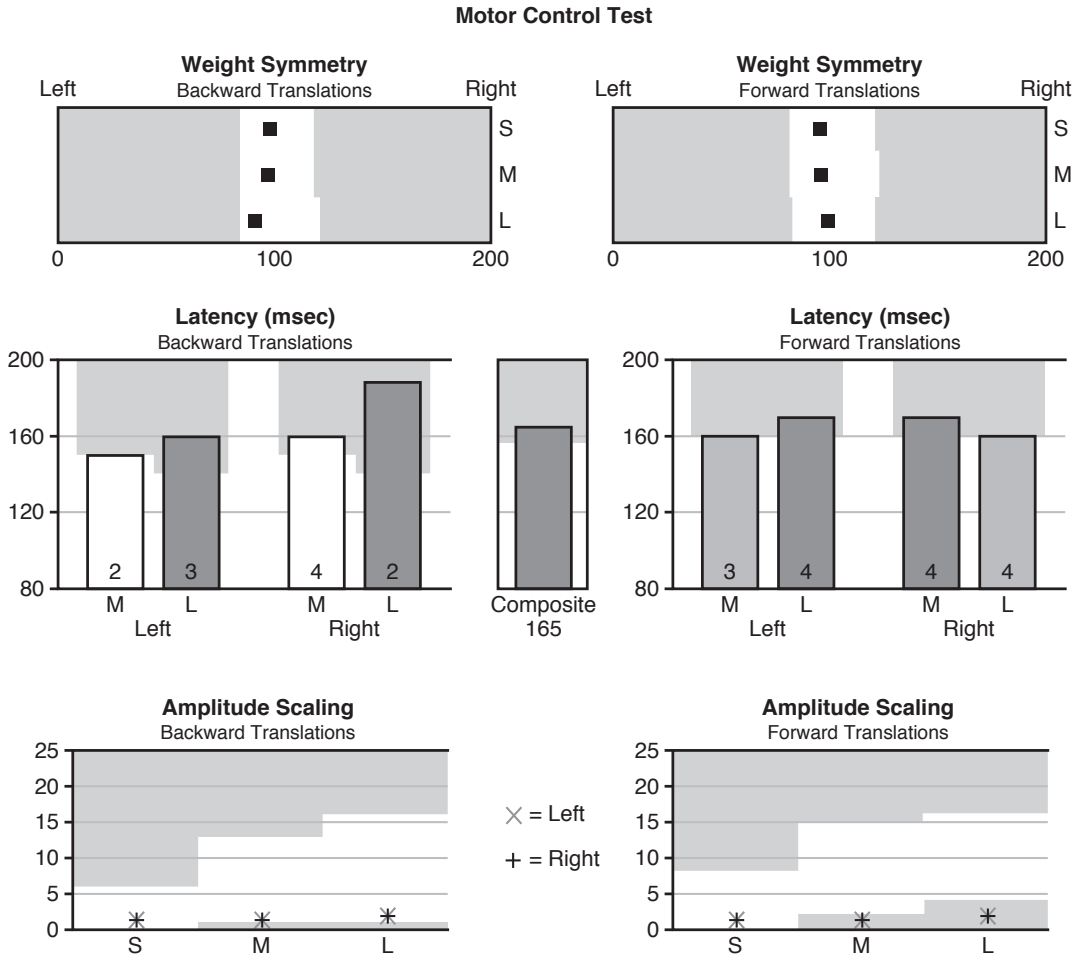


Figure 11.14 Motor control test report from the Equitest. Shaded regions represent performance outside normal function. Bars outside the shaded area denote reduced sway and normal performance; bars within the shaded area represent increased and abnormal sway. Weight is evenly distributed. The latency plots include those for medium and large translations only. The latency for large-amplitude translations are typically shorter than those for medium translations. Image courtesy of Natus Medical Incorporated.

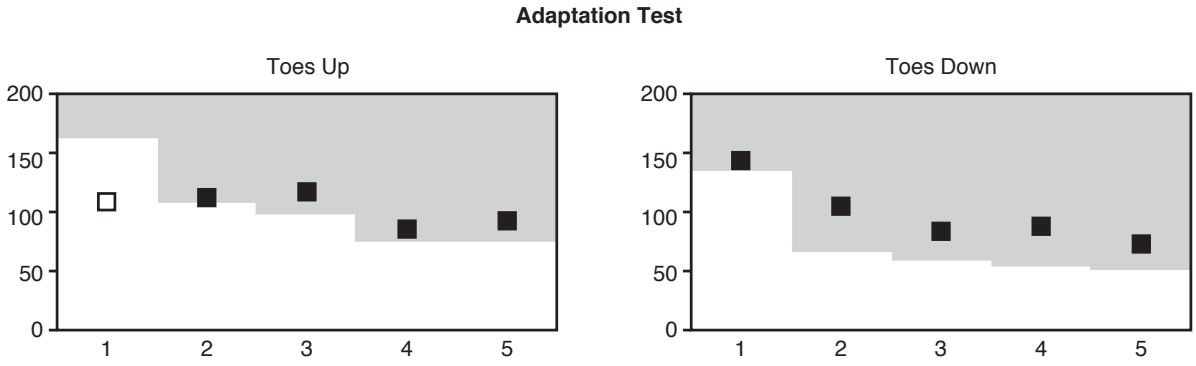
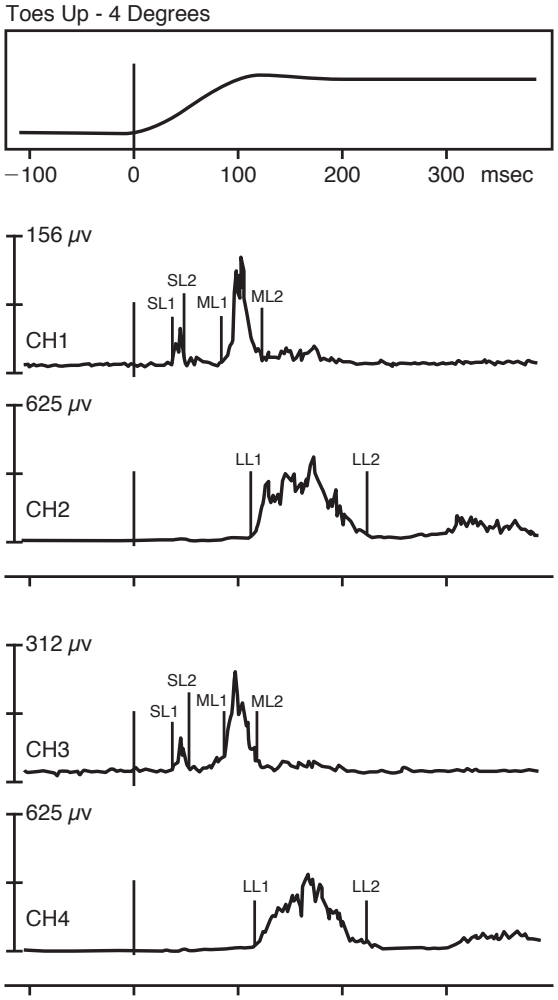


Figure 11.15 Adaptation test from Equitest. Shaded regions represent performance outside normal function. Trials are plotted along the x-axis. This patient is not able to reduce force production with repeated and identical toes up or toes down perturbations. Image courtesy of NatusMedical Incorporated.



CH1	SL1	SL2	ML1	ML2
Time (ms)	37	48	83	122
Amp (μv)	10	11	9	16
Peak (μv)	51		128	
IEMG (μvS)	0.3		1.9	

CH2	LL1	LL2		
Time (ms)	112	223		
Amp (μv)	16	17		
Peak (μv)	387			
IEMG (μvS)	19.8			

CH3	SL1	SL2	ML1	ML2
Time (ms)	36	50	85	118
Amp (μv)	20	24	44	32
Peak (μv)	97		246	
IEMG (μvS)	0.7		4.0	

CH4	LL1	LL2		
Time (ms)	113	220		
Amp (μv)	14	23		
Peak (μv)	342			
IEMG (μvS)	17.3			

Figure 11.16 Responses are averaged from surface EMG of the gastrocnemius (short and medium latencies) and anterior tibialis (long latency) muscles during 20 pitch up platform perturbations. SL – short latency; ML – medium latency; LL – long latency. Four channels are used to record data. Latency, amplitude, magnitude, and integrated EMG are presented in table format. (Integrated EMG is defined as the area under the curve of the rectified EMG signal.) Image courtesy of Natus Medical Incorporated.

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CHAPTER 12

Otolith Function Tests

G. Michael Halmagyi, MD ■ *Ian S. Curthoys, PhD*

The balance organs in the inner ear—the vestibular sensory regions—are the gyros of the human body, and unnoticed, they function continuously in an almost perfect fashion, providing the brain with information about head position and head movement. Until these organs fail, no one really appreciates their significance in daily life, and anyone who has experienced an attack of vertigo will readily verify their importance. The accelerations involved in movements generate forces, and the biological gyros of the inner ear detect these forces: both the force imposed by gravity and the forces generated when we move. The vestibular sensors are constructed in such a way that different regions detect rotational forces and linear forces in any direction. The brain synthesizes the information from these separate force detectors to provide a global integrated summary of where a person is and how the person is moving. This realization is of clinical importance, because it implies that disease affecting only one isolated region of the inner ear has consequences for the overall integration of all the vestibular sensory input. In this chapter, we deal only with the otoliths, the structures that sense linear forces, such as the force of gravity or the "straight line" acceleration experienced in a vehicle accelerating from a stop.

The peripheral vestibular system is sensitive to both linear and angular accelerations: the semicircular canals (SCCs) sense angular acceleration, whereas the otoliths—the saccule and the utricle—sense linear accelerations. Many different ways of testing otolith function have been proposed, including the measurement of horizontal, vertical, and torsional eye movements as well as psychophysical settings in response to linear accelerations produced on swings,[1] sleds,[2] centrifuges,[3] tilt-chairs,[4] and barbeque spits.[5] For an otolith function test to be clinically useful, it must be safe, practical, robust, and reproducible. The test also needs to be specific for, and sensitive to, otolith dysfunction, particularly unilateral otolith hypofunction. In our view, only two means of testing otolith function—the subjective visual horizontal or vertical (SVH or SVV) and the vestibular evoked myogenic potentials—fulfill these requirements. Both have been regularly used in our clinical laboratories for many years, and what follows is in part a distillation of our own experience with these tests and of their scientific basis. The SVV and SVH are perceptual tests that are well known. The newer VEMP tests have been the subject of intense recent interest, and so we devote detailed discussion of them below. Before considering these tests, however, one must have some familiarity with the structure and function of the otoliths.

Anatomy

The basic element of all vestibular transducers is the receptor hair cell, which is similar in both the angular and the linear force-sensing systems (Fig. 12.1 *A* to *E*). The SCCs and the otoliths detect these two different forces, not because of any differences in the intrinsic properties of the hair cells themselves but because of the way the

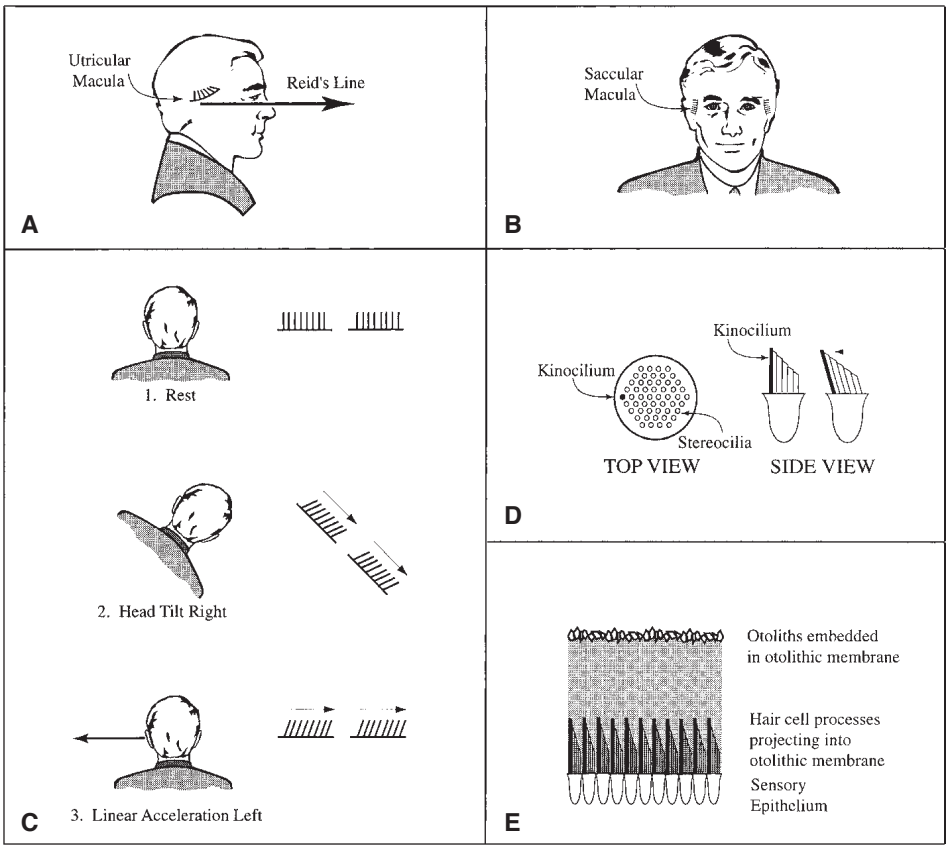


Figure 12.1 The structure and function of the otolithic system. (**A** and **B**) Schematic representation of the approximate orientation of the utricular and saccular maculae in the head. Reid's line is a standard reference line joining the center of the external ear and the lower edge of the bony orbit. (**C)** Three drawings show how different linear force stimuli affect the utricular receptor hair cells. The top diagram (1) shows the back view of a person at rest with the cilia upright. Head tilts to the right (2) cause the cilia to be deflected to the right; similarly, a linear acceleration to the left (3, *arrow)* causes a deflection of the cilia to the right. (**D**) Schematic top-down view of a single receptor hair cell to show the polarization pattern. The kinocilium is located at one extreme of the upper surface of the cell. If the stereocilia are bent toward the kinocilium, as shown at the right, the receptor cell activity increases. (**E***)* Schematic illustration of the organization of a macular surface. The crystals of calcium carbonate (the otoliths) are embedded in one side of the gelatinous otolithic membrane, and the tips of the cilia project into the other side of this membrane. Forces cause the membrane to slide so that the cilia are bent, thus stimulating the receptor cells.

structures that surround the hair cells are affected by the stimulating force. Each receptor cell has se veral, fine, hairlike *cilia* projecting from the cell body into a gelatinous overlying mass, the *otolithic membrane*. The longest cilium is called the *kinocilium;* it is located at one side of the receptor cell and has a specialized cross-sectional structure. The remaining cilia, the *stereocilia*, are of uniform cross-sectional structure and are arranged in a series of increasing height as the y approach the kinocilium. Linear accelerations such as tilts or translations cause the otolithic membrane to slide so that the cilia of the receptor hair cells are bent (Fig. 12.1*C*).

Each labyrinth consists of endolymph-filled tubes and sacs containing receptor structures. The two specialized sacs are the *utricular sac* and the *saccular sac*. A plate of specialized receptor hair cells and connecti ve tissue is contained within each of these sacs, and that plate is called the *macula*. Each macula takes its name from the sac in which it is located, so that in each inner ear there is a utricular macula and a saccular macula. The terms *utricle* and *saccule* are often used to refer to the utricular and saccular maculae, although strictly speaking, *utricle* and *saccule* refer to the membranous sacs that contain the maculae.

The two maculae have similar gross features. Dense crystals of calcium carbonate, the *otoconia* (Fig. 12.1*E*), are embedded on the outer (free) surface of the otolithic membrane. On the inner surface of this membrane, the cilia of the hair cells project into the membrane. The density of the otolithic membrane itself is similar to that of the surrounding endolymph, at 1.0 g/cm^3, but the density of the otoconia is almost three times greater, at 2.7 g/cm^3. These two receptor structures are together called the *otoliths*, because their construction is similar and because both detect forces according to the same physical principle—namely, that an imposed linear force displaces the relatively dense otoliths embedded in the otolithic membrane. The otolithic membrane then tends to slide across the surface of the macula, displacing the cilia of the receptor hair cells and producing a change in hair cell resting membrane potential. The displacements of the otolithic membrane generated in this way by natural head movements in a 1-*g* environment are less than one micron.

The utricular and saccular maculae are approximately perpendicular to each other. With the head erect, the utricular macula is tilted by about 30 degrees (open anterior) with respect to the horizontal plane of the head, whereas the saccular macula is almost vertical. These different orientations mean that the receptors on each macula respond maximally to forces in different directions. The surface area of the human utricular and saccular maculae is only about 4.2 mm^2 and 2.2 mm^2, respectively. There are about 33,000 receptor hair cells in the utricular macula and about 19,000 in the saccular macula. As in other sensory systems, there is convergence of receptors to primary afferent nerves, so that only about 6,000 primary afferent nerve fibers supply the utricle, and only about 4,000 supply the saccule.

Otolith Function

Intracellular recordings from single isolated otolithic receptor hair cells subjected to controlled forces in precisely defined directions have shown that each receptor hair cell is polarized [6]; that is, (1) it responds most strongly to forces in one direction, and (2) as the direction of the force deviates from that optimal direction, so the response of the receptor declines. This optimum direction is referred to as the cell's *polarization vector*. This physiological polarization corresponds to a morphological polarization, in that receptor cells respond optimally when the imposed force shears the stereocilia toward the kinocilium. Receptor hair cells are arranged in a regular fashion across each macula so that the direction of the kinocilium of each cell shifts in direction by only a small amount relative to its neighbors. The result is a highly ordered arrangement of polarization vectors across each macula (Fig. 12.2). For both maculae, the preferred directions of cells are opposite on each side of a line in each macula. This line, and the area around it, is called the striola. In the utricular macula, the polarization vectors of the receptors point toward the striola, whereas in the saccular macula the vectors point away from the striola (Fig. 12.2). At the band around the striola, there is a concentration of type I receptors, with large amphora-shaped cell bodies enveloped by a calyx ending. These receptors are in contrast to the other cylindrical (type II) receptors in the maculae periphery, which have a cylindrical cell body and small bouton, afferent nerve terminals. Afferents synapsing on type I receptors respond very strongly to *changes* in linear acceleration, and so play a major role in otolithic dynamic function: afferents synapsing on type II receptors faithfully signal maintained linear accelerations.[7]

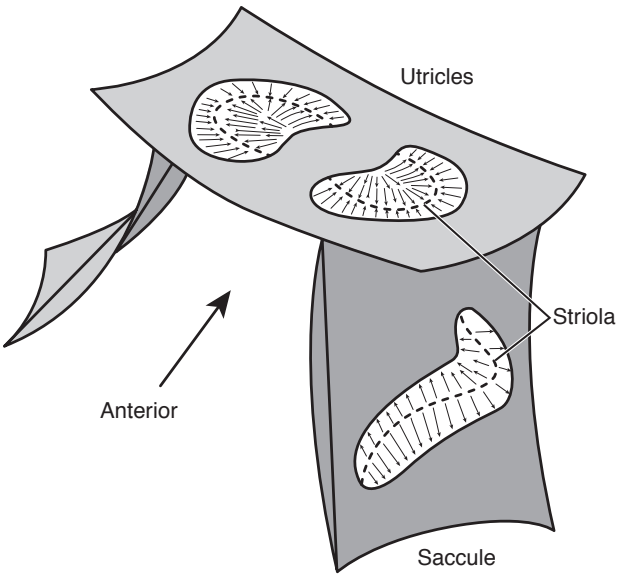


Figure 12.2 Schematic representation of the plates of receptor hair cells in each macula. The arrows show the preferred orientations of the receptor hair cells on each macula. On both maculae, hair cells have exactly opposite preferred orientation on either side of an imaginary line (or band) called the striola—shown as a dashed line. So, in each macula a linear acceleration will cause maximal activation of hair cells in one sector and maximal inhibition of hair cells with the opposite preferred orientation. Uchino's group has shown that central inhibitory interaction between afferents from the two opposing sides acts to enhance sensitivity—cross-striolar inhibition.[8] The amphora-shaped type I receptors are concentrated in the area of the striola, and afferents from these have an irregular resting discharge and a strong response to changes in linear acceleration[7] and appear to have a major role in generating otolithic responses to dynamic stimulation—such as VEMPs.

Primary afferent neurons synapse on a number of adjacent receptor hair cells, so that some of the precision of spatial tuning of the individual receptors is lost: afferent fibers respond to forces over a greater range of directions than the individual receptors. Each afferent fiber exhibits a preference for forces in a particular direction, reflecting the preferred orientation of the receptor hair cells on which the fiber synapses. For afferent fibers, just as for receptors, as the force direction deviates from the optimal direction, the fiber's firing rate declines. Utricular afferents in general show a preference for forces directed horizontally across the plane of the macula—in other words, for laterally directed forces—and similarly, saccular afferents show a preference for forces directed vertically across the plane of the macula.[9]

Primary Otolithic Afferents—Physiology

Recordings from primary otolithic afferent neurons in the vestibular nerve of experimental animals show a number of important characteristics (for a review see Goldberg and Fernandez.[10]

1. Primary otolithic afferents have a resting discharge rate. They fire at about 50 spikes per second even when there is no imposed force stimulus.
2. The spontaneous activity is highly regular in some afferents, whereas in others it is highly irregular. In addition, there is a continuum of regularity between these extremes. Physiological responses are closely related to this dimension of regularity. Irregular neurons show a strong response to changes in linear accelerations (jerks), whereas regular neurons show a faithful response to maintained linear accelerations.
3. Afferents have a *functional polarization vector*. This means that linear accelerations oriented in one particular direction will most effectively activate the afferent neuron. As the direction of the imposed acceleration is deviated from this optimum direction, so the response of the afferent neuron declines. If the stimulus is directed exactly opposite to the preferred direction, then the firing of the cell is maximally suppressed.
4. The afferents show a marked asymmetry in bidirectional sensitivity. This characteristic results in an increase in firing rate for acceleration in the excitatory direction that is larger than the decrease in firing rate for the same magnitude of acceleration in the opposite, disfacilitatory direction.
5. The spatial distribution of the directional preferences of all otolithic afferents is not uniform. In the squirrel monkey, for example, there is a directional preponderance so that more otolith afferents prefer ipsilaterally directed accelerations to contralaterally directed accelerations. There is 3:1 preponderance of utricular afferents preferring ipsilateral accelerations, whereas there is about a 1:1 distribution of saccular afferents preferring up-and-down accelerations.

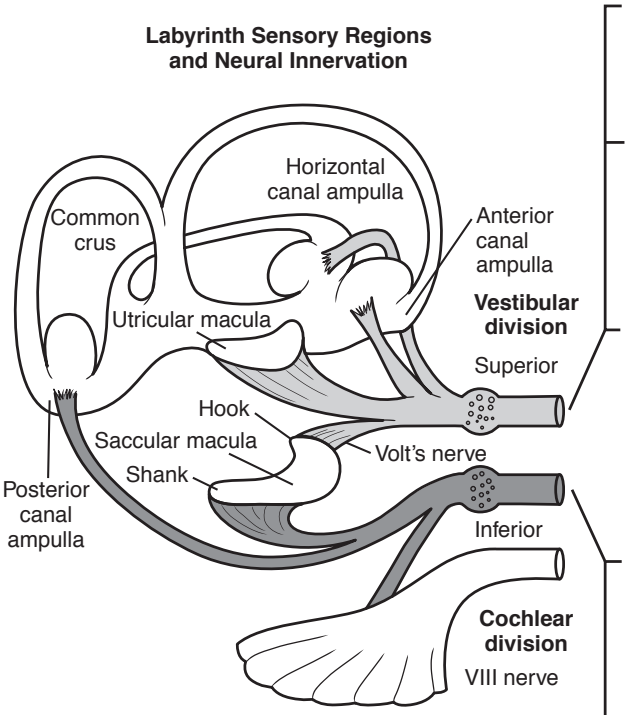
The dynamic response characteristics of regular and irregular otolithic afferents are quite different and indeed almost complementary. Regular neurons show a poor response to changes in linear acceleration and little adaptation to maintained accelerations. Irregular afferents fire vigorously during changes in linear acceleration but adapt rapidly (but not completely) for a maintained stimulus.[9,11,12] Bone conducted vibration (BCV) consists of many rapidly changing linear accelerations.[13,14] Recordings of primary vestibular neurons in guinea pigs show that low-intensity 500-Hz BCV activates a high proportion of otolith irregular neurons from the striolar region of *both* the utricular and saccular maculae, and many are activated with great sensitivity at very low stimulus values—thresholds of around 0.1*g*.[13-16]

BCV at 500 Hz activates very few semicircular canal neurons and then only at high intensities.[13,14] However, our recent results show that as the frequency is decreased to 100 Hz, canal neurons are also activated by vibration (Curthoys 2013, unpublished results). Air-conducted sound (ACS) evokes neural responses in many of these same neurons similar to those evoked by BCV, although the stimulus intensities required are very high (60 to 80 dB above auditory brainstem response [ABR] threshold; around 120 to 130 dB sound pressure level [SPL]).[14,15] It is still not known just how this transduction occurs, but it seems that the pumping of the stapes is sufficient to generate displacements of the cilia of the receptor cells. Most, but not all, neurons activated by BCV can be activated by ACS.[14] Because 500-Hz BCV and ACS are relatively specific stimuli for otolith irregular neurons,[13] so myogenic responses to ACS and BCV have been used to index dynamic otolith function in the clinic.[17,18] It is likely that responses to lower frequency stimulation may be contaminated by canal activation. This means that even at the level of the afferent neurons leaving the macula, functional specialization

has already taken place, in that different temporal and spatial aspects of the force stimulus are being signaled by these different parallel pathways.

Central Projections

The projections of otolith neurons are crucial in interpreting clinical otolith tests. In addition, the organization of the peripheral neural projections is especially important, because disease (neuritis) may be restricted to individual branches of the vestibular nerve. As we explain below, this differential vulnerability has been used to identify utricular as opposed to saccular function. The schematic figure (Fig. 12.3) shows that (1) afferents from the horizontal and anterior semicircular canals and the utricular macula course in the superior division of the vestibular nerve,[19,20] and (2) most saccular afferents course in the inferior vestibular nerve, except the branch from the small rostral area of the saccular macula (the "hook" of the saccular macula) called Voit's nerve,[21] which travels in the superior vestibular



Clinical Test	Healthy Subject	Superior Vestibular Neuritis	Inferior Vestibular Neuritis	Unilateral Vestibular Loss
Horizontal head impulse test to ipsilateral side—tests ipsilateral horizontal canal	✓	X	✓	X
Pitch head impulse test in the plane of the ipsilateral anterior canal, nose down—tests ipsilateral anterior canal	✓	X	✓	X
oVEMP n10 beneath the contralateral eye to bone conducted vibration at Fz, or air-conducted sound of one ear—tests utricular macula of the ear opposite the measured eye	✓	X	✓	X
cVEMP p13-n23 over the ipsilateral sternocleidomastoid (SCM) muscle to bone conducted vibration at Fz, or air-conducted sound of one ear—tests saccular macula of the ear on the same side	✓	✓	X	X
Pitch head impulse test in the plane of the ipsilateral posterior canal, nose up—tests ipsilateral posterior canal	✓	✓	X	X

✓ = Normal response X = Abnormal response

Figure 12.3 Schematic representation of the course of the afferents from each sensory region of the vestibular labyrinth. The text in the figure describes new clinical tests. Tests of each sense organ of the vestibular labyrinth and the columns show the pattern of responses associated with each major disease category. oVEMP is the ocular vestibular-evoked myogenic potentials; cVEMPs is the cervical vestibular-evoked myogenic potentials (described below). Fz is the location on the forehead in the midline at the hairline. Reprinted from Curthoys[22] "The interpretation of clinical tests of peripheral vestibular function." *Laryngoscope*, 122(6):1342-1352, DOI 10.1002/lary.23258, with permission from John Wiley and Sons.

nerve. In primates, Voit's nerve constitutes only about 10% of all the afferents from the saccular macula.[23] So, there are some saccular fibers coursing in the superior nerve, although the evidence is that this is not a large contingent.

Primary otolithic afferent neurons project to secondary vestibular neurons mainly in the lateral, medial, and descending vestibular nuclei. In some regions, these otolithic projections show considerable overlap with the projections of horizontal SCC afferents. The predominant response of lateral vestibular nucleus neurons is an increase in firing rate in response to ipsilateral tilts. Just as horizontal SCC neurons in the medial vestibular nucleus are interconnected to the contralateral medial vestibular nucleus via commissural fibers, which play a major role in the neural operation of the system, so otolith-responsive neurons are interconnected to similar neurons in the opposite vestibular nucleus.[8]

However, the interconnections in the otolithic system are indirect, and the functional mode of the bilateral interconnections is predominantly excitatory rather than inhibitory, as is the case with the horizontal SCC system. Commissural interaction occurs in the otolithic and the horizontal canal system.[8] Some cells in each vestibular nucleus can be activated both by SCC stimulation and by otolithic.[24,25] This is not too surprising—nodding the head causes activation of both semicircular canals and sectors of both otoliths, and the cooperative interaction of canal and otolith neurons will generate a smooth compensatory response. These convergent neurons integrate linear and angular acceleration input and thus help maintain posture and equilibrium. However, the existence of these convergent neurons means that pathways conveying otolith signals are not specific; many neurons can be activated by either canal or otolith stimulation.

The pathways from the otolithic regions of the vestibular nuclei to the ocular motor nuclei are poorly understood. Trochlear motoneurons can be disynaptically activated by electrical stimulation of the utricular nerve, and increasing attention is being paid to two midbrain regions close to the oculomotor nuclei—the interstitial nucleus of Cajal and the rostral interstitial nucleus of the medial longitudinal fasciculus—that integrate inputs for torsional and vertical eye movement responses produced by otolithic stimulation. In particular, it seems that the interstitial nucleus of Cajal has a major role in integrating otolithic input for coordinated eye and head responses to the linear forces detected by the otoliths. The neural substrate for the compensatory postural movements required by a linear acceleration are mediated by otolith-spinal projections, the lateral vestibulospinal tract arising in the lateral vestibular nucleus and the medial vestibulospinal tract arising in the medial vestibular nucleus.

Function of Otolithic Input

Electrical stimulation of the utricular nerve in the cat causes a distinct pattern of eye movement: a torsion of both eyes so that the upper poles of the eyes roll away from the side being stimulated[26] because of activation of the contralateral inferior oblique and ipsilateral superior oblique muscles. Complementing that finding, studies have shown that unilateral section of the vestibular nerve causes a torsion of both eyes toward the operated side.[5,27] Bone conducted vibration elicits eye movements in guinea pigs and humans that are analogous to those produced by electrical stimulation of the utricular nerve.[28,29] In natural head movements, the otoliths are activated and generate compensatory eye and postural responses. For example, tilting the head toward one shoulder causes the gravity vector to activate particular regions of the utricular and saccular maculae, and as a consequence, the eyeball *torts* (or *rolls*) around the visual axis in a compensatory direction. At the same time, there is a complex pattern of activation of neck and trunk muscles acting to oppose this challenge to the equilibrium of the head. The degree of this countertorsion or ocular *counter-rolling* is only about 10% of the head tilt, but it does depend on otolith function because subjects without otoliths do not show such counter-rolling.[6]

Clinical Tests of Otolith Function

Static Otolith Tests

Otolith primary afferents respond to maintained linear accelerations (such as head tilts re gravity) and a number of clinical tests examine the responses of patients to maintained linear accelerations.[30,31] We refer to these as tests of "static" otolith function to contrast them to the tests of "dynamic" otolith function, such as the VEMP tests described below. Maintained head tilt activates otolith receptors and causes both eyes to roll around the line of sight and maintain that rolled position during the maintained (i.e., static) head tilt. Conversely, if there is unilateral loss of otolith function, both eyes roll around the line of sight and adopt a maintained rolled-eye position—rolled toward the lesioned ear.[32] Unilateral otolith loss was probably the cause of the ocular tilt reaction because of an inadvertent vestibular lesion in a patient undergoing stapes surgery.[33] On recovery, the patient showed a distinctive pattern of responses that have been taken to indicate otolith function—the ocular tilt reaction: (1) both eyes adopted a maintained rolled position around the line of sight toward the affected side; (2) there was a small maintained roll-tilt of the head toward the affected side; and (3) there was a maintained skew deviation between the two eyes—the visual axis of

the eye on the side of the lesion was lower in the orbit than the visual axis of the eye on the intact side.

In human subjects with the eyeball in a rolled position, visual perception changes in accordance with this new torsional position[34] e.g., when a patient with maintained ocular torsion is asked to set a visible line—in an otherwise darkened room—to where they perceive gravitational horizontal (or vertical) to be, they set it in a direction that is aligned with their rolled eye position. This test is called the subjective visual horizontal (SVH) or subjective visual vertical (SVV).[3,29,34-38] Systematic deviations in SVV or SVH greater than about 2 degrees, in patients with peripheral vestibular dysfunction, are taken to indicate static otolith dysfunction.

Subjective Visual Horizontal or Vertical Testing of Otolith Function

Peripheral Vestibular Lesions

A normal subject sitting upright in a totally darkened room can accurately set a dimly illuminated bar to within 1 degree of the true gravitational vertical or horizontal. Friedmann[35,36] was the first to show that patients with various unilateral vestibular lesions set such a bar so that it was no longer aligned with the gravitational vector but was consistently tilted toward the side of the lesion (see Case Study 21-1). We studied the ability of patients with Ménière's disease to set such a light-bar to the visual horizontal before and after unilateral vestibular neurectomy (UVD).[3,27] Before UVD, the patients' settings were within the normal range. After UVD, patients invariably set the gravitationally horizontal bar so that it was actually tilted toward the lesioned side, by 15 degrees or more; they did this because they actually saw the gravitationally horizontal bar as being tilted toward the intact side. Although their settings of the bar gradually returned toward the true or gravitational horizontal, the settings were still tilted by a mean of 4 degrees 6 months or more after UVD. Thus a slight ipsilesional tilt, or *offset*, of the SVH was a permanent legacy of a UVD procedure for treatment of Ménière's disease. These findings keep being confirmed.[39-50]

What could be the cause of this perceptual error? Is it an offset of the internal representation of the gravitational vertical as a result of the profound asymmetry in otolithic input to the vestibular nuclei that must occur after UVD? Arguing against this mechanism is the observation that despite the UVD, the patients do not feel that their own bodies are tilted, but on the contrary, feel themselves to be normally upright, even in the dark. In other words, although they tilt the bar toward the UVD side, it is not to null a perceived tilt of the bar *with* the body, toward the intact side. Another possible mechanism of the SVH offset is a torsional deviation of the eyes as a part of the ocular tilt reaction. The *ocular tilt reaction* is a postural synkinesis consisting of head tilt, conjugate ocular torsion, and skew deviation, all toward the same side. Some patients demonstrate a florid, temporary, ipsilesional tonic ocular tilt reaction after a unilateral peripheral vestibular lesion,[33-51] and others just a partial one with ocular torsion and skew deviation.[51-55]

We measured torsional ocular position and SVH before and after UVD[27] and found that after UVD, there was invariably an ipsilesional deviation of torsional ocular position so that the 12 o'clock meridian of each eye was invariably rotated toward the side of the UVD (Fig. 12.4). One week after UVD, there was up to 15 degrees of ipsilesional ocular torsion and a close correlation ($r = 0.95$) between the magnitude of the ocular torsion and the offset of the SVH (Fig. 12.5). Furthermore, the ocular torsion gradually resolved with a temporal pattern identical to that of deviation of the SVH. One month after UVD, both the ocular torsion and the tilting of the SVH were at half the 1-week value. A slight but significant ipsilesional ocular torsion of 4 to 5 degrees is a permanent legacy of UVD for Ménière's disease.[27] After acute UVD in frogs,[56] dogs,[57,58] cats,[57] and horses,[59] there is head torsion, which follows the same time course as conjugate eye torsion in humans.

The offset of the SVH is caused by ocular torsion as part of the ocular tilt reaction, so that maintained ocular torsion can be seen as the equivalent in the otolithic system of the spontaneous nystagmus in the semicircular canal system. Torsional ocular position and, therefore, the setting of the SVH, depend on relative resting activity in the left and right vestibular nuclei. So, as the brainstem compensates for the UVD, the SVH returns toward normal, although a small offset of the SVH appears to be a permanent stigma of UVD.[27] Therefore, in analogy with spontaneous nystagmus, a return of the SVH toward normal is inevitable whether or not the labyrinth recovers. It should be noted that offsets of torsional ocular position and of the SVH can also occur with SCC stimulation—that is, together with horizontal or torsional nystagmus.[60,61] Nonetheless, ocular torsion in the absence of spontaneous nystagmus is likely to be otolithic, because it represents a tonic offset of the dynamic ocular counter-rolling mechanism, which is under utricular control (Fig. 12.6).[4,62]

Central Vestibular Lesions

Patients with acute brainstem,[63,64] cerebellar,[65] and cortical[66] lesions (ischemic or demyelinating,[63,66]) can show offsets of the SVH or SVV and of torsional eye position.[67]

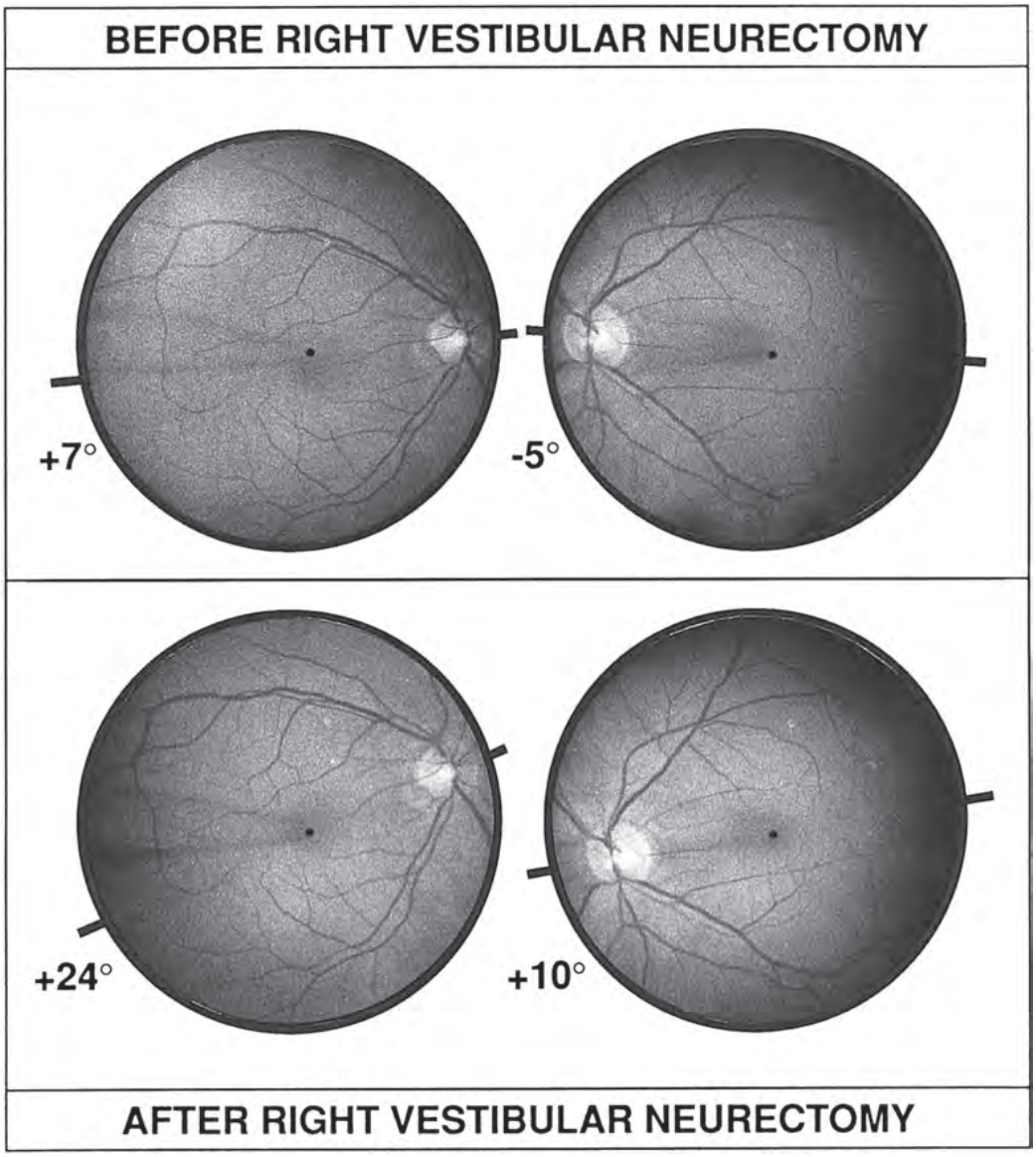


Figure 12.4 Ocular torsional position before and after unilateral vestibular deafferentation (uVD). Fundus photographs of the left and right eyes of a patient before *(top row)* and one week after *(bottom row) right* vestibular neurectomy. After the operation, there is tonic rightward torsion of the 12 o'clock meridian of each eye toward the patient's *right side*. The torsion measures 17 degrees in the right eye and 15 degrees in the left eye. When the patient was asked to set a luminous bar to the perceived visual horizontal in an otherwise darkened room, he set the bar tilted down on his right side by 14.2 degrees when viewing with the right eye and 15.1 degrees when viewing with the left.

Patients with lower brainstem lesions involving the vestibular nucleus (e.g., lateral medullary infarcts) offset the SVV toward the side of the lesion, whereas patients with upper brainstem lesions involving the interstitial nucleus (medial thalamic infarcts) or with cerebellar lesions involving the dentate nucleus have offsets of torsional eye position and of the SVV away from the side of the lesion. In most patients, there is a deviation of torsional ocular position (also called *cyclotorsion*) in the same direction as the offset of the SVV. The relationship between the SVV and ocular torsion is not as tight as with peripheral lesions but is nonetheless present.[67] In patients with peripheral vestibular lesions, the ocular torsion and the consequent setting of the SVV are almost the same in each eye; in contrast, there can be significant left eye–right eye differences in both ocular torsion and the SVV in patients with central vestibular lesions. For example, in patients with lateral medullary infarcts, the excyclotorsion of the ipsilesional

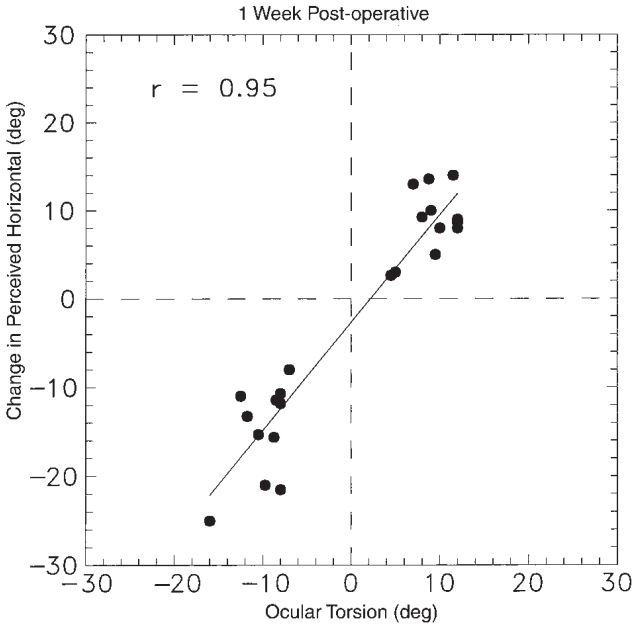


Figure 12.5 The relationship between ocular torsional position and the subjective visual horizontal. The average value 1 week after unilateral vestibular deafferentation of the change in ocular torsional position was calculated for each patient and correlated with that patient's average change in the visual horizontal. The correlation (0.95) is statistically significant. (From Dai et al, 1989.[3])

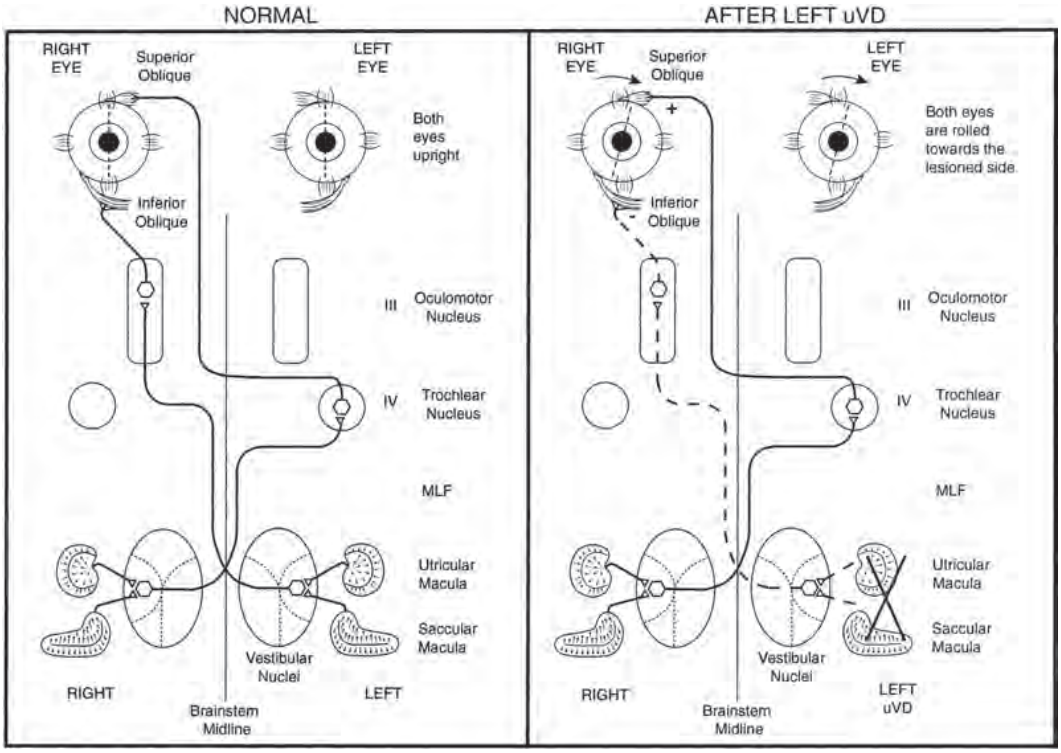


Figure 12.6 Diagrammatic explanation of the possible causes of the changes in torsional eye position after unilateral vestibular deafferentation (uVD). Normally, second-order afferents from the vestibular nucleus send excitatory projections to the contralateral inferior oblique and ipsilateral superior oblique muscles, as well as to the contralateral inferior rectus and ipsilateral superior rectus muscles (*not shown*). Left uVD, as shown here, produces reduced tonic activity in the contralateral inferior oblique (and inferior rectus, *not shown*) muscles, so that the contralateral eye intorts; it also produces reduced activity in the ipsilateral superior oblique (and superior rectus, *not shown*) muscles, so that the ipsilateral eye extorts. One presumes that, through commissural disinhibition, the left uVD increases tonic activity in the contralateral, right vestibular nucleus, and therefore increases tonic activity of the contralateral superior oblique muscle, which also produces intorsion of the contralateral eye. IV = trochlear nucleus; MLF = medial longitudinal fasciculus. (Courtesy of Ms. Agatha Brizuela.)

eye can be larger than the incyclotorsion of the contralesional eye.

Clinical Significance

Standardized measurement of the SVH, with a dim light-bar in an otherwise totally darkened room, can give valuable diagnostic information. In some laboratories, patients are asked to set a bar to the SVV, but we find that most patients have a better intuitive understanding of the horizontal than of the vertical and that the settings of the vertical might not be the same as those of the horizontal.[69,70] In any case, in order for the test to be valid, either the room must be totally dark apart from the light bar or there must be some other way, such as with a Ganzfeld stimulator or a rotating dome,[63] or even a simple bucket, to exclude all visual cues.[70,71] A significant offset of the SVH or the SVV indicates acute unilateral otolithic hypofunction from a lesion of the end-organ, the vestibular nerve, or the vestibular nucleus on the side to which the patient offsets the bar, above the vestibular nucleus on the side opposite to which the patient offsets the bar. The greater the offset, the more acute the lesion; a small permanent deviation of the SVV might be a permanent legacy of both central [72] and peripheral[46] vestibular lesions. The SVH test is the single most useful investigation in the acute phase of suspected vestibular neuritis: SVH is offset, sometimes by more than 20 degrees, always toward the side of the lesion.[43,44] SVH testing can also be used to follow the progress of vestibular loss and compensation after intratympanic gentamicin treatment for Ménière's disease.[42,45] Offsets of the SVH seem to correlate better with abnormalities in oVEMPS than in cVEMPs.[73]

CASE STUDY 12-1

A 61-year-old, previously well male business executive experienced sudden intense vertigo and nausea while driving home from work. He had to stop his car, vomited, and called for help. He was taken by ambulance to a hospital emergency room. On admission, he was distressed by vertigo, retching, and vomiting. He was unable to stand. There was no spontaneous or positional nystagmus with or without visual fixation; the head impulse test result was negative vertically and horizontally. A CT scan of the brain was normal. He was admitted to the hospital with the provisional diagnosis of cerebellar infarct. The following day he felt better and could stand without support. At that time, he noted that he could not hear in his left ear. Over the next 3 days his balance continued to improve, but his hearing did not. Investigations at that time revealed the following:

Audiogram: Severe flat sensorineural hearing loss left ear; slight conductive loss right ear (Fig. 12.7*A*).
Electronystagmogram: Minimal left-beating, gaze-evoked nystagmus in the dark; normal caloric test results (Fig. 12.7*B*).
Subjective visual horizontal: More than 6 degrees to the left (Fig. 12.7*C*).
Cervical vestibular-evoked myogenic potentials: Absent from left ear to clicks and to taps (Fig. 12.7*D*).

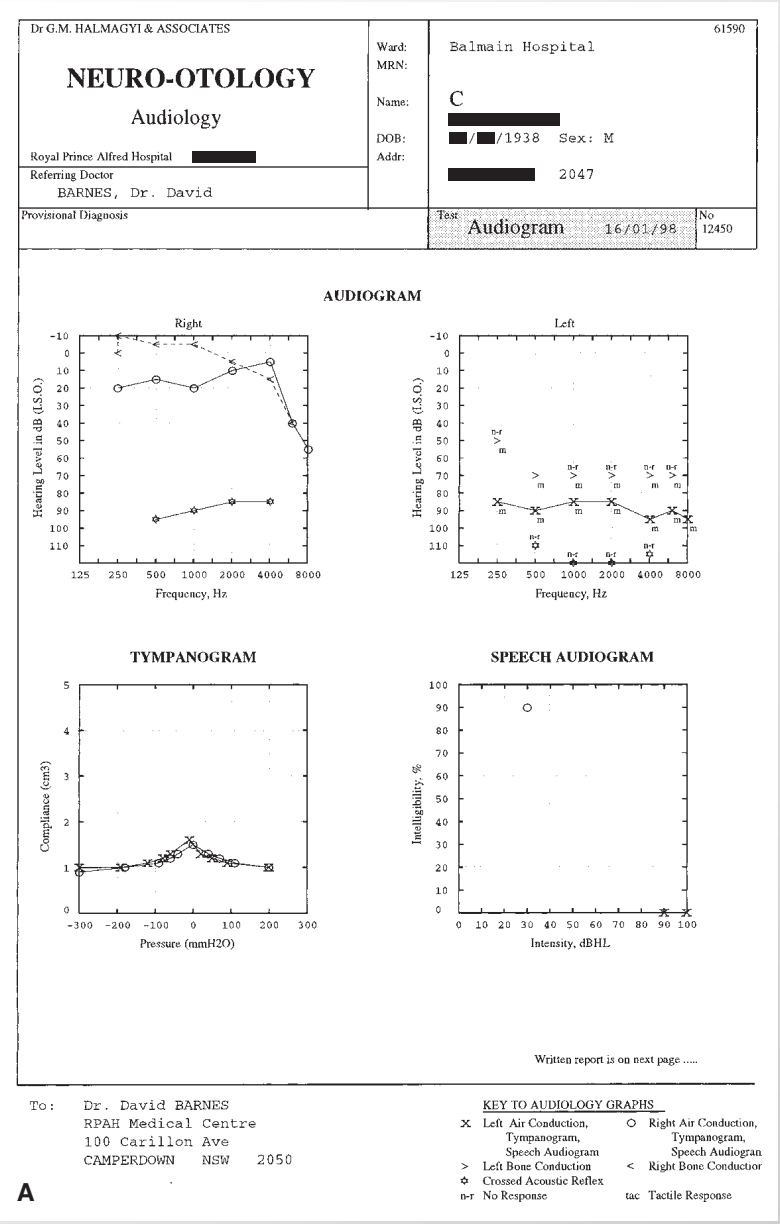
Magnetic resonance imaging with contrast showed no abnormality—in particular, neither cerebellar infarction nor contrast enhancement of the inner ear.

Despite the absence of pain and vesicles, the patient was given a tapering course of prednisone and acyclovir on the chance that this episode was caused by herpes zoster. His balance continued to improve, but even 2 weeks later, he still rotated to the left on the Unterberger (Fukuda) test and fell on the matted Romberg test. Within 1 month his hearing also improved but not quite back to normal, and SVH and cVEMP values both returned to normal.

Comment

This patient had acute neurolabyrinthitis affecting the cochlea, the saccule, and the utricle but sparing the SCCs. In some cases of acute neurolabyrinthitis, as in this one, the inner ear can recover most or all function. However, even if the ear had not recovered (as would have been indicated by persistent loss of hearing and VEMP), the SVH would have returned to normal through central vestibular compensation (see Chapter 8).

CASE STUDY 12-1

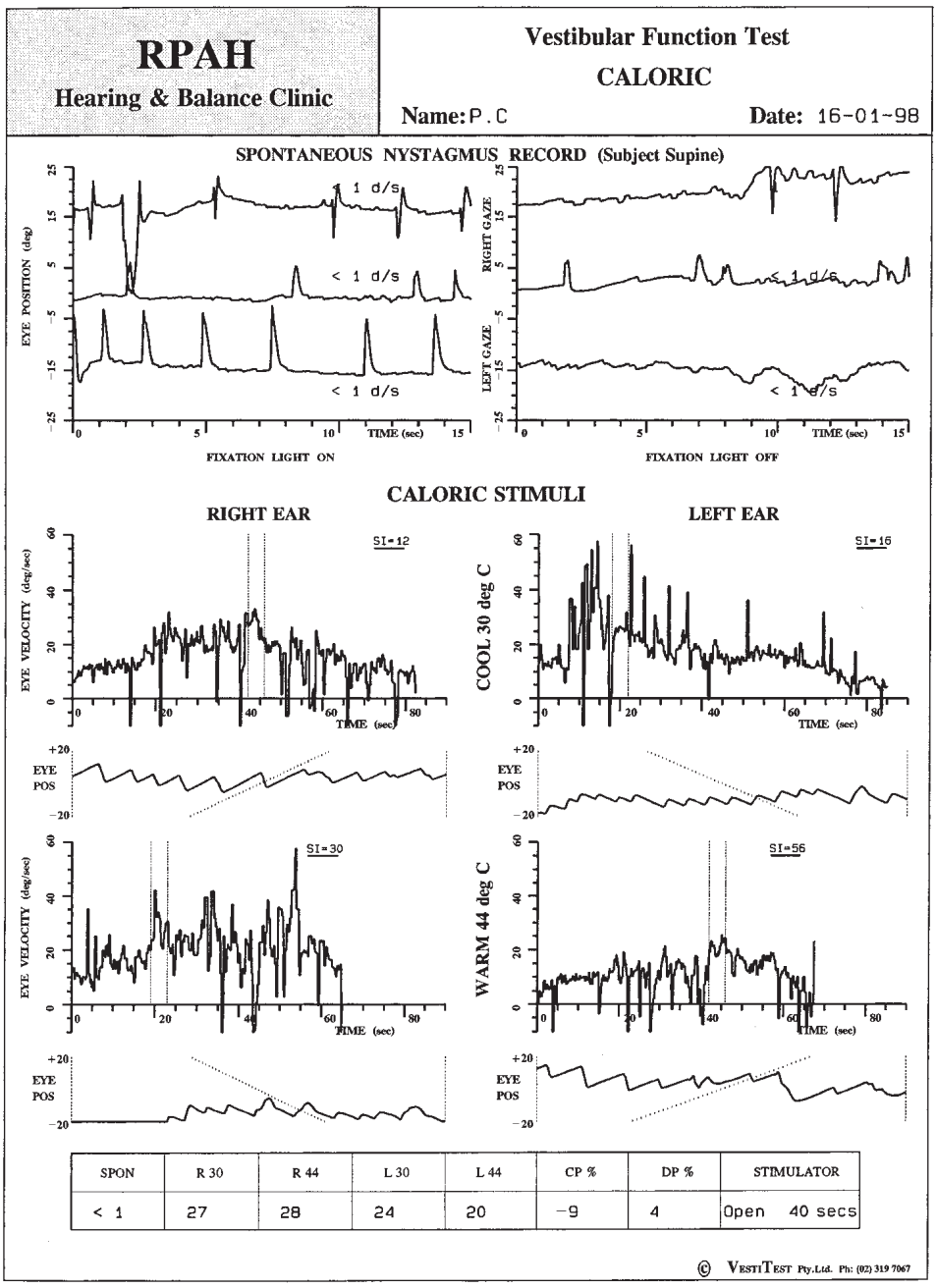


A

Figure 12.7 **(A)** Audiogram from a 61-year-old man with left neurolabyrinthitis. There is a severe sensorineural hearing loss on the left with absence of acoustic reflexes and zero speech discrimination, all findings suggestive of a retrocochlear loss. On the right, there is a slight conductive loss, which is asymptomatic and unrelated to the present problem.

Continued

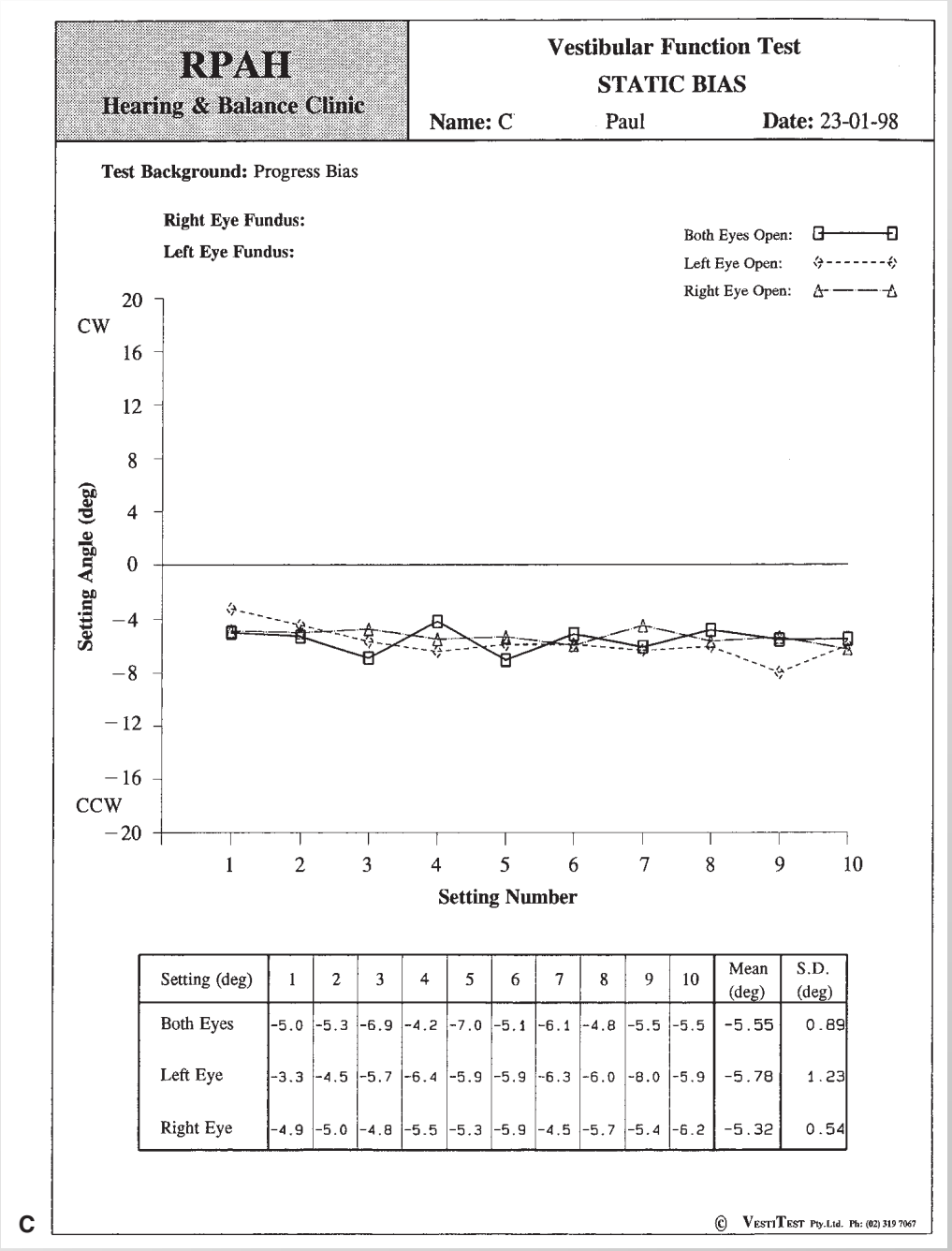
CASE STUDY 12-1



B

Figure 12.7—cont'd **(B)** Electronystagmogram and caloric test from the same patient. There is minimal left-beating gaze-evoked nystagmus in darkness (slow-phase velocity <1 deg/sec). Bithermal caloric tests show symmetrical slow-phase velocities from each ear. These findings indicate normal lateral semicircular canal function.

CASE STUDY 12-1

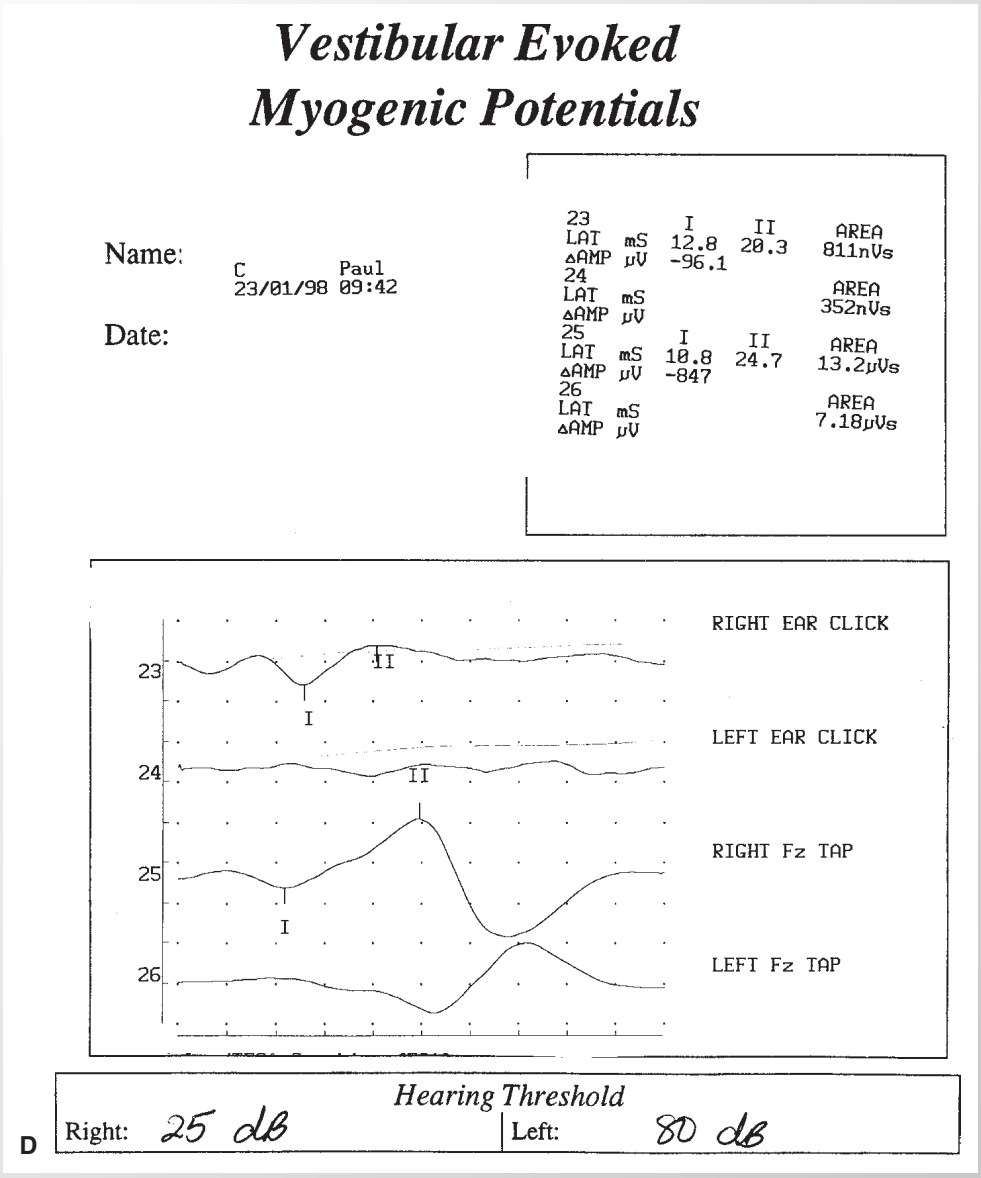


C

Figure 12.7—cont'd **(C)** Subjective visual horizontal (SVH) from the same patient. The patient makes 10 settings of the SVH at his own speed, with each eye open and then with both eyes open. There is a highly significant offset of more than 5 degrees to the left (the side of the hearing loss). Normal subjects can set the horizontal to within 2 degrees of the gravitational horizontal.

Continued

CASE STUDY 12-1



D

Figure 12.7—cont'd **(D)** Cervical vestibular-evoked myogenic potentials (cVEMPs) from the same patient. Channels 23 and 24 show the responses to 95-dB clicks; channels 25 and 26 show the response to forehead taps. The responses are absent from the left. The absence of tap responses shows that the absence of click response is caused not by some conductive hearing loss in the left ear (that is being masked by the severe sensorineural loss), but by loss of saccular function. The cVEMP response to clicks has nothing to do with the audibility of the stimulus—it is present even if the click is not heard.

Dynamic Otolith Tests

In the early 1990s, Colebatch and Halmagyi[74,75] showed that by measuring myogenic potentials evoked by putative vestibular stimulation—vestibular-evoked myogenic potentials (VEMPs)—picked up by surface electrodes on the skin over particular muscle groups in response to vestibular stimuli, that these VEMPs could identify vestibular deficits. The vestibular stimulus used was air-conducted sound (ACS) and later bone-conducted vibration (BCV) to evoke these VEMPs.[75] Evidence from physiology then and since supports the conclusion that these stimuli predominantly activate otolith neurons. Now a host of VEMPs have been reported because vestibular afferent input projects to so many different muscle groups. The two VEMPs that have solid physiological bases and strong evidence supporting their interpretation are cervical VEMPs (cVEMPs) and ocular VEMPs (oVEMPs), both generated in response to ACS and BCV. The cVEMP is recorded by EMG electrodes over the tensed sternocleidomastoid (SCM) muscles, and consists of a short-latency (13 msec from stimulus onset to response peak) positive (i.e., *inhibitory*) EMG potential called the cVEMP p13-n23. The ocular VEMP (oVEMP) is a small (5 to 10 μV) negative (i.e., excitatory) potential recorded by electrodes on the skin beneath the eyes while the person is looking up. Initially, ACS was widely used, and the misapprehension arose that ACS stimulates only the saccular afferents. The more recent physiological evidence shows that is not correct: ACS causes activation of both saccular and utricular afferents.

These new tests rely on the physiological evidence that otolith neurons are activated by both sound and vibration. But questions remain: are these VEMPs a result of vestibular or cochlear stimulation? If vestibular, are the VEMPs caused by otolith or canal stimulation? And is it possible to probe utricular and saccular function separately? We address these matters below.

The Cervical VEMP—cVEMP

The physiological evidence is that saccular afferents in cats and guinea pigs are activated by ACS and BCV.[14,15,76-81] Sound-evoked saccular neurons project to and synapse on neurons in the ipsilateral vestibular nuclei,[82] and inhibitory neurons in the vestibular nuclei project ipsilaterally to spinal motoneurons and inhibit them.[8] In healthy subjects, short tone bursts of 500 Hz of either high-intensity ACS or moderate BCV result in a stimulus-locked, short-latency, positive myogenic potential recorded by electrodes over the *ipsilateral* tensed sternocleidomastoid muscles (SCM). This cVEMP p13-n23 is an *uncrossed*, descending, inhibitory, sacculo-collic response. In patients with complete unilateral vestibular loss following vestibular schwannoma removal, there is a reduced or absent cVEMP p13-n23 from the ipsilateral SCM in response to Fz BCV or ACS stimulation of the affected ear.[75] The absolute values of the cVEMP depend on many variables such as stimulus intensity, neck muscle tension, and electrode placement. Examples of oVEMPs and cVEMPs are shown in Figure 12.8.

The diagnostically valuable information is the relative amplitude of the potentials on the left and right sides in response to symmetrical stimulation of both ears. It has been convenient to use an indicator called the asymmetry ratio (AR), which is defined like the Jongkees formula for caloric asymmetry, but here it becomes:

$$\text{Asymmetry Ratio (AR)} = \frac{(\text{Left VEMP} - \text{Right VEMP})}{(\text{Left VEMP} + \text{Right VEMP})} \times 100$$

where VEMP refers to the amplitude of the VEMP potential (either cVEMP or oVEMP) in response to approximately equal stimulation of both ears. An AR of 100% shows total asymmetry.

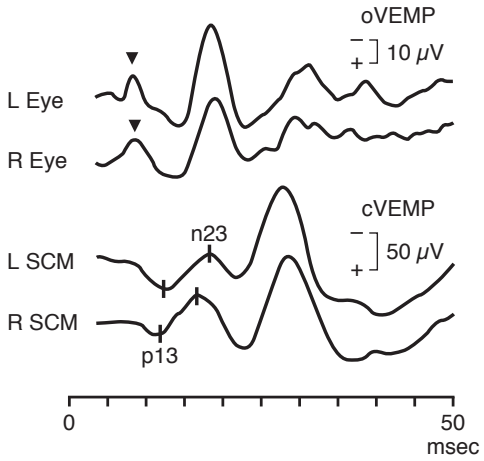


Figure 12.8 Examples of the averaged oVEMP and cVEMP responses from a healthy individual. The oVEMP is recorded by electrodes beneath each eye as the subject looks up, and the cVEMP by electrodes over the tensed SCM muscles, both in response to bone-conducted vibration, here repeated gentle taps at Fz by a tendon hammer. For the oVEMP, there is an initial negativity of about 10 μV (arrowheads) at a latency of about 10 msec, and for the cVEMP there is an initial positivity (vertical bars) of about 50 μV re baseline with a peak at about 13 msec. Note that for both VEMPs the amplitude is about equal on both sides. (Reprinted from *Neurology*, Vol 68, Iwasaki et al,[83] "Head taps evoke a crossed vestibulo-ocular reflex," with permission from Wolters Kluwer Health.)

The Ocular VEMP—oVEMP

In healthy subjects, short tone bursts of 500 Hz of either high-intensity ACS or moderate BCV result in a stimulus-locked, short-latency, negative myogenic potential—the oVEMP—to be recorded by surface electrodes beneath the eyes. The first negative (excitatory) component of which at a latency of about 10 msec, is called oVEMP n10.[83-86] This component probably indicates primarily the myogenic potential of the inferior oblique.[87] Stimulation of one ear by ACS also elicits oVEMP n10 responses beneath the contralateral eye; however, the sound intensities needed are very high, and the n10 potentials to ACS are typically small.[88-90] In addition to the contralateral n10, in some subjects ACS also produces small potentials beneath the ipsilateral eye,[89] but it is the amplitude of the oVEMP n10 beneath the *contralateral* eye that is of diagnostic value. ACS has the major disadvantage that conductive hearing loss renders the test results meaningless. The oVEMP is a *crossed*, excitatory, ascending, utriculo-ocular response.[83]

The Interpretation of oVEMP and cVEMP Results

The oVEMP and cVEMP tests are vestibular tests, because patients who are totally deaf and cannot hear the stimulus show the myogenic potentials to ACS or BCV. Conversely, patients tested after systemic gentamicin with probably absent vestibular function bilaterally, but with residual hearing, can hear and feel the stimuli but do not show the myogenic potentials.[84,85] In patients with complete unilateral vestibular loss following vestibular schwannoma removal, there is a reduced or absent oVEMP n10 from beneath the contralateral eye (as the subject looks up) in response to Fz BCV or ACS stimulation of the affected ear[88,91-93] and reduced or absent cVEMP p13-n23 from over the ipsilateral SCM in response to Fz BCV.

In light of the evidence that 500-Hz sound and vibration selectively activate otolithic afferents rather than canal afferents, together with the evidence of the differential strengths of the neural projections of the utricle and saccule, Curthoys put forward the hypothesis that measuring oculomotor responses to 500-Hz ACS and 500-Hz Fz BCV probes predominantly utricular function, whereas measuring neck muscle responses to these stimuli probes predominantly saccular function (Fig. 12.9).[18] In other words, it is possible to probe utricular and saccular function separately because of their differential neural projections, *not* because the stimuli (ACS and BCV) are *specific* selective stimuli for different otolithic sensory regions, because they are not.[15] The physiological evidence shows that utricular neurons respond to *both* 500-Hz BCV and 500-Hz ACS[14-16,81,94] and that saccular neurons also respond to *both* these stimuli.[15,16] So, the response to ACS may indicate either saccular or utricular function depending on which *response*, oculomotor or cervical, is being measured. Recently, we have found that the stimulus frequency is important for this otolithic specificity—if low-frequency (100-Hz) stimuli are used, canal afferents and otolithic afferents can be activated with high intensity vibration. However, at 500 Hz only otolith neurons are activated (Curthoys et al 2013, unpublished data). So, for clinical testing of otolith function, 500 Hz is an almost ideal choice.

The evidence for differential projections of utricular and saccular maculae is overwhelming. Uchino's research has shown that saccular neurons have a strong projection to neck muscles[95-98] and a weak projection to the oculomotor system.[99-101] Utricular afferents have a strong projection to eye muscles.[101,102] On the basis of many years of research on otolith-ocular and otolith-spinal projections, Uchino and Kushiro summarize the differential projections of utricular and saccular afferents as follows:

"Consequently, the neural connections in the sacculo-ocular system are relatively weak compared to the neural connections in the utriculo-ocular and sacculo-collic systems."[103]

Differentiating Utricular from Saccular Function

One important source of data confirming that oVEMPs to 500 Hz probe utricular function and cVEMPs to 500 Hz probe saccular function comes from the pattern of responses of patients with vestibular neuritis, in which hearing is not affected but vestibular function is affected.[104-109] These patients fall into three main categories: neuritis may affect the entire vestibular nerve, just the superior division, or just the inferior division. The consequence of neuritis is that afferents have reduced or absent function, and that loss of function manifests itself as a distinctive pattern of deficits on stimuli that activate various vestibular sense organs. The results can be understood in terms of the anatomical projections of the various vestibular sense organs within the vestibular nerve shown in Figure 12.9 and their differential projection to oculomotor or cervical regions.

If the neuritis affects the entire vestibular nerve, one would expect a pattern of VEMP deficits matching those of a patient after surgical removal of one entire vestibular nerve—loss of ipsilesional cVEMPs and contralesional oVEMPs (see Fig. 12.9; see also Fig. 12.3). Patients showing such a pattern akin to that of total unilateral vestibular

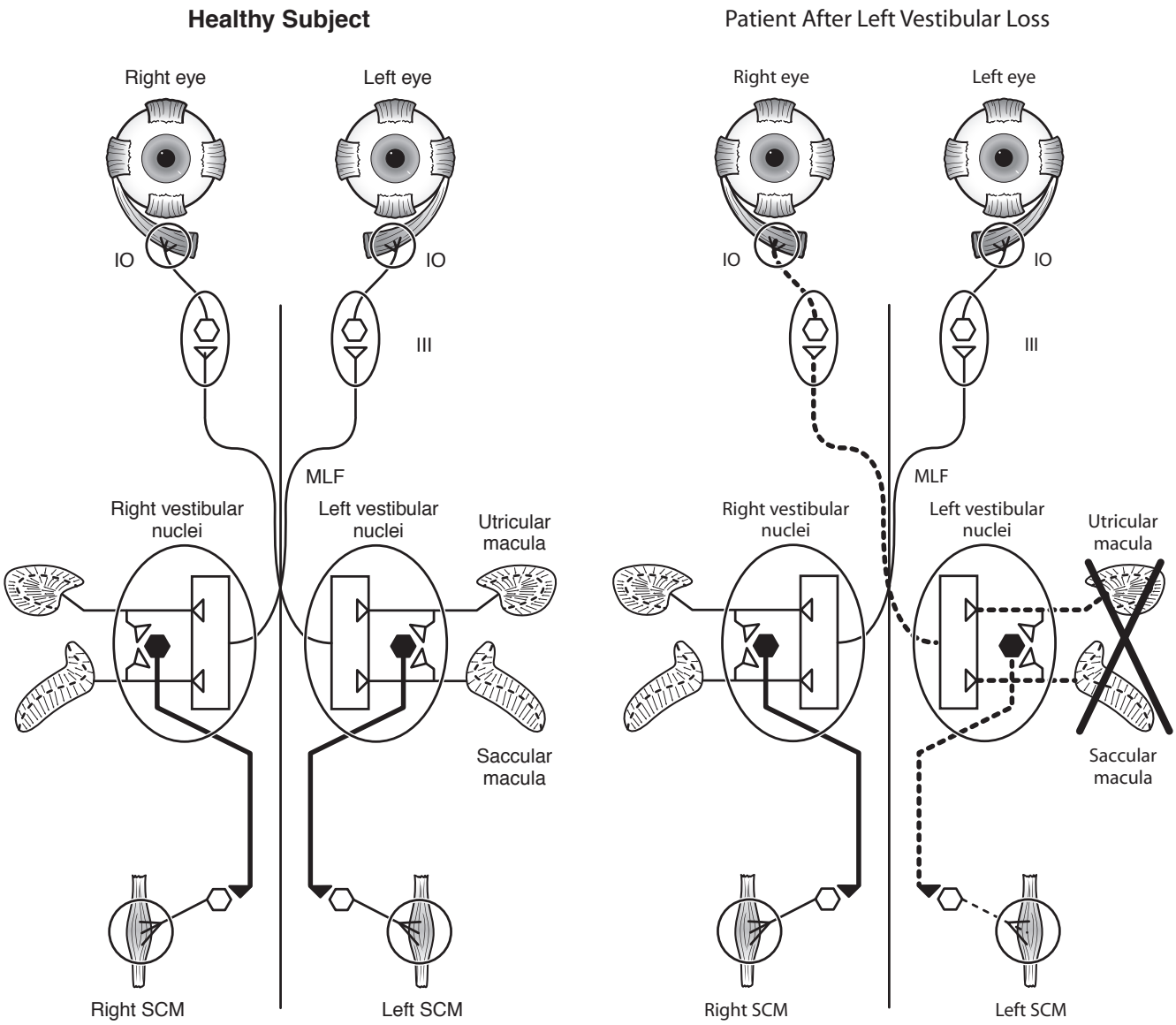


Figure 12.9 Schematic illustration of the possible pathways responsible for the asymmetric oVEMP and cVEMP after unilateral vestibular loss. Afferents from the utricular macula course in the superior vestibular nerve and have strong projections to the contralateral inferior oblique.[26] So, loss of the utricular macula results in a reduced or absent oVEMP n10 beneath the contralesional eye. Afferents from the saccular macula course predominantly in the inferior vestibular nerve and have strong projections to the ipsilateral SCM. So, loss of the saccular macula results in reduced or absent cVEMP p13-n23 on the ipsilesional SCM. If the entire vestibular nerve is affected, as shown here, then both contralesional oVEMP n10 and ipsilesional cVEMP p13 are reduced or absent (see also Fig. 12.3). (Reprinted from Iwasaki et al, 2008,[92] with permission of S. Karger AG, Basel).

loss are diagnosed as a total unilateral vestibular neuritis (TVN).[88,110,111]

The schema shown in Figure 12.9 implies that patients with probable unilateral superior vestibular neuritis (SVN) would have normal cVEMPs, because the saccular afferents in the inferior vestibular nerve will not be affected. However, these patients should have reduced or absent oVEMP n10 potentials beneath the contralateral eye, because all afferents from the utricular macula project in the affected superior vestibular nerve,[19] and the oVEMP n10 is a *crossed* response.[83,92] This was demonstrated by Iwasaki et al[92] (Fig. 12.10) and confirmed in a major study of 133 patients with SVN in response to 500-Hz Fz BCV.[111,112] In response to 500-Hz Fz BCV, SVN patients had symmetrical cVEMPs

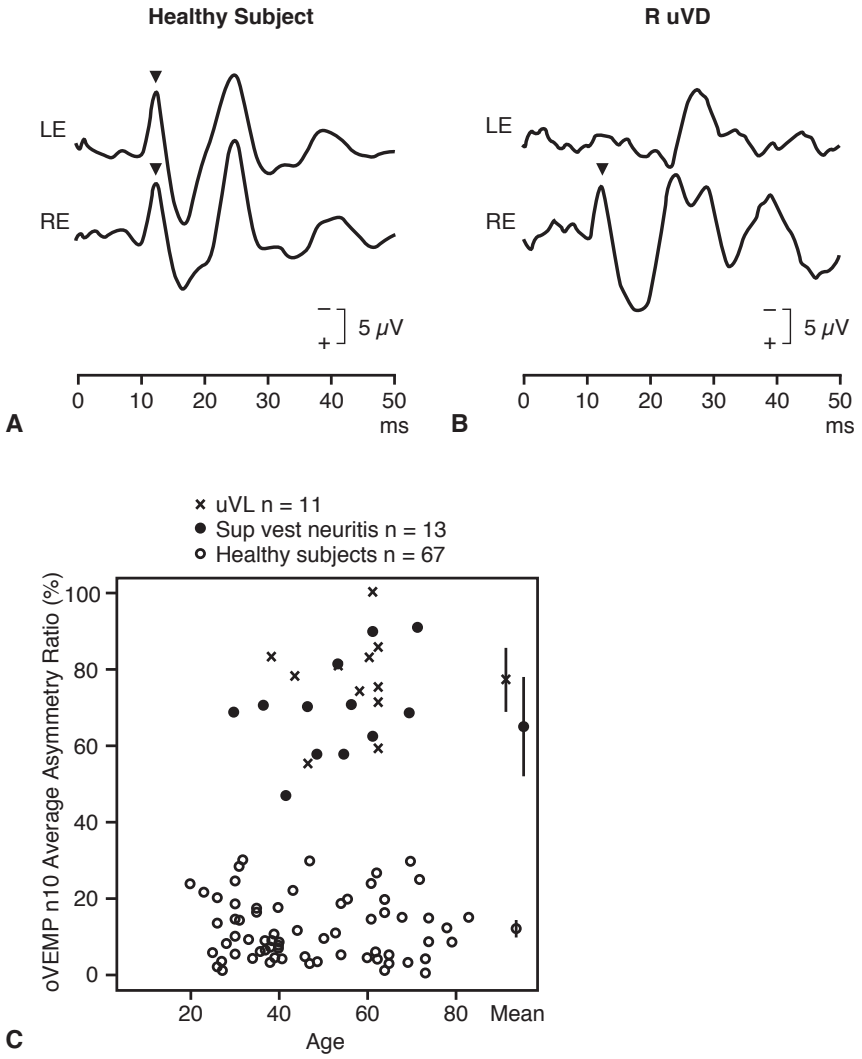


Figure 12.10 Examples of oVEMP responses to bone-conducted vibration at Fz in healthy subjects **(A)** and a patient with a complete unilateral vestibular loss **(B)**. In the patient, there is no detectable oVEMP n10 beneath the contralesional eye, whereas the oVEMP n10 beneath the ipsilesional eye is of normal amplitude. **(C)** The asymmetry ratios (ARs) for Fz stimulation for healthy subjects (open circles), patients after unilateral vestibular loss (uVL) (crosses), and patients diagnosed with unilateral superior vestibular neuritis (closed circles). The average AR for healthy subjects is about 11%, and all 67 healthy subjects have ARs less than 40%. On the other hand, all patients had ARs greater than 40% with no systematic difference between unilateral loss and superior vestibular neuritis. **(A,B)** Reprinted from *Clinical Neurophysiology*, Vol 121, Curthoys[18] "A critical review of the neurophysiological evidence underlying clinical vestibular testing using sound, vibration and galvanic stimuli," 2010, with permission from Elsevier. **(C)** Reprinted from *Clinical Neurophysiology*, Vol 120, Iwasaki et al,[114] "The role of the superior vestibular nerve in generating ocular vestibular-evoked myogenic potentials to bone conducted vibration at Fz," 2009, with permission from Elsevier.

but asymmetrical oVEMP n10s with the contralateral n10 being reduced as predicted.[113,114]

Patients with probable unilateral inferior vestibular neuritis (IVN) as shown by reduced or absent ipsilateral cVEMPs in response to 500-Hz Fz BCV, but with normal superior nerve function as shown by normal calorics and by horizontal head impulse testing, would have contralateral oVEMP n10 responses that are not detectably affected. It has been argued that the utricular afferents, coursing in the superior vestibular nerve, will not be affected by IVN. This prediction has recently been demonstrated in a major study of 59 patients with probable inferior vestibular neuritis in which, to exactly the same stimulus, the cVEMPs were asymmetrical but the oVEMPs were symmetrical.[115] That result confirms other independent evidence of the effect of IVN on VEMPs.[111,116] This is strong evidence that saccular dysfunction does not affect oVEMPs.

There is a proviso: in patients with probable inferior vestibular neuritis, the superior vestibular nerve is probably intact. So, it is possible that the function of the afferent fibers from the "hook" region of the saccular macula is preserved, because these fibers travel in Voit's nerve and merge with the superior vestibular nerve.[20-22] Could these "hook" afferents be responsible for the oVEMP response? That is not a likely possibility (1) because of the relatively small contingent of afferent fibers from the hook region of the saccular macula,[23] (2) because sacculo-ocular pathways are polysynaptic,[95,103] and (3) the oVEMP n10 is a sharply defined potential at a very short latency and appears to be caused by the EMG generated by a synchronous volley of action potentials arriving at the extraocular muscles. It is unlikely to be caused by action potentials down a weak polysynaptic pathway[95] from the very small bundle of afferent fibers innervating the hook region of the saccular macula.

Methods—Recording VEMPs

For details, see Rosengren et al.[116] Brief (0.1 msec), loud (95 db above normal hearing level [nHL]), monaural clicks[74,118] or short tone-bursts[119,120] produce a large (60 to 300 μV), short-latency (8 msec), inhibitory potential in the tonically contracting ipsilateral sternocleidomastoid muscle. The initial positive-negative potential, which has peaks at 13 msec (*p13*) and at 23 msec (*n23*), is abolished by selective vestibular neurectomy but not by profound sensorineural hearing loss. In other words, even if the patient cannot hear the clicks, there can be normal *p13-n23* responses. Later components of the evoked response do not share the properties of the *p13-n23* potential and probably do not depend on vestibular afferents. Failure to distinguish between these early and late components could explain why earlier work along similar lines was inconclusive.

For the preceding reasons, we called the *p13-n23* response the *vestibular-evoked myogenic potential* (VEMP)—more recently known as the cVEMP (cervical VEMP)—to distinguish it from other VEMPs. Unlike a neural-evoked potential such as the brainstem auditory-evoked potential, which is generated by the synchronous discharge of nerve cells, the cVEMP is generated by synchronous discharges of muscle cells or, rather, motor units. Being a myogenic potential, the cVEMP can be 500 to 1,000 times larger than a brainstem potential,[121] for example, 200 μV rather than less than 1 μV. Single motor unit recordings in the tonically contracting sternocleidomastoid muscle show a decreased firing rate synchronous with the surface cVEMP.[122]

The amplitude of the cVEMP is linearly related to the intensity of the click and to the intensity of sternocleidomastoid muscle activation during the period of averaging, as measured by the mean rectified electromyography (EMG) value.[118,123] Inadequate sternocleidomastoid contraction produces spurious results by reducing the amplitude of the cVEMP.[124] A conductive hearing loss abolishes the response by attenuating the intensity of the stimulus[125] (see Fig. 12.9). In such cases, the cVEMP can be elicited by a tap to the forehead,[75] by a bone vibrator,[126-128] or by a direct current applied to the mastoid bone.[129]

Methods—cVEMPs

Any equipment suitable for recording brainstem auditory potentials will also record VEMPs. Because the amplitude of the cVEMP is linearly related to the intensity of both the click and of sternocleidomastoid activation during the period of averaging, it is essential to ensure that the sound source is correctly calibrated and that the background level of rectified sternocleidomastoid EMG activation is measured. Two reasons why the cVEMPs could be absent or less than 50 μV in amplitude are a conductive hearing loss and inadequate contraction of the sternocleidomastoid muscles.

For clinical testing, three superimposed averages of 128 stimuli for each ear in response to clicks of 100-dB intensity are usually sufficient. The test cannot be done on uncooperative or unconscious patients. The patient lies down and activates the sternocleidomastoid muscles for the averaging period by keeping the head raised from a pillow. An alternative method—useful, for example, in patients with painful neck problems—is to ask the patient to turn the head, which continues to rest on the pillow, to one side. It is then possible to measure the VEMP in the sternocleidomastoid muscle on the side opposite to the rotation.

The peak-to-peak amplitude of the *p13-n23* potential from each side can be expressed relative to the level of background mean-rectified EMG to create a ratio that largely removes the effect of differences in muscle activity. More accurate, but more time-consuming, correction can be made by making repeated observations with differing levels of tonic activation.[118] One ear is best evaluated through comparison of the amplitude of its cVEMP with that of the cVEMP from the other ear. We take asymmetry ratios of 2.5:1 to be the upper limit of normal—a value similar to that obtained by others.[130,131] Minor left-right differences in latency commonly occur and might reflect differences in electrode placement over the muscle or differing muscle anatomy. VEMP amplitude declines after age 60 years.[132]

Methods—oVEMPs

For details, see Iwasaki et al.[84] Subjects lie supine on a bed with the head supported on a pillow but positioned so that the head was horizontal or pitched slightly nose down. After thorough cleaning of the skin beneath the eyes with alcohol wipes, surface EMG electrodes are applied to record the responses from beneath both eyes, as shown in Figure 12.11. For each eye, the active (+) electrode is placed on the infra-orbital ridge about 1 cm below the lower eyelid, and the reference electrode is placed about 2 cm below that first electrode. The electrodes are aligned with the center of the pupil as the subject looks up at a distant target in the midline. The self-adhesive pads around the electrode are cut to allow this very close electrode placement, taking care that no electrical bridge forms between the two closely juxtaposed electrodes.

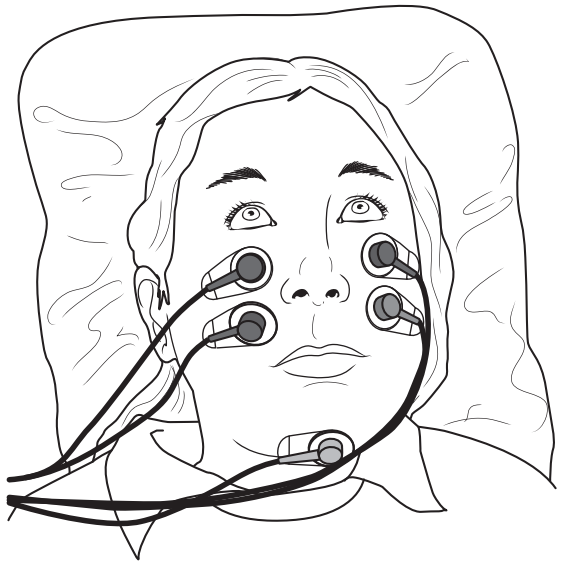


Figure 12.11 The placement of the electrodes for recording oVEMP n10. Each pair of electrodes is connected to a differential amplifier. The ground electrode is placed on the chin. Most importantly, the subject or patient must be looking up as high as possible, as shown. Voluntary vertical saccades should produce approximately equal potentials from both eyes. Reprinted from *Clinical Neurophysiology*, Vol 119, Iwasaki et al,[84] "Ocular vestibular evoked myogenic potentials to bone conducted vibration of the midline forehead at Fz in healthy subjects," 2008, with permission from Elsevier.

Recording

The surface potentials (predominantly EMG) are amplified by AC-coupled differential amplifiers (bandwidth 20 to 500 Hz), and the unrectified signals are averaged (n = 30 or 50 presentations) using an averager with a sampling rate of 20 kHz. The adopted electrical convention is that negative potentials at the active electrode (the electrode closest to the eye) cause an upward trace deflection. A ground electrode is placed on the chin or sternum. The potentials recorded are small, so the electrode leads may need to be shielded, with the shields connected to the ground electrode on the subject. Electrode impedance is maintained below 5 kohms in all trials. The subject's jaw muscles should be relaxed, and the person should be looking straight up at a target fixation dot in their midline about 30 degrees above the subject's visual straight ahead. Subjects tend to pitch their head up to do this but that should not happen—it is important the eye be elevated as high as possible *in the orbit*. The fixation point must be in the subject's midline, because lateral fixation results in asymmetric EMG recordings and, thus, asymmetric oVEMPs.

The stimulus is either a light tap to Fz with a tendon hammer or a modest 500-Hz vibration (about the intensity delivered by a body massager or an electric toothbrush) delivered by a Bruel and Kjaer minishaker 4810[83,84,133] at the rate of 3 to 5 per second. Fz is a special site for BCV stimulation, because stimulation there produces approximately equal stimulation of both ears.[84] Mastoid BCV stimulation (by linear accelerations) is sometimes used for clinical testing. It can elicit responses but does not permit the clinically important direct comparison of the level of utricular function of the two sides, because it is difficult to produce equal BCV stimulation of the two labyrinths by mastoid stimulation. Unequal oVEMP n10 responses may thus be a result of unequal stimuli.

The stimuli are tone bursts of 500 Hz lasting 7 ms at the rate of 3/sec or 5/sec delivered by a handheld Bruel and Kjaer (Naerum, Denmark) Mini-Shaker 4810, fitted with a short bolt (2 cm long, M4) and terminated in a bakelite cap 1.5 cm in diameter, which is the contact point for the stimulator on the subject's forehead at the junction of the midline forehead with the hairline (a location called Fz). The optimum stimulus is one with a very short rise time (ideally zero), which our recent research shows gives the largest oVEMP n10 (Curthoys et al, unpublished research 2013). The Mini-Shaker weighs approximately 1 kg, and the weight of the shaker is used to roughly equate the force used in all subjects. It is handheld, but the operator simply maintains its near-vertical orientation and does not force the shaker against Fz. To minimize electrical artifacts, the case of the Mini-Shaker is shielded, the Mini-Shaker signal leads are shielded, flexible shielded cable is used for the electrode leads, and all shields are led to the ground electrode on the subject.

The size of oVEMP n10 increases as the subject looks up,[84,85] so during clinical testing it is essential that the subject maintains upward gaze. The oVEMP n10 is not caused by a blink.[134] A standard audiometric bone oscillator (e.g., a Radioear B-71) placed at Fz does not deliver enough power to generate an oVEMP n10 reliably.[83] Also at Fz, small changes in Mini-Shaker location have relatively little effect on oVEMP n10 amplitudes or symmetry. However, with BCV stimulation at other stimulus sites, small changes in stimulator location can have major effects on oVEMP n10 amplitude. The amplitude of the oVEMP n10 potential is highly variable between individuals, probably because of the very different skull sizes and masses. But when any healthy subject is given the Fz BCV stimulus, which stimulates both labyrinths about equally, it is found that the amplitude of the oVEMP n10 is similar beneath both eyes.[84] This same stimulus arrangement can be used to measure cVEMPs to BCV.

For both cVEMPs and oVEMPs, there are a number of crucial parameters that have to be set correctly for the results to be interpretable. For example, inappropriate

electrode placement or low gaze can negate the oVEMP; insufficient neck muscle tension can negate the cVEMP. With ACS stimulation for either test, conductive hearing loss can nullify the results. Also, some healthy subjects simply do not have oVEMP n10 to ACS stimuli.[135] For these reasons, ACS is a "vulnerable" stimulus that can be easily affected by factors apart from the state of the vestibular receptors. Consequently, BCV is preferable for clinical testing.

Clinical Applications

Superior Semicircular Canal Dehiscence

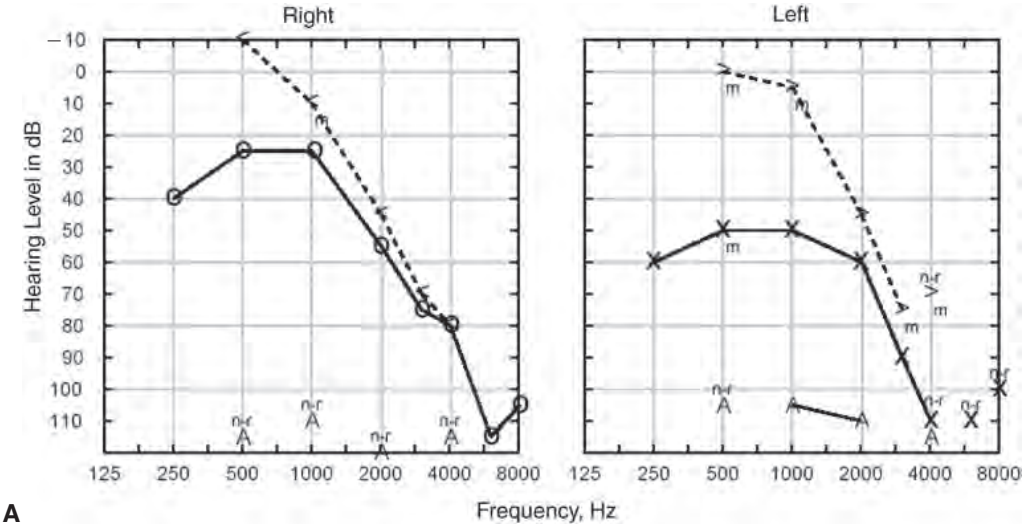
A third window into the bony labyrinth allows sound to activate the vestibular system in animals and in humans (for a review, see Halmagyi et al[136]). Patients with a bony dehiscence from the superior semicircular canal into the middle cranial fossa (Fig. 12.12 *C*) not only have sound- and pressure-induced vestibular nystagmus but also abnormally large, low-threshold cVEMPs and oVEMPs, to both air-conducted (AC) and bone-conducted (BC) stimulation (Fig. 12.12; Fig. 12.13). It is not yet certain whether the threshold or the amplitude of the oVEMP or cVEMP to AC or BC, or low- or high-frequency stimulation has the greatest sensitivity and specificity for the physiological diagnosis of superior semicircular canal dehiscence (SCD).[137-142] Air-bone gaps caused by SCD or large vestibular aqueduct[143] can be differentiated from those caused by middle ear disease using cVEMPs.[144] After successful SCD surgery, both oVEMPS and cVEMPs tend to normalize.[145,146] SCD patients also have abnormally large, low-threshold, click-evoked vestibulo-ocular reflexes.[147]

Ménière's Disease

In Ménière's disease[143,148-152] and in delayed endolymphatic hydrops,[153,154] and unlike in acute low-tone hearing loss,[155] VEMPs can be abnormal: too small[156] or too large,[157] and can have altered tuning properties.[148] In some cases, glycerol dehydration or furosemide loading can reduce VEMPs that are too large and enlarge the VEMPs that are too small.[158-163] VEMPs can be used to monitor intratympanic gentamicin therapy in patients with Ménière's disease.[164-166] In Ménière's patients with Tumarkin-type falls, VEMPs might be abnormal,[167,168] perhaps oVEMPs more than cVEMPs.[169] The amplitude of the oVEMP n10 from the affected ear appears to be selectively enhanced during attacks in early Ménière's Disease.[170,171]

Vestibular Neuritis

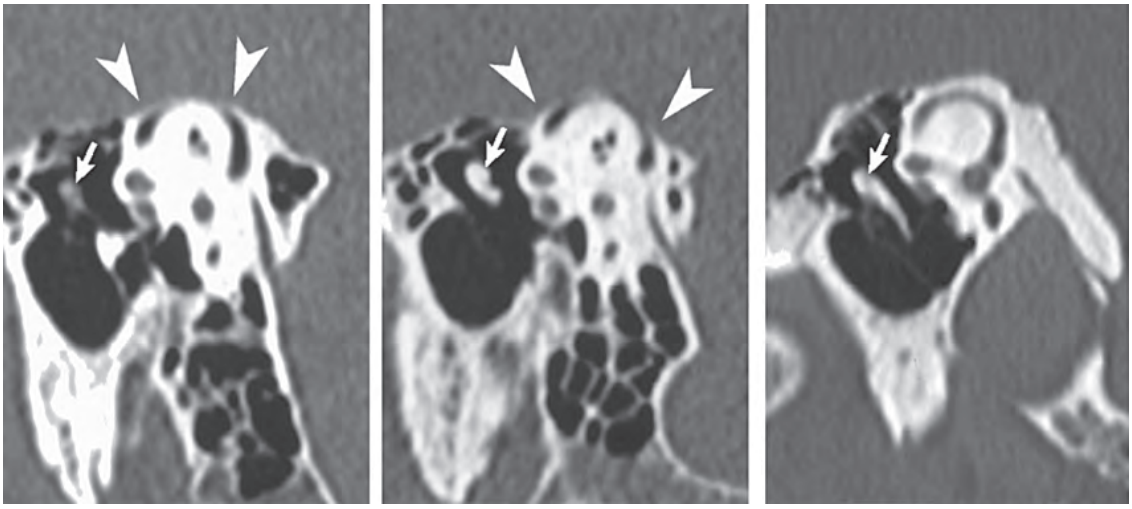
This subject has been comprehensively covered from the physiological viewpoint in the section above "Differentiating utricular from saccular function." Here we deal with diagnostic aspects. Usually, vestibular neuritis selectively



A

Figure 12.12 **(A)** Audiogram. Following the left stapedectomy and two revisions, there is still a large air-bone gap at 500 and 1,000 Hz on the left and on the unoperated right side. The contralateral acoustic reflex, the sound stimulus in the operated left ear and the volume probe in the unoperated right ear, is present at 1 kHz and 2 kHz (A—A) and absent at 0.5 and 4 kHz. It is absent at all frequencies with the stimulus in the unoperated right ear and the volume probe in the thrice-operated left ear. There is also a severe high-frequency loss caused by noise damage. Speech comprehension at 65 dB was 95% on the right and 85% on the left; m = masked; n-r = no response.

Continued



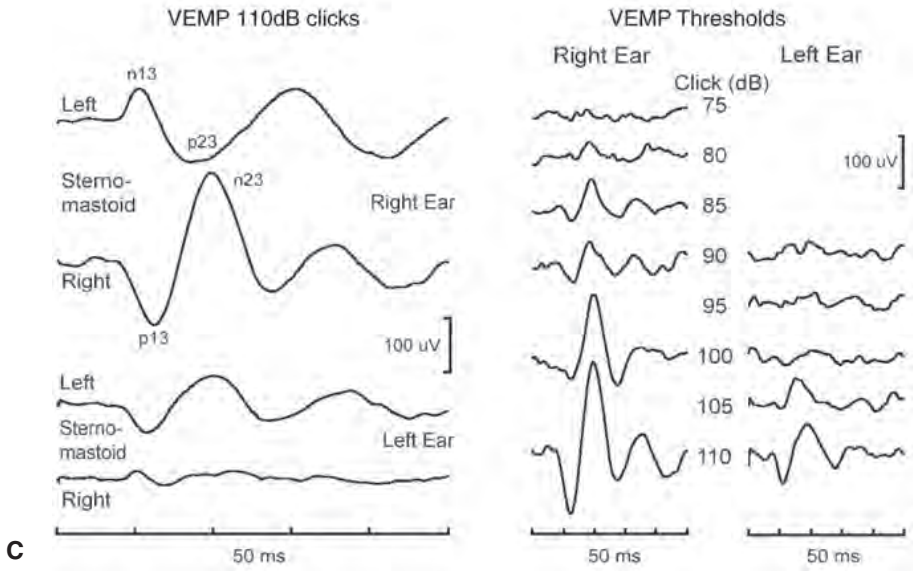


Figure 12.12—cont'd **(B)** Vestibular-evoked myogenic potentials (VEMPS). (*Left*) Despite the significant air-bone gap, p13-n23 VEMP responses to 110 dB clicks are still present. The VEMP from the left ear is normal in amplitude but smaller (115 μV) than from the right (307 μV), indicating that there is some conductive loss on the left (as a result of the 3 operations) and a conductive gain. The response from stimulating the right ear is abnormally large and is accompanied by a contralateral inverse response, *n13-p23*. (*Right*) The VEMP threshold is normal from the left ear (with a conductive loss, the VEMP should be absent) and abnormally low (80 dB; normal > 95 dB) from the right ear. **(C)** *Left* and *center*, High-resolution spiral computed tomography (CT) scan of the temporal bones, reconstructed in the plane of each superior semicircular canal, shows a bilateral superior semicircular canal dehiscence (SCD; *arrowheads*). CT scan from a normal subject is shown for comparison. The TORP device is clearly seen abutting the oval window on the *left*. (Courtesy of Dr. John Harding-Smith, Central Sydney Imaging.)

affects the superior vestibular nerve. These patients have absent or reduced vestibulo-ocular reflexes from the lateral and anterior (i.e., superior) semicircular canals as shown by impulsive or caloric stimulation, and absent or reduced utricular function as shown by absent or reduced oVEMPs in response to AC or BC stimulation, but preserved saccular function as shown by intact cVEMPs to AC or BC stimulation.[83,84,92,104,106-107,111-112,170-173] These patients can later develop posterior canal positional vertigo,[174] because the inferior vestibular nerve carries posterior canal

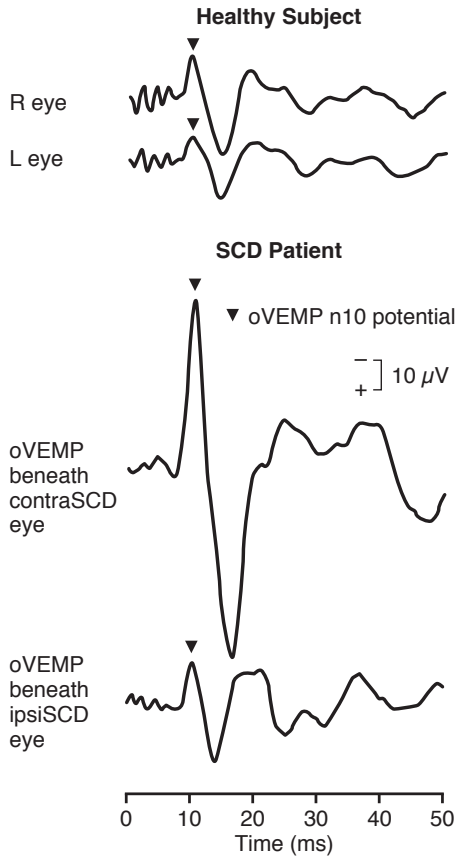


Figure 12.13 Examples of oVEMPs (arrowheads) to brief 500Hz Fz BCV in a healthy subject (a) and a patient with a CT-verified semicircular canal dehiscence (SCD). The oVEMP n10 in the patient is extremely large (few healthy subjects have n10s greater than 10 μV), and this enhanced oVEMP n10 is a fast simple indicator of probable SCD. Reprinted from Manzari et al,[137] "Ocular and cervical vestibular-evoked myogenic potentials to 500-Hz Fz bone-conducted vibration in superior semicircular canal dehiscence," (*Ear and Hearing*, 2012;33(4):508-520 by permission of Wolters Kluwer Health).

and saccular fibers. Rare patients have selective inferior vestibular neuritis with intact lateral and anterior canal function as shown by normal calorics, lateral and anterior canal impulses, and intact utricular function as shown by normal oVEMPs, but reduced or absent posterior canal and saccular function shown by impaired posterior canal head impulses and cVEMPs.[104,115,116,175-179] Both cVEMPs[180,181] and oVEMPs[182] can recover after vestibular neuritis.

Benign Positional Vertigo

In contrast to patients with posterior canal benign positional vertigo (BPV) after vestibular neuritis who generally have normal cVEMPs,[174] patients with idiopathic posterior canal BPV, especially recurrent BPV, not only have a higher incidence of abnormal cVEMPs than controls[174,183,184] but also a higher incidence of abnormal oVEMPs.[183]

Vestibular Schwannoma

Although most patients with vestibular schwannoma (formerly "acoustic neuroma") present with unilateral hearing loss, some present with vestibular ataxia. This fact is not entirely surprising, because most of these tumors arise not from the acoustic nerve but from one of the vestibular nerves, usually the inferior.[185] The cVEMP, which is transmitted via the inferior vestibular nerve, is abnormal—delayed,[186] of low amplitude, or absent—in perhaps four of every five patients with vestibular schwannoma.[91,187-192] Because the cVEMP does not depend on cochlear or lateral SCC function, it can be abnormal even when caloric test results are normal or when brainstem auditory-evoked potentials cannot be measured because the hearing loss is too severe. In patients with bilateral vestibular schwannoma caused by neurofibromatosis type 2, loss of cVEMP is unrelated to tumor size.[193] The cVEMP can be preserved after vestibular schwannoma surgery,[194] suggesting preservation of the saccular nerve. The oVEMP is reduced or absent slightly more often than the cVEMP.[92] With other cerebello-pontine angle tumors, such as meningiomas, cVEMPs are usually normal.[195]

Brainstem Lesions

Unilateral focal brainstem lesions caused by stroke and multiple sclerosis, especially those that affect the vestibular nuclei, the accessory nerve nuclei, and the medial vestibulospinal tract or its rostral equivalent, the medial longitudinal fasciculus, can disrupt ipsilateral cervical[195-201] and contralateral ocular VEMPs.[202,203]

Testing Otolith Function in Children

oVEMPs can be used to test utricular function in very young children.[204] The BCV stimuli can be delivered at a high rate (23/sec); thus, the trial is complete within a few seconds. This is valuable for assessing vestibular function in children for whom it is very difficult to give caloric tests.

Other Conditions

Initial experience with this technique suggests that the VEMP test can provide valuable information in addition to that obtained by tests of lateral SCC function, such as caloric or rotational tests, and by tests of utricular function, such as the SVH test.[128,205] We have seen patients with symptoms of unilateral or bilateral vestibulopathy (e.g., vertigo, ataxia) in whom SCC function test results

were normal, but VEMP test results were unequivocally abnormal.

Summary

The subjective visual horizontal test and vestibular-evoked myogenic potential measurements are simple, robust, reproducible, and specific tests of otolith dysfunction that can provide clinically useful diagnostic information in patients with vertigo and other balance disorders. Although they appear to have high specificity for otolith dysfunction, further clinical research is required to establish their sensitivity. The physiological and behavioral evidence underpinning these tests continues to grow; however, the exact transduction mechanism by which BCV and ACS cause hair-cell deflections is not known.[206]

Even seemingly minor methodological factors can affect the results. So, like others, we have (rarely) found individuals who have good oVEMPs to 500-Hz BCV but have very small oVEMPs to 500-Hz ACS (with no conductive hearing loss). Unusual patients such as these may be reflecting the exact mode of transduction by BCV as opposed to ACS and the changes taking place within the labyrinth during disease.

Just how good are oVEMPs at diagnosing dysfunction? In a study of 160 healthy subjects and patients with superior vestibular neuritis who had received both caloric testing and oVEMP testing, the oVEMP Asymmetry Ratio (AR)—analogous to the canal paresis score—has been shown to have a sensitivity of 0.9, a specificity of 0.8, and a diagnostic accuracy of 94% compared with the canal paresis (CP) score.[112] In other words, for SVN patients, at least, the AR of the oVEMP n10 is almost as good as the canal paresis score at identifying which is the affected side.

In conclusion, the oVEMP n10 has been in use for 5 years at Sydney, Melbourne, Tokyo, Zurich, and Cassino, and well over 3,000 patients have been tested without any adverse incident. The oVEMP to 500-Hz Fz BCV is a clinically practical way of measuring predominantly utricular function, and the cVEMP to 500-Hz Fz BCV is a clinically practical way of measuring predominantly saccular function—simply, safely, and quickly. Both are built on sound scientific foundations. When the results of oVEMP and cVEMP tests are combined with the results of other vestibular tests, the clinician can obtain a picture about the probable functional state of each sense organ of the labyrinth.

The tests described above and listed in Figure 12.14 and Table 12-1 are based on published evidence. Figure 12.14 is an integration of the anatomical, physiological, and clinical evidence from

a) anatomical evidence about the neural projections for each vestibular sense organ;[19,20]
b) physiological evidence concerning patterns of vestibular neuronal activation to sound and vibration; and
c) clinical evidence about the response of patients with known or probable vestibular losses.[22]

Table 12-1 shows the references used to justify the statements in each cell of Figure 12.14. It is emphasized that these are not meant to be comprehensive but to provide key evidence justifying each statement in the table.

For clinicians trying to evaluate the function of the various otolithic sensory regions, the new evidence and the dissociations discussed above reinforce the conclusion that for both ACS and 500-Hz Fz BCV, the oVEMP n10 is predominantly caused by the contralateral utricular macula, whereas the cVEMP p13-n23 is predominantly caused by the ipsilateral saccular macula.

■ Table 12-1 SELECTED PAPERS JUSTIFYING THE STATEMENTS IN EACH TABLE ENTRY OF FIGURE 12.14

- A.[4,27,30,31,33,35-38,208]
- B.[4,27,30,31,33,35-38,208]
- C.[4,27,30,33,35,36]
- D.[4,27,30,33,35,36]
- E.[107,110]
- F.[105,178]
- G.[209,210]
- H.[18,83-86,88-90,94,106,134,211-215]
- I.[18,83,85,88-90,94,117,134,211,212,214,215]
- J.[18,83,88,89,91-93,211,215]
- K.[18,83,88,89,91-93,115,211,215]
- L.[18,106,110-114,182,211,213,215]
- M.[216]
- N.[145,217]
- O.[17,18,74,75,117,118,133,218]
- P.[17,18,74,75,117,118,133]
- Q.[17,18,74,75,118,133]
- R.[17,18,74,75,118,213]
- S.[18,116,182,213]
- T.[115,174,178,219]
- U.[17,137,145,177,209,210,216,217,220]

Vestibular sense organ being tested	Test of vestibular sensory function	Normal result in a healthy subject ✓	Abnormal result in a patient with a vestibular loss X	Profile of a patient with unilateral vestibular loss UVL	Profile of a patient with superior vestibular neuritis SVN	Profile of a patient with inferior vestibular neuritis IVN	Profile of a patient with superior canal dehiscence SCD
Static otolith function	A. ***Subjective Visual Vertical*** (SVV) (or Subjective Visual Horizontal (SVH))–To set a line of light in an otherwise dark room to vertical or horizontal respectively. ***Roll head tilt*** The maintained position of the head, tested without visual cues. ***Skew Deviation*** (relative height in the orbit of the two eyes when the head is erect and subject is looking straight ahead)	B. ***SVV-SVH*** Settings are within ± 2 degrees of the true vertical or horizontal. ***Roll head tilt*** Head upright. ***Skew Deviation*** Both eyes are in horizontal alignment as subject looks straight ahead.	C. ***SVV-SVH*** The visual line is set toward the affected side; either to left or right if SVV, or down on the affected side if SVH ***Roll head tilt*** Head has a small maintained roll toward the affected ear ***Skew Deviation*** Ipsilesional eye is down in the orbit relative to the contralesional eye	D. Reduced otolith function X	E. Reduced utricular function X	F. Reduced saccular function X *(very few IVN patients tested on these tests)*	G. Normal result ✓
Dynamic otolith function: **Utricular macula**	H. **Ocular vestibular evoked myogenic potential (oVEMP)** beneath the contralateral eye; the n10 component in response to either bone conducted vibration at Fz or ACS to ipsilateral ear, as the subject looks up	I. oVEMP n10 beneath the contralateral eye of normal amplitude (5–10μV) and equal amplitude to n10 beneath ipsilateral eye in response to Fz BCV stimulation or ipsi ACS	J. Reduced or absent n10 of the oVEMP beneath the contralateral eye in response to Fz BCV stimulation or ipsilateral ACS stimulation	K. Reduced or absent n10 of the oVEMP beneath the contralesional eye in response to Fz BCV stimulation or ipsilesional ACS X	L. Reduced or absent n10 of the oVEMP beneath the contralesional eye in response to Fz BCV stimulation or ipsilesional ACS X	M. Normal n10 of the oVEMP beneath the contralesional eye in response to Fz BCV stimulation ✓	N. ***Enhanced*** n10 of the oVEMP beneath the contralateral eye to Fz BCV or ipsi ACS; also reduced threshold for n10 ✓
Dynamic otolith function: **Saccular macula**	O. **Cervical vestibular evoked myogenic potential (cVEMP)** over ipsilateral SCM muscle; the p13-n23 component in response to either bone conducted vibration at Fz or ACS to ipsilateral ear as the subject tenses their SCM	P. The p13-n23 of the cVEMP over the ipsilateral tensed SCM of normal amplitude and equal amplitude to p13-n23 over the contralateral SCM in response to Fz BCV stimulation or ipsilateral ACS stimulation	Q. Reduced or absent p13-n23 of the cVEMP over the ipsilesional SCM in response to Fz BCV stimulation or ipsilateral ACS stimulation	R. Reduced or absent p13-n23 of the cVEMP over the ipsilesional SCM in response to Fz BCV or ipsilateral ACS stimulation X	S. Normal p13-n23 of the cVEMP over the ipsilateral SCM in response to Fz BCV or ipsi ACS stimulation ✓	T. Reduced or absent p13-n23 of the cVEMP of the ipsilesional SCM in response to Fz BCV or ipsi ACS stimulation X	U. ***Enhanced*** p13-n23 of the cVEMP over the ipsilesional SCM to Fz BCV or ipsi ACS stimulation; also reduced threshold for p13-n23 ✓

Figure 12.14 Table showing tests of utricular function and their typical results in healthy subjects and in patients with vestibular loss. The references for each box are keyed to the letter and the adjacent table gives the actual references. Reprinted from Curthoys,[22] "The interpretation of clinical tests of peripheral vestibular function," *The Laryngoscope*, 122(6):1342-1352, DOI 10.1002/lary.23258, with permission from John Wiley and Sons.

Acknowledgments

Work described in this chapter is supported by the National Health and Medical Research Council (grants 632746, 1046826), the Garnett Passe and Rodney Williams Memorial Foundation, and the Royal Prince Alfred Hospital Neurology Department Trustees.

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CHAPTER 13

Assessment and Management of Auditory Disorders and Tinnitus

Sharon H. Polensek, MD, PhD ■ *Steven L. Benton, AuD*

Many patients presenting to a clinician with a possible vestibular disorder have underlying auditory impairment. Because the ear houses the sensory organs for auditory and vestibular input, not surprisingly many otologic disorders (e.g., Ménière's disease, perilymphatic fistula, labyrinthitis, acoustic neuroma, and others) give rise to both auditory and vestibular symptoms. An understanding of clinical methods of assessment and management of hearing loss and tinnitus is beneficial for the clinician who must diagnose and treat patients with vestibular impairment.

History and Physical Examination

A thorough assessment of a patient's auditory system requires obtaining information from a careful history and physical examination, as well as from audiological testing. A patient whose chief complaint is one of disabling dizziness may not volunteer information about a mild hearing difficulty, particularly one with an insidious onset. At the time of the initial clinical assessment, the examiner should grossly assess hearing acuity of any patient presenting with possible vestibular impairment, especially if audiological testing has not yet been performed. Obtaining a formal audiological assessment on patients with vestibular hypofunction would be a high priority. Inquiry should be made as to whether the patient has subjective hearing difficulties, and if so, in what environments is hearing most challenging. Information such as whether the patient can still talk on the telephone with either ear or whether they frequently need to ask for speakers to repeat themselves can be indicative of worsening hearing. For more subtle hearing losses, the patient may report problems only in certain situations such as listening in church or communicating in a noisy environment such as a restaurant.

The simplest method for identifying gross hearing impairment is for the examiner to occlude one external canal by pressing inward on the tragus and assessing whether the patient can hear a vibrating tuning fork, a whisper, or the examiner's fingers rubbing together near the other ear. An asymmetric hearing loss can sometimes be identified by the Weber tuning fork test, which entails placing a vibrating 512-Hz tuning fork firmly on the patient's forehead or teeth and determining whether the patient perceives the sound emanating from the midline (Fig. 13.1). If sound perception lateralizes to one side, then hearing loss is present. To determine which ear has the loss, the examiner can next perform a Rinne test with the fork (Fig. 13.2). To do this test, the examiner first places the vibrating fork behind the patient's ear, and when the patient can no longer hear the tone, the tines of the vibrating fork are placed in front of the external auditory canal of the same ear. A normal result is when the patient can still hear the tone when the fork is placed by the ear canal. If the Rinne test is abnormal on

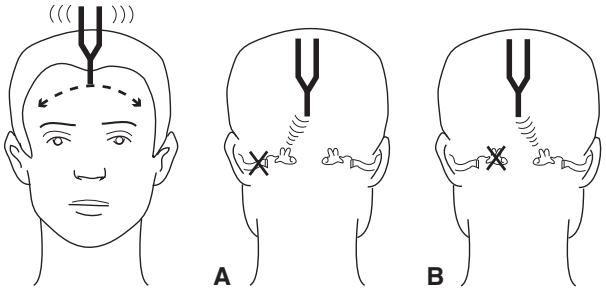

Figure 13.1 The Weber Test. **(A)** The sound of the vibrating tuning fork is perceived in the ear with a conductive hearing loss or **(B)** in the ear contralateral to the ear with a sensorineural loss.

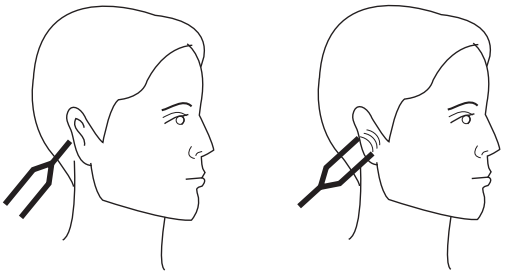

Figure 13.2 The Rinne Test. See text for explanation.

the side where the tone perception lateralized with the Weber test, then the patient likely has a conductive hearing loss in that ear. On the other hand, when the Weber test result is abnormal and the Rinne test yields a normal result, the patient probably has a sensorineural hearing loss in the nonlateralized ear. The Weber and Rinne tests are screens for hearing loss only. For example, if the patient has a mixed hearing loss, these tests can yield inconclusive results.

Otoscopic examination is frequently helpful to visualize the outer ear canal and tympanic membrane when evaluating a patient presenting with dizziness. The pinna and external auditory canal should be examined for any signs of erythema, edema, or mass and should be free from any foreign bodies, vesicles, or discharge. If the canal is totally occluded with material, such as wax or a foreign body, a conductive hearing loss will result in that ear.

Next, the tympanic membrane should be inspected for any signs of perforation, bulging, retraction, or evidence of underlying fluid in the middle ear cavity that could contribute to a conductive hearing loss. Pneumatic otoscopy should be performed by insufflating the ear canal while otoscopically examining the tympanic membrane for normal mobility. Visualization of normal drum mobility confirms the absence of a perforation and makes the presence of a middle ear effusion less likely.

Otoscopy should always be performed before performing a caloric test for vestibular function. If a tympanic membrane perforation is suspected in a patient being evaluated for vestibular loss, water caloric testing should be deferred in favor of alternative tests of vestibular function. Similarly, caloric tests are contraindicated in patients who have undergone a mastoidectomy. In the case of a painless middle ear effusion, a caloric test may be performed. However, if the test result is abnormal, the test may need to be repeated after the effusion has resolved to accurately interpret the result.

Audiological Evaluation and Management

Evaluative Procedures

Tests for Hearing Sensitivity

The most common screen for hearing acuity is the measurement of auditory thresholds to pure tone stimulation across frequencies from 250 Hz to 8 KHz. An *auditory threshold* corresponds to the lowest intensity of a sound stimulus that is detectable some percentage of time.[1] The intensity of the presented sound is typically measured in units of decibels of Hearing Level (dB HL), in which 0 dB HL is calibrated to the sound pressures that represent hearing sensitivity of young adults with normal hearing when tested in a reasonably quiet environment.[2] Audiometric thresholds of hearing sensitivity across a specified frequency range are typically plotted as an individual's *audiogram* (Fig. 13.3*A*). The abscissa of the audiogram is in units of dB HL of the presented sound stimulus, and the ordinate corresponds to the frequency of the tone presented.

An audiogram typically shows an individual's hearing thresholds to pure tone stimuli presented via *air conduction* (AC). Air-conducted sounds are presented using either binaural earphones or a loudspeaker, and thus the energy is transmitted through all parts of the ear. For this reason, measuring auditory thresholds to air-conducted sounds alone may be sufficient for diagnosing a hearing loss, but provides insufficient information for determining from which part of the ear the impairment originates. In contrast, hearing thresholds to tones presented via *bone conduction* (BC) are obtained by measuring hearing sensitivity to pure tones presented with an oscillator placed on a bony prominence of the skull, typically the mastoid process behind the auricle. The bone-conducted sounds are transmitted directly to the bone-encased cochlea, and thus bypass transmission through the outer and middle ear

Thus, if a hearing loss is identified by bone conduction audiometry, it is secondary to a cochlear or retrocochlear problem.

Pure Tone Audiogram Interpretation and Types of Hearing Loss

When AC thresholds are consistent with a hearing loss, but BC thresholds are normal, an *air-bone gap* is present, as shown in the audiogram depicted in Figure 13.3 *B* of Figure 13.3. The presence of an air-bone gap indicates that the hearing loss may be caused by a problem either in the outer or middle ear. Such a hearing loss is called a *conductive loss*. Causes for conductive losses include fluid in the middle ear, disruption or diminished mobility of the ossicular chain, or abnormal tympanic membrane compliance because of a perforation or excessive scarring. A conductive hearing loss can be caused by anything that occludes the ear canal, such as a mass, impacted wax or debris, or even a finger! Etiologies for a conductive hearing loss include otitis media, otosclerosis, ossicular chain disruption from trauma or chronic otitis media, cholesteatoma, hemotympanum (blood in the middle ear cavity), neoplasm in the outer or middle ear, superior semicircular canal dehiscence, and barotrauma.

On the other hand, if AC thresholds indicate a hearing loss, and the BC thresholds are the same as those obtained by AC, then no air-bone gap is present and the loss is of sensorineural origin (Fig. 13.3*A*, Fig. 13.3*C*). A *sensorineural loss* is therefore a result of a problem either in the cochlea or further proximal in the auditory nerve

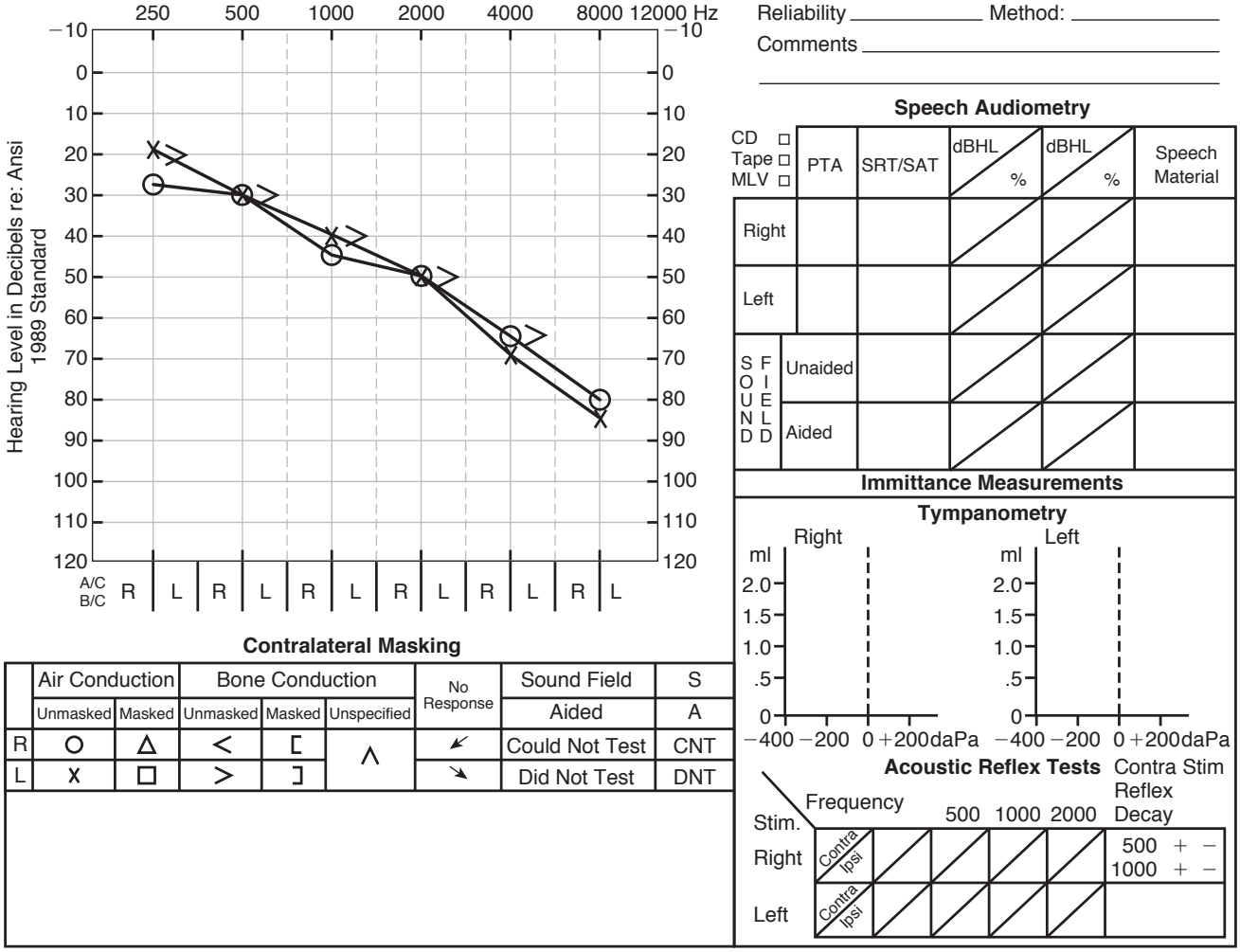

Figure 13.3 **(A)** An example of an audiogram. This is from a patient with a mid- to high-frequency sensorineural hearing loss consistent with presbycusis. Hearing loss is most severe at the highest frequencies and is less at lower frequencies.

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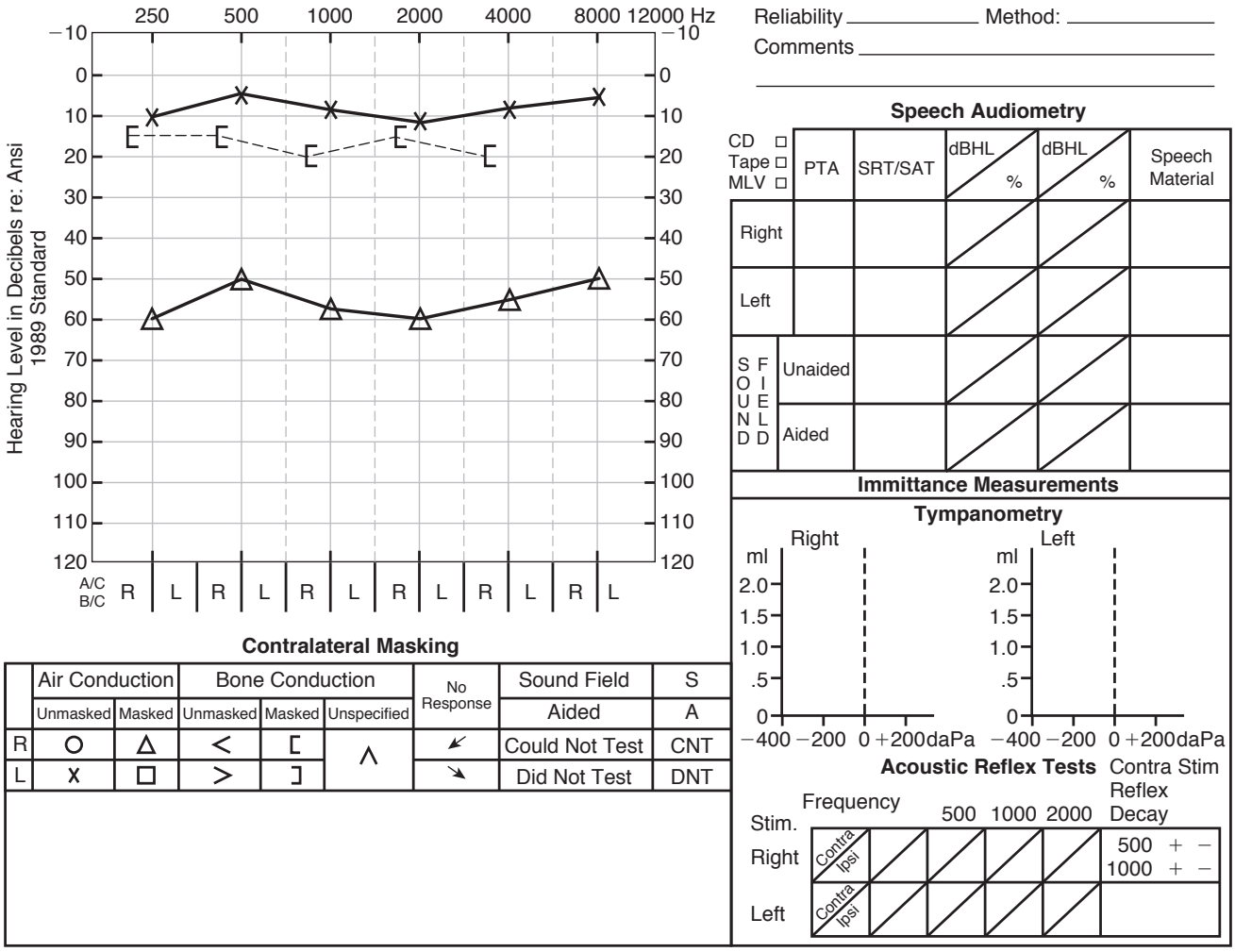

Figure 13.3—cont'd **(B)** Audiogram illustrating moderate conductive hearing loss characterized by air-bone gap in the right ear.

or CNS. Frequent etiologies of sensorineural hearing losses include presbycusis, ototoxic drugs, congenital losses, labyrinthitis, ischemia, acoustic neuroma, temporal bone fracture, and Ménière's disease. Some metabolic or infectious disorders also can cause sensorineural loss such as meningitis, congenital syphilis, toxoplasmosis, cytomegalovirus, rubella, and herpes. Birth complications such as hypoxia, prolonged mechanical ventilation, and low birth weight also increase the risk for development of sensorineural loss.

When BC thresholds show hearing loss, and AC thresholds indicate an air-bone gap, then a *mixed-type loss* is present, meaning the loss has both conductive and sensorineural components (Fig. 13.3*B*). Mixed-typed losses may be seen in advanced otosclerosis, severe chronic otitis media, and some genetic forms of auditory impairment.

In addition to classifying a hearing loss as either conductive, sensorineural, or mixed, hearing losses can also be further classified according to severity, frequencies predominantly affected, or according to the threshold shape configuration on the audiogram.

The severity of the hearing loss may be described as mild, moderate, moderately severe, severe, or profound based on the amount of threshold elevation compared with normal range (Table 13-1). In general, hearing thresholds are considered normal in adults if less than 20 dB HL. For children under the age of 18, thresholds less than 15 dB HL are considered within normal limits. Hearing loss is considered "mild" if thresholds are less than 40 dB, "moderate" between 41 and 55 dB, "moderately severe" between 56 and 70 dB, "severe" between 71 and 90 dB, and "profound" if greater than 90 dB HL.

Many sensorineural hearing losses disproportionately affect higher frequencies, such as those usually associated with aging or noise exposure, and thus may be described as being "down-sloping" on the audiogram. Classically,

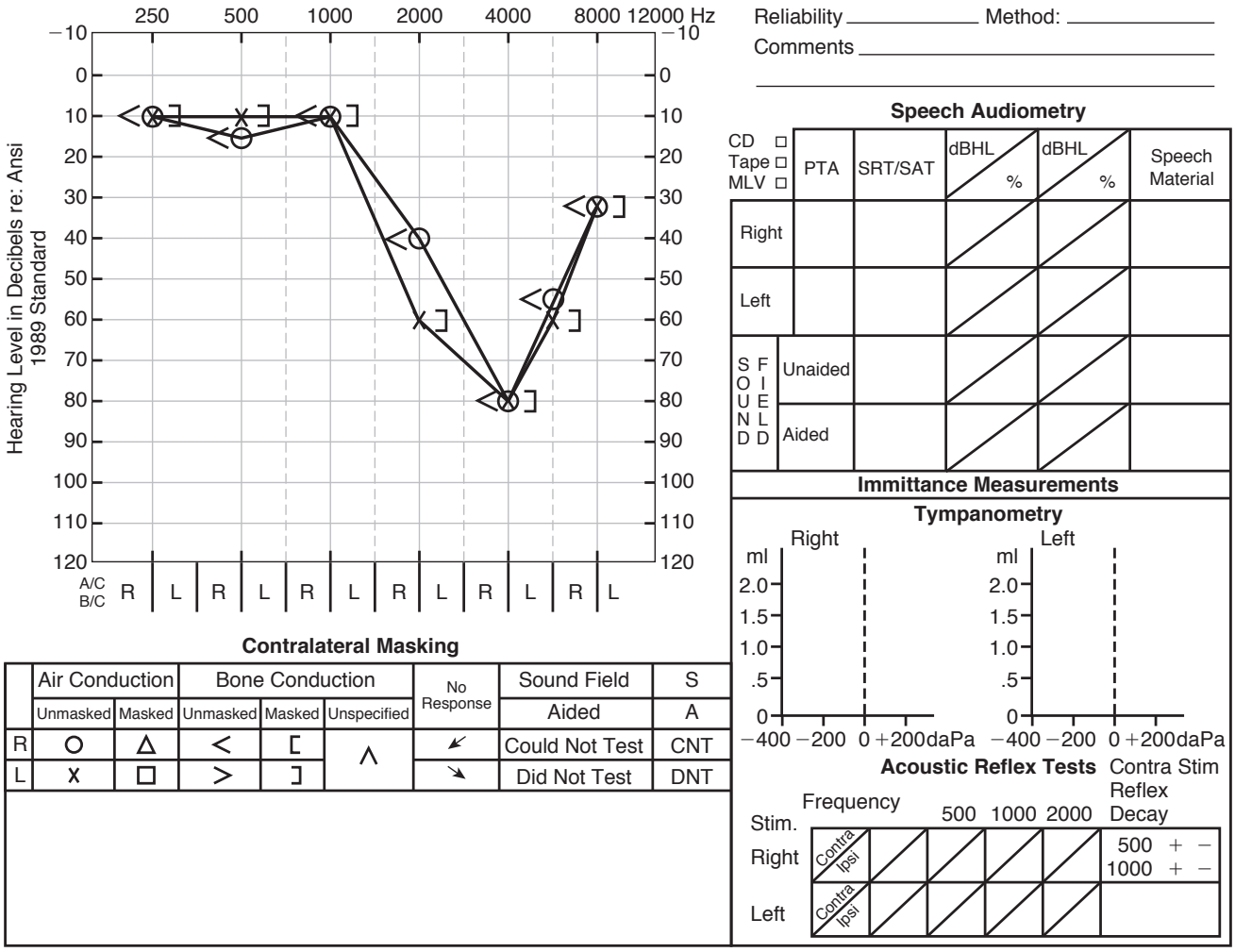

Figure 13.3—cont'd **(C)** Audiogram of bilateral noise-induced sensorineural hearing loss. Typically, it is a high-frequency hearing loss, with normal sensitivities at lower frequencies.

■ Table 13-1 CLASSIFICATION OF HEARING LOSS

Pure-Tone Audiogram Result (dB)	Classification
<20 (adults) <15 (children)	Normal hearing
16–25	Slight loss
26–40	Mild loss
41–55	Moderate loss
56–70	Moderately severe loss
71–90	Severe loss
>91	Profound loss

noise trauma leads to hearing loss in which sensitivity to 4,000 Hz is most affected. This kind of hearing loss will form a V-shape on the audiogram, or what is sometimes referred to as a "4 KHz notch." Individuals with high-frequency hearing loss have the most difficulty with hearing high-pitched consonants within words, although they may hear the lower frequencies of vowels. Therefore, English-speaking patients may complain that they can hear speech, but have the most difficulty understanding what is being said. Languages in which the words end in vowels do not produce the same problems for people with high-frequency hearing loss.

Low-frequency sensorineural hearing losses are typically seen in patients with Ménière's disease and in some forms of inherited deafness. Audiograms may also be described as being "U-shaped" or being a "cookie-bite" audiogram. Thresholds in these audiograms form a "U shape" by having either a hearing loss with either the

least or most severely affected frequencies being in the mid-portion of the hearing range.

Speech Audiometry

Another component of a routine hearing evaluation is the determination of an individual's ability to detect and recognize speech sounds. The intensity of a speech stimulus that is detectable by a listener 50% of the time is called the *speech detection threshold*, or SDT. The SDT should closely coincide with the average threshold of hearing sensitivity to pure tones at 500, 1,000, and 2,000 Hz, an average referred to as a *pure tone average*. The intensity of a speech stimulus necessary for a patient to recognize a word is typically around 8 dB greater than the SDT. The intensity level at which a listener can repeat 50% of the speech material is referred to as the *speech reception threshold*, or SRT.

The ability to recognize words presented at a comfortable listening level is assessed by determining a patient's *speech recognition score* (also referred to as the speech discrimination score in some texts). This score is obtained by presenting a list of words well within the patient's audible range and calculating the percentage correctly recognized. A score that is within normal limits is dependent on the number and type of words in the list, and normative ranges are available based on the method used. Speech discrimination scores are more likely to be abnormal in sensorineural losses and frequently are severely affected in retrocochlear losses. In addition to providing information that may be helpful in determining whether a loss is of sensorineural origin, discrimination test results provide information about the listener's ability to communicate effectively and whether hearing aids would be effective management.

Measurements of Acoustic Immittance

A routine audiological evaluation frequently includes measurements of acoustic impedance, or resistance to acoustic energy transmission. The most common tests of acoustic impedance are tympanometry and stapedial reflex measures. *Tympanometry* measures the change in the compliance with changes in air pressure in the external auditory canal. Compliance peaks when air pressure in the outer ear is equivalent to that in the middle ear. Thus, when negative pressure applied to the ear canal gives rise to the peak compliance, it may be inferred that negative pressure exists in the middle ear cavity. Such would be the case with eustachian tube dysfunction. On the other hand, when a middle ear effusion is present, changing ear canal pressure will have no significant effect on compliance. Hence, the compliance will remain constant at all pressure levels, thereby giving rise to a flat tympanogram (without a peak).

For routine immittance testing, several types of tympanograms exist, as depicted in Figure 13.4. Type A refers to normal compliance with normal middle ear pressure. Type A deep (or A_D) is consistent with eardrum compliance, which peaks with normal middle ear pressure, but with peak compliance being greater than normal. Type A_D pattern is typical for ossicular chain disruption or for a flaccid eardrum. A shallow (or A_S) pattern of tympanogram is seen when compliance peaks with normal middle ear pressure, but with a reduced peak compliance, such as would be expected for ossicular chain fixation or otosclerosis, tympanic membrane thickening, or with a middle ear mass that dampens ossicular chain mobility. Type B tympanograms have compliance measures that stay constant as the ear canal pressure is changed and are consistent with a middle ear effusion, tympanic membrane perforation, or cerumen impaction. Negative middle ear pressure gives rise to a Type C tympanogram.

Another assessment of acoustic impedance is the measurement of the acoustic reflex. The *acoustic reflex* is the contraction of the stapedius muscle in response

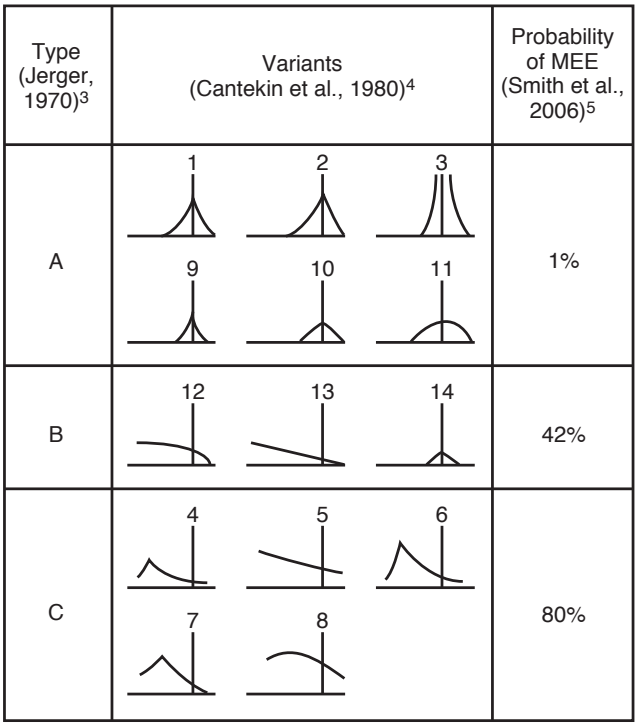

Figure 13.4 Tympanometric patterns in otitis media. The vertical height of the pressure line is the same throughout only for comparison, as the units are arbitrary. Note that the shape of the curve (peaked or gradual), the height of the curve (proportional to the compliance of the middle ear), and the middle-ear pressure at peak compliance are all related to type of tympanogram classification and to likelihood of middle-ear effusion (MEE).

to loud sound stimulation, which results in a decrease in tympanic membrane compliance leading to a reduction in the transduction of acoustic energy across the tympanic membrane. For the reflex to occur, residual hearing must be present in the ear being stimulated, and the efferent fibers to the stapedius muscle must be intact. The reflex in normal-hearing individuals typically occurs when sound is presented 70 to 100 dB above auditory threshold either in the ipsilateral or contralateral ear.

The acoustic reflex test battery evaluates for the presence of an acoustic reflex and the threshold at which it occurs. If the reflex threshold is less than 70 dB greater than the hearing threshold, a cochlear lesion is suspected because of the evidence of abnormal loudness growth or sensory recruitment.[6] Practically, the presence of the acoustic reflex at lower levels above auditory threshold confers a reduced dynamic range of comfortable hearing, which is useful information in the fitting of appropriate hearing aids.

Middle ear pathology also will inhibit the acoustic reflex, although in mild serous otitis media or in some forms of ossicular chain disarticulation, the reflex may remain intact. In most cases, both ipsilateral and contralateral reflexes are absent when the patient has a conductive hearing loss with an air-bone gap of 20 dB or more in the stimulated ear.

The pattern of abnormal acoustic reflex responses to ipsilateral and contralateral stimulation helps to differentiate impairment of the seventh or eighth cranial nerve. Abnormal elevation of the reflex threshold (i.e., more than 95 dB above the auditory threshold) or absence of the reflex in the face of normal hearing threshold measurements should raise suspicion of eighth cranial nerve pathology. Reflex response amplitude that decays to less than half of the original amplitude within 10 seconds for 500-Hz and 1-kHz tones at 10 dB above reflex threshold is also consistent with neural impairment. Finally, the absence of contralateral reflexes, with ipsilateral reflexes intact, may be seen in patients with brainstem pathology.

The acoustic reflex threshold should never be less than the pure tone threshold. If such is measured, it is most likely caused by either a nonorganic hearing loss or technical error.

Auditory Evoked Potentials

Electrical activity from the cochlea, auditory nerve, and auditory centers within the central nervous system in response to presentation of an acoustic stimulus form the auditory evoked potentials (AEP). AEPs are most commonly recorded using noninvasive scalp electrodes and normally range in humans from one one-thousandth to several tenths of a second. The latency of the responses from points along the pathway is dependent on the neuronal conduction velocity and the delay as the activity passes through neuronal synapses. For this reason, auditory evoked potentials are frequently classified by order of their latencies, which correspond to the duration of time between the presentation of the stimuli and the onset of the electrical potential.

Electrocochleography

Neuroelectric events generated by the cochlea and auditory nerve in response to acoustic stimulation are measured using electrocochleography. The electrocochlear response consists of the cochlear microphonic, the summating potential (SP), and the whole-nerve action potential generated by the auditory nerve (AP) (Fig. 13.5). The cochlear microphonic mimics the waveform of the sound stimulus, and the shift in its baseline (or DC, direct current) is called the SP. The AP corresponds to Wave I of the auditory brainstem response (ABR). Ideally, electrocochleography is recorded with an electrode placed as close as possible to the round window of the cochlea. A transtympanic needle electrode may be used referenced to another electrode placed on the forehead or tragus, or an electrode may be placed on the surface of the tympanic membrane or in the ear canal.

The electrocochleogram is particularly useful in identifying abnormal cochlear function and has been used extensively to suggest possible endolymphatic hydrops or Ménière's disease. Endolymphatic hydrops changes the elasticity of the basilar membrane, causing an increase in amplitude of the SP relative to that of the AP. The SP/AP ratio normally ranges from 10% to 50%, with higher ratios in many patients with Ménière's disease.

Auditory Brainstem Response

Measurement of the auditory brainstem response, or ABR, provides another means of assessing the integrity of the auditory pathways from the outer ear to the level of the midbrain. The ABR is a surface-recorded, averaged response representing the activity of the distal portion of the auditory pathway in response to a sound stimulus. Practical uses for ABR measurement include estimation of auditory threshold of patients unable to participate in routine audiometric testing, such as infants or the cognitively impaired, and documentation of an audiological threshold in those feigning hearing loss. Information regarding site of a lesion along the auditory pathway may be inferred by the ABR, as waveform morphology may be selectively abnormal at the site of lesion. ABR monitoring is also useful intraoperatively when surgery is performed that otherwise could put the auditory nerve at risk, such as during the resection of an acoustic neuroma.

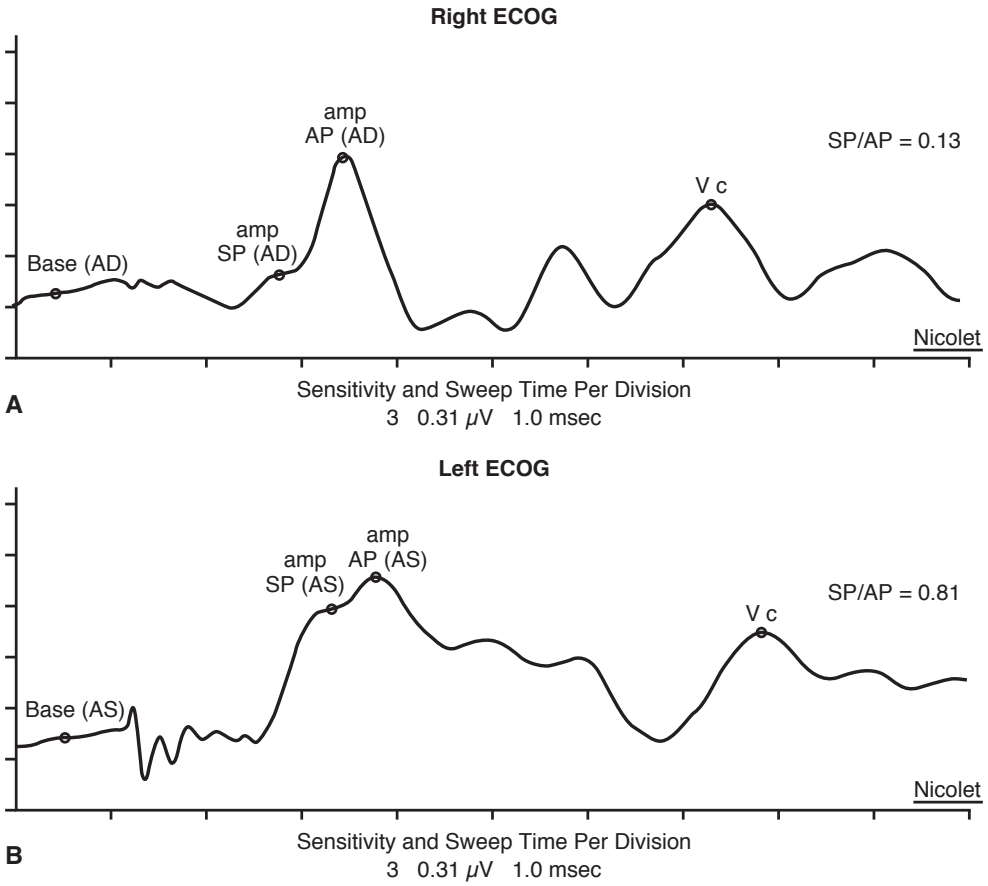

Figure 13.5 **(A)** Electrocochleogram obtained with a tympanic membrane surface electrode from a normal ear: the summating potential/action potential ratio is normal at 0.13. **(B)** Electrocochleogram obtained from the ear of a patient with Ménière's disease: the summating potential/action potential ratio is elevated (0.81).

To elicit the ABR, both clicks and brief frequenc y-specific tonal stimuli may be used. Electrodes are placed on the vertex, forehead, and behind each earlobe. The electrode on the earlobe ipsilateral to the ear stimulated is used as a reference electrode and the other as a ground electrode. The first five positive polarity peaks within 10 msec are routinely analyzed. Typically, waveforms of 1,000 to 3,000 sweeps are averaged to obtain the ABR.

The morphology of the ABR is sho wn in Figure 13.6*A*. Wave I originates from the portion of the auditory nerve within the internal auditory canal. Wave II is from the cochlear nucleus and proximal portion of the eighth nerve in the cerebellopontine angle (CP A); wave III is from the superior olivary complex; wave IV is from the lateral lemniscus, and wave V is from the inferior colliculus. Of note, w aves III to V have bilateral crossed inputs and no longer represent a solely ipsilateral response. Response latencies normally increase as stimulus amplitude decreases, but interwave intervals remain relatively constant regardless of stimulus intens ity; hence, waves I to V and III to V intervals are routinely measured.

Norms exist for waveform response latencies, and abnormally long latencies or widened interw ave intervals may be seen with lesions of the auditory nerv e or brainstem. An example of abnormal ABR waveform from a patient with multiple sclerosis is shown in Figure 13.6*B*.

Middle Latency and Cortical Event-Related Auditory Evoked Potentials

Middle latency responses (MLR) are thought to be generated by the thalamus and auditory corte x, and unlik e electrocochleography and ABR, are apparently affected by subject attention and arousal levels. Latencies of MLRs range from approximately 12 to 65 msec in humans. Responses occurring beyond 75 msec are referred to as slow vertex potentials or late component potentials. Late component potentials, which are strongly dependent on the arousal and attention of a subject, are sometimes referred to as e vent-related

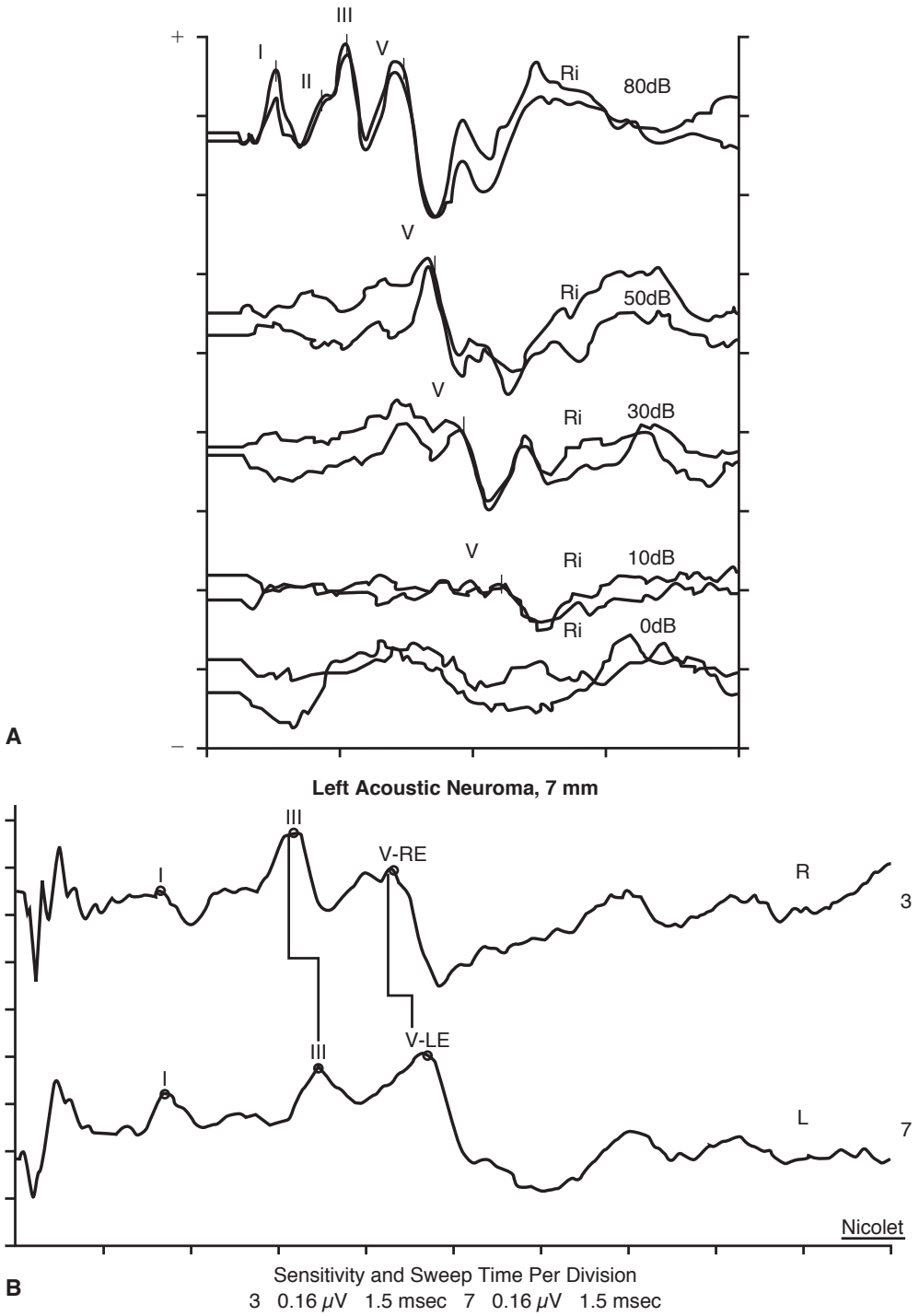

Figure 13.6 **(A)** A series of auditory brainstem responses (ABRs) recorded across a range of stimulation levels from a normally hearing young adult. At high levels, all five peaks of the ABR are labeled. As the stimulation level decreases, wave V shows decreasing amplitude and increasing latency. These responses are plotted showing vertex positive potentials as upward deflections. The scale used for the vertical axis is 0.4 μV/division. The scale used for the horizontal axis is 4 msec/division. **(B)** ABR from a patient with an acoustic neuroma in the left ear. Note the decrease in the wave amplitudes and delayed response (increased latency) on the left.

potentials (ERP). An ERP called the P300 (named for being a positive waveform peak at 300-msec latency), may be elicited only when a subject is attending to an infrequent acoustic stimulus.

Auditory steady-state responses (ASSR) are brain potentials evoked by steady-state stimuli. Steady-state stimuli are usually presented as pure tone bursts and must have frequency or amplitude modulation or both.[7] ASSR has the advantage of yielding information that can predict more frequency-selective hearing thresholds than ABR, but be less susceptible to arousal state than MLR or late component potentials if the modulation rate is high enough. Currently, ASSR is used primarily for estimating hearing thresholds in those at risk for hearing loss, especially in infants and young children.[8]

Otoacoustic Emissions

Otoacoustic emissions (OAEs) are sounds that are generated from healthy cochlear outer hair cells. Outer hair cells are motile and are thought to function as an amplifier of cochlear partition displacement during acoustic stimulation, and so generate acoustic by-products, or *cochlear echoes*. OAEs are obtainable from essentially all patients with normal hearing but may be reduced or absent in ears with mild hearing loss. Intrasubject variability is minimal in OAEs, and emissions will remain stable over several years for any one ear. OAEs are highly useful in screening for hearing loss in neonates, because the test is quick, noninvasive, and highly sensitive.

Otoacoustic emissions may be divided into spontaneous emission (SOAE), which occur without acoustic stimulation of the ear, and evoked otoacoustic emissions (EOAE), which represent a response to an acoustic stimulus. Evoked otoacoustic emissions may be further subdivided into transient evoked otoacoustic emissions, elicited by transient, brief stimulus such as a click or a brief tone burst; stimulus-frequency otoacoustic emissions, elicited by a pure tone; and distortion-product otoacoustic emissions, generated by pure tones separated by a specific frequency difference.

Otoacoustic emissions are recorded by inserting a probe tip containing both a miniature loudspeaker and a sensitive microphone into the ear canal. The loudspeaker delivers the stimulus for eliciting evoked OAEs, and the microphone samples the emission for approximately 20 msec. Recorded emissions are then signal-averaged and subsequently delivered to a sophisticated signal processor. The transient evoked otoacoustic emissions correspond to a delayed echo of the stimulus and span a frequency range of 0.4 to 6 kHz with a latency of 5 to 20 msec in humans. Emission amplitude typically decreases as frequency increases.

Distortion-product otoacoustic emissions are elicited by two stimulus tones delivered at a 55- to 85-dB sound pressure level, separated by a particular frequency interval. The most prominent distortion-product otoacoustic emission occurs at the cubic difference frequency described by the expression 2f1-f2, in which f1 represents the lower-frequency stimulus and f2 the higher-frequency primary tone. The resultant emission typically has an amplitude that is 60 dB lower than the primary tones.

Tests of Central Auditory Processing

Audiological assessment of the central auditory nervous system is not yet routine, but as increasingly more research frequently finds association of learning difficulties with auditory perceptual problems, more tests for central auditory processing disorders are being applied and advocated. Central auditory processing disorder (CAPD) was first officially described in 1992 by the American Speech and Hearing Association (ASHA). Individuals so affected had particular difficulties in retrieving, transforming, analyzing, organizing, and storing information from auditory stimuli.[9] Later interpretations of CAPD included subsequent functional deficits involving communication, language, and learning.

Screening for CAPD is not uncommon for audiologists and speech pathologists, particularly those who work with populations at high risk for having CAPD, such as children with learning disabilities and attentional disorders and patients who may have sustained a traumatic brain injury. Results of screening assessments may lead to possible referral for more comprehensive CAPD evaluation.

Auditory tasks administered to assess for CAPD consist of monotic (stimuli presented separately to each ear), diotic (same stimulus presented to both ears simultaneously), and dichotic (different stimuli presented to each ear simultaneously) tests. Many tests use measures of speech (e.g., dichotic digits, staggered spondaic words) and nonspeech (e.g., temporal gap detection, tone frequency discrimination) reception.

Audiological Management

Hearing Loss

Hearing Aids

The majority of individuals with hearing loss are treated with hearing aids (Fig. 13.7). Hearing aids receive sounds and convert them either into electrical signals, which are then amplified (analogue conversion), or to digital signals. Selection of proper amplification systems for hearing loss depends on many factors including type and extent of the loss, and whether the loss is bilateral and symmetric. In

Figure 13.7 Types of hearing aids. *Left,* in-the-ear; *center,* behind the ear; *right,* in-the-canal.

addition, personal and practical factors such as patient age, cognitive and physical health status, auditory needs, and cosmetic concerns also can play a role. Most hearing aid devices are worn either in the ear (ITE) or behind the ear (BTE); rarely a hearing aid may be worn as a device combined with eyeglasses. Recent developments in hearing aids have allowed high-powered aids for more severe hearing losses to be smaller in size and less conspicuous. The smallest hearing aids are worn in the ear canal (ITC) or completely in the ear canal (CIC) so that they are minimally visible, but these are typically not powerful enough to amplify for severe hearing loss. Some individuals with severe to profound hearing loss use body aids. A body aid consists of a microphone and power supply within a small box connected to the earmold with a lead. The power supply may be worn in a shirt pocket. Because the power supply may be larger in body aids, they can be quite powerful. These aids are not as popular now as they were in the past, but they are still useful in some patients with severe loss or in those patients with vision or dexterity limitations, which otherwise make it difficult to use a conventional hearing aid.

For patients with conductive hearing loss, bone-conduction hearing aids may be of benefit. These hearing aids use an oscillator that contacts skin overlying the mastoid bone, which results in sound being transmitted via bone conduction and bypassing the middle and outer ear. They may be worn either attached to a headband or eyeglasses, or similar to a behind-the-ear hearing aid. An alternative to traditional bone conduction aids is an implanted bone-anchored hearing aid (BAHA). The BAHA consists of a permanent titanium fixture or implant, which is surgically inserted into the mastoid bone. It has a separate directional microphone and detachable external sound processor (Fig. 13.8).

Lastly, some hearing aids are specifically manufactured to benefit those with unilateral hearing losses. The CROS hearing aid delivers sound from an ear with normal hearing to the contralateral ear with hearing loss. For those with marked asymmetric bilateral hearing loss, a BICROS hearing aid may be more appropriate. The BICROS amplifies sounds from both sides and delivers it to the ear with the better hearing.

The technology of hearing aids has significantly advanced over the past 20 years. Most modern hearing aids are digital and have the advantage of being able to selectively amplify certain frequencies to suit an individual's needs and hearing loss characteristics. Today's hearing aids may have multiple programs that a wearer may select based on the listening environment (e.g., quiet vs. noisy, speech or music), or be configured such that the programs may change automatically based on the type of acoustic signal received. Such programs reduce acoustic feedback (whistling), and background noise and may transpose frequencies (shift high frequencies that a wearer may not

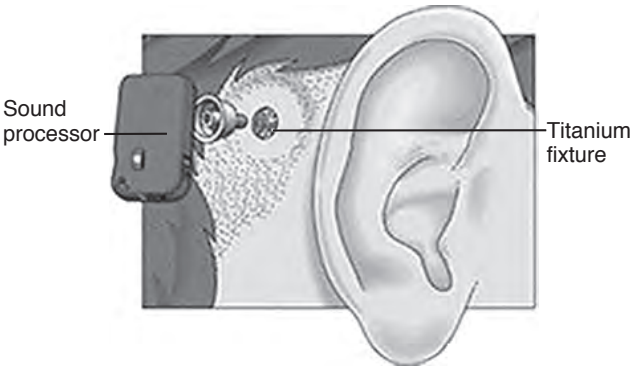

Figure 13.8 The BAHA aid. (Diagram copyright © Entific Medical Systems.)

hear to lower frequency regions where hearing may be better). Some hearing aids use directional microphones that selectively amplify sounds originating from in front of the user, which may enhance signal-to-noise ratio, particularly in noisy environments. Many hearing aids are equipped with a data logging feature that records the time a hearing aid has been used in certain environments (e.g., quiet, noisy, musical) and can be useful when an audiologist is counseling a wearer on optimal hearing aid usage.

Aural Rehabilitation

Aural rehabilitation involves the administration of services to assist people in adjusting to their hearing loss and making the best use of residual hearing and hearing aids. Exploring possible assistive devices and providing education on ways to optimally manage conversations and enhance communication skills may also be included in aural rehabilitation. Sometimes spouses or other family members can participate in aural rehabilitation to gain information about hearing loss and the attendant issues of communication. In some aural rehabilitation programs, patients are assisted in learning lip reading skills.

Tinnitus

Up to 50 million Americans experience tinnitus (pronounced either tin-NYE-tis or TINN-i-tis),[10] which can be defined simply as ear or head noises (ringing, buzzing, hissing, etc.) perceived in the absence of any external sound. Approximately 10 to 12 million Americans with tinnitus are so disturbed by it that they seek professional help and 1 to 2 million report being debilitated by their tinnitus.[10]

Tinnitus can be *objective*, heard by outside examiners, or *subjective*—inaudible to outside examiners. Subjective tinnitus secondary to a serious underlying disorder is rare, with only 0.4% of patients with either unilateral or bilateral tinnitus being diagnosed with acoustic neuroma in one study.[11] Patients reporting subjective tinnitus should be referred to an audiologist for a comprehensive hearing evaluation. Audiological findings then may determine any need for referral to an ENT specialist to rule out retrocochlear pathology.

Objective tinnitus is more rare and may be *pulsatile* or *non-pulsatile*. Pulsatile tinnitus typically arises from vascular abnormalities near the ear. Non-pulsatile tinnitus typically arises from less-threatening muscular abnormalities such as temporomandibular joint (TMJ) dysfunction or tensor tympani muscle spasms. Because pulsatile and non-pulsatile objective tinnitus may respond to medical treatment, referral to an ear, nose, and throat (ENT) specialist is strongly encouraged.

Common risk factors for tinnitus include noise, age, ear disease, and hearing loss,[12] among others. Noise exposure is a well-known cause of cochlear sensory cell damage,[13] and as a result, noise exposure is considered a primary risk factor for tinnitus.[14] The prevalence of tinnitus among those with high lifetime noise exposure (20.7%) is nearly three times greater than that of those with low lifetime noise exposure (7.5%).[15] Veterans are particularly at risk for developing tinnitus, with 49% of Iraq and Afghanistan veterans sustaining blast-related injuries having it[16] and being more than twice as likely to have chronic tinnitus (12%) as non-veterans (5%).[17] For one large audiology clinic with the Department of Veterans Affairs, 26% of referrals were for the primary complaint of tinnitus.[18]

The normal effects of aging can result in similar hair cell damage,[19] and the risk of tinnitus increases with age up to 65 or 70 years, after which the risk decreases.[20] Although the prevalence of tinnitus increases with increasing levels of hearing loss, approximately 20% of persons with tinnitus have normal hearing,[14] likely because of areas of hair cell damage being too small to be detected on a traditional hearing test.[21]

Research findings indicate that tinnitus is generated by reduced sensory input related to damage to sensory cells in the cochlea.[22] The discordant dysfunction theory[23] postulates that this aberrant nerve activity is propagated throughout the auditory nervous system resulting in cortical perception of tinnitus. Based on the *Neurophysiological Model*,[23] subawareness centers of the brain classify sound as either neutral, positive, or negative, and the specific classification results in specific activation of the limbic and autonomic nervous systems. In those who merely experience tinnitus, the tinnitus is classified as a neutral sound, resulting in minimal, if any, activation of the limbic and autonomic nervous systems with no emotional or physiological reaction to the tinnitus. For those who experience tinnitus as a significant problem, the tinnitus is classified as negative, as it elicits significant negative limbic and autonomic nervous system reactions, and subsequent emotional reactions assigned by the limbic system (such as anxiety, anger, and/or frustration) and physiological reactions assigned by the autonomic nervous system (such as increased muscle tension, release of stress hormones, increased heart rate, and/or increased blood pressure). These emotional and physiological reactions cause tinnitus to be a problem for the sufferer, because the negative limbic and autonomic nervous system reactions create physical arousal and alertness. As a result, sleep is often adversely affected: 50% of tinnitus patients may report sleep problems.[24] Sleep deprivation can cause mood swings, irritability, and problems with concentration and attention,[21] further reducing overall quality of life.

Although multiple herbal, mineral, and homeopathic compounds are marketed as tinnitus treatments, there is no cure for tinnitus. Although there is no cure, tinnitus can be managed successfully, reducing the emotional (limbic system) and physiological (autonomic nervous system) reactions to tinnitus. Successful management results in the patient feeling less stressed about tinnitus, almost never thinking about tinnitus, and feeling like tinnitus is not much of a problem.[25]

Many tinnitus patients can be helped by demystification and environmental sound enrichment. Fear of the unknown increases negative limbic system activation. *Demystification* is specific counseling directed toward providing the patient with an understanding of the mechanisms by which tinnitus is generated and by which tinnitus becomes a problem. Counseling focuses both on the patient's main tinnitus concerns and on information to correct common misperceptions and negative thoughts about tinnitus. Demystification often is sufficient to reduce limbic and autonomic system engagement and can therefore reduce tinnitus disturbance to acceptable levels.[26]

Environmental sound enrichment is the use of sound to manage tinnitus. The perceived loudness of a sound is based on a comparison of the level of the target sound to the level of other sounds in the environment.[21] Weak signals in a quiet environment are perceived as louder than the same weak signal in an enriched sound environment. Ordinarily, excited neurons reduce the activity of neighboring neurons. *Lateral inhibition* is reduced in quiet environments. The fewer nearby neurons that are stimulated, the more strongly excited neurons respond. In short, the auditory system enhances weak tinnitus signals in a quiet environment. As a result, tinnitus patients universally report that their tinnitus seems much louder in quiet environments.

The strength of the negative limbic and autonomic nervous system activities is related to the strength of the tinnitus signal. In an enriched sound environment, lateral inhibition reduces the strength of the tinnitus signal, thereby reducing the strength of negative limbic and autonomic nervous system activity: tinnitus is perceived as less loud and is therefore less disturbing.

Tinnitus and Hearing Aids

When best practices are used, hearing aids provide some degree of tinnitus relief for up to 85% of hearing-impaired tinnitus patients and moderate to complete tinnitus relief for up to 67%.[27] The tinnitus signal typically is generated in the region of the cochlea where outer hair cell damage, and resulting hearing loss, is greatest. As a result, the tinnitus signal occurs in a hearing region that is always quiet. There is minimal, if any, lateral inhibition of the tinnitus signal, and the hearing-impaired patient perceives the tinnitus as loud. Hearing aids restore more normal awareness of ambient environmental sounds, thus reducing abnormal auditory gain, increasing lateral inhibition and reducing the perceived loudness of the tinnitus.

Progressive Tinnitus Management, or PTM, is a hierarchical system designed to assist audiologists in identifying the least intensive tinnitus management strategy adequate to meet a specific patient's needs.[28-33] Tinnitus patients proceed to higher PTM levels only if the preceding level failed to result in adequate tinnitus relief. Table 13-2 summarizes the various levels of PTM.

Tinnitus Retraining Therapy (TRT) combines directive counseling and sound therapy to induce habituation to the tinnitus and has shown to result in significant improvement in 85% of patients.[34] Regular directive counseling sessions focus on demystification and education on the mechanisms of tinnitus generation and tinnitus distress. Over time, the tinnitus is reclassified as a neutral, rather than negative, stimulus. Full-time use of in-ear low-level broadband sound (white noise) generators decrease the strength of the tinnitus signal within the brain, further facilitating habituation. Successful TRT outcomes may take up to 24 months, although significant improvements may be observed in as few as 3 to 4 months.[21]

Neuromonics Tinnitus Treatment (NTT) is a modification of TRT in which the broadband stimulus is embedded in a relaxing music carrier signal. Both the broadband stimulus and the music are customized using a proprietary algorithm based on the individual patient's hearing profile. The musical selections provided with the device were chosen specifically to create positive emotional reactions and to elicit relaxation. Because the broadband treatment signal is customized to the individual, the NTT device can be used as few as 2 to 4 hours per day with successful NTT outcomes possible in as few as 6 to 9 months. Benton [18] reported that among a group of patients who had used NTT for 5 to 8 months, tinnitus awareness was reduced an average of 72% and tinnitus-related distress was reduced an average of 69%.

Central Auditory Processing Disorders

Many interventions may be implemented to assist children and adults with central auditory processing disorders (CAPD). Compensatory strategies may include consciously attending to speakers' faces (i.e., speechreading) and having talkers employ "clear speech," in which the goal is to provide the clearest possible sample of the spoken utterance.[35] Classroom interventions for students with CAPD include techniques such as preferential seating, having students preview notes before lectures, providing deliberate clear instructions, listening breaks, and allowing

■ Table 13-2 **PROGRESSIVE TINNITUS MANAGEMENT**

Level 1	Triage	Identifies those patients who may require tinnitus-specific services, most often through the use of screening questionnaires or surveys.
Level 2	Audiological Evaluation	Allows the audiologist and patient to distinguish whether tinnitus and/or other medical issues require intervention. Routine clinical procedures combined with an in-depth case history and validated surveys can provide this valuable information. Hearing loss may require amplification; patients with mental health needs may require referral to appropriate professionals; other health issues that may impact perceived tinnitus severity, such as hypertension, also may require appropriate referral.
Level 3	Group Education	Provides the patient instruction in various tinnitus management strategies, such as sound enrichment, relaxation/stress reduction, and cognitive restructuring in a support group atmosphere. Benton (2010)[18] reported that 83% of patients with primary complaint of tinnitus were able to exit PTM after Level 3 Group education.
Level 4	Tinnitus Evaluation	Consists of assessments to determine which patients are likely to benefit from specific types of treatment, to provide treatment guidelines (e.g., spectrum and/or loudness characteristics of broadband desensitization or masking sounds) and to determine if any treatment has had an effect.[36] Measurements most often obtained are quality (e.g., noise- or tone-like), pitch, loudness, perceptual location (e.g., right or left ear, both ears, midline), and minimum masking levels.[37] Level 4 may include other disciplines (e.g., mental health, otolaryngology, neurology) depending on evaluation findings and individual patient characteristics.
Level 5	Individualized Management	Provides intensive one-on-one sessions for greater patient support. Individualized management is reserved for those patients with the most problematic tinnitus that does not respond to environmental sound enrichment. In some cases, special devices may be used. Benton (2010)[18] reported that fewer than 6% of tinnitus patients progressed to PTM Level 5.

extra time to complete class work. For students, oftentimes such compensatory strategies may be included in an individualized education plan (IEP). The speech-language pathologist frequently is involved in management of language comprehension and expression difficulties, which frequently accompany CAPD, and may work together with the audiologist in an interdisciplinary team.

Different techniques for direct auditory therapy are now being developed to treat the speech perception difficulties of CAPD. Some clinicians specifically address decoding difficulties by teaching individual speech sounds and incorporating words or nonsense syllables to teach accurate speech-sound perception and discrimination. Speech-in-noise sensitization training addresses the common problem that patients with CAPD encounter understanding speech in the presence of background noise. Such direct auditory therapies, although not yet mainstream, are now showing significant promise because results show that skills learned in therapy do translate into daily skills that patients can successfully employ in their natural environments.

Medical Testing in the Evaluation of Hearing Loss

The majority of hearing losses may be attributed to genetics, age-related, or both, and may be optimally treated by conventional audiological evaluation and management. However, with some clinical presentations, a closer evaluation to uncover other potential etiologies for the hearing loss may be necessary. The clinician needs to recognize

common medical causes underlying hearing impairment, and in addition, may need a low threshold of suspicion to look for uncommon, yet potentially treatable, causes of hearing loss.

Laboratory Testing

Some systemic disorders present with hearing loss, such as autoimmune or infectious disease. For patients presenting with sensorineural hearing loss, either the fluorescent treponemal antibody absorption test (FTA-ABS) or the microhemagglutination test for *Treponema pallidum* (MHA-TP) should be obtained, particularly as hearing loss related to syphilis is potentially treatable. Routine screening for autoimmune disorders is not routinely warranted, however, unless disease is suggested from information obtained from the history and physical examination. In particular, if other cranial neuropathies are present, lumbar puncture is warranted to check the Venereal Disease Research Laboratory (VDRL) test for neurosyphilis, and also to evaluate cerebrospinal fluid (CSF) cytology, cell count, levels of glucose and protein, and cultures. In endemic geographic areas, Lyme's titers should be checked. If the history or physical exam findings are otherwise suggestive of systemic disorders, serological tests for possible autoimmune etiologies, such as erythrocyte sedimentation rate, rheumatoid factor, ACE level, c-ANCA, p-ANCA, and ANA may be helpful.

Radiological Imaging

Radiographical imaging is warranted in selected patients with hearing loss. Magnetic resonance imaging (MRI) with gadolinium enhancement is presently the "gold standard" in evaluating hearing losses of possible retrocochlear origin and particularly in making the diagnosis of a tumor on the eighth cranial nerve or in the CPA region. Computed tomography (CT) is useful in patients with suspected labyrinthine congenital anomalies, such as large vestibular aqueduct syndrome or Mondini dysplasia. CT also is useful in cases with suspected labyrinthine fistula or temporal bone fractures.

Clinical Presentations of Auditory Impairment

Presbycusis

An estimated 40% to 50% of adults greater than 75 years of age have hearing loss. The loss associated with aging, or presbycusis, is typically of gradual progression and affects both ears symmetrically. Hearing loss for high frequencies is associated with presbycusis and occurs secondary to age-related loss of outer hair cells and spiral ganglion cells greatest in the basal turn of the cochlea.[38-40] Other structures in the auditory pathway are also vulnerable such as the striavascularis, which atrophies with age; cochlear neurons, which decrease in number; and the middle ear, auditory brainstem, and cortex, which exhibit degenerative changes. The outcome of age-related changes in the auditory system is characteristically a gradual and progressive loss of hearing in both ears, which initially reduces the ability to hear high pitches. Such hearing loss leads to a listener's complaints of having difficulty discriminating speech and problems hearing in noisy environments. Frequently, tinnitus also accompanies presbycusis.

Hearing Loss Associated with Noise Exposure

Noise exposure can cause either a temporary or permanent hearing loss. A noise-induced temporary threshold shift (NITTS) is a temporary hearing loss that follows exposure to loud sound and is characterized by a subjective aural fullness caused by reduction in the high-frequency sensitivity, and tinnitus. The symptoms of NITTS may be on the order of minutes or may last several hours or days. Stimulus frequencies from 2,000 to 6,000 Hz are most effective at producing NITTS[41,42] as are longer stimulus durations and greater stimulus intensities.[2,43,44] Noise-induced temporary threshold shift is associated with swelling of cochlear hair cells and reduction in stiffness of hair cell cilia.[45]

Recurrent or chronic exposure to loud sound may lead to noise-induced permanent threshold shift (NIPTS). Swelling of hair cells after noise exposure can lead to cell rupture and disruption and permanent loss. In addition, stereocilia may become fused[46] and become unable to effectively transmit energy to the hair cells.[47] Progressive hair cell damage may subsequently lead to degeneration of auditory nerve fibers and to changes within the central auditory system.[47]

Typically, noise-induced hearing loss associated with workplace noise is characterized by having the greatest loss of hearing from 2,000 to 6,000 Hz, with maximum hearing loss occurring at 4,000 Hz. This leads to the classic "noise-induced 4 KHz notch" seen on an audiogram. However, with continuing noise exposure, the loss will expand to include other frequencies outside the 2,000 to 6,000 Hz range.

A single instance of intense loud sound, such as occurs with an explosion, may cause physical trauma to the ear resulting in permanent hearing loss. Acoustic trauma may also cause rupture of the tympanic membrane and/or fracture of the ossicular chain. Such hearing loss is noticeable immediately.[47]

Sudden Sensorineural Hearing Loss (SSNHL)

The cause of sudden sensorineural hearing loss (SSNHL) is not completely understood and has been hypothesized to be a result of a variety of etiologies: viral, vascular, hypoxia, intralabyrinthine membrane rupture, inflammatory, metabolic, and others. SSNHL is considered an otologic emergency, because accepted guidelines for treatment are to institute pharmacological agents within 2 weeks to achieve maximum benefit. Many physicians use the criteria for SSNHL as minimum loss of 30 dB in three contiguous frequencies measured in routine audiometric evaluation over a period of 3 days or less. The rate of spontaneous resolution is relatively high at about 65%, but variables such as severity of loss, flat or downsloping audiograms, age extremes, elevated erythrocyte sedimentation rate, time from onset to diagnosis, and the presence of vertigo all portend a poorer prognosis. In general, patients with lower frequency hearing loss have a better chance for recovery.[48] Steroids administered either orally or intratympanically is currently the mainstay of treatment for SSNHL and have been shown in some studies to be of value.[48,49] A recent literature review cautiously concluded that application of hyperbaric oxygen therapy may significantly improve hearing in cases of acute idiopathic SSNHL, but that hyperbaric oxygen was not likely to be of benefit in longstanding SSNHL. Various reviews of the literature have failed to identify clear benefit with other treatments for SSNHL, for example, vasodilators,[50] antivirals,[51] or addition of an antiviral agent to steroid treatment.[52] Whether other combinations of agents may be more effective remains unclear and is an area of ongoing research.

Hearing Loss from Infectious Disease

Labyrinthitis

An infectious or inflammatory process within the labyrinth can take two forms pathologically: serous or suppurative. *Serous labyrinthitis* is defined as an abnormal process within the labyrinth as a result of the degradation of the tissue-fluid environment within the inner ear caused by bacterial toxins or contamination of perilymph with blood, products of tissue injury, or air at surgery. The principal abnormal finding in serous labyrinthitis is endolymphatic hydrops, with temporary or permanent hearing loss and vestibular dysfunction. Commonly, labyrinthitis is clinically diagnosed when patients present with a relatively sudden onset of sensorineural hearing loss and acute vertigo. Most commonly, serous labyrinthitis is of viral origin.

Suppurative labyrinthitis is caused by bacterial invasion of the inner ear and is manifested by profound hearing loss and acute vertigo. The route of invasion can be from otitis media, via a fistula between the middle ear and the labyrinth. Alternatively, the route of invasion can be meningogenic, through the cochlear aqueduct or internal auditory canal. Suppurative labyrinthitis is the most common etiology of deafness associated with meningitis.

Measles and mumps rarely cause hearing and vestibular loss in developed countries because of widespread vaccination. The hearing loss in measles is usually bilateral and moderate to profound, and vestibular function is similarly affected. In contrast, mumps typically causes a unilateral hearing loss. Cytomegalovirus (CMV) may cause hearing loss as well, but is typically associated with an immunocompromised patient, such as one with acquired immunodeficiency syndrome (AIDS).

Ramsay Hunt Syndrome

If skin vesicles are noted on the pinna or external auditory canal of an ear with new onset hearing or vestibular loss, herpes zoster infection should be suspected. If accompanied by an ipsilateral facial weakness, the condition is referred to as *Ramsay Hunt Syndrome* caused by a herpes virus. Facial paralysis is most commonly seen; however, hearing loss and vertigo can occur alone or in combination as well. The infection probably represents the reactivation of a latent virus, such that, after the primary infection, the virus travels to the dorsal root ganglion of a cranial nerve, where it remains dormant until reactivated. Incidence and severity is increased with advancing age associated with an age-related decrease in cellular immune response to herpes zoster virus.[53]

Syphilis

The incidence of hearing loss is as high as 80% in cases of symptomatic neurosyphilis and 29% in those with asymptomatic neurosyphilis.[39] The mechanism of hearing loss in syphilis is either a meningolabyrinthitis or an osteitis of the temporal bone with secondary involvement of the labyrinth. Pathologically, a resorptive osteitis is seen in the temporal bone, with endolymphatic hydrops noted within the labyrinth. Clinically, hearing loss caused by syphilis is frequently indistinguishable from Ménière's disease, as it may fluctuate and be associated with tinnitus, aural fullness, or episodic vertigo. Hennebert's sign and Tullio phenomenon may be seen in otosyphilis. Treatment consists of antibiotics and corticosteroids.

Rocky Mountain Spotted Fever (RMSF)

Rapidly progressive sensorineural hearing loss has been associated with Rocky Mountain Spotted Fever (RMSF), which is thought to be secondary to a vasculitis involving the auditory system. The hearing loss may be transient. RMSF is caused by the tick-borne pathogen *Rickettsia*

rickettsii. Diagnosis is made on the basis of presentation and confirmed by serologic titers. Treatment is with broad-spectrum antibiotics.

Lyme Disease

Lyme disease is caused by the tick-borne spirochete *Borrelia burgdorferi*. Most commonly, it gives rise to facial paralysis, but it can cause hearing or vestibular loss and should be considered as a possible etiology in endemic areas. Recommended antimicrobial therapy for Lyme disease with neurological manifestations is third-generation cephalosporins, if no allergy exists.

Pharmacological Toxicity

Like the vestibular system, the auditory system is vulnerable to the toxic effects of a number of pharmacological agents. Other than removing the offending agent, no reliable treatment is known for hearing loss from ototoxic drug therapy.

Aminoglycosides

Probably the most common ototoxic agents encountered in clinical practice are the *aminoglycoside* antibiotics, which are lethal to hair cells in the inner ear. Kanamycin, tobramycin, amikacin, neomycin, and dihydrostreptomycin are more cochleotoxic than vestibulotoxic, whereas gentamycin and streptomycin cause disproportionately more vestibular damage than cochlear.[54] Hearing loss may be asymmetric and progress even after cessation of the therapy. Some auditory recovery may occur weeks to months after treatment. Risk factors for aminoglycoside ototoxicity include renal impairment, longer duration or high doses of drug exposure, advanced age, and combined administration of other ototoxic drugs, particularly loop diuretics.[55]

Aspirin

Aspirin toxicity may present with tinnitus and reversible sensorineural hearing loss, and the toxic effects are typically dose dependent. The mechanism of injury is most likely alteration of the turgidity and motility of the outer hair cells, with resultant loss of OAEs and reduction in cochlear action potentials.[56] Caloric responses can also be reduced by salicylates.[57] Nonsteroidal anti-inflammatory drugs (NSAIDs) such as naproxen, ketorolac, and piroxicam have been known to cause reversible sensorineural hearing loss, but this occurs far less frequently than that seen with salicylates.

Chemotherapeutic Agents

Numerous cancer chemotherapeutic agents have been associated with ototoxicity. Cisplatin (*cis*-diamminedichloroplatinum) is associated with irreversible, bilateral, high-frequency hearing loss. If ultra-high frequencies are tested, then virtually all patients who have received cisplatin probably have at least some degree of ototoxicity.[58] Occasionally, the hearing loss is accompanied by vertigo or tinnitus. Other chemotherapeutic agents known to cause ototoxicity include the vinca alkaloids vincristine, and to a lesser extent, vinblastine.

Ototopical Medications

Ototopical preparations containing neomycin, gentamicin, and tobramycin are widely used for the treatment of otitis externa and chronic otitis media. However, in experimental animals and patients, these drugs are now known to cause auditory and vestibular loss when they are instilled in the middle ear cavities of normal healthy ears. The toxic effect of gentamycin administered transtympanically is the mechanism of action of chemical labyrinthectomies performed to treat patients with refractory Ménière's disease.[59] Thus, the use of aminoglycoside-containing topical preparations in uninflamed ears with tympanic membrane perforations should be avoided. Other ingredients in ototopical preparations that have ototoxic potential include polymyxin B, propylene glycol, acetic acid, and antifungal agents.[60,61]

Other drugs that have been reported to cause hearing loss include deferoxamine, an iron-chelating agent, and eflornithine, used for the treatment of *Pneumocystis carinii* pneumonia, trypanosomiasis, cryptosporidiosis, leishmaniasis, and malaria. Quinine has long been known to cause sensorineural hearing loss, tinnitus, and visual disturbances.[55] Loop diuretics alone can cause reversible hearing loss, which is usually bilateral and symmetric.

Surgical Management of Hearing Loss

Although a thorough review of this topic is beyond the scope of this chapter, many forms of ear disease can give rise to hearing loss amenable to surgical treatment. Most often, surgical approaches are used to treat conductive hearing loss, because frequently the loss is secondary to a structural problem in the middle or outer ear. When sound transduction cannot take place because either disease or trauma has disrupted the ossicular chain, for example, this can frequently be surgically corrected either by clearing the middle ear cavity of the structural disease and/or replacing the ossicles with a prosthetic device. Disruptions in the tympanic membrane can also be treated by a tympanoplasty procedure. Rarely do etiologies of conductive hearing loss without a sensorineural component give rise to vestibular symptoms.

In contrast to management for conductive hearing loss, surgical treatment for sensorineural hearing loss is considerably less common, but notable approaches include cochlear implants and interventions for CPA tumors, semicircular canal dehiscence, and other perilymphatic fistulas (PLFs). CPA tumors and PLFs commonly give rise to vestibular symptoms, and patients with cochlear implants may have vestibular *and* hearing loss. Thus, clinicians managing vestibular disorders may frequently encounter patients with these lesions.

Cochlear Implants

Cochlear implants are an option for an increasing number of hearing-impaired patients. Although the first attempt to electrically stimulate the auditory system occurred nearly two centuries ago, the development of a cochlear prosthesis to restore hearing to patients with sensorineural hearing loss took place only in the past four decades. A deafened auditory nerve was first electrically stimulated by Djourno and Eyries in 1957,[62] but it was not until 1972 that a commercial device was developed for this purpose. Traditional criteria for implantation include bilateral profound-to-total sensorineural hearing loss, inability to benefit from conventional hearing aids, good physical and mental health, and the motivation and patience to complete a rehabilitation program.[63]

All cochlear implants have several elements in common (Fig. 13.9*A*). A microphone, usually at ear level, detects acoustic energy, which is then encoded into an electrical signal by the external sound processor. The electrical stimulus is then transmitted to the implanted electrode array either in the middle ear or inner ear through some form of signal coupler (Fig. 13.9 *B*). The most commonly used commercially available implants in the United States are those with 22 active electrodes.

The criteria for candidacy for cochlear implantation has become less rigorous over the years. Specifically, cochlear implants are now implanted more commonly bilaterally[64] and have now been used in patients with unilateral deafness[65] and in children less than 2 years of age. Children even younger than 12 months of age have been implanted with good results when hearing loss can be reliably confirmed.[66] Cochlear implants that are either shorter than standard lengths or partially surgically inserted into the cochlea have been used with success in patients in which preservation of low-frequency hearing was a goal.[67-69] Cochlear implants have been advocated as treatment for hearing loss associated with enlarged vestibular aqueduct syndrome.[70,71] Implantation criteria continue to broaden as studies now show success with cochlear implants in patients with genetic disorders associated with hearing loss, such as connexin 26 mutation,[72] Usher syndrome,[73,74] mitochondrial disorders,[75] Waardenburg syndrome,[76] and Jervell and Lange-Nielsen syndrome.[77,78]

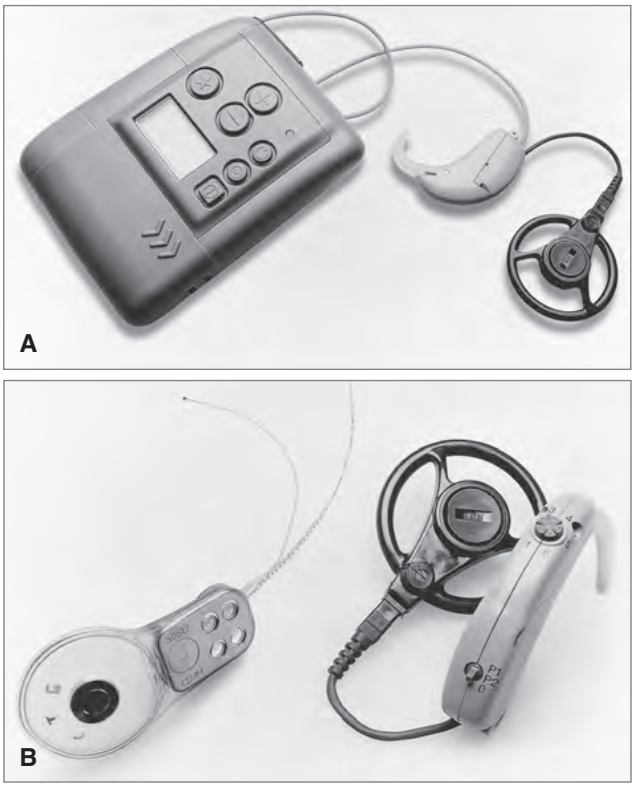

Figure 13.9 Cochlear implant. **(A)** A multichannel cochlear implant processor, microphone, and magnet coil. **(B)** Cochlear implant internal device electrode array and ear-level cochlear implant. (Photographs courtesy of Cochlear Corporation.)

The technology of the cochlear implant continues to improve, particularly in the area of speech processors with increasingly more sophisticated coding strategies. Patient selection criteria have continued to broaden as increasing numbers of patients are found to benefit from the procedure. The rehabilitation process continues to be lengthy, however, because it involves adjustments of the speech processor and extensive aural training.

Cerebellopontine Angle (CPA) Tumors

Although the acoustic neuroma (acoustic neurinoma, vestibular schwannoma, acoustic neurilemmoma) is the most common lesion found in the cerebellopontine angle, a variety of other benign and malignant lesions may be found in this region. Patients with these lesions may also present with vertigo, unsteadiness, headache, twitching, weakness, or numbness of the face. Unilateral progressive sensorineural hearing loss is the most common first symptom, and

predominantly high-frequency sensorineural hearing loss with poor word discrimination ability is characteristic, although other hearing loss patterns have been described. Little relationship exists between the size of the tumor and the audiometric results.[79]

Brainstem evoked response audiometry is the single most reliable audiometric diagnostic procedure in the diagnosis of CPA lesions. Specificity and sensitivity of the ABR ranges from 92% to 96% in the diagnosis of acoustic neuromas of approximately 1 cm in diameter.[80] However, as many as 1/3 of patients with smaller tumors on MRI will have normal ABRs. Vestibular hypofunction is demonstrated in 82% to 96% of patients with CPA lesions,[81] and positional nystagmus is a very frequent finding.[82] MRI of the brain with gadolinium contrast enhancement is the optimal test for diagnosing acoustic neurinomas (Fig. 13.10), which may be purely within the internal auditory canal, and are particularly useful for monitoring tumor growth and postoperative recurrence.

The acoustic neuroma arises most commonly from the inferior division of the vestibular nerve and next most commonly from the superior division. In rare cases, it arises from the cochlear nerve.[83] Acoustic neuromas are a feature of neurofibromatosis type 2 (NF2). A partial deletion in the long arm of chromosome 22 has been found in patients with NF2.

Clinically, symptomatic acoustic neuromas are seen in 1.5 per 100,000 population, which constitute only 0.2% of the acoustic neuromas found in postmortem studies of temporal bones. Because the growth rate of these tumors is generally slow, and malignant transformation is rare, in patients who are otherwise poor operative candidates, the most appropriate management may be monitoring the lesion only. Patients with serviceable hearing with unilateral tumors that are less than 2.0 cm in diameter may be offered either surgical removal with an attempt to save hearing or radiographic monitoring, reserving surgery for an enlarging tumor. For small tumors with poor or no hearing, either surgical removal or radiographic follow-up may be considered. For larger tumors in healthy young and middle-aged adults, surgical removal is generally recommended, because the morbidity of surgical removal is directly correlated with the size of the tumor, and enlargement in healthy patients under 65 years of age is likely. For large tumors in older patients without evidence of brainstem compression, radiographic monitoring is recommended, reserving surgery for enlarging tumors and progressive symptoms. In NF2 cases with bilateral tumors, the goal is to avoid bilateral deafness and vestibular hypofunction, so if hearing is good in both ears, the goal is to proceed with surgical excision of one tumor in the attempt to preserve hearing.[84]

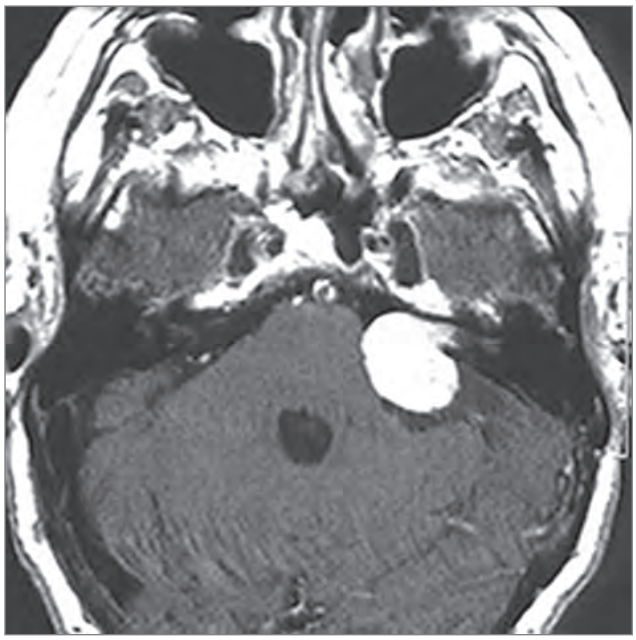

Figure 13.10 Post contrast T1-weighted axial image demonstrating an intense contrast enhancing extraaxial mass at the left cerebellopontine angle close to the left internal auditory canal (IAC) consistent with an acoustic neuroma.

Superior Semicircular Canal Dehiscence

Superior semicircular canal dehiscence is caused by a disruption in the bony capsule of the temporal bone overlying the superior semicircular canal, which renders the vestibular labyrinth sensitive to loud sounds or pressure changes. The condition is probably congenital, because it is frequently bilateral but oftentimes may be asymptomatic. Diagnosis can be made by demonstration of characteristic torsional nystagmus induced by application or positive or negative ear canal pressure or by presentation of loud sound.

Frequently, these patients first present with a conductive hearing loss. Moreover, patients with this condition may actually be hypersensitive to bone-conducted sounds, resulting in measurements of audiometric air-bone gaps in which BC thresholds are lower than normal 0 dB hearing level and AC thresholds are within normal threshold range. This hypersensitivity to bone-conducted sounds may generate patient complaints of hearing their own pulse or eye movements. Treatment is surgical and may involve blocking the affected superior semicircular canal.[85,86]

Perilymphatic Fistula

Perilymphatic fistulas (PLFs) are associated with a number of conditions including barotrauma, head injury, heavy lifting or straining, invasive cochlear procedures, stapedectomies, congenital defects of the middle ear, and labyrinthine

erosion from mastoiditis or chronic granulomatous disease. The existence of PLFs of the oval and round windows is controversial, although some clinicians believe PLFs may occur spontaneously as well.[87] The onset of PLF frequently presents as sudden onset of either hearing loss, vertigo, or both. Unsteadiness or dizziness is present in most cases, and the dizziness is usually positional in nature. Seventy-five percent of patients will complain of tinnitus, regardless of hearing status. Hearing loss is present in 53% of patients and usually does not fluctuate. Moreover, patients do not typically present with episodic vertigo, tinnitus, or an aural fullness, features which would be more consistent with Ménière's disease. Hearing loss with PLF is usually sensorineural but may be predominantly conductive in the case of a stapes abnormality.

Physical findings of PLF are not always present, and identification of the involved ear may not be straightforward if no hearing loss is present. Examination findings, if present, may include positional nystagmus occurring in several head positions, but especially with the suspect ear down. Hennebert's sign may be present with PLF, which consists of a few beats of nystagmus or ocular deviation induced with application of negative or positive pressure to the external auditory canal of the involved ear with an intact tympanic membrane. This sign may be secondary to stimulation of the utricular macula as a result of deformation of the utricular wall from the pressure change.

No specialized audiometric or ENG result, including caloric, has been of any value in identifying PLF or the side of the lesion. Black and coworkers[88] reported posturography as being a highly specific and sensitive test for PLF, but this finding has not been corroborated well by others.[89] Other than rarely documenting intralabyrinthine air in association with perilymphatic leakage, neither CT nor MRI can positively identify a PLF.

In animals, it has been shown that most PLFs will heal spontaneously, and some physicians will elect for nonoperative management for at least several days, giving time to complete the evaluation and allow for possible spontaneous recovery. Conservative management comprises bed rest, with the head of the bed elevated to 30 to 40 degrees, and a stool softener. Lifting, stooping, nose-blowing, or any kind of activity that could elevate cerebral or middle ear pressure is not permitted. Hearing loss is monitored with interval audiograms. In cases of sudden hearing loss, worsening hearing or dizziness, or if hearing fails to improve, surgery may be recommended. Of note, however, is that in contrast to PLF secondary to superior semicircular canal dehiscence, which may be more readily diagnosed, identification of PLF of the oval or round window is not always possible, even intraoperatively, unless an obvious anatomical malformation is present.

Other Etiologies

Many other disorders may exhibit hearing loss as a symptom, although rarely are auditory symptoms the sole manifestation of the disorder. For example, ischemic stroke and multiple sclerosis may give rise to hearing loss, as may most any intracranial process that disrupts the central auditory pathways. Paget's disease (osteitis deformans), although a rare disorder of bone, frequently manifests with hearing loss and can be treated with calcitonin or etidronate disodium.[90] Rarely, hearing loss may accompany blood dyscrasias, acquired immunodeficiency disease, autoimmune disease, or paraneoplastic disease.

Summary

This chapter provides an introduction to the evaluation and management strategies for auditory disorders. Components of the history and physical examination that are pertinent in the evaluation of this patient population are discussed, and standard medical and audiological testing procedures and their indications are reviewed. The clinical presentations of the more common etiologies of hearing loss are outlined by their relevance to the clinician who assesses and treats patients with vestibular disorders, because the pathophysiology underlying vestibular disease also frequently gives rise to auditory symptoms. Familiarization with the clinical presentation and audiological features for disorders of the auditory system significantly assists the clinician in the formulation of differential diagnoses in these patients, which in turn provides the foundation for the choice of the appropriate management and therapy.

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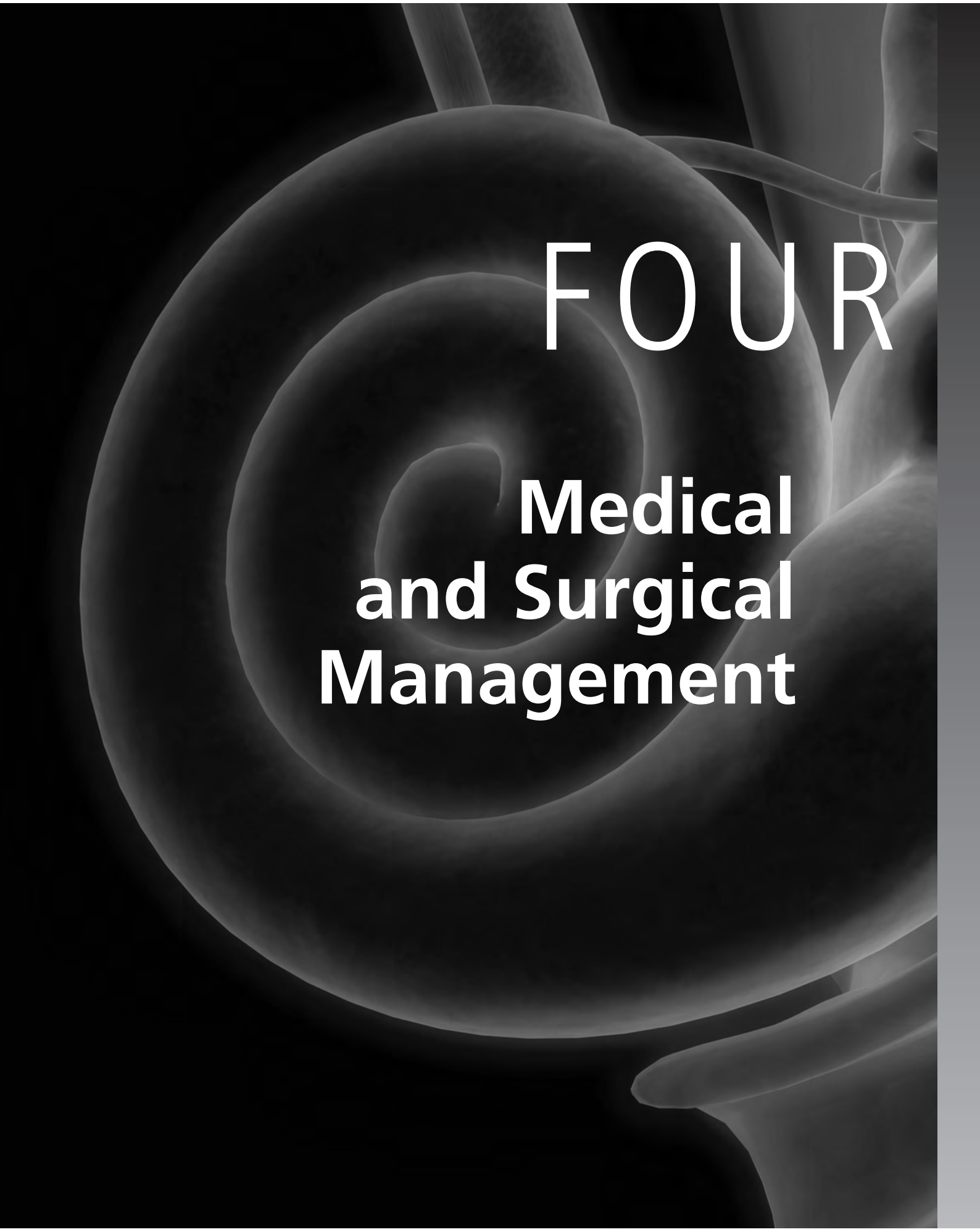

FOUR

Medical and Surgical Management

CHAPTER 14

Pharmacological and Optical Methods to Treat Vestibular Disorders and Nystagmus

Rosalyn Schneider, BA ■ *R. John Leigh, MD*

Patients with vestibular disease may complain of vertigo, oscillopsia, or the visual consequences of nystagmus.[1] *Vertigo* means whirling or spinning, but the term is also used to describe other illusions of motion, and implies vestibular imbalance. *Oscillopsia* consists of illusory, oscillatory movements of the seen environment. When oscillopsia occurs with head movements, it usually implies loss of vestibular function. Oscillopsia also may occur when the head is stationary in patients with spontaneous *nystagmus*. In this chapter, we review current treatments for vertigo, oscillopsia, and the visual consequences of nystagmus from the standpoint of known pathophysiology.

In interpreting and treating symptoms resulting from vestibular disorders, it is helpful to consider the nature of the demands placed on the vestibular system *during natural activities, especially locomotion*. The purpose of the vestibulo-ocular reflex (VOR) is to maintain clear and stable vision during natural head movements. A major threat to clear vision is posed by the head perturbations occurring during locomotion. This fact was pointed out by the anonymous physician J.C. who had lost vestibular function because of aminoglycosides[2]; he wrote, "During a walk I found too much motion in my visual picture of the surroundings to permit recognition of fine detail. I learned that I must stand still in order to read the lettering on a sign."

The range of peak velocities and predominant frequencies of head rotations measured in 20 normal subjects as they walked or ran in place are summarized in Figure 14.1. Note that although peak head velocity is generally below 150 deg/sec, the predominant frequencies range from 0.5 to 5 Hz.[3,4] The latter value exceeds the frequencies that vestibular physiologists have conventionally used to test patients in the laboratory but generally corresponds to the bedside head-impulse test.[5] Besides head rotations, linear movements or "translations" occur during locomotion.[6] However, recent studies indicate that head translations are less of a threat to vision than are rotations, unless subjects view near targets during locomotion.[7,8] This information about the range and nature of head perturbations that occur during natural activities such as locomotion is useful in formulating strategies to rehabilitate patients with vestibular disorders, especially if the functional goal of therapy is the ability to walk normally, without aids.

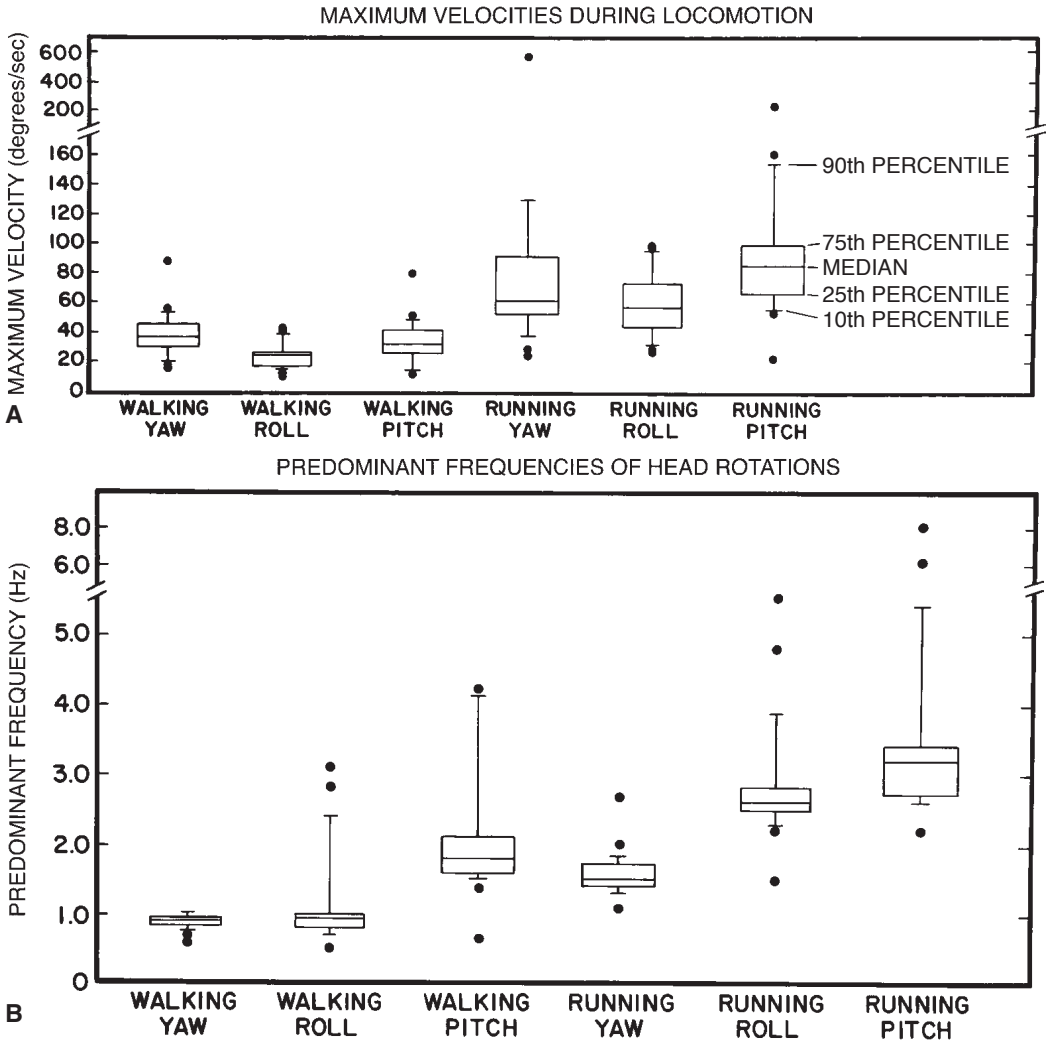

Figure 14.1 Summary of the ranges of **(A)** maximum velocity and **(B)** frequency of rotational head perturbations that occur during walking or running in place. Distributions of data from 20 normal subjects are displayed as Tukey box graphs, which show selected percentiles of the data. All values beyond the 10th and 90th percentiles are graphed individually as points. Reproduced with permission.[1]

Vertigo

Pathophysiology of Vertigo

During locomotion, our multisensory inputs—visual, vestibular, and somatosensory—allow a unified perception of self-motion that is consistent with our expectations. When vestibular inputs are disturbed, vertigo results—a distressing, illusory sensation of turning or other unexpected movement. Thus, it is the mismatch between the abnormal vestibular and other sensory inputs that causes vertigo.[9,10] Although rotational vertigo connotes disturbance of the semicircular canals or their central projections, sensations of body tilt or impulsion (e.g., lateropulsion) imply otolithic disturbance.[1]

Vertigo should be differentiated from other causes of "dizziness," such as pre-syncopal faintness, loss of stable balance, lightheadedness, and psychological disorders (such as agoraphobia, acrophobia, and phobic vertigo syndrome).[11] A careful, unhurried history and a systematic physical examination are required to distinguish these various causes of the complaint of dizziness.[1,9,10] Accurate identification of symptoms is essential before therapies are begun. Vertigo caused by disease of the vestibular system should be differentiated from "physiological vertigo" that some normal individuals experience with motion or at height.[12]

Besides causing vertigo, sudden loss of tonic neural input from one labyrinth or vestibular nerve causes

nystagmus and unsteadiness.[1,10] The nystagmus is typically mixed horizontal-torsional with quick phases beating away from the side of the lesion. The nystagmus is more marked when visual fixation is prevented (e.g., with Frenzel goggles) and when the patient looks in the direction of the quick phases, a phenomenon known as Alexander's law.[1] Past-pointing to the side of the lesion reflects imbalance of vestibulo-spinal reactions.[1] Patients with rotational vertigo caused by acute, peripheral vestibular lesions are often uncertain as to the direction of their vertiginous illusions. This is because there is a conflict between vestibular sensation indicating self-rotation in one direction and retinal image motion (consequent on slow phases of vestibular nystagmus) that, when self-referred, connotes self-rotation in the opposite direction. It is therefore worthwhile to evaluate patients' vestibular sense alone by asking them to report their perceived direction of self-rotation with the eyes closed, which will aid localization of the side of the lesion. Although most patients with acute peripheral lesions recover to become independent for most activities within a month or two, some are left with recurrent vestibular symptoms, and others experience benign paroxysmal positional vertigo. These topics are discussed in detail in other chapters.

Neuropharmacology of the Vestibular System and Nystagmus

A broad range of neurotransmitters have been identified within the vestibular system.[13] *Vestibular hair cells* express muscarinic acetylcholine receptors.[14] *Primary vestibular afferents* in the eighth cranial nerve, which synapse in the vestibular nuclei, express glutamate and histamine as transmitters.[13,15,16] *Secondary excitatory projections* from the vestibular nuclei to ocular motoneurons express glutamate and aspartate as transmitters.[17] *Secondary inhibitory projections* from the vestibular nuclei to the vertical ocular motoneurons express gamma-aminobutyric acid (GABA), whereas those to horizontal motoneurons are glycinergic.[13,17,18] These findings have led to the development of drugs with a range of pharmacological actions as treatment for acute vestibular imbalance and its associated nystagmus; they are discussed in the next section.

In addition to the pathways serving the "elementary" vestibulo-ocular reflex, other systems continuously monitor and optimize the reflex's performance (see Chapters 1 and 2). The gain (magnitude of the vestibulo-ocular response) depends on a form of "memory encoding" in the cerebellum due to metabotropic glutamate receptors and $GABA_B$ receptors on Purkinje cells in the cerebellum.[19] The phase of the vestibulo-ocular reflex (timing of eye rotations with respect to head rotation) depends on the "velocity storage mechanism," by which the peripheral vestibular signal is prolonged. This perseveration of the raw vestibular signal is achieved by a vestibular commissure that is governed by the cerebellar nodulus and uvula.[20] The metabotropic $GABA_B$ receptor, which is a mediator of slow inhibitory postsynaptic potentials, is important in down-regulating velocity storage. Thus, the $GABA_B$ agonist baclofen suppresses the velocity-storage phenomenon in normal monkeys.[21] These findings provide an explanation for the effectiveness of baclofen in treatment of a central form of vestibular nystagmus (periodic alternating nystagmus)[22] and possibly also in motion sickness.[21]

The vestibular system also contributes to the gaze-holding mechanism by which the eyes can be held steady in an eccentric position (e.g., far right gaze). Thus, the medial vestibular nucleus and adjacent nucleus prepositus hypoglossi, along with reciprocal connections with the vestibular cerebellum, constitute a "neural integrator" for ocular motor signals. This network of neurons receives raw vestibular signals encoding head velocity and integrates them into the eye position signals required to hold the eyes steady in eccentric gaze. The network also integrates pre-motor signals for saccades and smooth pursuit; thus, it is the common neural integrator for all classes of eye movements. For vertical eye movements, the interstitial nucleus of Cajal also contributes to this neural integrator function. Several neurotransmitters contribute to the neural integrator, and nitric oxide appears to perform a neuromodulatory stabilizing function within this neural network.[23] Some vestibular inputs to nucleus prepositus hypoglossi are GABAergic and are facilitated by nitric oxide.[24,25] The projections of nucleus prepositus hypoglossi (the output of the neural integrator) to the abducens nucleus are cholinergic.[13,26] Further insights into the neuropharmacology of the neural integrator have been gained using the technique of pharmacological inactivation.[27,28] Microinjection of the weak GABA antagonist bicuculline and the strong GABA agonist muscimol into the MVN-NPH region may cause either gaze-evoked nystagmus (caused by a "leaky neural integrator") with centripetal slow-phase drifts or, occasionally, centrifugal, increasing velocity waveforms (caused by an "unstable neural integrator"). These paradoxical findings may reflect inactivation of either the integrator itself (causing leakiness) or cerebellar control of the integrator (causing instability).[28] Nystagmus with characteristics implying a vestibular imbalance may also be produced during these inactivation experiments. Agents with glutamatergic effects may also

compromise the neural integrator, but glycinergic agents do not.[28] Inactivation of the interstitial nucleus of Cajal with muscimol causes neural integrator failure, implying that GABA also contributes to vertical gaze holding.[29] Taken together, these findings indicated that GABA and glutamate are not just an important neurotransmitter for the horizontal and vertical vestibulo-ocular reflex but are also involved in the gaze-holding mechanism, malfunction of which may lead to nystagmus.[30]

Clinical evidence has also supported a role for nicotinic *acetylcholinergic mechanisms in vertical eye movements*. Thus, upbeat nystagmus may be induced in darkness in normal subjects by nicotine.[31-33] Functional imaging has indicated activation by nicotine of the nucleus reticularis tegmenti pontis (NRTP),[34] suggesting that nicotinic acetylcholinergic mechanisms contribute to vertical smooth-pursuit, although other mechanisms probably also contribute. These findings suggest that drugs with actions on nicotinic cholinergic receptors might influence nystagmus.

Treatment of Vertigo

The general goals in treatment are to eliminate vertigo and accompanying neurovegetative symptoms (nausea, vomiting, anxiety) and to promote (or, at least, not hinder) the normal process of vestibular compensation. In certain cases, such as viral neurolabyrinthitis and vertigo caused by migraine, it may be possible to treat the underlying cause. Often, however, treatment is symptomatic.

In *acute vertigo* caused by a peripheral vestibular lesion, recovery from symptoms and resumption of normal activities is the rule in the ensuing weeks.[35] However, if viral etiology is suspected and the patient is evaluated within three days of onset, a short course of methylprednisolone may speed recovery from symptoms; antiviral agents such as valacyclovir offer no therapeutic advantage.[36] There is a consensus that drugs exerting a "sedative effect" on the vestibular system should be used for only the first 24 hours.[10] Some drugs commonly used for treatment of vertigo, nausea, and vomiting are summarized in Table 14-1 and are discussed more fully in reviews.[37,38] Ondansetron, a serotonin 5-hydroxytryptamine$_3$ (5-HT$_3$) antagonist, seems especially helpful in the management of nausea and vomiting in acute vestibular disorders.[39] Most agents probably affect more than one neurotransmitter system, and in intractable cases, a combination of different types of agent may be more effective than one alone.

After the first 24 hours, drugs should be used sparingly and patients should be encouraged to get out of bed and increase their activities, because there is evidence that failure to do so limits recovery of symptoms. During this period, a course of specific vestibular exercises may be indicated (see Chapter 22). Patients in whom enduring vestibular symptoms develop, such as vertigo with head movement, may have an underlying central nervous system disorder, typically of the cerebellum,[40] and imaging studies are indicated. Other patients who complain of persistent symptoms may have either a phobic disorder[11] or the potential for secondary gain as a consequence of their injury.

Treatment of *recurrent vertigo* depends on the nature of the underlying disorder. For example, vertigo caused by migraine (including migraine without a headache) can usually be successfully managed medically (see Chapter 15). Recurrent brief episodes of vertigo—vestibular paroxysmia—that may be a result of vascular compression of the eighth cranial nerve, are often effectively treated with carbamazepine.[41] Recurrent vertigo caused by a perilymphatic fistula or superior canal dehiscence syndrome may recover spontaneously, but some patients require surgical repair (see Chapters 4 and 17). Less commonly, recurrent attacks of vertigo are a feature of episodic ataxia type 2 (Case Study 14-1), along with ataxia, vegetative symptoms and, sometimes, headache.

Vertigo caused by Ménière's syndrome is often difficult to manage, although a low-salt diet and diuretics help some patients with the disorder.[38] In Europe, betahistidine, which is an H1-agonist and H3-antagonist that may improve the labyrinthine microcirculation, is given as prophylactic treatment[37]; it is not available in the United States. Because the vestibular imbalance may be in a continuous state of flux, long-term use of "vestibular sedatives" such as meclizine is justified in some affected patients. Treatment with intratympanic injection of gentamicin, which is toxic to labyrinthine hair cells, can be beneficial when vertigo persists, especially when it arises from a deaf ear.[38,42] Patients with Menière's disease tend to improve with time, although the second ear may become involved.[43]

Central neurological conditions, such as multiple sclerosis, vertebrobasilar ischemia, and posterior fossa mass lesions, may cause severe, recurrent vertigo. When treatment of the underlying condition does not produce improvement, "vestibular sedatives" are justified. Ondansetron helps some patients with vertigo caused by brainstem stroke or multiple sclerosis.[44,45] Sometimes a combination of agents such as an anticholinergic (e.g., scopolamine) and an antidopaminergic agent (e.g., prochlorperazine) are required.

Benign paroxysmal positional vertigo is effectively treated in most cases by specific vestibular exercises or maneuvers (see Chapter 20); drugs are seldom indicated in this condition.

■ Table 14-1 SOME COMMONLY USED VESTIBULAR SEDATIVES[10,37,38]

Drug	Class	Dosage	Comments	Precautions
Dimenhydrinate (Dramamine)	Antihistamine Phosphodiesterase inhibitor	Oral: 50 mg every 4–6 hr IM: 50 mg; maximum of 200 mg in 24 hr	Mild sedative Causes dryness Moderate antiemetic	Asthma, glaucoma Prostate enlargement
Diphenhydramine (Benadryl)	Antihistamine	Oral: 25–50 mg every 6 hr IM: 50–100 mg; maximum of 200 mg in 24 hr	Mild sedative Tachycardia Urinary retention	Asthma, glaucoma Cardiac conditions Prostate enlargement
Promethazine (Phenergan)	Antihistamine Anticholinergic Phenothiazine	Oral: 25 mg, every 6 hr Supp: 50 mg, every 12 hr IM: 25 mg; maximum of 75 mg in 24 hr	More sedative Moderate antiemetic	Asthma Glaucoma Prostate enlargement Epilepsy
Meclizine (Antivert, Bonine)	Antihistamine Anticholinergic	Oral: 25 mg or 50 mg every day or twice daily; maximum of 150 mg in 24 hr	Peak effects 8 hr after ingestion Less sedative	Asthma Glaucoma Prostate enlargement
Prochlorperazine (Compazine)	Antihistamine Anticholinergic Phenothiazine	Oral: 5–10 mg every 6 hr Supp: 25 mg every 12 hr IM: 5–10 mg every 6 hr; maximum of 60 mg in 24 hr	Sedative and antiemetic Can cause extrapyramidal reactions	Can cause liver disease when used in combination with CNS depressants, propranolol, phenytoin, anticoagulants, levodopa, diuretics
Scopolamine (Transderm Scop)	Anticholinergic (nonselective muscarinic)	Transdermal patch, every 3 days; peak effect 4–8 hr after application	Less sedative More antiemetic Suitable for motion sickness Can cause confusion, mydriasis, "dependency"	Asthma Glaucoma Prostate enlargement
Ondansetron (Zofran)	Serotonin 5-hydroxytryptamine$_3$ (5-HT$_3$) receptor antagonist	Oral: 4–8 mg every 8 hr IV: 4 mg	Antiemetic; developed for patients receiving cancer chemotherapy May be effective for nausea caused by CNS disease	Headache Constipation
Lorazepam (Ativan)	Benzodiazepine	0.5 mg every 12 hr IM: 1 mg; maximum of 6 mg in 24 hr	GABA modulator May be habit-forming	Glaucoma Additive with sedative drugs, scopolamine

CNS = central nervous system; GABA, gamma-aminobutyric acid; IM, intramuscular; IV, intravenous; supp = suppository.

Oscillopsia

Pathogenesis

Oscillopsia brought on or accentuated by head movement is usually of vestibular origin and reflects an inappropriate VOR gain or phase.[1] Vision becomes blurred because of excessive retinal image motion so that, for example, fine print on grocery items can be read only if the patient stands still in the store aisle. Oscillopsia is usually caused by excessive motion of images of stationary objects on the retina (Box 14-1). Oscillopsia with head movements may also occur as a result of weakness of an extraocular muscle (e.g., abducens nerve palsy) or internuclear ophthalmoplegia (medial rectus weakness). Oscillopsia caused by nystagmus and other ocular oscillations occurs when the head is stationary. Patients with a head tremor do not complain of oscillopsia (*i.e.*, they do not have excessive motion of images on their retina) unless they have also lost their vestibulo-ocular reflex.[46,47]

An abnormal VOR may lead to oscillopsia during head movements in three possible ways: abnormal gain (inappropriate size of eye movement), abnormal phase shift (a timing lag between eye and head rotations), and a directional mismatch between the vectors of the head rotation and eye rotation (such as vertical eye movements in response to horizontal head rotations). Disease of either the vestibular periphery or its central connections may be the cause.

Typically, oscillopsia is worse during locomotion, but it may be noticed during chewing food and, in the severest cases, may occur from transmitted cardiac pulsation.[2] In addition, visual acuity declines during head movements, although the relationship with oscillopsia is variable. The clinician can easily demonstrate this behavior at the bedside by documenting a decline of visual acuity while rotating the head from side to side at 1 to 2 cycles/second compared with visual acuity with the head still. Normal subjects show a decline of 1 to 2 lines on the visual acuity card, but patients with loss of vestibular function often report difficulty in seeing even the largest optotypes.

Oscillopsia may also occur with disorders of the central nervous system that change the gain, phase, or

Box 14-1

ETIOLOGY OF OSCILLOPSIA*

Oscillopsia with Head Movements: Abnormal Vestibulo-ocular Reflex

Peripheral vestibular hypofunction:[48]
- Aminoglycoside toxicity
- Ménière's syndrome
- Meningitis/encephalitis
- In association with cerebellar degenerations[49]
- Autoimmune disorders affecting the labyrinth (e.g., Cogan's syndrome)
- Surgical section of eighth cranial nerve
- Congenital ear anomalies
- Hereditary vestibular areflexia
- Cisplatin therapy
- Idiopathic

Central vestibular dysfunction (often with associated cerebellar disease):
- Decreased vestibulo-ocular reflex (VOR) gain
- Increased VOR gain
- Abnormal VOR phase

Oscillopsia with Head Stationary: Abnormal spontaneous eye movements

Acquired nystagmus (especially pendular, upbeat, downbeat, see-saw, dissociated nystagmus)

Congenital nystagmus (uncommon under natural illumination)

Saccadic oscillations (ocular flutter (including psychogenic) and opsoclonus

Superior oblique myokymia (monocular oscillopsia)

Oscillopsia with Head Stationary and Normal Eye Movements:
- Central oscillopsia
- With cerebral disorders: seizures, occipital lobe infarction

*Adapted from Leigh and Zee, 2006.[1]

direction of the VOR. Some patients with hereditary spinocerebellar degeneration may have associated impaired vestibular responses (Case Study 14-2). Other patients with disease of the vestibulocerebellum show vestibular hyper-responsiveness; this occurs in patients with the Arnold-Chiari malformation, and occasionally patients with cerebellar disorders are reported with increased gain of both the horizontal and vertical VOR.[50] In some patients with vestibulocerebellar dysfunction, the gain of the VOR is normal but the phase relationship between head and eye movements is abnormal, causing retinal image slip.[51] Lesions of the medial longitudinal fasciculus (internuclear ophthalmoplegia in multiple sclerosis) may cause a low gain of the vertical VOR and produce oscillopsia with vertical head movements.[52,53]

Treatment of Oscillopsia

With time, compensation takes place in some patients with oscillopsia caused by vestibular loss,[2] although many others remain more permanently disabled.[54] When compensation occurs, it is a result of a variety of factors, including potentiation of the cervico-ocular reflex, preprogramming of compensatory eye movements, and perceptual changes (see Chapters 8 and 9).[55-58] In general, drugs have little to offer. Patients should be encouraged to resume activities, especially walking, and enroll in exercise programs (see Chapters 22 and 23). Rarely, oscillopsia caused by a hyperactive VOR can be treated pharmacologically.[50] Paradoxically, patients who lack a VOR can read head-fixed visual display during locomotion better than normal subjects can.[59] The reason is that clear vision of a head-fixed display requires that vestibular eye movements be suppressed or canceled. Because such patients have little or no VOR, it is easy to suppress vestibular eye movements. This behavior suggests that head-fixed video displays of images obtained with cameras might be able to compensate for head perturbations in patients who have lost vestibular function; the technology required for such devices has been developed but has not yet been applied to patients with deficient vestibular function.[60,61]

The prospects for treating patients with loss of peripheral vestibular function by implanting a vestibular prosthesis are improving, with some success reported in restoring vestibular responses in a macaque model.[62,63] An alternative approach is to present tactile sensory stimuli that can substitute for the missing vestibular sense.[64,65] Future studies will determine how well these devices can overcome the disability following loss of vestibular function, particularly in elderly patients.

Nystagmus and Its Visual Consequences

Pathogenesis

As indicated previously, acquired nystagmus commonly causes impaired vision and oscillopsia (illusory movement of the environment). These symptoms, which are a result of excessive drift of images of stationary objects on the retina, interfere with reading and watching television, and are often distressing to the patient. The relationship between retinal image velocity and visual acuity is a direct one: For higher spatial frequencies (which correspond to text), image motion in excess of about 5 deg/sec impairs vision.[66] On the other hand, the relationship between retinal image velocity and the development of oscillopsia is less consistent; the intensity of oscillopsia is usually less than the magnitude of nystagmus. For example, in patients with downbeat nystagmus, oscillopsia, on average, was estimated to be about 1/3 the magnitude of the nystagmus.[67] This latter finding suggests that the brain compensates for the excessive retinal image motion and so partly maintains visual constancy, although the mechanism is debated. In practice, it is often not necessary to reduce retinal image slip to less than 5 deg/sec for patients to report appreciable benefit, and this can be achieved at drug doses that do not produce side effects.

Treatments

Drugs

Some of the drugs reported to suppress a range of forms of nystagmus are listed in Box 14-2. Here we review medicines that have been evaluated in controlled clinical trials. Perhaps the most visually disabling ocular oscillations are those of acquired pendular nystagmus, which occurs in two common settings: associated with multiple sclerosis (Case Study 14-3) or as a component of the syndrome of oculopalatal tremor, which follows brainstem stroke (Case Study 14-4). Two medicines have proven useful in the treatment of both forms of acquired pendular nystagmus, though the site of action of both is uncertain. The first is *gabapentin*, which was initially thought to be GABAergic but exerts its effect via a subunit of voltage-dependent calcium channels.[68] Gabapentin was introduced as an anticonvulsant but is now widely used for the control of chronic pain. Gabapentin suppresses nystagmus and affords visual improvement in many patients with acquired pendular nystagmus, perhaps more frequently when the cause is multiple sclerosis.[69,70] However, not all such patients

Box 14-2

TREATMENTS FOR NYSTAGMUS AND ITS VISUAL CONSEQUENCES[37,71,72,73]

Drugs

- Gabapentin
- Memantine
- Baclofen
- 4-aminopyridine and 3,4-diaminopyridine
- Acetazolamide
- Clonazepam
- Valproate
- Trihexyphenidyl and benztropine
- Scopolamine
- Isoniazid
- Carbamazepine
- Barbiturates
- Alcohol

Optical Devices

- Base-out prisms
- Spectacle lens–contact lens combination (for retinal image stabilization)
- Electro-optical devices for retinal image stabilization

Invasive Procedures

- Operative treatment of Arnold-Chiari malformation
- Botulinum toxin

respond, and worsening of ataxia may be a troublesome side effect. The second treatment is *memantine*, an uncompetitive *N*-methyl-D-aspartate (NMDA) antagonist, which is used to treat memory failure in Alzheimer's disease.[74] Memantine is often effective for acquired pendular nystagmus.[70,75,76] It is generally well tolerated, but there is some evidence that it may cause recurrence of symptoms because of prior attacks of multiple sclerosis.[77] In both multiple sclerosis and oculopalatal tremor, a worthwhile strategy is to slowly increase the dose. For gabapentin, the daily dose can be increased by 300 mg per week to a target dose of 300 mg four times daily, or perhaps higher.[70,71] In the case of memantine, the daily dose can be increased by 10 mg per week to a target dose of 10 mg four times daily.[70,76] To date, there have been no controlled studies that have evaluated combination therapy with gabapentin and memantine.

Perhaps the most common form of acquired nystagmus is downbeat nystagmus. Controlled trials have identified two chemically related potassium channel–blocking agents, 4-aminopyridine and 3,4-diaminopyridine to be therapeutically effective in some patients.[78,79] These drugs appear to restore precise "pacemaking" by Purkinje cells, by prolonging the after-polarization of action potential.[80] Of the two, 4-aminopyridine (fampridine or dalfampridine) is more effective and has fewer side effects and is now available in a sustained-release preparation (Ampyra), which is FDA-approved for treatment of gait disorder in patients with multiple sclerosis.[81] Potential side effects are seizures and cardiac dysrhythmia.[82] Thus, the drug should not be given to patients with a history of epilepsy, and a normal electrocardiogram is recommended before starting the medicine. The drug is conventionally given at a dose of 10 mg twice per day. Not all patients with downbeat nystagmus respond to 4-aminopyridine, but many do, especially when their head is semiprone (as when bending forward during reading)[83]; thus, this drug appears to have specific effects on downbeat nystagmus via otolithic mechanisms. Fampridine is also effective treatment of the syndrome of episodic ataxia type 2 (see Case Study 14-1), reducing the frequency of attacks, although *acetazolamide* remains the mainstay of therapy for this condition.[84]

Other drug therapies for nystagmus are less reliable, with the exception of *baclofen*, which is usually effective treatment for the rare disorder, periodic alternating nystagmus;[22] Patients who remain unresponsive to baclofen may be treated with memantine.[85] Other GABAergic agents, such as *clonazepam*, are sometimes effective in the treatment of acquired pendular nystagmus or downbeat nystagmus.[86,87] However, *anticholinergic agents* such as oral benztropine or transdermal scopolamine are not reliable as treatment for acquired nystagmus and may actually make the nystagmus worse in some patients.[88,89]

Optical Devices

A number of optical devices have been suggested as treatment of nystagmus, with the goals of reducing nystagmus and achieving more sustained foveal vision. One simple approach—convergence prisms—often benefits patients whose nystagmus dampens while they are viewing a near target. An arrangement that is often effective is 7.00 diopter base-out prisms with –1.00 diopter spheres added to compensate for accommodation.[90] The spherical correction may not be needed in presbyopic individuals. Especially in some patients with congenital nystagmus,

the improvement of visual acuity through nystagmus suppression as a result of wearing base-out prisms may be sufficient to qualify them for a driving license. Some patients with acquired nystagmus also benefit.[91] Occasionally, in patients in whose nystagmus is worse during near viewing, base-in prisms help.[92] It should be theoretically possible to use prisms to deviate both eyes so that they lie at a position in which nystagmus is suppressed (the "null region"), which is common in patients with congenital forms of nystagmus. However, in practice, patients use head turns to bring their eyes to the quietest position, and only rarely are prisms that produce a conjugate shift helpful.

Another approach has been the development of optical systems to hold images of the environment steady on the retina during eye movements. One system consists of a high-plus spectacle lens worn in combination with a high-minus contact lens.[93] Stabilization can be achieved if the power of the spectacle lens focuses the primary image close to the center of rotation of the eye. A contact lens is then required to extend the focus back onto the retina. Because the contact lens moves with the eye, it does not negate the effect of retinal image stabilization produced by the spectacle lens. With such a system, it is possible to achieve up to about 90% stabilization of images on the retina. There are several limitations to the system, however. One is that it disables all eye movements (including the VOR and vergence), so that it is useful only while the patient is stationary and views monocularly (to avoid double vision). Another is that some patients with ataxia or tremor (such as those with multiple sclerosis) have difficulty inserting the contact lens. In practice, using lens-spectacle combinations that impose lesser degrees of stabilization helps a few selected patients.[94]

A second approach is the use of an electro-optical device that measures ocular oscillations and moves prism devices to negate the effects of the nystagmus.[95] This approach is best suited for pendular nystagmus, which can be electronically distinguished from normal eye movements, such as voluntary saccades that are required for clear vision. The development of lightweight video eye monitors is making this approach more possible. Figure 14-2 summarizes the image-shifting optics that are used in one such device.[96,97]

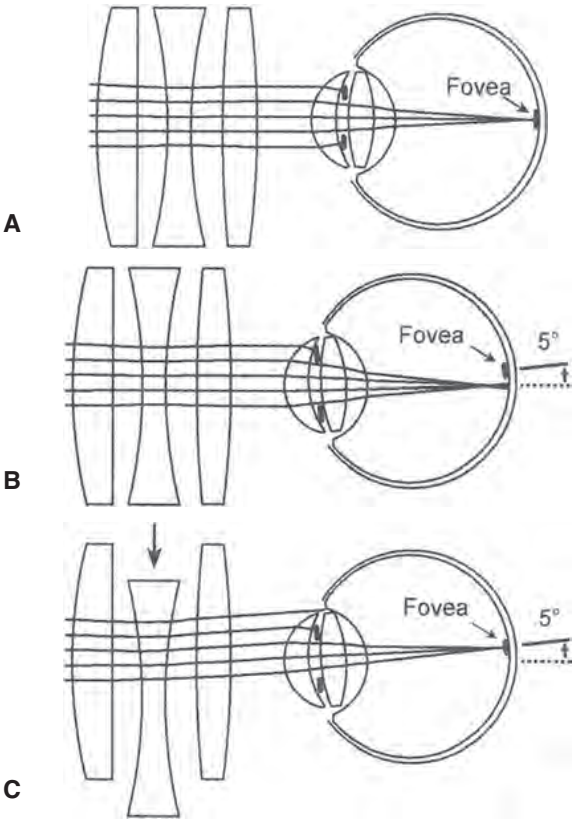

Figure 14.2 Demonstration of how a three-lens, image-shifting device can null the visual effects of ocular oscillations. Starting with the eyes and optics in a neutral position **(A)**, light from a distant target is brought to the fovea of the retina. If the eye is rotated down **(B),** the image is displaced from the fovea. However, if the central lens is moved downward by the appropriate amount **(C)**, the image is once again brought onto the fovea. Reproduced with permission.[96]

Surgery

A range of operative procedures have been developed for treatment of congenital forms of nystagmus,[71] but only occasionally have they been applied successfully as treatments for acquired nystagmus.[98-100] There is still a need for controlled trials to evaluate these surgical procedures, and they are reviewed in more detail elsewhere.[71,72,73] Injection of botulinum toxin either into selected extraocular muscles or into the retrobulbar space can temporarily abolish or suppress nystagmus, but side effects of diplopia and ptosis may limit the therapeutic value, except in carefully selected patients.[101-105] Finally, neurosurgery (suboccipital decompression) may help resolve downbeat nystagmus and other symptoms caused by the Arnold-Chiari syndrome.[105]

CASE STUDY 14-1

A 32-year-old man presented with the complaint of recurrent episodes of vertigo and unsteadiness. The attacks had started in childhood. Often they would be precipitated by stress or exercise. The attacks of vertigo were severe, being accompanied by nausea and sweating. Often the patient would "sleep off" attacks, with resolution in an hour or two. His hearing was normal. He remained able to perform martial arts between attacks. His mother and sister were similarly affected. His examination (between attacks) showed gaze-evoked nystagmus with a down-beating component (Fig. 14.3*A*) and mild gait ataxia. An audiogram was normal. An MRI scan of his head showed mild cerebellar atrophy (Fig. 14.3 *B*) but no abnormal white-matter signals. Based on genetic testing (courtesy of Dr. Robert W. Baloh) a diagnosis of episodic ataxia type 2 (mutation of *CACNA1A* gene) was made. He was started on acetazolamide, and his attacks subsided; he successfully completed a marathon race. Four years later, he developed renal calculi—a known complication of acetazolamide. Accordingly, he was switched to fampridine, which has suppressed his attacks.

Comment

The patient was initially referred as "a case of either Ménière's disease or multiple sclerosis." His normal hearing, despite many attacks, militated against a diagnosis of Ménière's disease. His normal neurological examination, aside from his nystagmus, and MRI scan made multiple sclerosis unlikely. The nature of his attacks, and family history of similar disorder, suggested a genetic disorder—episodic ataxia.[106] The duration of his attacks and lack of myokymia made episodic ataxia type 2 most likely, and this was diagnosed genetically. Acetazolamide suppresses attacks in most patients, but fampridine is an effective alternative if side-effects to acetazolamide develop.[84]

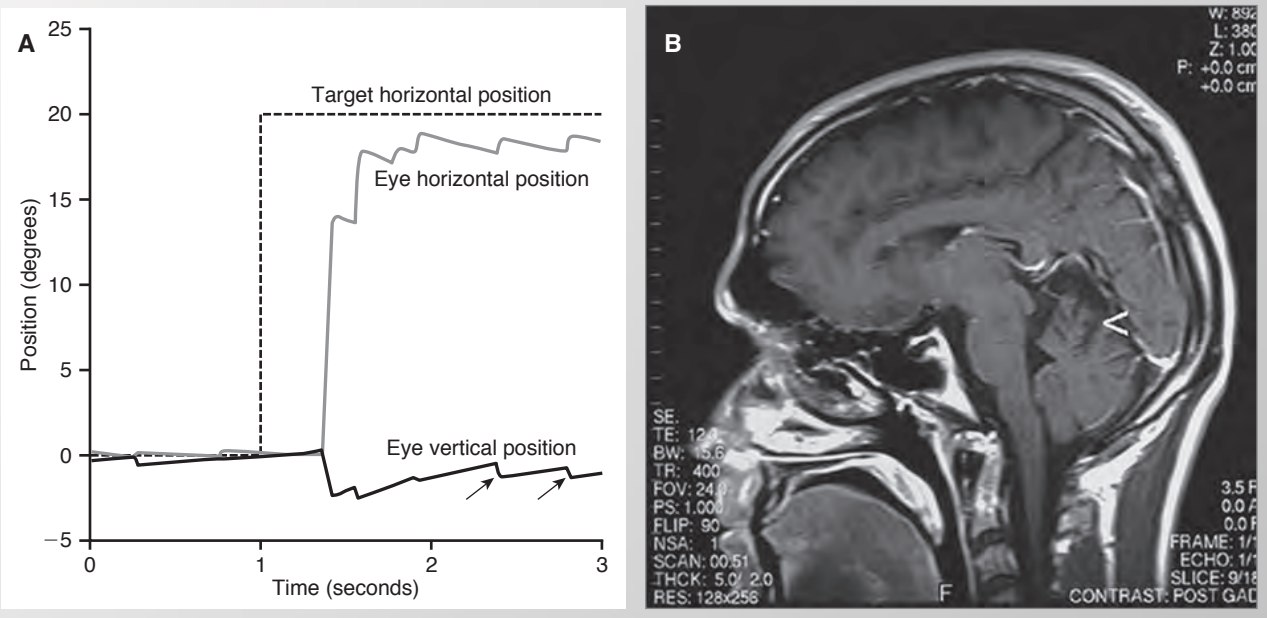

Figure 14.3 Disturbances of eye movements with cerebellar disease. Positive values indicate rightward or upward movements. **(A)** Case History 14-1: A patient with episodic ataxia type 2 (EA-2) developed gaze-evoked nystagmus with a down-beating component (*arrows*) as he shifted gaze to a target jump to right 20 degrees. **(B)** Midsagittal MRI showing some atrophy (*arrowhead*) in this patient with EA-2.

CASE STUDY 14-2

A 68-year-old woman presented with the complaint of progressive unsteadiness of gait and blurred and double vision. She had been a healthy individual until about 6 years previously, but then her gait started to deteriorate. She had been losing her balance, although she had not fallen. Her visual complaints consisted of vertical blurred double vision on lateral gaze and further blurring of vision when she was in motion. She had stopped driving because of poor vision and, when questioned, agreed that she had more difficulty reading street signs when she was in motion. Her examination was notable for gaze-evoked nystagmus that developed a down-beating component on lateral gaze. Using a Maddox rod, it was possible to identify an alternating skew deviation, such that the abducting eye was higher that the adducting eye. In response to rapid, horizontal head turns (head impulse test), she made diagonal corrective saccades that were directed opposite to the direction of head rotation and downward (indicated by arrows in Fig. 14.4). These saccades indicated that her horizontal VOR was reduced, and she also made an inappropriate upward eye movement. She had normal hearing and other cranial nerves, with no dysarthria or limb ataxia. However, she was unable to walk in tandem. Sensory examination was normal. Fampridine was unavailable at the time when she was evaluated.

Comment

This is a fairly common presentation of late-onset gait ataxia. Such patients often show downbeat nystagmus that causes oscillopsia, along with an alternating skew deviation. Not only may they show an inappropriate vertical component to their vestibulo-ocular responses, but these responses are also reduced, accounting for additional oscillopsia during self-motion. In some cerebellar degenerations, vestibular responses are notably diminished.[49] Downbeat nystagmus in some patients may be suppressed by fampridine,[79,83] but gait ataxia may not benefit.[107] Unfortunately, despite therapy aimed at preserving gait, progression occurs, and the cerebellar disease limits their ability to adapt. Nonetheless, such patients usually do better with gait therapy, or regular walking with a companion, than if they resign themselves to a life of inactivity.

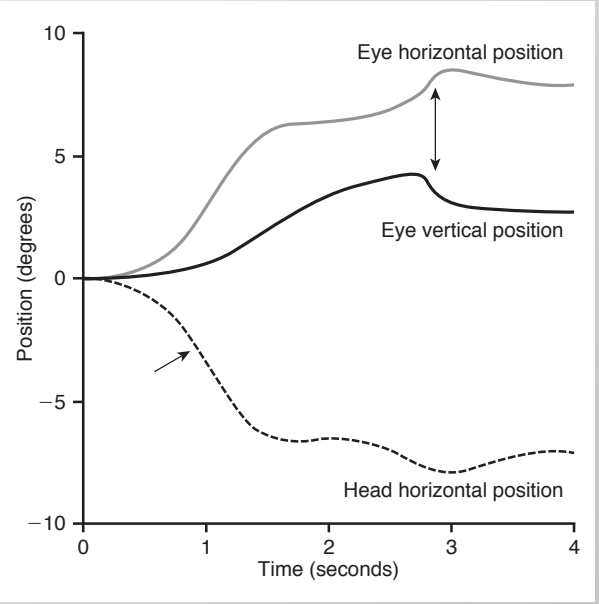

Figure 14.4 Case History 14-2: Head-impulse test (*initiation indicated by arrow*) in a patient with late-onset cerebellar degeneration, who complained of oscillopsia that was worse during locomotion. As her head is turned to the left, a slower, smaller rightward eye movement is generated, along with an inappropriate upward movement. This is followed by a corrective saccade that is directed downward and to the right (*double-headed arrow*).

CASE STUDY 14-3

A 50-year-old woman who had suffered from multiple sclerosis since the age of 25 presented complaining of constant motion of her visual world. She had suffered several episodes of optic neuritis affecting each eye leaving her visual acuity as 20/100 OD and 20/50 OS, with pale optic discs and impaired color vision. She had been troubled by oscillopsia caused by pendular nystagmus for 7 years. Besides her visual complaints, she could only walk with support. On examination, her pendular nystagmus had an elliptical trajectory, with a predominant vertical component (Fig. 3*A*); the frequency was about 5.5 Hz, and there was a superimposed upbeat component. She also had bilateral internuclear ophthalmoparesis. She gained some benefit from gabapentin, but her best response was to memantine (Fig. 14.5*A* and *B*), improving her visual acuity to 20/50 OD and 20/32 OS.

CASE STUDY 14-3

Comment

This patient illustrates several common characteristics of acquired pendular nystagmus in association with multiple sclerosis. Such patients often have some impairment of vision as a result of coexistent disease of the visual system (consequent to prior attacks of optic neuritis) that limits the improvement of vision that can be achieved by abolishing their nystagmus.[108] They may also show internuclear ophthalmoparesis, pointing to brainstem lesions that may cause their pendular nystagmus. Either gabapentin or memantine may suppress their nystagmus, and it is worth trying both agents so that each patient's optimal responses can be determined.[70]

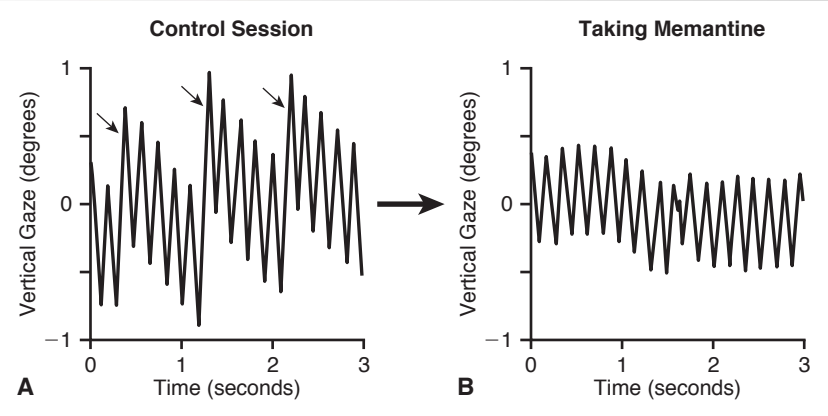

Figure 14.5 Effects of drug treatments on acquired pendular nystagmus; vertical components are shown. **(A)** Case History 14-3: Patient with multiple sclerosis shows a small, high-frequency pendular oscillation with a superimposed upbeat jerk nystagmus (*arrow*). **(B)** Measurements were repeated when she was taking memantine 40 mg/day. The amplitude of the pendular nystagmus is reduced and the upbeat component is absent.

CASE STUDY 14-4

A 37-year-old man suffered a pontine hemorrhage that caused a horizontal gaze palsy, right facial palsy, left hemiparesis, left hemiataxia, and gait ataxia. He made a partial recovery and was able to take care of himself following discharge. About 16 months after his stroke, he noticed vertical oscillopsia, which progressed. His visual acuity was 20/70 OD and OS. His pendular nystagmus was predominantly vertical at about 2.5 Hz (Fig. 14.6 *A* and *B*); oscillations of his soft palate at a similar frequency were observed. His ocular oscillations were suppressed with gabapentin (visual acuity improved to 20/30 OD and OS), although his asymptomatic palatal movements persisted.

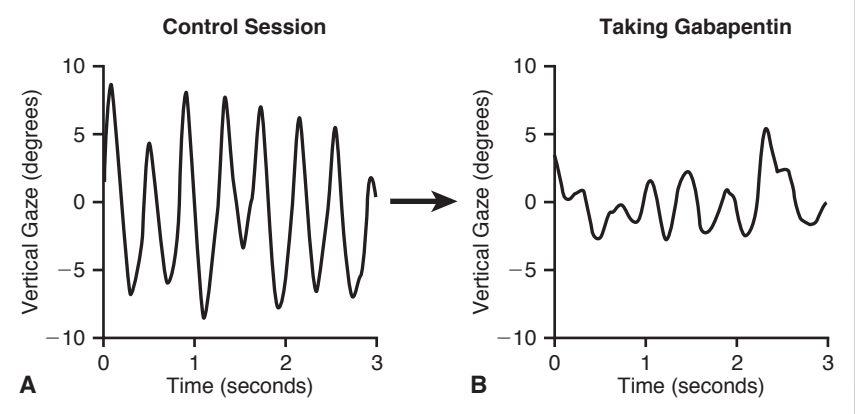

Figure 14.6 **(A)** Case History14-4: Patient with oculopalatal tremor shows large amplitude, somewhat irregular oscillations at about 2 Hz. **(B)** Measurements were repeated when he was taking gabapentin 1,200 mg/day. The amplitude of the nystagmus is reduced.

Continued

CASE STUDY 14-4

Comment

The syndrome of oculopalatal tremor develops following brainstem or cerebellar stroke and is caused by development of abnormal connectivity between neurons in the inferior olivary nucleus.[109] Thus, the syndrome appears several months after the stroke. In health, the inferior olivary nucleus sends "error signals" to the cerebellum, making motor learning possible. In the syndrome of oculopalatal tremor, the inferior olivary nucleus becomes unable to relay signals necessary for motor learning but, instead, acts as a "mindless pacemaker," producing maladaptive motor learning in the cerebellum (oculopalatal tremor) and preventing the ability to learn new motor skills (and limiting adaptive recovery). Nonetheless, affected patients may benefit from gabapentin, memantine, or clonazepam, which sometimes suppress nystagmus and improve vision.[70,86]

Summary

Disruption of the peripheral or central vestibular system often results in vertigo, oscillopsia, and nystagmus. Acute vertigo from peripheral vestibular lesions usually recovers spontaneously and vestibular suppressant medications, although appropriate during the first 24 hours, should be used sparingly after that initial period. The use of medications in recurrent vertigo depends on the specific disorder affecting the vestibular system. Oscillopsia caused by loss of the vestibular sense often improves spontaneously, and medications do not aid the recovery; vestibular prostheses may eventually provide a new therapeutic strategy. Oscillopsia caused by acquired pendular nystagmus may be suppressed by gabapentin or memantine, and downbeat nystagmus by fampridine. Several different optical devices have been developed as treatment for the visual consequences of nystagmus, and electro-optical devices hold promise in the future. No medications or optical devices can be applied uniformly to all patients; careful diagnosis and patient selection is essential for treatments to be effective.

Acknowledgments

The author's work described in this chapter is supported by NIH grant EY06717, the Department of Veterans Affairs, and the Evenor Armington Fund.

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CHAPTER 15

Medical Management of Migraine, Ménière's Disease, and Motion Sensitivity

Ronald J. Tusa, MD, PhD

Migraine is a common cause of episodic vertigo and disequilibrium in children and adults. In practices treating patients with headaches, 27% to 33% of patients out of a population of 700 patients with migraine report episodic vertigo.[1,2] Thirty-six percent of these patients experience vertigo during their headache-free period; the others experience vertigo either just before or during a headache. The occurrence of vertigo during the headache period is much higher in patients with migraine headaches with aura (classic migraine) as opposed to migraine without aura (common migraine).[3] In this chapter, we describe the incidence of migraine; current classification and criteria used for diagnosing migraine, neuro-otological syndromes, and genetics related to migraine; and the management of migraine.

Incidence of Migraine

Migraine is an extremely prevalent disorder. An epidemiological study involving over 20,000 individuals between 12 and 80 years of age found that 17.6% of all adult females, 5.7% of all adult males, and 4% of all children had one or more migraine headaches per year.[4] This study used the diagnostic criteria recommended by the International Headache Society[5] (IHS, which is now referred to as ICHD[6]), which will be described later. Of those individuals with migraine, approximately 18% experienced one or more attacks per month. In both males and females, the prevalence of migraine was highest between the ages of 35 and 45 years. The type and severity of migraine often varies within the same individual. Migraine with or without aura frequently begins between 12 and 30 years of age. After the age of 50, migraine is much less common, and it frequently presents as migraine aura without headache.[7]

Migraine and International Classification of Headache Disorders (ICHD)

Migraine disorders are usually subdivided into several types. The classification of headache disorders was initially published by an Ad Hoc Committee of NIH.[8] This included short descriptions of the headache disorder types

without a description of the criteria. In 1988, an international committee developed a more comprehensive classification with criteria for each headache disorder termed the International Classification of Headache Disorders (ICHD).[5] In most cases, the criteria for headache disorder was based on expert opinion. In 2004, the second version, the ICHD-II, was published, which was based on clinical research trials published since the 1988 study.[6] The ICHD-III version should be published in 2013 and will be accessible on the IHS website (http://www.ihs-headache.org/). The classification and criteria for headaches pertinent to neuro-otological disorders is summarized in Box 15-1.

Migraine without Aura

Migraine without aura (ICHD 1.1), which replaces "common migraine," consists of periodic headaches that are usually throbbing and unilateral, e xacerbated by

Box 15-1

INTERNATIONAL HEADACHE SOCIETY CLASSIFICATION OF HEADACHE*

1.1 Migraine without aura (replaces common migraine, hemicrania simplex):

A. At least 5 attacks fulfilling criteria B–D.
B. Headache attacks lasting 4–72 hours (untreated or unsuccessfully treated)
C. Headache has at least two of the following characteristics:
   1. Unilateral location.
   2. Pulsating quality.
   3. Moderate or severe intensity that inhibits or prohibits daily activities.
   4. Aggravation by walking stairs or similar routine physical activity.
D. During headache, at least one of the following:
   1. Nausea and/or vomiting.
   2. Photophobia and phonophobia.
E. Not attributed to another disorder

1.2 Migraine with aura (replaces classic migraine, ophthalmic, hemi-paraesthetic, hemiplegic or aphasic migraine, migraine accompagnee, complicated migraine):

A. At least 2 attacks fulfilling B:
B. Migraine aura fulfilling criteria B and C for one of the subforms 1.2.1–1.2.6.
C. Not attributed to another disorder

1.2.6 Basilar-type migraine (replaces basilar artery migraine, basilar migraine):

A. At least 2 attacks fulfilling criteria B–D:
B. Aura consisting of at least two of the following fully reversible symptoms, but no motor weakness:
   1. dysarthria
   2. vertigo
   3. tinnitus
   4. hyperacusis
   5. diplopia
   6. visual symptoms simultaneously in both temporal and nasal fields of both eyes.
   7. Ataxia
   8. Decreased level of consciousness
   9. Simultaneously bilateral paresthesias
C. At least one of the following:
   1. At least one aura symptom develops gradually over 5 min or longer, and/or different aura symptoms occur in succession over 5 min or longer.
   2. Each aura symptom lasts 5 to 60 min.
D. Headache fulfilling criteria B–D for 1.1 Migraine without aura begins during the aura or follows aura within 60 min.
E. Not attributed to another disorder

1.3.3 Benign paroxysmal vertigo of childhood:

A. At least 5 attacks fulfilling criterion B.
B. Multiple episodes of severe vertigo, occurring without warning and resolving spontaneously after minutes to hours.
C. Normal neurological examination and audiometric and vestibular functions between attacks.
D. Normal electroencephalogram.

Adapted from the International Headache Society Classification Subcommittee, 2004.[6]
*A beta version of the "The International Classification of Headache Disorders, 3rd edition" is now published. *Cephalalgia*. 2013 Jul;33(9):629-808.

activity, and associated with nausea, photophobia, and phonophobia. These headaches are frequently referred to as "sick" headaches (because of the nausea) or "sinus" headaches (because of their location). Patients usually prefer to lie down in a quiet, dark room during the headache and feel better after sleep. A family history for migraine can usually be obtained in the immediate family.

Migraine with Aura

Migraine with aura (ICHD 1.2), which replaces "classic migraine," is associated with transient neurological symptoms consisting of sensory, motor, or cognitive disorders. These neurological disorders usually precede the headache, but may develop during or following the headache. The neurological disorder usually lasts 5 to 20 minutes, but can last as long as 1 hour. There are three relevant subtypes. The first is migraine with prolonged aura, in which neurological symptoms can last up to 7 days. The second is called basilar migraine, which replaces "basilar artery migraine," and presents with symptoms in the distribution of the basilar artery including vertigo, tinnitus, decreased hearing, and ataxia. The third is called migraine aura without headache, which replaces "migraine-equivalent spells" or "acephalgic migraine." This presents with the neurological disorders found in migraine with aura, except there is no headache.

Basilar-Type Migraine

Basilar-type migraine (ICHD classification 1.2.6) was first described by Bickerstaff,[9] and has been subsequently reported by a number of individuals.[10,11] This disorder consists of two or more neurological problems (vertigo, tinnitus, decreased hearing, ataxia, dysarthria, visual symptoms in both hemifields of both eyes, diplopia, bilateral paresthesia or paresis, decreased level of consciousness) followed by a throbbing headache. The majority of these occurs before 20 years of age, but can occur up until age 60. Vertigo typically lasts between 5 minutes and 1 hour. In the majority of cases, audiograms are normal. Many of these patients eventually develop more typical migraine headaches with aura, and there is frequently a positive family history for migraine. Transient ischemic attacks (TIAs) need to be considered before basilar migraine is diagnosed. TIAs within the vertebral-basilar circulatory system (vertebrobasilar insufficiency) may cause the same symptoms as basilar migraine, although TIAs usually last less than a few minutes.[12]

Childhood Periodic Syndromes

There are two neuro-otological disorders in children as a result of migraine. First, is benign paroxysmal vertigo of childhood (ICHD classification 1.3.3), which was first described by Basser.[13] This disorder consists of spells of vertigo and disequilibrium without hearing loss or tinnitus.[14-18] The majority occurs between 1 and 4 years of age, but can occur anytime during the first decade. Vertigo and disequilibrium typically last for minutes, but can last up to several hours. Patients may experience visual disturbance, flushing, nausea, and vomiting. In the majority of cases, audiograms and caloric tests are normal. These patients also have a normal electroencephalogram (EEG), and a normal physical examination in between spells. Initially, headache is usually not a major feature of these spells. Many of these patients eventually develop migraine with aura and there is frequently a positive family history for migraine. The differential diagnosis includes vestibular epilepsy, perilymphatic fistula, posterior fossa tumors, and psychogenic disorders. The second childhood periodic syndrome, is benign paroxysmal torticollis (ICHD A1.3.5. This disorder was first described by Snyder,[19] and has now been reported by a number of individuals.[20-22] Benign paroxysmal torticollis consists of spells of head tilt and rotation without vertigo, hearing loss, or tinnitus. These usually occur in the first 5 years of life and typically last between 10 minutes and several days. They may be associated with nausea, vomiting, pallor, agitation, and ataxia. In the majority of cases, audiograms and caloric tests are normal between the spells. This syndrome is believed to be caused by migraine auras without headache. Some individuals complain of headache when they become older. The differential diagnosis includes posterior fossa tumors and torticollis.

Vestibular Migraine

This disorder was not formally classified by the ICHD. Based on the symptoms, it may overlap basilar-type migraine.[1,2,6] This was first described by Slater,[23] who labeled it benign recurrent vertigo in adults. This disorder consists of spells of vertigo, and in some individuals, jerk nystagmus may occur during the spell.[24,25] (see Case Studies 15-1 and 15-5 at end of the chapter). Vertigo typically lasts from minutes to 72 hours with or without headache. There is no vestibular defect or hearing loss noted on electronystagmography (ENG) and audiogram from the spell, which rules out vestibular neuritis and Ménière's disease. The spells usually occur between the ages of 20 and

60. Another type of vestibular migraine occurs in some individuals who develop exercise-induced spells from a variety of physical activity including sit-ups, heavy lifting, intercourse, and other strenuous aerobic exercises[26] (see Case Study 15-2). One needs to rule out Ménière's disease, benign paroxysmal positional vertigo (BPPV), TIAs, vestibular epilepsy, and perilymphatic fistula before making a diagnosis of migraine-induced vertigo. The diagnostic criteria for vestibular migraine has recently been defined by a joint committee of the Baran y Society and subcommittee of the International Headache Society[27] (Box 15-2). The criteria will appear as an appendix of the third edition of the ICHD (still in discussion; publication date unknown).

Box 15-2

DIAGNOSTIC CRITERIA FOR VESTIBULAR MIGRAINE

1.1 Vestibular Migraine

A. At least 5 episodes with vestibular symptoms (vertigo that occurs spontaneously, or with change in head position, or visually induced, or head motion induced) of moderate or severe intensity, lasting 5 min to 72 hr.
B. Current or previous history of migraine with or without aura according to the ICHD.
C. One or more migraine features with at least 50% of the vestibular episodes:
   a. Headache with at least 2 of the following characteristics: one-sided location, pulsating quality, moderate or severe pain intensity, aggravation by routine physical activity,
   b. photophobia and phonophobia,
   c. visual aura.
D. Not better accounted for by another vestibular or ICHD diagnosis.

1.2 Probable Vestibular Migraine

A. At least 5 episodes with vestibular symptoms of moderate or severe intensity, lasting 5 min to 72 hr.
B. Only one of the criteria B and C for vestibular migraine is fulfilled (migraine history or migraine features during the episode).
C. Not better accounted for by another vestibular or ICHD diagnosis.

Adapted from consensus document of the Barany Society and the IHS.[11]

Disorders Associated with Migraine

Increased motion sickness and anxiety disorders are frequent in individuals with migraine. Both can cause different types of dizziness that need to be recognized by the clinician.

Motion Sickness

Motion sickness consists of episodic dizziness, tiredness, pallor, diaphoresis, salivation, nausea, and occasional vomiting induced by passive locomotion (e.g., riding in a car) or motion of the visual surround while standing still (e.g., viewing a rotating optokinetic stimulus or large screen motion picture). Motion sickness is partially caused by a visual-vestibular conflict or mismatch.[28] Twenty-six to sixty percent of patients with migraine have a history of severe motion sickness compared with 8% to 24% of the normal population.[1,3,29] The cause for this relation is not clear.

Anxiety Disorders (DSM IV)

Anxiety disorders are diagnosed using the DSM IV criteria. More detail of these disorders and management can be found in Chapter 18.

Generalized Anxiety Disorder

This is characterized by generalized and persistent unrealistic worry with motor tension, autonomic hyperactivity, apprehensive expectation, and vigilance for over 6 months.

Panic Attacks

These are discrete spells of intense fear or discomfort and greater than 4 symptoms that develop abruptly and peak in 10 minutes.

- Dizziness, unsteady feelings or faintness
- Nausea or abdominal distress
- Shortness of breath (or smothering sensations)
- Palpitations or tachycardia
- Trembling or shaking
- Sweating
- Choking
- Depersonalization or derealization
- Numbness or paresthesias
- Flushes (hot flashes) or chills
- Chest pain or discomfort
- Fear of dying
- Fear of going crazy or doing something uncontrolled

Summary of Clinical Presentation of Migraine-related Dizziness

Case: Patient is a 50-year-old with dizziness. Several months ago while standing and looking out the window, the patient developed severe vertigo, N/V, and fell backwards. This was followed by a mild headache. The vertigo lasted 10 minutes, the headache lasted for 1 day, and the patient felt washed out for days. Since then, he has had five more spells of vertigo, photophobia, and phonophobia followed by headache. Feels better after a short nap when spell occurs. He also notes increased motion sensitivity while wife drives car, while he plays games on his computer, or while he is driving a motorboat. In addition, he now has daily dizziness consisting of light-headedness and the sense that his head is bobbing. There is a sense of helplessness and increased stress now because of overwhelming amount of work, and he is unable to return to work as a mechanic. He had a normal neurological and otological exam. He already had an audiogram, ENG, and MRI of head, all of which were normal. He was told to come in next time he had a spell. When he came in during a spell, he had a sustained horizontal jerk nystagmus, but the VOR gain was normal based on head thrust.

Individuals with vestibular migraine often have several overlapping causes for their symptoms (Fig. 15.1). This case example illustrates the overlapping symptoms often found in patients with vestibular migraine. He started with vertigo with a migraine headache consistent with vestibular migraine. These attacks of vertigo are migraine aura and can occur with or without headache. In addition, he had increased motion sensitivity. This became a chronic problem whenever there was a visual-vestibular mismatch (watching computer games and as a passenger in a moving vehicle). Finally, he developed anxiety and depression. The three causes of dizziness in Figure 15.1 can occur separately, but frequently they occur together in individuals with vestibular migraine.

Symptoms during Vestibular Migraine Aura

In an unpublished study, we gave a questionnaire to 38 consecutive patients with vestibular migraine to determine the type of symptoms and incidence (Table 15-1). The symptoms were divided into different types of dizziness,

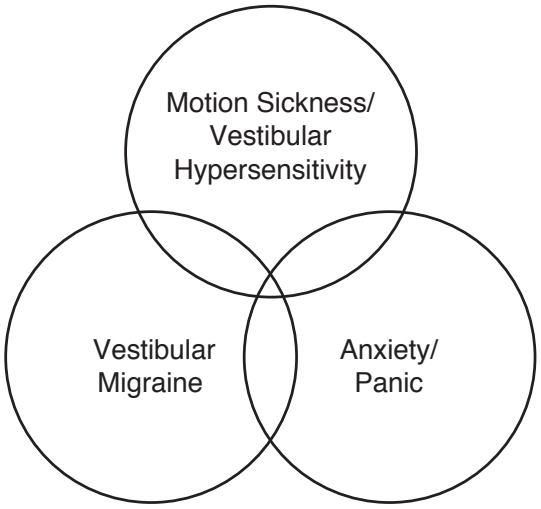

Figure 15.1 Causes of symptoms in patients with migraine.

non-dizzy symptoms, and chronic symptoms consisting of increased motion sensitivity and anxiety disorders. The majority of patients had dizziness during their aura that primarily occurred when they moved through space (dynamic). Some patients had spontaneous vertigo (static) that occurred while seated or lying down (positional vertigo).

Pathophysiology of Migraine

There are four phases in migraine, which includes the prodrome phase, aura, headache, and postdrome phase. Each of these phases includes symptoms that are induced by different neurotransmitters.

Dopamine D2 Receptor

There is a clinical overlap between dopamine stimulation and migraine.[30,31] As dopamine increases, yawning begins to occur. With further dopamine increase, more symptoms occur that include mood changes, nausea, gastrokinetic changes, hypotension, and vomiting. These are similar symptoms as the prodrome phase in patients with migraine. Patients with migraine have D2 receptor hypersensitivity based on apomorphine response. During migraine, D2 is activated. The NcoI C allele in the *DRD2* gene is higher than the NcoI T allele in patients with migraine, generalized anxiety, and panic disorders.[32,33] Perhaps this common allele in individuals with migraine and anxiety is the reason why these two entities frequently coexist in the same individual. Dopamine stimulation may also be the cause of GI symptoms in the headache phase and the postdrome phase, and the "hung over or washed out feeling" in the postdrome phase.

■ Table 15-1 **SYMPTOMS DURING VESTIBULAR MIGRAINE IN 38 PATIENTS**

Symptom	No. of Cases (%)
Dizzy Symptoms	
Vertigo:	21 (55)
Static	5 (13)
Dynamic	12 (32)
Non-vertigo:	35 (92)
Imbalance	30 (79)
Lightheadedness	26 (68)
Gait disorder	23 (61)
Non-Dizzy Symptoms	
Tinnitus	10
Bilateral paresthesia	6
Blurred vision	8
Bilateral weakness	5
Scintillating scotoma	4
Diplopia	2
Decreased consciousness	2
Chronic Symptoms	
Motion sickness/vestibular hypersensitivity	78%
Generalized anxiety disorder (GAD)	57%
GAD + panic attacks	47%

Calcium Channel Receptor (CACNA1A)

The discovery of the genetic cause of a migraine aura phase (hemiplegic migraine) has been one of the most promising breakthroughs in understanding and potentially treating this disorder. This aura causes transient hemiparalysis. Familial hemiplegic migraine is an autosomal dominant disorder. In 50% of all families, this disorder is mapped to chromosome 19p13 in the gene called the *CACNA1A*. This gene codes for a subunit of the P/Q voltage-gated neuronal calcium channel.[34] This same chromosome locus may be involved also in other forms of migraine aura.[35] Defects involving this gene are involved with other autosomal dominant disorders that have neuro-otological symptoms (Table 15-2). The symptoms of these disorders overlap extensively.[36] Only 50% of families with familial hemiplegic migraine map to chromosome 19p13. Other families with this disorder map to chromosome 1 (1q21-q21).51. Other chromosome defects are likely to be found in the future.

Noradrenergic System

Altered sensory processing mediated by serotoninergic midbrain raphe nucleus and noradrenergic locus ceruleus may be the cause of increased sensitivity to sensory stimuli in individuals with migraine to levels that would not trouble a normal individual.[37,38] This may be the cause for increased motion sensitivity in individuals with migraine. Twenty-six to sixty percent of patients with migraine have a history of severe motion sickness compared with 8% to 24% in the normal population.[1,3] The noradrenergic system may also be the cause of the hypersensitivity to sensory stimuli during the aura phase, headache phase, and postdrome phase of migraine.

Serotonin 5HT$_1$ Receptor and the Headache Phase

According to the neurogenic hypothesis, the headache phase may be mediated by neurons containing serotonin (5-HT) within the trigeminal nucleus.[39] 5-HT is an intracranial vasoconstrictor; it rises during the migraine aura and falls during the headache. Platelet 5-HT levels drop rapidly during the onset of migraine. Through autoregulation, blood flow is reduced to this area of neuronal dysfunction. Agonists of these receptors block neuropeptide release and alter neurotransmission in trigeminovascular neurons.[40] Serotonin 5-HT$_1$ receptor agonists such as sumatriptan, zolmitriptan, frovatriptan, almotriptan, eletriptan, rizatriptan, and naratriptan are effective drugs in aborting migraine headache.

Management

Management begins with identifying to what extent the patient is suffering from spells as a result of vestibular migraine, or more chronic symptoms caused by chronic anxiety and increased motion sensitivity.

■ Table 15-2 *CACNA1A* GENE DEFECTS THAT CAUSE AUTOSOMAL DOMINANT DISORDERS

Gene Defect	Syndrome	Symptoms and Signs
Point mutation	Familial hemiplegic migraine	Episodic hemiparesis for up to 60 minutes followed by headache GEN and DBN may persist after spells
Point mutation	Episodic ataxia-2	Episodic ataxia and vertigo GEN DBN Decreases in VOR cancel and pursuit Normal VOR
CAG repeats	SCA 6	Progressive ataxia GEN DBN Decreases in VOR cancel and pursuit Normal VOR

DBN = downbeat nystagmus; GEN = gaze-evoked nystagmus; VOR = vestibulo-ocular reflex.

Treatment of Vestibular Migraine

Spells of vertigo and disequilibrium secondary to migraine usually respond to the same type of treatment as that used for migraine headaches. Migraine is triggered by a number of factors including stress, anxiety, hypoglycemia, fluctuating estrogen, certain foods, and smoking.[41-43] Treatment of migraine can be divided into (1) the reduction of risk factors, (2) prophylactic medical therapy, and (3) abortive medical therapy.

Reduction of Risk Factors

Sometimes, in practice, patients with migraine are given a management schedule to follow, which is explained at the time of their first visit (Box 15-3). It may be pointed out that there are several triggers for migraine, which can be avoided by following the schedule.

Stress

All patients are started on an aerobic exercise program to help reduce stress. This program is gradually increased until the individual is exercising 3 to 5 times per week for at least 30 minutes at the end of the day (jogging, swimming, fast walk, racquetball, tennis, etc.). Several good aerobic exercise programs can be found in a paperback book by Cooper.[44] If patients are reluctant or unable to participate in an exercise program, other stress reduction programs can be very helpful. These include biofeedback and relaxation programs, which have been shown to significantly reduce the frequency of recurrent migraine disorders in clinical trials.[45] Patients are urged to avoid hypoglycemia by eating something at least every 8 hours. Many individuals skip breakfast; the need to eat breakfast at the same time each morning, including weekends, is emphasized. Finally, maintenance of a regular sleep schedule (going to bed and getting up at the same time each day) is strongly recommended.

Nicotine

Patients who smoke cigarettes are urged to stop smoking.

Estrogen

If patients are taking estrogen supplements (other than vaginal creams), work with their gynecologist to either eliminate the supplement or reduce the estrogen to the lowest level possible for a 3-month trial.

Diet

All patients are placed on a diet schedule, which is given to them in written form (Table 15-3). This diet eliminates foods containing high levels of tyramine and other substances known to exacerbate migraine.[40,41,45] Some of these foods cause migraine almost immediately (red wine, MSG); most cause migraine the next day (chocolate, nuts, cheese). Aspartame, found in NutraSweet, can also provoke migraine.[46]

Box 15-3

SCHEDULE TO TREAT MIGRAINE DISORDERS

1. Reduction of stress:
   a. Aerobic exercise at end of day (3 to 4 times/week). Get heart rate above 100 and sustain it for at least 20 minutes.
   b. Eat something at least every 8 hours to avoid hypoglycemia. Eat breakfast at same time each morning (breakfast on weekends should be at the same time as on weekdays).
   c. Maintain a regular sleep schedule.
2. Do not smoke or chew any products that contain nicotine.
3. Avoid exogenous estrogen (oral contraceptives, estrogen replacement).
4. Follow diet.
5. Keep a diary:
   a. Note time and date of all headaches and/or spells that interfere with daily routine.
   b. Write down any foods that you had that are listed on the other side of this sheet during the 24 hours before the headache and/or spell.
   c. Bring diary in with you on your next visit!
6. Medications.

Diary

Finally, all patients are asked to keep a careful diary, noting the time and date of all spells or headaches that interrupt their daily activities. They are asked to write down any foods from the list of "foods to avoid" (see Table 15-3) that they had during the 24 hours before the headache or spell. This forces the individual to become more aware of the association of diet with migraine and potentially identifies certain foods they should avoid.

Prophylactic Medical Therapy

When migraine occurs several times a month, prophylactic daily medical therapy designed to prevent migraine should be used. Evidence-based guidelines for preventive therapies of migraine have recently been updated for both prescription medications (Table 15-4) and non-prescription medications (Table 15-5).[47,48] Tables 15-4 and 15-5 include only Level A and B, Class I or II studies. The criteria used to establish the Levels and Classes of studies is listed in Box 15-4.[49] All these medications have been found to be effective in reducing the frequency and severity of headache with or without aura. Some of these medications have also been found to be useful in preventing migraine-associated dizziness.[50,51]

Based on personal observations, this author has found that propranolol is quite effective in preventing auras, including vertigo; therefore, it is the first drug used to treat patients with frequent migraine auras. Contraindications include congestive heart disease, cardiac block, asthma, diabetes, and orthostatic hypotension. Patients start on 40-mg tablets, 1/2 tablet bid and increase this drug in 20-mg increments every 3 to 7 days, depending on patient tolerance of the drug. The effective dose is usually 80 to 200 mg/day. As this drug is increased, heart rate and blood pressure are monitored. Once the therapeutic dose is found, long-acting propranolol (80- to 120-mg capsules) may be prescribed. If they remain relatively symptom free for a few months, the medication is tapered every 1 to 2 weeks to the lowest effective dose.

Finally, acetazolamide (Diamox) 250 mg bid has been shown to decrease spells in patients with episodic ataxia-2. It may also be helpful in patients with familial migraine with vertigo.[52]

Abortive Medical Therapy

Several medications are used to treat acute migraine (abortive therapy).[53] For mild to moderate headache, NSAID, aspirin, or similar medications are used. If these fail, then triptans (sumatriptan, zolmitriptan, frovatriptan, almotriptan, eletriptan, rizatriptan, and naratriptan) and DHE are effective. Triptans or DHE can also be used for moderate to severe migraine headaches.

Migraine versus Ménière's Disease

There is a great deal of confusion between migraine aura without headache and vestibular hydrops (vestibular Ménière's disease). There is a higher than expected incidence of both migraine and Ménière's disease in the same individual and among individuals with Ménière's disease, 55% also have migraine headaches.[1,54-57] Both can present with transient vertigo, ear fullness, or occasional tinnitus, but without any decrease in hearing (Table 15-6). A history of headaches associated with the spells of vertigo may help to distinguish these two

■ Table 15-3 DIET FOR PATIENTS WITH MIGRAINE

Food Category	Foods to Reduce or Avoid	Instead Use
Beverages	Chocolate and cocoa. Alcoholic beverages aged in wood containers (red wine, port, sherry, scotch, bourbon, gin, chardonnay). Sugar-free, nonalcholic drinks with aspartame or Nutrasweet)	All other beverages including: wine aged in metal containers (including chardonnay and red wines of the "Naked brand"); sugar-free, nonalcoholic with saccharin or Splenda
Aged Cheese	Cheeses: Stilton, bleu, cheddar, mozzarella, cheese spread, Roquefort, provolone, gruyere, muenster, feta, parmesan, emmenthal, brie, brick, camembert types, cheddar, gouda, romano	Cheeses: American, cottage, farmer, ricotta, cream, Canadian, processed cheese slice
Nuts	Almonds, walnuts, cashews	Peanuts
Beans	Garbanzo or chick peas	All others
Smoked Meats	Ham, turkey, hot dogs, etc. (look for nitrates on the package)	All meats without nitrates listed in ingredients
Desserts	Any product containing chocolate and nuts	Any sweets without chocolate and nuts
Miscellaneous	Monosodium glutamate (MSG) in Chinese, Mexican, and seafood. Also found in soups, Accent seasoning, meat tenderizer, seasoned salt, yeast, yeast extract	Salt in moderation, lemon juice, butter or margarine, cooking oil, whipped cream, white vinegar, commercial salad dressings

Modified from Diamond, 1991,[41] and Shulman et al, 1989.[46]

■ Table 15-4 EVIDENCE-BASED GUIDELINES FOR PREVENTIVE THERAPIES AVAILABLE IN USA OF MIGRAINE

	Antiepileptics	β-Blockers	Triptans	Antidepressants
Level A, Class I or II	Divalproex sodium, sodium valproate, Topiramate	Metoprolol, Propanolol, Timolol	Frovatriptan	
Level B, Class I or II		Atenolol, Nadolol	Naratriptan, Zolmitriptan	Amitriptyline, Venlafaxine

*Modified from Silberstein et al, 2012.[48]

■ Table 15-5 EVIDENCE-BASED GUIDELINES FOR COMPLEMENTARY PREVENTION OF MIGRAINE

	Herbal Preps, Vitamins, Minerals	NSAIDs	Histamines
Level A, Class I or II	Petasites (butterbur)		
Level B, Class I or II	Magnesium, MIG-99 (feverfew, Riboflavin)	Fenoprofen, Ibuprofen, Ketoprofen, Naproxen, Naproxen sodium	Histamine SC

*Modified from Holland et al, 2012.[49]

Box 15-4

EVIDENCE-BASED CRITERIA

Class I Evidence provided by a prospective, randomized, controlled clinical trial with masked outcome assessment, in a representative population. The following are required: (a) primary outcome(s) is/are clearly defined; (b) exclusion/inclusion criteria are clearly defined; (c) adequate accounting for drop-outs and crossovers with numbers sufficiently low to have minimal potential for bias; and (d) relevant baseline characteristics are presented and substantially equivalent among treatment groups or there is appropriate statistical adjustments for differences.

Class II Evidence provided by a prospective matched group cohort study in a representative population with masked outcome assessment that meets a–d above OR a randomized control trial in a representative population that lacks one criteria a–d

Class III All other controlled trials (including well-defined natural history controls or patients serving as own controls) in a representative population, where outcome assessment is independent of patient treatment.

Class IV Evidence from uncontrolled studies, case series, case reports, or expert opinion.

Level A Established as effective, ineffective, or harmful for the given condition in the specified population. (Level A rating requires at least two consistent Class I studies.)*

Level B Probably effective, ineffective, or harmful (or probably useful/predictive or not useful/predictive) for the given condition in the specified population. (Level B rating requires at least one Class I study or two consistent Class II studies.)

Level C Possibly effective, ineffective, or harmful (or possibly useful/predictive or not useful/predictive) for the given condition in the specified population. (Level C rating requires at least one Class II study or two consistent Class III studies.)

Level U Data inadequate or conflicting; given current knowledge, treatment is unproven.

Class of evidence for therapy: *"Class"* refers to the quality of research methods employed in the reviewed literature.

Recommendation Level: *"Level"* refers to the strength of the practice recommendation based on the reviewed literature.

*Adapted from AAN therapeutic classification of evidence scheme.[50]

■ Table 15-6 **MIGRAINE VERSUS MENIERE'S DISEASE**

Migraine	Ménière's Disease
Tinnitus: high-pitched	Tinnitus: low-pitched, roar
May have ear fullness (ache), phonophobia, and photophobia	Usually ear fullness or hearing loss
True spontaneous vertigo is rare; can occur for minutes	True spontaneous vertigo is common; can occur for hours
Short nap usually helps	Short naps usually do not help
Visual auras are common	Visual auras are uncommon
Motion sickness is common	Motion sickness is uncommon

syndromes, but occasionally the diagnosis is only made following the patient's response to a therapeutic trial (see Case Study 15-3). Patients with well-documented Ménière's disease may later develop migraine aura without headache. Therefore, they may initially do well with treatment for Ménière's disease and then appear to fail to respond to treatment, when in fact they have developed spells of vertigo as a result of migraine aura without headache (see Case Study15-4). Where there is no fluctuation in hearing, one should lean toward a diagnosis of migraine, as that disorder is five time more common then Ménière's disease.[58-60]

Patient Information

Glaxo Wellcome (1-800-377-0302) and http://www.healthylives.com/. Obtain free pamphlets including "Chart Your Route to Relief: A Personal Migraine Management Program."

For free newsletters, contact the National Headache Foundation at 1-800-843-2256 and http://www.headaches.org/.

CASE STUDY 15-1

A 46-year-old owner of a blacktop paving company is referred for spells of vertigo, nausea, vomiting, oscillopsia, and diaphoresis for the past 2 years, each lasting approximately 30 minutes. He had sustained tinnitus and hearing loss on the right side, which did not fluctuate with his spells of vertigo. In addition, he had a lifelong history of sinus pressure discomfort in the forehead, eyes, and behind the nose, for which he took a decongestant. His last sinus discomfort was 3 years ago. In addition, he has recently had episodic flashes of light lasting 10 to 15 minutes. He has a history of hypertension and angina but a normal EKG and coronary angiography. In the last 2 months, the frequency of his spells of vertigo increased to 1 per week, and the last few spells were associated with left arm paresthesia and dysarthria. One spell was witnessed, and the patient was found to have a sustained left-beating nystagmus for 20 minutes.

Comment

Migraine and Ménière's disease can be frequently difficult to distinguish. An important feature in this patient's history that points to migraine is that his tinnitus and hearing loss did not fluctuate with his spells. This feature applies only to patients who have sufficient hearing to notice fluctuation. Sinus headaches and migraine are also frequently confused; both can be located in the same area of the face and head. Episodic flashes of light are a helpful tip pointing to migraine aura. Basilar artery migraine is suggested by the other signs and symptoms during the spell, including paresthesias, dysarthria, and left-beating nystagmus. A diagnosis of basilar artery stroke was considered. A four-vessel cerebral arteriogram was normal, as was an MRI of the head with contrast. In summary, this patient was thought to be having impending brainstem stroke with possible ischemia to the right brainstem resulting in vertigo, left-beating nystagmus, and left arm paresthesia. Of interest was that he also had angina with normal coronary arteries. He had a remote history of "sinus headaches" and recently has been experiencing scintillating scotomas. A diagnosis of basilar-type migraine (ICHD classification 1.2.6) was made, and his spells of vertigo stopped after he was placed on a diet and propranolol.

CASE STUDY 15-2

A 28-year-old real estate developer is referred for thirty 5 to 10 minute spells of disequilibrium, vertigo, 15 degrees tilt of world, and diplopia (vertical and horizontal) over the past 5 years. Many of these spells occurred during a variety of physical activities including running, weight lifting, intercourse, and strenuous aerobic exercises (rowing machine, stair-climber, and stationary bicycle).

Comment

Because of the exercise-induced nature of these spells, they were believed to be caused by a perilymphatic fistula, and surgery was initially recommended. Features more characteristic of migraine included the development of a dull soreness over his left occiput following each spell, the association of certain foods with spells (Chinese food, ice cream, cream cheese), and the frequent omission of breakfast. Migraine can be caused by exercise. Other features not consistent with a perilymphatic fistula were a normal audiogram and no history of barotrauma, ear surgery, or ear infection. A diagnosis of basilar-type migraine (ICHD classification 1.2.6) was made. These spells stopped after he was placed on the antimigraine schedule listed in Box 15-3.

CASE STUDY 15-3

A 47-year-old medical transcriptionist is referred for a 10-year history of spells of nausea, disequilibrium, and occasional vomiting and ear fullness without hearing loss or tinnitus. Her audiogram was normal. She was diagnosed with probable Ménière's disease and treated with chlorothiazide (Diuril), dimenhydrinate (Dramamine), and no caffeine or nicotine. Because she continued to have bad attacks, she was then treated with scopolamine patches. She then began to develop headaches (usually left frontal), and ear pressure with some of the spells, worse in the summer. She had a history of severe headache with her menses since the age of 30. She had a normal caloric, CT scan of the head, and rotary chair test.

Comment

This is another case that illustrates the difficulty in distinguishing migraine from Ménière's disease. This patient started off with spells without headache. Because of a lack of headaches, she did not initially satisfy the criteria for migraine. Although rare, vestibular hydrops without hearing loss can occur. Usually, this entity eventually also affects hearing. Vestibular hydrops versus migraine aura without headache can present with identical symptoms. Until the diagnosis is secured, both entities are treated. Headaches eventually occurred. She was diagnosed with migraine with aura (ICHD classification 1.2) and migraine aura without headache (ICHD classification 1.1). She was placed on an antimigraine schedule and treated with isometheptene at the onset of the spell, which did not help. She was then placed on an increasing dose of amitriptyline and eventually reached a dose of 50 mg each night. For the next year, she continued to get a headache a few days before her menses but no dizzy spells. Because of the complaint of difficulty getting up in the morning and a dry mouth, the amitriptyline was tapered, and she was placed on an increasing dose of propranolol. She has had no headaches or spells of vertigo for the past 9 months.

CASE STUDY 15-4

A 35-year-old biochemist is referred for spells of vertigo, nausea, and vomiting lasting for less than 1 hour during the past year. As a teenager, she recalled having occasional bad "sinus headaches." Between the ages of 22 and 32 she had spells of vertigo, nausea, vomiting, fluctuating hearing loss, and tinnitus in the left ear. At that time, she was diagnosed with Ménière's disease and treated with diuretics, antihistamines, and a low-salt diet. Her current spells of vertigo were not associated with fluctuating hearing loss or tinnitus and were not altered by the use of a diuretic and a low-salt diet. Two years ago, she had a visual scintillation that lasted

Continued

CASE STUDY 15-4

for a few minutes. She had a normal neurological examination, normal caloric test, and normal rotary chair test. She had moderate to severe low-frequency sensorineural hearing defect and decreased speech discrimination on the left side. Hearing was normal on the right side. An MRI of the head with gadolinium was normal.

Comment

This patient started off with Ménière's disease that responded well to treatment. Migraine and Ménière's disease are very common, and patients frequently can have both. The peak incidence for migraine is between the ages of 35 and 45. She was diagnosed with migraine with aura (ICHD classification 1.2). She was placed on an antimigraine schedule. Over the next year, her spells of vertigo stopped, hearing in the low frequencies became normal, and she developed normal speech discrimination.

CASE STUDY 15-5

A 33-year-old professor of history is referred for five spells of vertigo beginning several years ago. These spells usually lasted a few minutes and occurred in the morning around the time of her menses. It was unclear whether head movement triggered them. She usually had disequilibrium for up to 1 hour following the vertigo. She also noted minor right-sided headaches during her menses with queasiness but denied vomiting, hearing loss, and tinnitus. She recalls having ear infections when she was young. She wondered about anxiety attacks; she lost her husband 2 ½ years previously to colon cancer. She had normal neurological and neuro-otological examinations. She was reassured that no serious problem was found and was told to come in if she developed another attack. One month later, she called and stated the spells returned. She stated that she had just finished her menses and had a minor right-sided headache. On examination, she had sustained geotropic nystagmus during the Hallpike-Dix maneuver.

Comment

Migraine is most common during the week before menstrual periods. During the migraine aura, head movements usually provoke dizziness. In about 30% of patients with migraine-provoked dizziness, true vertigo with the head still occurs. A variety of types of nystagmus can be found, including spontaneous and position induced. Sustained geotropic nystagmus is central. Transient geotropic nystagmus can be caused by canalithiasis of the horizontal semicircular canal and sustained ageotropic nystagmus can be a result of cupulolithiasis of the horizontal semicircular canal. This patient was also placed on an antimigraine schedule. Her spell and positional nystagmus resolved in a couple of days, and she did not have any recurrence of headache or vertigo during the 6 months she was followed.

Summary

It is becoming increasingly recognized that migraine is a common cause of episodic vertigo and disequilibrium in children and adults. It may present as benign paroxysmal vertigo of childhood, paroxysmal torticollis of infancy, and benign recurrent vertigo in adults. In addition, migraine is associated with motion sickness, Ménière's disease, and BPPV. Migraine is triggered by a number of factors including stress, anxiety, hypoglycemia, fluctuating estrogen, certain foods, and smoking. Episodic vertigo and disequilibrium from migraine should be treated by reducing these risk factors and, if necessary, by medical therapy.

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CHAPTER 16

Medical Management of Mal De Debarquement Syndrome

Timothy C. Hain, MD ■ *Janet O. Helminski, PhD, PT*

Mal de débarquement syndrome (MDDS), literally, "sickness of disembarkment," refers to prolonged and inappropriate sensations of movement after exposure to motion. The syndrome typically follows a 7-day sea voyage, but it has also been observed following extended airplane travel, train travel, and space flight.[1] Symptoms include rocking and swaying accompanied by imbalance. MDDS is distinguished from ordinary motion sickness, seasickness (mal de mer), and "land-sickness" by persistence of symptoms for a month or longer. Additionally, unlike disorders of the inner ear, most individuals with MDDS report that their symptoms remit with re-exposure to motion, such as driving a motor vehicle.[2] A typical case history is as follows: A 50-year-old woman went on her first ocean cruise. She had some motion sickness on the cruise, which responded to transdermal scopolamine. Immediately after returning from the cruise and getting onto solid ground, she developed imbalance and a rocking sensation, accompanied by fatigue and difficulty concentrating. Her description was "Imagine feeling like you are on rough seas 24 hours a day, 7 days a week."

Table 16-1 summarizes the available literature about MDDS. It is a disorder that mainly affects women (87%) in their mid-40s. Symptoms last at least 1 month and most frequently abate before 6 months have elapsed (median 4.1 mo).

Table 16-2 lists the features that distinguish MDDS from simple land-sickness. Land-sickness is common, and between 41% and 73% of persons disembarking from seagoing voyages experience a brief unsteadiness syndrome.[3-5] Common land-sickness typically persists for 2 days or less. Persons with land-sickness are also likely to have seasickness,[5] although persons with MDDS generally are untroubled by seasickness. Males and females do not appear to differ significantly in the incidence, intensity, or duration of land-sickness symptoms.[3] Land-sickness (or LDS), confusingly, is also termed "mal de debarquement" by some. Table 16-1 does not include reports or data concerning subjects whose symptoms last less than 1 month (i.e., potential land-sickness), except for the work of Cha, in whom the duration of symptoms in patients with "classic" MDDS could not be determined due to study design.[2]

MDDS also has some similarities to *motion sickness* (mal de mer). However, MDDS again is easily distinguished by the relatively short duration of motion sickness and gender distribution. Persons with MDDS reliably have relief of symptoms when in motion, such as driving a car, but experience recurrence of rocking once motion has stopped.[2,6] In motion sickness, many persons find driving very difficult. This is also often true for persons with vestibular disorders.

MDDS also overlaps with a little-studied group of patients called "rockers," who develop similar symptoms

■ Table 16-1 **CHARACTERISTICS OF MDDS**

Authors	Subjects	Age	Females	Males	Median Duration (mo)
Cha et al, 2008[2]	64	39	48	12	Variable
Hain et al, 1999[6]	27	49.3	26	1	3.5 (6 mo to 10 yr)
Brown and Baloh, 1987[7]	6	50.8 (33–71)	5	1	4 mo
Murphy, 1993[8]	4	40 (36–48)	4	0	1–12 mo
Mair, 1996[9]	10	37.5 (15–55)	9	0	1–6 mo
Total or (average)	111	(43.3)	93	14	(4.1)

■ Table 16-2 **FEATURES DISTINGUISHING MDDS FROM LAND-SICKNESS**

	MDDS	Land-Sickness
Duration	1 or more months	2 days maximum
Gender	About 90% female	Equal distribution
Motion-sick on boat	No	Yes
Relieved by driving	Yes	No

to MDDS without a preceding motion exposure.[10] Often, these patients develop head or trunk rocking, which is called "titubation."[11] In our clinical experience, the age, gender, and pattern of medication responsiveness of this group are similar to those of MDDS. Although titubation is associated with cerebellar disturbance, evidence of cerebellar damage is generally not found in "rockers."

Cause of MDDS: Persistent Adaptation to Swaying Environments?

MDDS syndrome does not have the features of a "pathological" disease, in the sense that it does not follow an injury or stressful event. Rather, it is provoked by exposure to motion that does not trouble most individuals in a persistent manner. While on the ship, the brain must adjust leg and body motion so that they counter the rhythmic pattern of shipboard motion. Adaptation to such movement is sometimes called "gaining sea legs." A common explanation for MDDS is that persons with MDDS are good at adapting to unusual motion situations, such as ocean travel, but slow to give up their adaptation when they return to the stable ground.[9]

Let us consider the visual, somatosensory, and vestibular consequences of boat motion in the anterior-posterior plane and how the brain might develop adaptive "rules" to handle them. Traveling on a boat exposes a person to angular and linear movement, some of which is predictable and some of which is not. For small rotations of the boat under the person, there is no vestibular consequence because bodily inertia tends to keep the person upright in space. Vision is accurate on the deck but inaccurate inside. Although there is rotation around the ankle joint, and thus somatosensory input, there should be no "righting" response from the person, because the body is upright in space. As vision is unreliable, a "rule" about using visual cues cannot be made. The rule, then, for pitch rotation of the boat is that one should ignore somatosensory information signaling rotation. Thus, for pitch of the boat, a selective "downweighting" of somatosensory information, or both somatosensory and visual information according to context, would be reasonable.

For linear acceleration of the boat under the person, or "surge" as it is called in nautical contexts, inertia attempts to keep the person still in space, but due to shear force at the feet, the person becomes destabilized and rotates at the ankles. Then vision, vestibular, and somatosensory senses are activated by the bodily rotation with respect to the boat, and an active response is needed to prevent a fall. Thus, for surge of the boat, no relative sensory reweighting would be needed, although increased responses to all types of input might be helpful.

As different weightings would be useful for different types of boat motion, no single weighting rule would be

optimal. A potential solution that also explains the persistence of symptoms is that, in MDDS, there is prediction of boat motion through an internal model of boat motion—an internal oscillator. One should ignore ankle inputs that are entrained with boat rocking, but one should compensate for input that is uncorrelated with boat rocking. An internal model of periodic boat motion—an internal oscillator that is entrained by boat motion—might allow one to select out salient sensory input (boat surge) and ignore the non-salient input (boat pitch). In support of this idea, some animals exhibit persistent oscillations in central neurons after periodic movement ends.[12] Also, post-movement illusions of rocking can be induced by sinusoidal rotation in some individuals.[13] Such an internal oscillator might result in persistent rocking when there is no ongoing periodic motion.

With respect to the hypothesis that MDDS is caused by reweighting of visual, vestibular, or somatosensory input, the data so far is contradictory. Nachum and associates used posturography to study young males aged 18 to 22 with motion sickness and land-sickness (they considered land-sickness to be equivalent to mal de debarquement in their paper). They reported that these young men developed increased reliance on somatosensory input after motion exposure, and reduced weighting of vision and vestibular input.[14] Although the accuracy of visual input depends on whether one is inside the boat or on the deck, semicircular canal input is accurate on boats, and somatosensory input is intermittently accurate. Accordingly, it is difficult to understand a rationale for this adaptation. An intrinsic problem with this study is that the study group was young men with motion sickness and land-sickness, not middle-aged women with the month or greater MDDS syndrome.

A more reasonable possibility is that individuals with MDDS may develop an increased reliance on visual and vestibular information (and thus *decreased* somatosensory weighting). This occurs in normal subjects who are exposed to situations in which somatosensory feedback is distorted and would also be a reasonable adaptation to boat pitch.[15] Either adaptation might result in inaccurate land sensorimotor integration. Nevertheless, neither of these adaptations explains the rocking sensation of MDDS or the characteristic improvement on driving a car.

Treatment of MDDS

The usual treatment strategy for MDDS is to attempt to make the patient comfortable, while waiting for the MDDS to end by itself (typically within 6 months, see Table 16-1). Conventional vestibular suppressants that affect anticholinergic pathways such as meclizine and transdermal scopolamine are not helpful in MDDS.[6] Benzodiazepines, such as clonazepam, are of the most benefit,[6,10] and selective serotonin reuptake inhibitor (SSRI)-type antidepressants are also suggested as being potentially helpful.[10] There are also anecdotal reports of good responses to gabapentin, amitriptyline, and venlafaxine—all medications that are also helpful in migraine.

After 6 months have gone by, if the MDDS patient is no better, there is more pressure to find another intervention. While vestibular physical therapy would seem reasonable, Cha commented in passing that "only rare patients seem to be cured by vestibular therapy."[10] In fact, the only literature describing physical therapy treatment for MDDS is a single case report.[16] Of course, it is not known how this case would have done without intervention. In general, although many individuals with MDDS undergo vestibular rehabilitation, again because of a lack of controls, it is not possible to determine whether they did any better than persons who were not treated.[6] Thus the efficacy of vestibular rehabilitation for MDDS is unknown.

Our view is that MDDS was acquired from motion, and it should be possible to find an approach that extinguishes MDDS too. But what should this procedure include? Because no compelling or controlled studies have been published to date, conjectures regarding treatment are speculative.

Motion sickness has been treated successfully with habituation,[17] and one might reasonably argue that if MDDS is a motion sickness variant, it might also respond to a similar approach. Habituation entails a down-weighting of motion input and can reduce the long-duration vestibular responses commonly associated with motion sickness susceptibility.[18] Although there are well developed self-directed motion habituation protocols, such as the PUMA exercises,[19] there are presently no reports of their efficacy in MDDS. Because patients with MDDS initially became ill after repetitive motion exposure, the possibility exists that more motion exposure as is required for habituation might worsen MDDS.

Based on the present theories of mechanism, there are several treatment strategies that might be attempted in MDDS. As outlined in the section above, two theories applicable to MDDS differ in the conjecture regarding weighting of somatosensory input (one too much, one too little), and a third proposes that a central internal oscillator is the source. Accordingly, if during one's assessment of the patient, one determines that somatosensory weighting is inappropriate, one might attempt a specific intervention.

For example, if the individual disregards proprioceptive information in sitting, the individual should practice detecting joint position sense and kinesthetic awareness of the toes and ankles with the eyes closed. Once there is heightened awareness of joint position sense, the individual should use the sensory information during a dynamic task. While standing on a firm surface with the eyes closed, the individual may dynamically shift weight anterior to posterior for 1 to 2 minutes and then side to side for 1 to 2 minutes. The individual may then be asked to walk with eyes closed on mats with objects placed randomly beneath the mat to increase focus on the somatosensory information.

For persons who overweight motion information, one's efforts might be reasonably directed toward down-weighting the inputs. Here, abundant movement stimuli bringing the person to the edge of their tolerance, such as visual or vestibular habituation protocols, might reduce the size and duration of their motion responses and thus reduce symptoms.

If MDDS is instead caused by an internal oscillator developed to predict boat motion, one's treatment strategy should be aimed at manipulation of psychological variables rather than somatosensory integration. Patients need to ignore their aberrant internal signal, in the same way that most persons with tinnitus eventually develop an ability to ignore abnormal internally generated sounds. Treatments that decrease vigilance, obsessiveness, and anxiety, as well as "tincture of time," would be the optimum strategy. If this conjecture is correct, therapy that focuses attention on the rocking sensation could even be counterproductive.

Summary

MDDS is an uncommon disorder in which individuals, mainly women in their mid-40s, develop an inappropriate sensation of rocking, paradoxically relieved by actual motion such as driving. Research is needed to establish the role of physical therapy in the treatment of MDDS.

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CHAPTER 17

Surgical Management of Vestibular Disorders

Yew Ming Chan, FRCS Edinburgh, FAMS ■ *Douglas E. Mattox, MD*

The diagnosis of vestibular disorders is complicated by overlapping symptoms among the various disorders and the lack of pathognomonic diagnostic tests. At times, determining which inner ear is causing the symptoms may even be difficult. Most patients' symptoms can be managed with the medical and physical therapy measures described elsewhere in this book. However, surgical intervention may be appropriate when the symptoms have failed to respond to aggressive nonsurgical medical management.

With the exception of acoustic tumors, vestibular disorders are a matter of lifestyle and comfort and are not life threatening. Therefore, it must be the patient living with the symptoms who makes the decision whether or not to proceed with surgery. The physician should discuss the likelihood of a successful outcome and the nature and likelihood of potential complications, and must leave the ultimate decision up to the patient. In the authors' experience, patients have a broad spectrum of responses to their symptoms of vertigo. Some patients want immediate intervention; others consider surgery only when life becomes unbearable.

Vestibular Schwannoma (Acoustic Neuroma)

Acoustic neuromas are nerve sheath tumors occurring in the internal auditory canal (IAC) or cerebellopontine angle (CPA).[1] They are the third most common intracranial tumor, accounting for 8% to 10% of all intracranial tumors. Most patients with acoustic neuromas present with progressive unilateral sensorineural hearing loss. However, some patients first complain of vestibular symptoms, sudden hearing loss, or occasionally trigeminal symptoms.[2]

An acoustic neuroma should be suspected in any patient with an unexplained unilateral sensorineural hearing loss, particularly if the patient has discrimination scores inconsistent with pure tone audiometry, abnormal brainstem auditory responses, or hypoactive/absent caloric responses. Magnetic resonance imaging (MRI) with gadolinium contrast has become the "gold standard" for the diagnosis of these tumors. Although there are rare instances of false-positive results, usually from arachnoiditis, an enhancing mass in the cerebellopontine angle extending into the internal auditory meatus on MRI is almost always an acoustic neuroma. Meningiomas occasionally occur in the CPA or IAC. Radiologically, they frequently have a dural "tail" and are acentric to the IAC.

Once the diagnosis is established, there are three therapeutic options: watchful waiting, microsurgical removal, and stereotactic radiosurgery (SRS). Watchful waiting is indicated only in patients with small intracanalicular tumors for which the diagnosis is inconclusive, and in patients who are elderly or in poor medical condition. Several recent series have reported the results of this approach. Wiet and colleagues[3] found that in 40% of 53 patients, continued growth of tumors required intervention over a mean follow-up of about 3 years. More frightening is an experience reported by Charabi and associates.[4] In their series, 34% of 123 patients followed for a mean of 3.4 years

required intervention for enlarging tumor and 7 died of brainstem compression secondary to the tumor. Therefore, it may be concluded that there is a significant risk of tumor enlargement, and if a "wait-and-see" approach is taken, the importance of serial scanning at 6- to 12-month intervals must be emphasized to the patient.

Surgical Approaches

Surgical removal of acoustic tumors has been the treatment of choice since described by Harvey Cushing[5] in 1917. The three basic approaches used for removal of acoustic neuromas are (1) middle fossa craniotomy, (2) translabyrinthine approach, and (3) suboccipital craniotomy.[6,7] The choice of approach is based on the size and location of the tumor and whether any attempt will be made to preserve hearing.[6,7]

Middle Cranial Fossa

The middle cranial fossa approach is used for tumors confined to the internal auditory canal in patients who have usable hearing (Fig. 17.1). A vertical incision is made in the scalp superior to the external auditory canal. The soft tissues are elevated from the bone and a 3 × 4 cm temporal craniotomy is performed. The dura is elevated from the floor of the middle cranial fossa. The internal auditory canal is identified by drilling the bone overlying it. The facial nerve, mastoid air-cell system, and superior semicircular canal are important landmarks. The bone is thinned over the entire extent of the internal auditory canal, and then the dura of the canal is incised. Care is taken not to damage the facial nerve in the anterior superior quadrant of the internal auditory canal. The cochlear nerve is anterior-inferior in the canal and safely out of harm's way.

The advantage of the middle cranial fossa approach is that it does not destroy the inner ear and hearing. Care must be taken to avoid damaging the facial nerve, because it is superficial in the dissection. Managing tumors that extend through the porus acusticus into the posterior fossa with use of the middle cranial fossa approach is very difficult. This approach, therefore, is indicated only for tumors limited to the internal auditory canal.

Recovery after middle fossa craniotomy is prompt. Unless the patient has significant vestibular symptoms, he or she should be up and about the next day. Cerebrospinal fluid (CSF) leakage is unlikely, but the patient should be checked for both external leak and a leak down the eustachian tube into the nasopharynx after undergoing surgery via this approach and all other approaches described in this section.

Translabyrinthine Approach

The translabyrinthine approach is the procedure of choice for tumors up to 3.0 cm in diameter when hearing preservation is not a consideration (Fig. 17.2). The tumor is

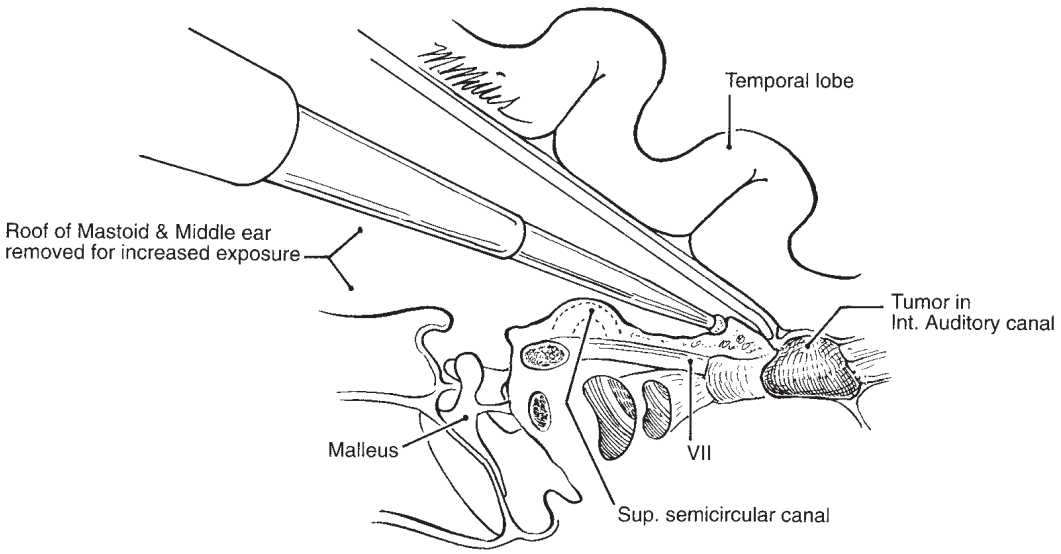

Figure 17.1 Middle fossa. This coronal section through the internal auditory canal and middle ear shows the exposure of the internal auditory canal through the middle fossa. A drill is shown passing through a small temporal craniotomy. The temporal lobe is elevated extradurally. The superior semicircular canal is identified, and the internal auditory canal is exposed by removal of the bone over the auditory canal medial to the superior semicircular canal.

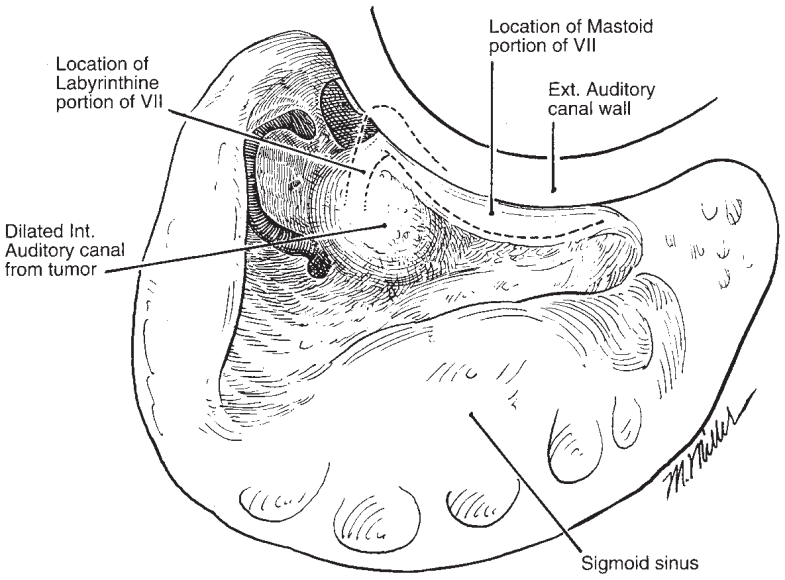

Figure 17.2 Translabyrinthine approach for removal of acoustic neuroma. A translabyrinthine approach to the internal auditory canal is shown in the surgical position with anterior-top, superior-left. The tumor is exposed after completion of a labyrinthectomy. The internal auditory canal has been dilated by the tumor. Removal of the last shell of bone over the tumor and the bone from the posterior fossa allows complete removal of the tumor.

approached as in a standard mastoidectomy. The cortical and pneumatized bone of the mastoid are drilled away to expose the sigmoid sinus, posterior fossa dura, and middle fossa dura. The facial nerve is protected by identification within its bony canal. A complete labyrinthectomy is performed to expose the internal auditory canal. Once the intracanalicular portion of the tumor is mobilized, the dura of the posterior fossa is incised, and the remainder of the tumor is removed. The translabyrinthine approach can be extended by combining it with a temporal/retrosigmoid craniotomy for larger tumors.

Recovery after translabyrinthine removal of acoustic tumors is generally prompt, and patients can be out of bed in 2 to 3 days. This rapid recovery is attributable to the lack of pressure or retraction of the cerebellum during the procedure. The disadvantage of the translabyrinthine approach is that hearing is automatically sacrificed. The exposure is excellent for small and medium-sized tumors but is inadequate for tumors more than 3 cm in diameter and for those that are adherent to the brainstem. In the ideal case for translabyrinthine removal, computed tomography (CT) or MRI shows a clear separation between the tumor and the brainstem.

Suboccipital Craniotomy (Retrosigmoid Approach)

Suboccipital craniotomy is used for medium and large tumors (Fig 17.3). In rare cases in which there is good hearing preoperatively, the suboccipital approach offers the possibility of preserving hearing. Maintenance of hearing requires the preservation of both the cochlear nerve and the fragile capillary blood supply of the inner ear. Hearing can be spared in one-third to one-half of the patients in whom this approach is attempted.[8]

The suboccipital craniotomy differs from the translabyrinthine approach in that the angle of the approach is from behind rather than in front of the sigmoid sinus. The incision is placed 5 to 6 cm behind the ear, and a 5 cm opening is made in the occipital skull. This defect is reconstructed with prosthetic mesh at the end of the procedure.

The cerebellum lies between the craniotomy and the CPA; however, the operation is performed with the patient in the lateral position, allowing the cerebellum to fall away by itself without additional retraction.

After the tumor is identified, the capsule of the tumor is incised, and the central core of the tumor is removed with hand instruments, an ultrasonic aspirator or laser. After the tumor has been decompressed, the portion of the tumor within the IAC is removed by drilling away the posterior surface of the canal. The facial nerve is identified in the fundus of the IAC where its anatomy is constant. The tumor is mobilized from the brainstem, and the eighth nerve is identified medial to the tumor.

Recovery time after a suboccipital craniotomy is longer than from the other two approaches because of the magnitude of the procedure, but patients are usually ready for discharge from the hospital within a week. Occasionally, a patient experiences a postcraniotomy headache that requires long-term pain management.

Complications

Severe complications after acoustic neuroma surgery are relatively uncommon. Hearing loss always occurs in

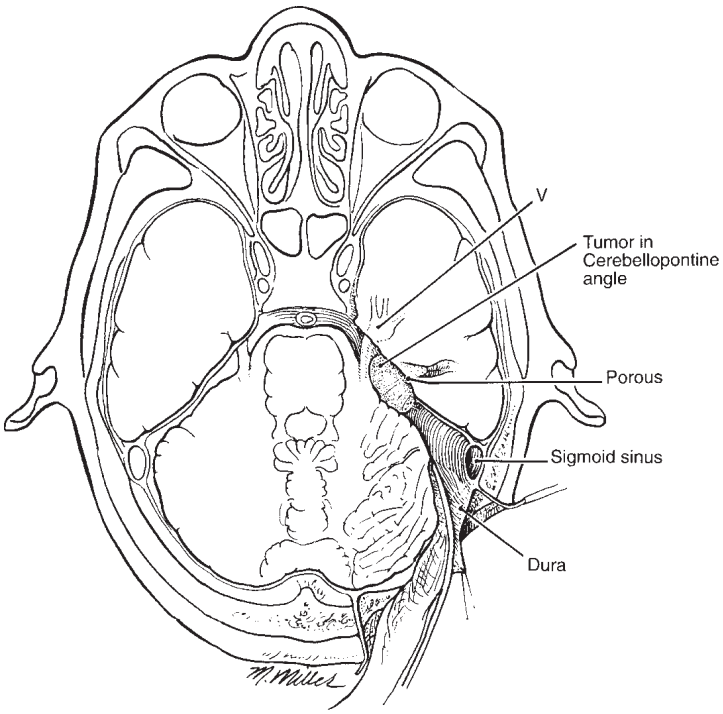

Figure 17.3 Suboccipital craniotomy. The axial section demonstrates a posterior fossa craniotomy behind the sigmoid sinus. The cerebellum drops away from the posterior surface of the temporal bone. The tumor is exposed in the cerebellopontine angle. The posterior lip of the internal auditory canal must be drilled away to remove the tumor extending into the canal.

translabyrinthine procedures and is common after suboccipital removal of tumors larger than 1.5 cm. Transient facial paralysis is common with larger tumors. Permanent facial paralysis is seen in fewer than 5% of patients. CSF leaks can occur postoperatively but are usually transient and rarely require secondary surgical correction.

Stereotactic Radiosurgery

SRS was introduced by Leksell[9] in 1971 and has gained wide popularity in the last few years. In this procedure, a single treatment of high-dose irradiation is administered by stereotactically focused multiple radiation sources or arced beams focused on the tumor. The tumor receives an extremely high radiation dose (currently 12 Gy for single treatment protocols), although the surrounding neural and vascular structures are spared.

The results of radiosurgery are encouraging. Tumor control is achieved in 70% to 80% of patients, and the procedure has a low incidence of complications, especially facial paralysis.[10,11] Pollock and coworkers[12] compared the results of SRS and microsurgical removal in 87 patients (40 surgery, 47 SRS). The results for surgery versus SRS, respectively, were as follows: tumor control, 98% versus 94%; preservation of hearing, 14% versus 75%; and normal facial function, 63% versus 83%. Longer-term follow-up is not as encouraging for hearing preservation. Lin and associates[13] found a drop in serviceable hearing from 69% pretreatment to 6.7% at a mean of 4 years after SRS.

Despite successful outcome in the majority of patients treated with SRS, a small fraction of patients have continued growth of the tumor and require surgical salvage. Several authorities have commented on the increased difficulty of surgical dissection after SRS. A case-matched study by Limb and colleagues[14] showed severely increased fibrosis of the tumor, more difficult dissection of the facial and lower cranial nerves from the tumor capsule, longer operative times, and poorer facial nerve outcomes for surgery after irradiation. The ideal indications for microsurgical removal versus stereotactic radiosurgery for acoustic tumors remain contentiously debated. I recommend surgical removal of tumors in younger patients, of medially placed tumors, which tend to grow faster, of tumors larger than 3 cm or with significant brainstem compression, and of small tumors in patients with a legitimated chance of hearing preservation. SRS is ideal for middle-aged and elderly patients and patients with medical problems that make them poor surgical risks.

Ménière's Disease

Although the underlying etiology of Ménière's disease is unknown, a consistent histopathological finding is hydrops (dilation) of the endolymphatic spaces.[15] The hydrops

presumably results from a malfunction of the resorptive function of the endolymphatic sac. The classic constellation of symptoms includes fluctuating hearing loss, episodic vertigo, tinnitus, and a sensation of fullness in the ear.[16] These symptoms do not necessarily develop simultaneously, however, and many patients do not experience all of them. Subcategories of Ménière's disease describe these other conditions—for instance, cochlear hydrops (fluctuating hearing loss alone) and vestibular hydrops (vestibular symptoms without hearing loss).

In most patients, Ménière's disease is ultimately self-limited; over time, the patient suffers deterioration of hearing and a gradual subsiding of the episodic dizzy spells. This evolution, however, may require 10 or 20 years. In the interim, the patient's lifestyle may be severely impaired.

Medical therapy of Ménière's disease rests on avoiding factors known to exacerbate the symptoms: stress, caffeine, alcohol, nicotine, and foods high in salt. Diuretics and vestibular suppressant drugs are usually prescribed. This regimen, known as the Furstenberg regimen, can adequately control the symptoms in up to three-quarters of patients.[17] A few patients, however, cannot be adequately managed by medical means alone, and surgical intervention must be considered. The surgical procedures for Ménière's disease may be categorized as those designed to improve the function of the endolymphatic sac and those that ablate the vestibular system, with or without preservation of hearing.

Surgical Management of Ménière's Disease

Transtympanic Steroids

Transtympanic steroid therapy has been advocated for the treatment of vertigo symptoms in Ménière's disease and idiopathic sudden sensorineural loss. An extensive review had difficulties making definitive recommendations because of the heterogeneous nature of the data related to individual drugs, drug doses, and frequency of injection.[18] Certainly, intratympanic steroid treatment has a role in Ménière's patients with bilateral disease or when the physician is treating the only hearing ear. The choice of type of steroid injection is between dexamethasone and methyl prednisolone. We prefer two to three injections per week for 2 weeks of dexamethasone 4 mg/ml. Usually, 0.4 to 0.5 mL can be injected into the middle ear space, and the patient is advised to rest in a supine position for 45 minutes with head turned 30 degrees to the unaffected side.

Although middle ear infusion of steroids may have a role in amelioration of Ménière's symptoms, our experience concurs with others that intratympanic gentamicin therapy achieves better long-term control of vertigo in Ménière's disease.[19]

Endolymphatic Sac Surgery

Endolymphatic sac procedures attempt to reestablish the function of the sac as the resorptive organ for the endolymph of the inner ear by draining the excess endolymphatic sac into the mastoid cavity.[20] A standard postauricular mastoidectomy is performed, and the sigmoid sinus, mastoid antrum and incus, facial nerve, and lateral and posterior semicircular canals are identified. The endolymphatic sac is found between the posterior surface of the temporal bone and the dura of the posterior fossa. The bone is thinned until the dura and the sac are identifiable through the last layer of bone. This bone is picked away to expose the dura and the overlying endolymphatic sac. The sac is opened, and polymeric silicone (Silastic) sheeting or another shunt device is inserted into the lumen of the endolymphatic sac and allowed to drape into the mastoid cavity. Care must be taken to open the endolymphatic sac without puncturing the underlying dura and thereby possibly causing a CSF leak. Any endolymph drained by the shunt is resorbed by the mucous membranes of the mastoid cavity.

It is an understatement to say that endolymphatic sac surgery is controversial. The fluid spaces involved are minuscule, and many investigators are skeptical about the ability of mechanical means to improve function of the sac. In a clinical trial, similar results were obtained with real and sham operations.[21] Nonetheless, the procedure seems to control the vertiginous attacks in one-half to two-thirds of patients and has the advantage of relative ease, safety, and preservation of hearing.

Ablative Procedures—Chemical and Surgical Labyrinthectomy

In all the ablative procedures described below, the implicit belief is that the disease is unilateral or, if disease is bilateral, the side producing the majority of symptoms can be determined. Surgical procedures are seldom, if ever, indicated in patients who have active bilateral disease.

Special care should be taken with any of the ablative techniques in patients with bilateral or contralateral labyrinthine hypofunction. A patient left with bilateral vestibular loss may have chronic disequilibrium, significant difficulty walking in the dark, and inability to keep the eyes fixed on a target during head movements (oscillopsia).[22]

Chemical Labyrinthectomy

Chemical labyrinthectomy with aminoglycosides has gained popularity in the management of Ménière's disease

and has almost replaced surgical intervention in some centers. Several protocols, doses, and dosing schedules have been reported, but in essence, a low dose of gentamicin is injected into the middle ear space, either directly through the tympanic membrane with topical anesthesia, through a myringotomy tube, or with a continuous-perfusion device.[23,24] This procedure appears to produce good control of vertigo attacks but poses a risk to hearing.[25] Cohen-Karem and associates[26] performed a meta-analysis of 15 studies totaling 627 patients treated with intratympanic gentamicin. Although the doses and dosing schedules varied widely among the studies, these researchers found that complete control of vertigo had been achieved in 75% of patients, and complete or substantial control in 93%. The vertigo control results did not appear to differ for the various treatment regimens. Among multiple studies, progression in hearing loss occurs in about 30% of treated patients.

Intratympanic administration of aminoglycosides has the great advantage of being a non-operative clinic procedure, thus avoiding the risks of anesthesia, surgery, and the cost of hospitalization. However, even though it is a simple procedure, it must be taken with the same seriousness as any other destructive procedure on the labyrinth. The risks of chronic post-labyrinthectomy disequilibrium and hearing loss must be carefully explained to the patient, and the same rules of informed consent apply to this procedure as a surgical intervention.

Vestibular Neurectomy and Labyrinthectomy

Although vestibular neurectomy and labyrinthectomy are more complex than endolymphatic sac surgery, control of vertigo is predictable and reliable in 90% to 95% of patients with either procedure. These procedures should completely relieve the vertiginous attacks, because vestibular input from the operated ear is completely eliminated. The loss of all vestibular function on one side can easily be compensated by an intact labyrinth on the opposite side.

When the hearing is worth preserving (the ability to detect speech—or speech reception threshold—is better than 60 dB, and the ability to understand speech—or discrimination score—is better than 50%), a vestibular neurectomy through either the middle cranial fossa or retrolabyrinthine space is the procedure of choice.

The middle fossa approach is the same as described previously for acoustic tumors. Once the internal auditory canal has been identified and exposed, it can be opened, and the superior and inferior vestibular nerves divided.

The vestibular nerve can also be sectioned through either a retrolabyrinthine or retrosigmoid (suboccipital) approach (Fig. 17.4). In the retrolabyrinthine approach, a complete mastoidectomy is performed as described for the endolymphatic sac procedure.[27] In addition, all the bone medial to the sigmoid sinus is removed to expose the posterior fossa dura. The dura is opened to expose the CPA. The vestibular and auditory branches of the eighth nerve are directly in the field of view, and the vestibular nerve is divided. A disadvantage of this approach is that the auditory and vestibular portions of the eighth nerve are fused as they exit the brainstem and may not have separated before they enter the internal auditory canal. Some surgeons have advocated a retrosigmoid approach to drill away the posterior lip of the internal auditory canal; this procedure permits identification of the vestibular nerve after it has separated from the auditory nerve.[28]

If hearing preservation is not a goal, for example, in patients with unilateral Ménière's disease and severely impaired hearing or discrimination, a labyrinthectomy is the most effective treatment. Labyrinthectomy can be performed either through the external auditory canal or through the mastoid. In the transcanal approach, the tympanic membrane is elevated to expose the middle ear. The stapes is removed, and the vestibule is opened between the oval and round windows. The saccule, utricle, and ampullae of the superior, lateral, and posterior semicircular canals are removed with an angled pick. Reaching the ampulla of the posterior semicircular canal is a blind maneuver and may leave neuroepithelium behind. For this reason, we prefer the transmastoid approach. A standard mastoidectomy is performed, and all three semicircular canals are identified. Each one is drilled away in turn, and the neuroepithelium is identified under direct vision and removed. The three semicircular canals lead to the vestibule, where once again the saccule and utricle are removed.

Post-traumatic Vertigo

Post-traumatic vertigo is managed in a manner identical to Ménière's disease, with either a hearing-preserving or hearing-sacrificing form of vestibular ablation. Most authorities, however, report less reliable control of recurrent attacks of dizziness after such treatment for this disorder.[29] The reasons for these results are unknown.

Benign Paroxysmal Positional Vertigo

Unlike patients with Ménière's disease, who experience spontaneous episodes of dizziness, those suffering from benign paroxysmal positional vertigo (BPPV) have transient symptoms only when they assume certain positions.[16]

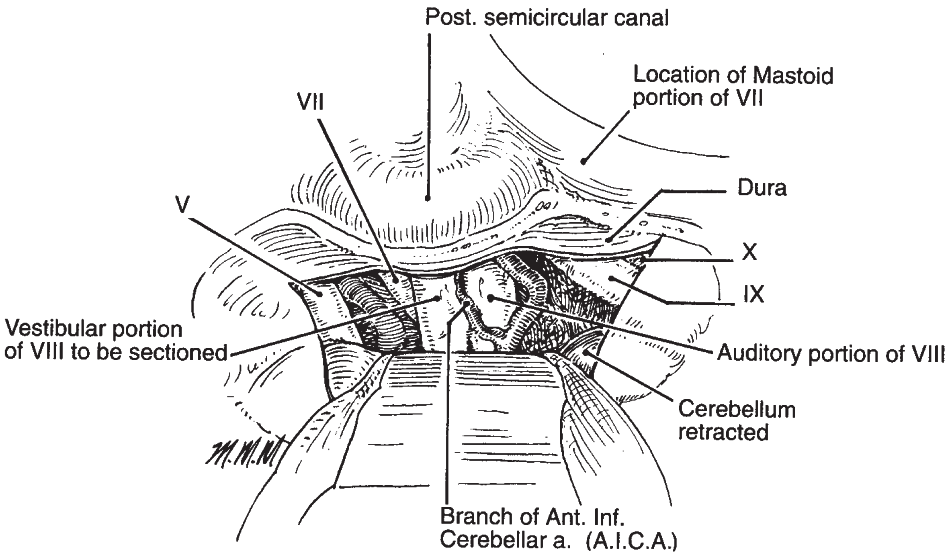

Figure 17.4 Retrolabyrinthine vestibular nerve section. This surgeon's view of a right ear in surgical position (anterior-top; superior-left) shows the exposure for a retrolabyrinthine nerve section. The cerebellopontine angle is exposed by removal of the posterior surface of the temporal bone between the sigmoid sinus and the posterior semicircular canal. The dura is opened, and the seventh and eighth nerves are identified. The demarcation between the vestibular and auditory portions of the nerve is usually marked by a small branch of the anterior-inferior cerebellar artery (AICA). The vestibular portion of the eighth nerve (superior half) is divided. Retrolabyrinthine vestibular nerve section. This surgeon's view of a right ear in surgical position (anterior-top; superior-left) shows the exposure for a retrolabyrinthine nerve section. The cerebellopontine angle is exposed by removal of the posterior surface of the temporal bone between the sigmoid sinus and the posterior semicircular canal. The dura is opened, and the seventh and eighth nerves are identified. The demarcation between the vestibular and auditory portions of the nerve is usually marked by a small branch of the anterior-inferior cerebellar artery (AICA). The vestibular portion of the eighth nerve (superior half) is divided.

The most common position is in a lateral or head-hanging position with the diseased ear down. The symptoms generally have a latency of a few seconds before onset, develop in a crescendo-decrescendo pattern, demonstrate torsional nystagmus, and habituate on repeated trials. The site of the pathology is generally thought to be in the posterior semicircular canal. Schuknecht[15] has described debris on the cupula of the posterior semicircular canal, which he has called "cupulolithiasis." The symptoms could arise from dislodged otoconia floating in the posterior semicircular canal (canal lithiasis) and from those physically attached to the cupula.

BPPV is commonly a self-limiting condition that resolves regardless of what treatment is given.[16] A number of different physical therapy measures have been designed to dislodge the otoconia from the posterior semicircular canal. These measures are effective in the vast majority of patients (see Chapter 20).

Rarely, a patient with BPPV has persistent symptoms despite physical therapy intervention and the passage of time. In these cases, two surgical procedures can be considered. The first is singular neurectomy—division of the branch of the vestibular nerve to the posterior semicircular canal.[30] This procedure is technically difficult and has been described as being performed at only a few centers. The singular nerve passes just medial to the round window niche before entering the ampulla of the posterior canal. The lip of the round window is drilled away, but the round window membrane must be violated. Bone is removed posterior and inferior to the round window with tiny diamond spurs to expose the singular canal. The canal is opened, and the nerve avulsed.

Surgical blockade of the flow of endolymph in the posterior semicircular canal has also been described.[31] In this procedure, the bony posterior semicircular canal is opened without violation of the membranous labyrinth. Flow within the membranous labyrinth is blocked by occlusion of the bony and membranous canals with a bone plug. Reports from several centers confirm this procedure as an effective and low-risk treatment for those rare cases

of BPPV that fail to respond to particle-repositioning maneuvers.[32,33]

Inflammation and Cholesteatoma

Complications of the middle ear and mastoid infections can lead to bacterial suppurative labyrinthitis and semicircular canal fistula. Management involves early intervention and treatment of bacteria otitis media with myringotomy and insertion of ventilation tube, cultures of the middle ear aspirate, and appropriate targeted antibiotics to the infection. The role of mastoidectomy surgery in acute otitis media is related to the associated symptoms of complications of acute mastoiditis with severe pain, facial nerve weakness, mastoid subperiosteal abscess, or intracranial complications. Surgical principles involve draining mastoid air cells and ventilation of the middle ear.

Surgery related to cholesteatoma matrix overlying the dehiscence of semicircular canal involves careful removal of the matrix, repairing of the fistula with careful plugging with bone pâté, and an overlay temporalis fascia. The potential of loss of hearing and labyrinthine function in large fistula defect is a real concern and risk.

Superior Semicircular Canal Dehiscence

Since Lloyd Minor[34] first described superior semicircular canal dehiscence syndrome in 1998, a larger group of similar discrete or diffuse lesions in the temporal bone have been described as part of the third-window lesions.

The symptoms are variable and range from vestibular complaints of vertigo, especially in response to external loud sounds or straining, autophony, pulsatile tinnitus, and mild conductive hearing loss.

The positive signs of the third window include sound- or pressure-induced nystagmus (Tullio's phenomenon or Hennebert's sign, respectively). A tuning fork at 128 Hz on the ankle or knee may lateralize to the affected ear. The audiogram will show either a normal hearing or conductive hearing loss mimicking otosclerosis with largest air-bone gap in the lower frequencies and supernormal bone conductions threshold below 2 kHz. Acoustic reflexes are usually present (contrary to otosclerosis), and vestibular evoked myogenic potentials (VEMPs) are present and show abnormally low thresholds.

Anatomically discrete lesions may be classified by location: semicircular canals (superior, lateral, or posterior canal dehiscence), bony vestibule (large vestibular aqueduct syndrome), or the cochlea (carotid-cochlear dehiscence, X-linked deafness with stapes gusher). Paget's disease may also behave with similar symptoms and present as a diffuse third-window lesion.[35]

Surgery to repair superior semicircular canal dehiscence is directed at the patient's needs, presence and severity of the symptoms, and confirmed by the typical hearing pattern, abnormal VEMPs and CT temporal bone (Fig. 17.5). Because there is no indication canal dehiscence is a harbinger of dire consequences in the future, surgical intervention is dictated by the patient's assessment of the severity of symptoms.

Various surgical approaches have been described to treat this condition. The middle fossa approach with plugging and resurfacing of the dehiscent canal have led to the most lasting results with improvement in the low-frequency conductive hearing loss.[36] Resurfacing alone has been less successful. The resurfacing approach with a bone flap results in recurrence of symptoms. This has led to the plugging technique with the same approach.A transmastoid approach has since been described as a quicker, safer surgical approach with less hospitalization stay.[37]

The risk of sensorineural hearing loss following various surgeries has been reported up to 12% and higher following revision surgery. A single largest report of 29 surgical repairs suggests that surgical hearing results did not differ according to method of canal repair (plugging versus resurfacing).[34] However, others mentioned delayed hearing loss in patients who had middle fossa approach following resurfacing of the canal.[38,39]

We generally prefer a middle fossa approach, repairing the defect carefully by gentle plugging of the dehiscent superior canal with hydro set or bone cement, because we believe that this has produced better hearing outcomes as had been reported by others.[40,41]

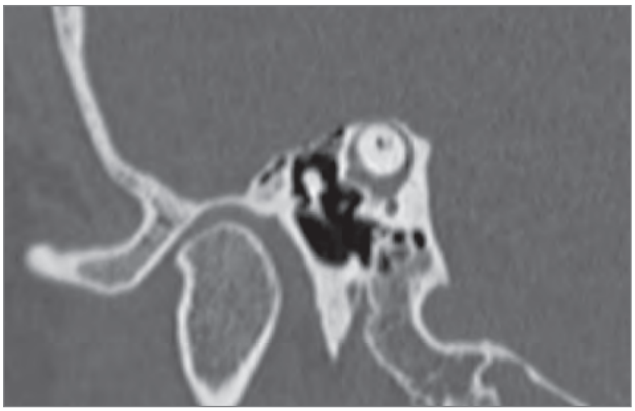

Figure 17.5 CT scan of superior semicircular canal dehiscence. This is a parasagittal non-contrast CT of the left superior canal in the plane of the canal. An open dehiscence can be seen in the top of the arch of the canal.

Perilymphatic Fistula

Perilymphatic fistula is a direct communication between the inner ear and the middle ear, usually through the round or oval window (Fig.17.6).[31] Such leaks were initially described in association with barotraumas. In the past decades, spontaneous leaks were described, although it is now clear that such occurrences are rare. Middle ear exploration for fistula has become uncommon. The symptoms of perilymph leak include hearing loss, usually sudden or episodic; vertigo associated with the hearing loss; and, more recently, generalized spatial disorientation with normal hearing.

The diagnosis of perilymphatic fistula, and the indications and timing of surgery, are controversial subjects in the otological literature. To date, no preoperative diagnostic test definitively confirms or excludes the presence of a perilymphatic fistula. Even upon surgical exploration, there may be disagreements among observers as to the presence or absence of a fluid leak. Biochemical and fluorescent tracer studies and protein analyses are being developed as potential markers for the presence or absence of the perilymph fistula.[42]

Middle ear exploration for perilymph fistula is straightforward. The middle ear is approached through the external auditory canal, and the tympanic membrane is elevated. Both the oval and round windows are carefully observed for the repeated accumulation of fluid. The leak may become more obvious with a Valsalva maneuver (increased intrathoracic pressure against a closed glottis). The leak is repaired with autogenous tissue. Clinicians generally believe that the patient should remain on bedrest for some time after closure of perilymph fistula to allow the graft to heal in place.[43]

In most cases, the patient feels better shortly after repair of a perilymphatic fistula. Patients with persistent symptoms present the physician with the difficulty of deciding whether the repair has failed or the diagnosis was wrong in the first place.

Vascular Loops

Vascular loops are elongated or tortuous vessels (arteries or veins) within the intracranial cavity that are thought to press on nerve roots as they exit from the brainstem (Fig. 17.7). The first well-described vascular syndrome

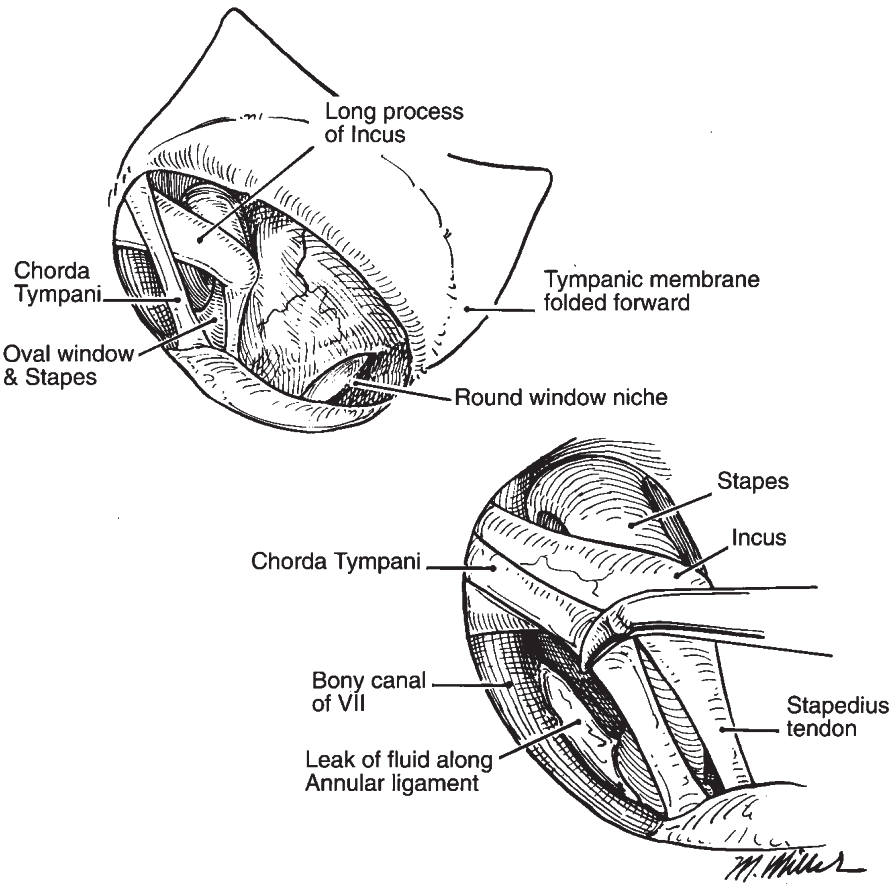

Figure 17.6 Perilymphatic fistula. A right middle ear exploration for perilymph fistula is shown in the surgical position (anterior-top; superior-right). The tympanic membrane has been reflected forward to expose the oval and round windows. Close inspection of the annular ligament of the oval window demonstrates a leak of perilymph. This will be closed with an autologous tissue graft. VIIth = seventh cranial nerve.

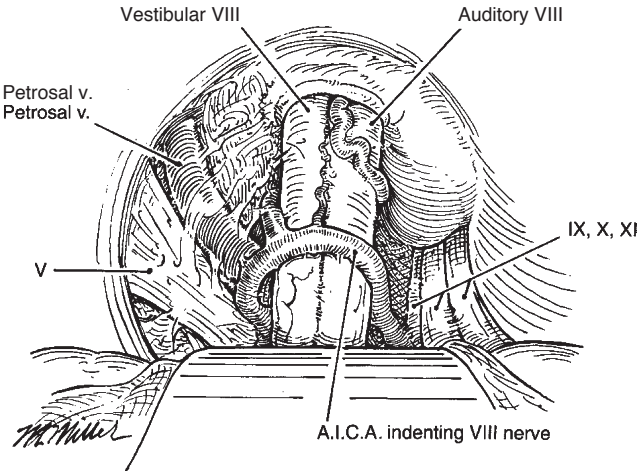

Figure 17.7 Retrolabyrinthine exposure of vascular compression. The exposure is the same as shown in Figure 17.4. The anterior-inferior cerebellar artery (AICA) is seen compressing the eighth nerve. This compression is treated by careful elevation of the vessel and interposition of muscle or sponge material between the vessel and the nerve. Roman numerals indicate cranial nerves.

was hemifacial spasm, an uncontrollable twitching of one side of the face. This was found by Janetta and colleagues[44] to be caused by an abnormal vessel pressing on the root-entry zone of the facial nerve. This concept has been expanded to include vestibular and auditory disorders.[45] The significance of these vascular loops is difficult to determine, because the symptoms o verlap other diagnostic categories, including Ménière's disease and perilymphatic fistula. Furthermore, tortuous vessels are common in the CPA of normal individuals, especially after middle age. Nonetheless, there are documented cases of vessels impinging on nerves and causing abnormal stretching or displacement. It has been suggested that radiological confirmation can be obtained with the combination of high-resolution CT with intravenous and air contrast.

Microvascular loop decompression is performed through a standard posterior craniotomy. The offending artery or vein is carefully dissected from the nerve, and a small piece of muscle or polytetrafluoroethylene sponge is interposed to keep the vessel from pressing on the nerve.

Summary

The development of surgical interventions for vertigo is a fascinating and challenging branch of neurotology. Unfortunately, at the moment, most of the procedures used are ablative rather than restorative. Future developments in this field will be directed to ward the rehabilitation and functional restoration of the diseased inner ear.

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CHAPTER 18

Management of Psychological Problems and the Dizzy Patient

Ronald J. Tusa, MD, PhD

> "It is more important to know what kind of patient has the disease than to know what kind of disease the patient has.
>
> The good physician will treat the disease, but the great physician will treat the whole patient."
>
> —*Sir William Osler*

Dizziness can cause extreme stress, which may in turn lead to anxiety (including panic attacks and agoraphobia), depression, and somatoform disorders. These psychological problems can also cause severe dizziness. At times, these psychological causes may become the primary cause of dizziness and may replace the initial organic cause of dizziness.

Two good longitudinal studies have assessed the role of psychological problems in dizziness. Kroenke and associates[1] examined 94 patients at onset of dizziness and then had the patients complete questionnaires at 4 months and 1 year later. Symptoms improved for 51 patients, stayed the same for 32, and worsened for 11. Etiology of the dizziness affected outcome. The majority of patients with benign paroxysmal positional vertigo (BPPV), neuritis, migraine, or presyncope experienced improvement. Less than half of those with Ménière's disease or psychiatric or nonvestibular disequilibrium showed improvement. The four multivariate predictors of poor outcome were (1) primary psychiatric etiology, (2) dysequilibrium, (3) daily dizziness, and (4) dizziness aggravated by walking.

Yardley and colleagues[2] examined 101 patients at onset of dizziness and 7 months later. The best longitudinal predictors of poor outcome were autonomic symptoms (heart pounding, excessive sweating, hot or cold spells, feeling faint or short of breath) and somatization (general tendency to complain of a diversity of unrelated health problems, ranging from pains in the back to difficulty concentrating). These symptoms had a better prediction of poor outcome than did the etiology of true vertigo, severity, duration, test results, or medication. High and persistent handicap arose from psychiatric or psychosocial problems unrelated to the vertigo.

This chapter summarizes the interaction between dizziness and psychological problems. It also discusses conversion disorders and malingering. Finally, it presents a practical clinical approach to these problems.

Psychological Disorders and Their Prevalence

Dizziness in Patients with Psychological Disorders

Prevalence of Psychological Disorders

The prevalence of psychological problems in the general population is very high. Table 18-1 lists the prevalence in the United States as of 2012.[3]

■ Table 18-1 **PSYCHOLOGICAL DISORDERS IN THE ADULT U.S. POPULATION (National Institute of Mental Health)[3]**

Disorder	No. Affected in U.S.	% Affected in U.S.
Anxiety disorders:	40 million:	18.1:
Phobia (Social and situational)	34.2 million	15.5
Post-traumatic stress disorder	7.7 million	3.5
Generalized anxiety disorder	6.8 million	3.1
Panic with and without agoraphobia	6 million	2.7
Obsessive-compulsive disorder	2.2 million	1.0
Agoraphobia without panic	1.8 million	0.8
Mood Disorder: (Major depressive disorder, dysthymic disorder, bipolar disorder)	20.9 million	9.5

Abnormal Results of Vestibular Tests in Patients with Psychological Problems

To what extent patients with panic disorder have a vestibular defect is controversial. Several authorities have reported abnormal results on a variety of tests used to assess dizziness, including caloric testing, rotary chair testing, vestibular autorotation, and posturography.[4-8] Few articles have sufficient detail to allow determination whether these deficits are truly a result of vestibular dysfunction (high vestibulo-ocular reflex [VOR] gain on rotary chair testing or decreased response on caloric testing). Of these more-detailed articles, one article suggests that as many as 14% of patients with panic but without agoraphobia and 39% of patients with panic and agoraphobia have compensated peripheral vestibular defects.[6] Another article found discrepancies in the VOR but no caloric deficits in patients with panic disorder.[5]

Psychological Problems in Patients with Dizziness

There is a high prevalence of unrecognized mood and psychological problems in dizzy patients, especially anxiety disorders.[9,10] Forty percent of all dizzy patients have psychological disorders.[1] Stein and coworkers[11] reported that 15% of all dizzy patients meet the *Diagnostic and Statistical Manual of Mental Disorders* (DSM-IV) criteria for panic disorder, agoraphobia, or both and that these patients rate themselves as much more disabled by their dizziness than patients with no psychiatric disorder. The prevalence of psychiatric disorders as the primary cause of dizziness declines with increasing patient age.[1,12] In patients older than 60 years, 38% have psychological diagnosis contributing to dizziness, but of these, only 6% were believed to have a primary psychological cause for the dizziness. Psychological problems often coexist with a significant balance disorder.

Patients with and without Peripheral Vestibular Deficits

Forty percent of patients with vestibular hypofunction presenting to an otolaryngology clinic have an additional panic disorder with and without agoraphobia.[13] Fifty percent of patients with vestibular hypofunction evaluated 3 to 5 years after the original referral still had a significant psychiatric disturbance (panic disorder or major depression).[14] In my experience, panic attacks do occur in a number of patients with vestibular deficits, but not to the same extent as reported in these studies conducted by psychiatrists. Chronic anxiety is much more prevalent. The level of anxiety in these patients is usually not high enough to warrant psychotherapy or medication. Patients without evidence of peripheral vestibular deficit have a greater mean number of lifetime psychiatric diagnoses, especially major depression and panic disorder, than those with a vestibular deficit.[10] Patients without vestibular deficits also more frequently have somatization disorders as well as more current and lifetime unexplained medical symptoms.

Disability

Clark and associates[15] evaluated disability in dizzy patients with and without vestibular defects. They found that severe impairment of the ability to function was more strongly associated with the presence of a psychiatric disorder than was the presence of a vestibular disorder. Nausea, vomiting, palpitations, weakness, and difficulty with speech in patients with complaints of dizziness were indicators of a psychiatric disorder and not of a peripheral vestibular disorder. This finding suggests that the clinician should look for comorbid psychiatric disorders in patients with persistent complaints of these symptoms.

Assessment

Scales

A number of questionnaires can be helpful in the diagnosis and assessment of psychological problems. They are discussed here.

Millon Behavioral Medicine Diagnostic

The Millon Behavioral Medicine Diagnostic (MBMD) is an inventory developed in the early 1970s and extensively revised during the 1990s.[16] It is a 165-item, self-report inventory with 29 clinical scales. It was designed to assess psychological factors that can influence the course of treatment of medically ill patients. The MBMD has been validated in physically ill patients and behavioral medicine patients 18 to 85 years old. It takes 20 to 25 minutes to complete and requires a sixth grade reading level (see Case Study 18-1 at the end of the chapter).

Positive and Negative Affective Scale

The Positive and Negative Affective Scale (PANAS) is a good screening scale for anxiety and depression.[17] The patient is given a paper form similar to the representation in Box 18-1. The patient's form does not contain the (P) and (N) labels at the ends of the items, which indicate positive and negative terms.

The numbers assigned to each (P) term are added up. The mean score for (P) is 35.0 ± 6.4.

Depression should be considered if the subject scores less than 22 (2 standard deviations below the mean). The numbers assigned to each (N) term are then added up.The mean score for this term is 18.1 ± 5.9. Anxiety should be considered if the subject scores more than 29.9 (2 standard deviations above the mean).

Dizziness Handicap Inventory

The Dizziness Handicap Inventory (DHI) is a measure of self-perceived disability attributable to vestibular disease. Twenty-five questions classified into physical, functional, and emotional domains are given (Table 18-2).[18] A

Box 18-1

POSITIVE AND NEGATIVE AFFECTIVE SCALE (PANAS)

This scale consists of a number of words that describe different feelings and emotions. Read each item and then mark the appropriate answer in the space next to it. Indicate to what extent you generally feel this way, that is, how you feel on the average. Use the following scale to record your answers.

1	2	3	4	5
very slightly	a little	moderately	quite a bit	extremely

_____interested (P)	_____irritable (N)	_____jittery (N)
_____distressed (N)	_____alert (P)	_____active (P)
_____excited (P)	_____ashamed (N)	_____afraid (N)
_____upset (N)	_____inspired (P)	_____hostile (N)
_____strong (P)	_____nervous (N)	_____enthusiastic (P)
_____guilty (N)	_____determined (P)	_____proud (P)
_____scared (N)	_____attentive (P)	

(N) = negative term; (P) = positive term. The patient's form does not contain these labels. See text for explanation of scoring. Adapted from Watson et al.[17]

■ Table 18-2 ITEMS CONSTITUTING THE DIZZINESS HANDICAP INVENTORY*

P1.	Does looking up increase your problem?
E2.	Because of your problem, do you feel frustrated?
F3.	Because of your problem, do you restrict your travel for business or recreation?
P4.	Does walking down the aisle of a supermarket increase your problem?
F5.	Because of your problems, do you have difficulty getting into or out of bed?
F6.	Does your problem significantly restrict your participation in social activities, such as going out to dinner, movies, dancing, or parties?
F7.	Because of your problem do you have difficulty reading?
P8.	Does performing more ambitious activities, like sports, dancing, and household chores such as sweeping or putting dishes away, increase your problem?
E9.	Because of your problems, are you afraid to leave your home without having someone accompany you?
E10.	Because of your problem, have you been embarrassed in front of others?
P11.	Do quick movements of your head increase your problem?
F12.	Because of your problem, do you avoid heights?
P13.	Does turning over in bed increase your problem?
F14.	Because of your problem, is it difficult for you to do strenuous housework or yard work?
E15.	Because of your problem are you afraid people may think you are intoxicated?
F16.	Because of your problem, is it difficult for you to go for a walk by yourself?
P17.	Does walking down a sidewalk increase your problem?
E18.	Because of your problem, is it difficult for you to concentrate?
F19.	Because of your problem, is it difficult for you to walk around your house in the dark?
E20.	Because of your problem, are you afraid to stay home alone?
E21.	Because of your problem, do you feel handicapped?
E22.	Has your problem placed stress on your relationships with members of your family or friends?
E23.	Because of your problem, are you depressed?
F24.	Does your problem interfere with your job or household responsibilities?
P25.	Does bending over increase your problem?

*E = emotional subscale items; F = functional subscale items; P = physical subscale items.
Adapted from Jacobson and Newman.[18]

"yes" response is scored 4 points, "sometimes" is scored 2 points, and no response is scored 0 points. Thus, the total score ranges from 0 (no perceived disability) to 100 (maximum perceived disability). The developers of the test, Jacobson and Newman,[18] gave the test to 106 consecutive patients seen for vestibular testing. The mean score and standard deviation were 32.7 ± 21.9.

The DHI can be used to identify specific functional, emotional, or physical problems associated with dizziness. No significant correlation has been found between DHI and the results of caloric or rotary chair testing.[18,19] Whether there is a correlation between the sensory aspect of posturography and DHI is controversial.[19,20] Nevertheless, this self-assessment inventory is reliable, requires little time to administer, has a high internal consistency, and may be useful to evaluate the efficacy of treatment.

Disability Scale

Shepard and colleagues[21] developed a disability scale that can be used as a screen to predict whether patients with vestibular defects will improve with vestibular rehabilitation. This scale has been validated for extent of perceived disability in patients with unilateral and bilateral vestibular loss (UVL and BVL, respectively). The patient is asked to pick one statement out of six that best fits how he or she feels (Table 18-3). A score of 4 or higher is correlated with poor outcome from vestibular rehabilitation.

Other Scales

Other standardized questionnaires are listed here. My colleagues and I have used some of them in our clinic, and they may be useful for assessing the effect of certain forms of therapy on clinical outcome.

- Structured Clinical Interview for DSM-IV: A structured interview in which a series of standard questions are asked in standard order, with options to focus on the most relevant points for that patient.[22]
- Beck Anxiety Inventory[23] and the Beck Depression Inventory[24]: Specific screens for anxiety and depression.
- Symptom Checklist-90 (SCL-90-R): A standardized questionnaire for medical, functional, and demographic data; it also contains anxiety, depression, somatization, and phobic anxiety subscales.

Clinical Examination

Examination for Psychogenic Stance and Gait Disorders

On the basis of a review of videotapes from 37 patients with psychogenic balance and gait, Lempert and associates[25] have identified six characteristic clinical features that are useful in the diagnosis of psychogenic stance and gait disorders. Table 18-4 lists these features and their prevalence

■ Table 18-3 **DISABILITY SCALE***

For the following, please pick the *one* statement that best describes how you feel.

Statement	Score
Negligible symptoms	0
Bothersome symptoms	1
Performs usual work duties but symptoms interfere with outside activities	2
Symptoms disrupt performance of both usual work duties and outside activities	3
Currently on medical leave or had to change jobs because of symptoms	4
Unable to work for over 1 year or established permanent disability with compensation payments	5

Patient is given a paper with the statements on it and asked, "Please pick the *one* statement that best describes how you feel." See text for scoring.

Adapted from Shepard et al.[21]

■ Table 18-4 **CLINICAL FEATURES OF PATIENTS WITH PSYCHOGENIC BALANCE AND GAIT DISORDERS**

Feature	Frequency (%)
Moment-to-moment fluctuations in the level of impairment	51
Excessive slowness or hesitation	51
Exaggerated sway on Romberg test, often improved by distraction	32
Uneconomical postures with waste of muscular energy	30
Extreme caution with restricted steps (walking on ice)	30
Sudden buckling of the knees, typically without falling	27

*Adapted from Lempert et al.[25]

in the 37 patients in the Lempert study. This review found psychogenic gait disorders in 9% of their neurological inpatients. In 5 years, 47% had favorable outcomes.[26]

Aphysiological Spasm of Convergence

Patients sometimes demonstrate convergence of the eyes (cross-eyed) while being examined. This is usually associated with pupillary constriction. It is aphysiological (functional) when it is episodic and there are no other ocular motor defects.[27] When other dorsal midbrain abnormalities are present (chronic pupillary changes, upgaze defect), an organic basis should be considered.

Voluntary Nystagmus

Voluntary nystagmus is a type of nystagmus that some individuals can learn to exhibit volitionally.[28] Historically, it was used by young American men to avoid military draft. It should not be confused with flutter. Voluntary nystagmus is commonly associated with vergence eye movements and pupillary constriction.

Dynamic Posturography

A number of studies have found characteristic features on dynamic posturography in patients with psychogenic balance disorders.[29-34] Box 18-2 lists the key patterns in such patients. Figure 18.1 compares the sensory test results of a patient with a psychogenic balance disorder with those of a patient with an acute UVL.

Box 18-2

KEY PATTERNS ON DYNAMIC POSTUROGRAPHY IN PATIENTS WITH PSYCHOGENIC BALANCE DISORDERS

- Substandard performance on sensory tests 1 and 2[29,30]
- Better performance on harder tests (sensory 5 and 6) compared with easier tests (sensory 1–3)[29]
- Large intertrial variability[29]
- Repetitive anterior-posterior sway without falling (voluntary sway)
- Large amplitude (>5 deg) anterior-posterior sway without falling on sensory tests 4–6[29]
- Large amplitude lateral sway (>1.25 deg) on sensory tests 4– 6[29]

Psychological Disorders

Anxiety

Panic Attacks with and without Agoraphobia

Dizziness is the most common symptom in patients with panic disorder, occurring in 50% to 85% of all such patients.[35,36] Panic attacks consist of discrete spells of intense fear or discomfort, in which at least four of the symptoms listed in Box 18-3 develop abruptly and reach crescendo within 10 minutes. Panic attacks can be unexpected (uncued) or situationally bound (cued). An example of the latter is spells induced when entering a car. Agoraphobia is the aversion of open spaces, including leaving the house. Agoraphobia is commonly found in patients with severe panic attacks.

Obsessive-Compulsive Disorder

An *obsession* is a repetitive intrusive thought, impulse, or image that causes marked anxiety. A *compulsion* is a repetitive ritualistic behavior or mental act that aims to reduce anxiety. Examples of the latter include hand washing and checking to see that doors are locked.

Generalized Anxiety Disorder

Generalized anxiety disorder is characterized by generalized and persistent anxiety with motor tension, autonomic

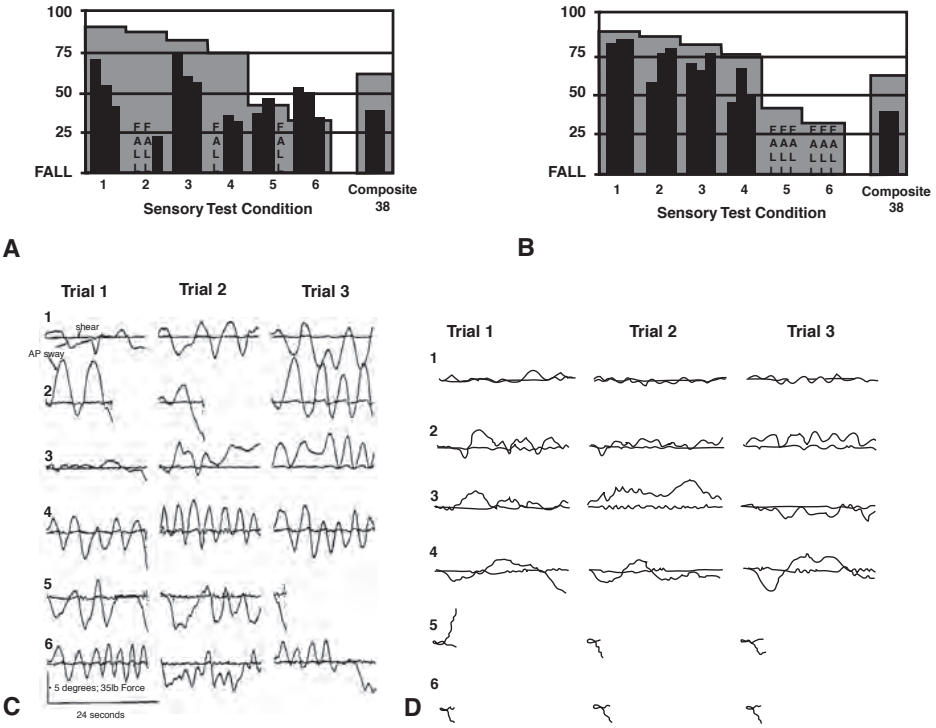

Figure 18.1 Sensory tests during dynamic posturography in a patient with psychogenic balance disorder **(A** and **C)** and a patient with an acute unilateral vestibular loss **(B** and **D)**. **A** and **B** show the mean sway on the 6 sensory test conditions repeated three times (100 indicates no sway; 0 indicates sway beyond level of stability or a fall). The patient with psychogenic balance disorder shows better performance for age on the more difficult tests (5 and 6) compared with easier tests (2 and 4). In addition, there is considerable variability from trial to trial within a given test. The patient with the psychogenic balance disorder also shows a regular frequency of sway on all trials suggesting voluntary control **(B)**. In contrast, the patient with the unilateral vestibular loss has increased difficulty performing the harder tests (5 and 6), has less variability from trial to trial, and does not have a regular periodicity of sway. The *shaded areas* in **A** and **B** indicate the regions where scores are abnormal for age. Note that the two subjects were in the same age bracket and that their overall composite scores were equal.

Box 18-3

SYMPTOMS OF PANIC ATTACK

- Dizziness, unsteady feelings or faintness
- Nausea or abdominal distress
- Shortness of breath (or smothering sensations)
- Palpitations or tachycardia
- Trembling or shaking
- Sweating
- Choking
- Depersonalization or derealization
- Numbness or paresthesias
- Flushes (hot flashes) or chills
- Chest pain or discomfort
- Fear of dying
  - Fear of going crazy or doing something uncontrolled

hyperactivity, apprehensive expectation, and vigilance. An essential feature is unrealistic worry. For the diagnosis, the duration of these symptoms should be at least 6 months (see Case Studies 18-2 and 18-3 at the end of this chapter).

Mood Disorders

Minor depressive disorders have the following criteria: fewer than five of the symptoms listed in Box 18-4 must be present daily for at least 2 weeks.[22] Minor depression is characterized by mood and cognitive symptoms rather than neurovegetative symptoms (anorexia, insomnia, and fatigue). In addition, the patient must have "Depressed mood" or "Loss of interest or pleasure in usual activities, including social contact." The disturbance causes significant impairment in social or occupational functioning, or marked distress. There can be no substance abuse and no manic episodes.

Box 18-4

SYMPTOMS CONSISTENT WITH DEPRESSION (DSM-IV)[22]

- Depressed mood most of the day, nearly every day, as indicated by either subjective report (e.g., feels sad or empty) or observation made by others (e.g., appears tearful)
- Markedly diminished interest or pleasure in all, or almost all, activities most of the day, nearly every day (as indicated by either subjective account or observation made by others)
- Significant weight loss when not dieting or weight gain (e.g., a change of more than 5% of body weight in a month), or decrease or increase in appetite nearly every day. *Note:* In children, consider failure to make expected weight gains.
- Insomnia or hypersomnia nearly every day
- Psychomotor agitation or retardation nearly every day (observable by others, not merely subjective feelings or restlessness or being slowed down)
- Fatigue or loss of energy nearly every day
- Feelings of worthlessness or excessive or inappropriate guilt (which may be delusional) nearly every day (not merely self-reproach or guilt about being sick)
- Diminished ability to think or concentrate, or indecisiveness, nearly every day (either by subjective account or as observed by others)
- Recurrent thoughts of death (not just fear of dying), recurrent suicidal ideation without a specific plan, or a suicide attempt or a specific plan for committing suicide

Somatoform Disorders

Somatization

Somatization is the propensity to experience and report somatic symptoms that have no pathophysiological explanation, to misattribute them to disease, and to seek medical attention for them. Much of general medical practice is devoted to the care of somatizing patients who are symptomatic but not seriously ill.[37]

Conversion

The development of a symptom or deficit suggestive of a neurological disorder that affects sensation (including vestibular) or voluntary motor function (including imbalance) is *conversion*. There is a temporal relationship between the symptoms or deficits and psychological stressors, conflicts, or needs. The patient has no conscious intention of producing the symptoms (i.e., factitious disorder or malingering). The symptoms or signs cause significant impairment in social or occupational functioning or marked distress, or require medical attention or investigation (see Case Study 18-4).

Factitious and Malingering Disorders

Factitious Disorder

Factitious disorder is intentional complaint of psychological signs and symptoms or intentional production of physical signs and symptoms motivated by the psychological need to assume a sick role. For this diagnosis, there must not be external incentives, such as economic gain and obtaining better care.

Malingering

Malingering is similar to factitious disorders, but the malingering patient has an external incentive (usually economic gain) (see Case Study 18-5).

Management

Success in treatment of the psychological problems related to dizziness depends on a positive discussion with the patient. Physicians can have this discussion during the first clinic visit. For therapists, the discussion may be delayed until the patient is well into the course of rehabilitation, when a good rapport has been established. An excellent approach has been outlined by Bursztajn and Barsky.[38]

If the opportunity arises, the clinician should:

- Discuss how emotions and stress can cause the same types of symptoms as a vestibular disorder
- Try to eliminate any stigma or low self-esteem by stating the prevalence of the psychological disorder
- Assure the patient of the clinician's continuing interest and involvement

A statement about a possible psychological problem should be included in the clinician's notes to the referring physician. The clinician should never merely tell a patient that he or she needs to see a psychiatrist. Patients object to the suggestion of a psychiatric referral for several reasons, including the social stigma of being a psychiatric patient, the creation of low self-esteem, a poor understanding of the role of emotions in causing symptoms, and a feeling

of rejection. Using the term *psychogenic* to describe the problem, which is diagnostically neutral, may be preferred to "functional" or "hysterical" (Box 18-5).

Medications

Physicians may want to start the patient on medication, as listed in Table 18-5. The agents listed as daily medication are nonaddictive, take up to 3 weeks to become effective, and can be taken for years. These medications are all antidepressants and are approved by the U.S. Food and Drug Administration (FDA) for treatment of anxiety. Paroxetine (Paxil) and sertraline (Zoloft) should be given in the morning, because they can impair sleep. Agents listed as intermittent medications are addictive, can cause sedation, act immediately, and are strictly for anxiety. For patients with severe chronic anxiety or panic attacks, I usually start two drugs, one from the intermittent medication group and one from the daily medication group. I usually use the lowest dose listed in Table 18-5. After 3 weeks, I either taper the intermittent medication completely or limit its use to 1 or 2 days per week at most.

Box 18-5

SUMMARY OF MANAGEMENT

- Recognize the type of psychological disorder to determine whether you can begin or recommend treatment.
- If you believe the patient does have a psychological disorder that will respond to treatment, do the following:
  - Discuss the problem in clinic, discuss model, other patients, and reassure the patient.
  - Have the patient start a home exercise program.
  - When appropriate, refer the patient for medication and/or counseling.

■ Table 18-5 **MEDICATION FOR CHRONIC ANXIETY**

Medication	Dose	Half-Life (hr)
Intermittent medication:		
Xanax (alprazolam)	0.2–1 mg tid	11–15
Ativan (lorazepam)	0.5–5 mg bid	10–20
Klonopin (clonazepam)	0.5–10 mg bid	18–50
Valium (diazepam)	2–10 mg bid	20–50
Daily medication:		
Paxil (paroxetine hydrochloride)	10–20 mg daily in morning	
Zoloft (sertraline hydrochloride)	50–100 mg daily in morning	
Tofranil (imipramine)	10–75 mg/day	
Norpramin (desipramine)	50–100 mg/day	

CASE STUDY 18-1

A 54-year-old woman is referred for evaluation before removal of a small left vestibular schwannoma totally within the internal auditory canal. Her major complaints are listed here in decreasing order of severity as judged by the patient: the most severe problem is poor balance. She still depends on a cane. She even sometimes uses a walker, when she is tired. She has fallen twice in the house without injury. Coupled with that is head fullness, which increases whenever she does vestibular enhancement exercises. She feels as though she has severe fullness in the head. A third complaint is of poor stamina.

Clinical examination shows decreased VOR for head thrust to left- and right-beating nystagmus after horizontal head shake. Her neurological findings are otherwise normal except for gait, which is very slow with weaving to the left and right. Caloric testing shows 80% decrease on the left; dynamic visual acuity (DVA) is normal for age. Rotary chair testing shows normal gain during constant velocity steps at 60 degrees per second (deg/sec) and decreased gain to the left at 240 deg/sec chair rotations. Rotary chair testing also shows low gain and high phase for low-frequency sinusoidal chair rotation but normal values

CASE STUDY 18-1

at high-frequency rotations. The patient's Millon Inventory is shown in Figure 18.2.

Comment

This patient shows evidence of good central compensation of a left vestibular defect based on DVA and rotary chair testing (see Chapter 11 for more detail).This is what is expected in the majority of individuals with vestibular schwannomas, because these tumors are slow-growing, allowing good central compensation. Her history reveals more subjective complaints than would be expected. The Millon Inventory result suggests that she may ha ve a functional defect (aphysiological) and may not be comfortable discussing her medical problems or how she is coping. Her gait is consistent with an aphysiological component.

This patient underwent tumor remo val, and her vestibular rehabilitation took three times longer than average. She was referred to counseling, but she did not believe she had a problem coping so did not gain much benefit from it.

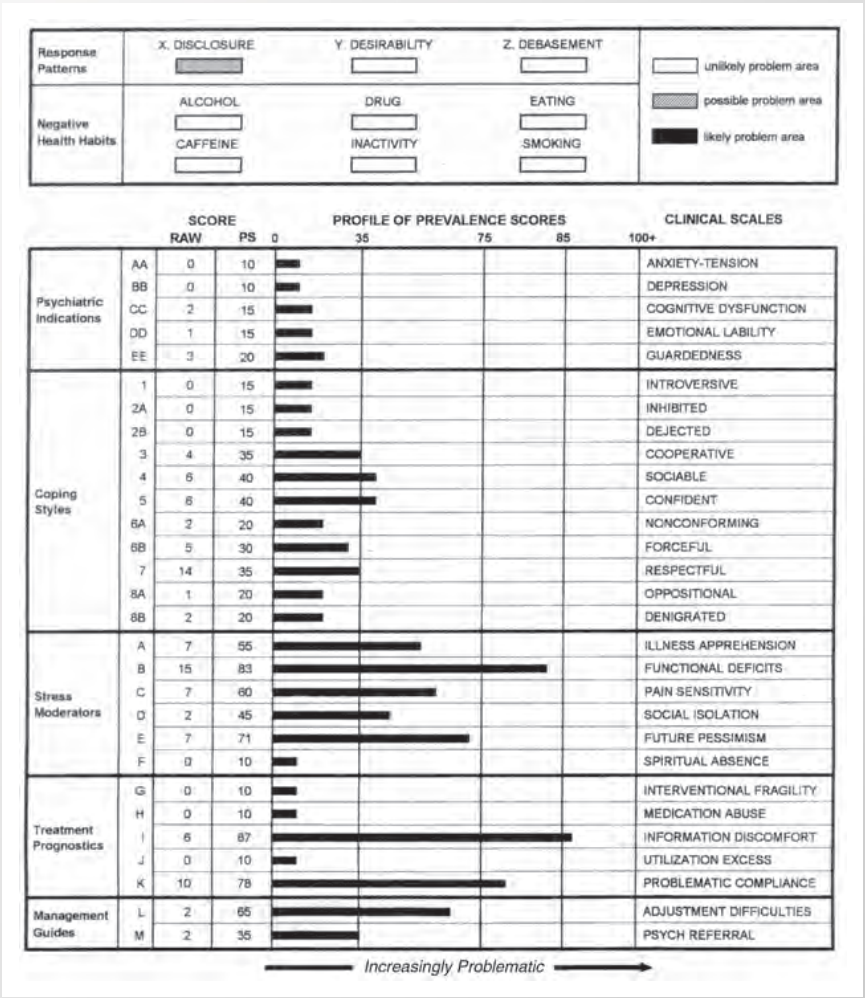

Figure 18.2 Sample of the Millon Inventory in a dizzy patient.

CASE STUDY 18-2

A 39-year-old man experienced severe nausea and vertigo 7 months ago when he put his head down in bed after a trip to a theme park. Vertigo persisted for a few days, resolved, and then reoccurred 2 weeks later accompanied by shortness of breath, palpitations, trembling, and chest pain. These symptoms were so incapacitating that he was hospitalized for a few days for "tests." MRI of the head and neurological examination results were normal. Since then, the patient has had chronic dizziness. He also now has head pressure, decreased energy, decreased weight, trouble sleeping, and apprehension. He stopped all exercise after the onset of dizziness. Findings of the current examination are entirely normal, as are all vestibular test results.

Comment

This patient exemplifies chronic anxiety with features of panic attacks. There was no evidence on clinical examination nor on vestibular function testing of vestibular hypofunction. His original problem was probably BPPV and anxiety. His management now focused on reassurance. He also was prescribed Xanax, 0.25 mg daily at bedtime × 3 weeks and Paxil, 10 mg daily in morning and told to restart his exercise program.

CASE STUDY 18-3

A 27-year-old electrician complains of "dizziness for the past 2 years." He uses trouble walking, poor balance, linear movement, tilt, floating, rocking, and blurred vision all as equal terms for his dizziness. The problem started while he was working on a high lift for 2 hours. He was at a height that caused a sense of rocking on the platform. In addition, he attributes his dizziness to inhalation of fumes of a floor sealant on the job. Since then, he has constant dizziness, which is severe when he first awakens in the morning. It is also severe when he is fatigued or is walking in a dark room. His head feels heavy. He denies vertigo, hearing loss, and tinnitus. Because of his symptoms, he has reduced his exercise program. In the last 6 months, he has experienced loss of strength and energy, memory loss, paresthesias, muscle and joint aches, trouble sleeping and speaking, tremor, incoordination, and headaches.

The patient's past medical history includes surgery to the knee years ago that required intravenous antibiotic therapy, gonorrhea treated with antibiotics, and anxiety and panic attacks 2 years ago. His mother has been on long-term benzodiazepine therapy for stress. In the last 6 months, the patient's dizziness has interfered with his activities 95% of the time, and currently, it is moderately intense. It has markedly changed his ability to work or do household chores. Dizziness has severely decreased the amount of satisfaction or enjoyment the patient finds from taking part in family-related or social activities. On the PANAS, he scores 34 on positive affect and 19 on negative affect, which are normal.

The physical findings are normal, including a visual acuity of 20/26-2 static, 20/30-1 dynamic, normal VOR gain to head thrust, no head-shaking or positional nystagmus, normal pursuit and saccades, normal hearing, and normal gait and balance. He has already undergone MRI with and without gadolinium that included eighth nerve "cuts" and a caloric test. Results of both were normal.

Comment

This patient has several features consistent with chronic anxiety. His complaints are vague, numerous, and out of proportion to the findings. Complaints of floating and rocking are typical in patients with anxiety or depression. He has a history of panic attacks 2 years ago. There is likely a family history of anxiety, as his mother has been on benzodiazepines for "stress." Patients with anxiety commonly have a family history of stress, anxiety, or nervousness. Exercise is an excellent stress reducer; the patient stopped all exercise 2 years ago. Even though the PANAS score is useful, it is not positive for anxiety in this patient.

A tentative diagnosis of chronic anxiety is made. The symptoms from stress and anxiety are discussed with the patient. He is told that these symptoms are very real and can be extreme. The role of his past medical and family history for anxiety is also discussed. He

CASE STUDY 18-3

is encouraged to restart a regular exercise program to help reduce stress. He is started on Paxil, 10 mg daily in the morning, and Klonopin, 0.5 mg daily in the evening; the side effects of each agent are explained. He is asked to return in 3 weeks.

When he returns to the clinic, most of his symptoms have resolved. He is exercising on a regular basis. The Klonopin dosage is tapered over a 3-week period, but the Paxil is continued for 1 year.

CASE STUDY 18-4

A 12-year-old girl is brought to the physician by her mother because of inability to walk for 2 weeks. She can take only a few steps before she has to sit down or her knees buckle. She started attending a new school 3 weeks before her illness. She was doing very well until the dizziness started. There is an older daughter who is excelling in the same school. The patient has undergone head CT and audiography; the results of both were normal. She had a positive tilt table response to isoproterenol, suggesting possible orthostatic hypotension, and was started on medication and salt tablets; this may have initially helped but for only a week. Her physical findings are normal except for her stance and gait; there is significant sway at the hips with eyes open and closed, but she does not fall. While walking, she has sudden buckling of the knees but is still able to walk. There is much side-to-side swaying and waste of muscular energy.

Comment

This patient has a conversion disorder. She has a deficit that suggested a neurological disorder, but no disorder was found. She has a psychogenic stance and gait disorder with several of the features described in Table 18-4. Her symptoms began temporally with the stress of starting a new school—the same school attended by her overachiever sister. There is no evidence of external economic gain, as one would expect for malingering. She does not have a history of assuming a sick role motivated by psychological need, as one would expect for a factitious disorder. As in several cases of conversion disorder, this case prompted extensive evaluations and an organic diagnosis (orthostatic hypotension) that proved later to be wrong. The tilt table test, especially with isoproterenol, has a number of false-positive results.

The diagnosis of conversion disorder is not discussed with the girl. It is discussed with the mother. The social problems with starting a new school attended by an overachieving sister are discussed. School counseling is recommended.

The patient's gait disorder slowly resolves after she is switched to a different school (i.e., one not attended by her sister). Medications are not used.

CASE STUDY 18-5

A 40-year-old man fell off a scaffold and hit the right side of his head 10 months ago. He is referred as part of a Workman's Compensation case. During the accident, he had brief loss of consciousness. Initially, he had positional vertigo while getting out of bed (BPPV), which was treated. Now he has poor balance. Clinical findings, including those of vestibular and neurological examinations, are normal except for gait. Dizziness interferes with his activities 100%, resulting in an extremely changed ability to work or do household chores. The PANAS score is normal, as is the caloric test result.

Comment

This patient has a functional (aphysiological) gait (see Table 18-4). He shows no evidence for anxiety or depression based on the PANAS scale. There is secondary gain, in that he does not have to work while his case is reviewed. The physician may want to give him limited gait and balance physical therapy (4 sessions) to see whether he responds, but there is a high probability that he will not improve with PT at this time.

Summary

Psychological problems, especially anxiety, are a major contributing factor in all patients with dizziness. The therapist needs to recognize and deal with this f act for a good outcome. Treatment usually requires simply patient education and reassurance by the referring physician or therapist. Some patients need medication or stress-reducing programs. If a patient is not progressing during therap y as expected, the therapist should consider a signif icant psychological problem. Reevaluation by the physician may be indicated.

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CHAPTER 19

Management of the Patient with Chronic Subjective Dizziness

Jeffrey P. Staab, MD, MS ■ *Neil T. Shepard, PhD*

For more than 150 years, the medical literature has contained descriptions of patients with chronic dizziness and motion sensitivity, accompanied by varying degrees of anxiety and phobic behaviors. In 1871, Karl Westphal, a German neurologist, coined the term agoraphobia to describe a syndrome of dizziness, spatial disorientation, and anxiety experienced by individuals in the busy open marketplaces of 19th century European villages. [1] Westphal considered postural control, spatial orientation, and threat assessment to be fully integrated components of locomotion, a view that was lost as clinical neuro-otologic and psychiatric practice diverged through the 20th century. The term agoraphobia was linked to panic attacks as a primary psychiatric diagnosis. In the 1980s and 1990s, neuro-otologists and psychiatrists renewed their interest in interactions among vestibular and anxiety disorders. Brandt and Dieterich described phobic postural vertigo (PPV) in the German medical literature in 1986 [2] and in the English medical literature a decade later.[3] They defined PPV as a syndrome of postural dizziness and fluctuating unsteadiness provoked by environmental or social stimuli (e.g., crossing bridges; descending staircases; traveling on busy streets or through crowds). They reported that PPV could be triggered by vestibular disorders, medical illnesses, or psychological stress. [4] They included two behavioral criteria in their original definition of PPV: (1) obsessive-compulsive personality traits, labile affect, and mild depression; and (2) anxiety and vegetative disturbance.[2] Brandt, Dieterich, and colleagues found PPV to be the second most common cause of vestibular symptoms in their university-based neuro-otology practice, behind only benign paroxysmal positional vertigo (BPPV).[3] Diagnostic studies differentiated PPV from panic disorder, agoraphobia as currently defined in modern psychiatric nomenclature, and other psychiatric disorders, suggesting that it is a primary neuro-otologic condition with behavioral elements.[3] Huppert and colleagues[5] found that PPV could be diagnosed quite reliably. They contacted 105 patients who were diagnosed with PPV 5 to 16 years earlier (average 8.5 years) and reaf firmed the diagnosis in all patients. No patients had been misdiagnosed, and none had developed other neurological or vestibular conditions. Two other studies showed that the long-term course of PPV was marked by chronic dizziness and unsteadiness that waxed and waned over time. Symptoms tended to decrease gradually, but rarely resolved.[5] Curiously, Westphal reported this same pattern in his reports in the 1870s. [1] Behavioral outcomes of PPV were not as good. Two-thirds of patients developed clinically significant anxiety or depression.[3] Finally, recent studies found that behavioral factors may affect the natural course of PPV and outcomes of treatment.[6,7]

In the early 2000s, Staab and Ruckenstein defined chronic subjective dizziness (CSD),[7,8] which had three features: (1) persistent nonvertiginous dizziness lasting 3 months or more; (2) hypersensitivity to motion stimuli, including a patient's own movement and motion of objects in the visual surround; and (3) difficulty with precision visual tasks such as reading or using a computer The definition of CSD was influenced not only by the original concept of PPV, but also by the work of Jacob and colleagues[9] on space-motion phobia and Bronstein and colleagues[10] on visual vertigo. Subsequent studies investigated the relationships between CSD and other neuro-otologic and psychiatric conditions.[7,11,12] This research found that the behavioral criteria included in the definition of PPV [2] (e.g., obsessive compulsive personality traits, mild depression and anxiety) were predisposing factors and behavioral comorbidities, respectively, not essential components of CSD. More specifically, anxious, introverted personality traits may predispose individuals to developing CSD, and anxiety or depressive symptoms may coexist with CSD, but they are not core elements of the syndrome. [13] Additionally, investigators in the United States and Japan completed successful open-label medication trials for CSD showing that selective serotonin reuptake inhibitors (SSRIs) could be used to treat the disorder.[7] Other investigators studied rehabilitative and psychotherapeutic treatments. They observed that the habituation form of vestibular rehabilitation could be an effective intervention for CSD. Psychotherapy yielded mixed results. [14] For patients with long-standing CSD, researchers in Europe and the United States were able to demonstrate short-term reductions in dizziness using cognitive-behavioral therapy (CBT),[15,16] but improvements in symptoms were not sustained in long-term follow-up. [17] For patients with new onset CSD-type symptoms, cognitive-behavioral therapy may have more enduring benefits.[18]

Current Formulation of Chronic Subjective Dizziness

The clinical elements of CSD are organized in Table 19-1. They are separated into primary symptoms, relationship to posture, exacerbating factors, precipitating events, examination and laboratory findings, and behavioral factors. A validation study of this structure is under way at Mayo Clinic.[19] Initial findings from that study support the description given in Table 19-1.

Core Symptoms

The primary symptoms of CSD are persistent, nonvertiginous dizziness or swaying/rocking unsteadiness. For most patients, these symptoms are present throughout the day, though they may wax and wane in severity. Asymptomatic periods may occur. CSD often lasts for months or years. It may remit spontaneously, but when it does, resolution is slow and gradual. [5] The unsteadiness experienced by patients with CSD is subjective. In cases in which unsteadiness is observable on clinical examination, the possibility of comorbid neuro-otologic illnesses (e.g., bilateral vestibular deficits, movement disorders) or behavioral disorders (e.g., phobia of falling, functional gait disorder) must be considered.

Relationship to Posture

A postural criterion was not part of the original definition of CSD,[8] but the core symptoms of dizziness and unsteadiness are usually worse when sitting, standing, or walking than when recumbent. Postural symptoms occur when patients are upright, but not when they are reclining. These are to be distinguished from orthostatic and positional symptoms. As defined by the International Committee for Classification of Vestibular Disorders,[20] orthostatic symptoms occur during or immediately after patients arise from a recumbent posture (e.g., during or immediately after standing up). Positional symptoms occur when the head or body moves into or through a specific orientation in space. Postural symptoms are so common in CSD that the diagnosis should be reconsidered (though not absolutely excluded) if patients report no increase in vestibular symptoms when upright. Patients with CSD may have orthostatic or positional problems, but these should be part of a generalized pattern of sensitivity to motion in all directions.

Provocative Factors

The third row of Table 19-1 lists additional factors that may provoke CSD symptoms. These include head or body motion that is not direction-specific and visual stimuli such as large-field visual flow, complex patterns, or performance of precision visual tasks. These provocative factors may not seem to be unique to CSD, but there are important distinctions between the responses of patients with CSD and those with other neuro-otologic disorders. Patients with Ménière's disease, vestibular migraine, and other vestibular disorders find head movements and environmental motion stimuli to be quite troublesome during acute attacks of their illnesses but much less problematic or not bothersome at all during interictal periods. Patients with most neuro-otologic conditions can reduce their symptoms by holding still or removing themselves from motion-rich environments. In contrast, patients with CSD are likely to

■ Table 19-1 CLINICAL FEATURES SHARED BY PHOBIC POSTURAL VERTIGO AND CHRONIC SUBJECTIVE DIZZINESS

Feature*	Description	Comments
Primary symptoms	Unsteadiness, dizziness, or both are typically present throughout the day but fluctuate in severity. Symptoms are present on most days for 3 months or more.	Symptoms of phobic postural vertigo (PPV) and chronic subjective dizziness (CSD) are usually quite persistent but wax and wane spontaneously and in response to provocative factors. Vertigo is not part of PPV or CSD, but CSD may coexist with other vestibular disorders. In those cases, patients may experience episodic vertigo superimposed on chronic unsteadiness and dizziness.
Postural relationship	Primary symptoms are related to body posture. Symptoms are most severe when walking or standing, less severe when sitting, and absent or very minor when recumbent.	Some patients with CSD prefer walking to standing still, although either is more troublesome than sitting or lying down. Postural and orthostatic symptoms are not the same. Postural symptoms are present while patients are in upright postures. Orthostatic symptoms occur as patients arise into an upright posture. Orthostatic tremor develops while standing and improves during walking.
Provocative factors (context-dependent symptom exacerbation)	Primary symptoms are present without specific provocation but are exacerbated by the following: 1. Active or passive motion of self that is not related to a specific direction or position 2. Exposure to large-field moving visual stimuli or complex (fixed or moving) visual patterns 3. Performance of small-field, precision visual activities (e.g., reading, using a computer, fine tasks with hands)	Symptoms of CSD exist without provocation but usually reflect the cumulative burden of exposure to provocative activities throughout the day. Context-dependent factors include motion of self, exposure to environments with challenging motion stimuli, or complex visual cues and performance of visual tasks that require precise, sustained focus, such as using a computer or reading.
Precipitating factors (triggering events)	Precipitating factors include: 1. Acute or recurrent neuro-otologic diseases that cause central or peripheral vestibular dysfunction 2. Acute or recurrent medical problems that produce unsteadiness or dizziness 3. Acute or recurrent psychiatric disorders that produce unsteadiness or dizziness	The most common triggers for CSD are: • Previous acute vestibular disorders (e.g., benign paroxysmal positional vertigo, vestibular neuritis) • Episodic vestibular disorders (e.g., vestibular migraine, Ménière disease) • Mild traumatic brain injury or whiplash • Panic attacks, generalized anxiety • Dysautonomias • Dysrhythmias • Adverse drug reactions and other medical events

Continued

■ Table 19-1 CLINICAL FEATURES SHARED BY PHOBIC POSTURAL VERTIGO AND CHRONIC SUBJECTIVE DIZZINESS—cont'd

Feature*	Description	Comments
Physical examination and laboratory findings	Physical examination and vestibular laboratory testing are often normal. Minor, nondiagnostic abnormalities occur frequently. Examination and testing may reveal diagnostic evidence of a neuro-otologic or other medical condition that may be active, treated, or resolved but cannot fully explain all of the patient's symptoms.	CSD may occur as an isolated condition or coexist with other neuro-otologic or medical illnesses. Positive examination findings do not necessarily exclude CSD. They may instead identify comorbid conditions. At present, there are no established biomarkers for CSD, but emerging data suggest that patients may have a unique pattern of sway on static or dynamic posturography with relatively poorer performance on simpler tasks.
Behavioral symptoms	Behavioral assessment may be normal. Low levels of anxiety or depression are common. Behavioral assessment also may find clinically significant psychological distress, psychiatric disorders, or adverse changes in activities of daily living.	Several carefully designed diagnostic studies have shown that CSD is a unique clinical entity, not a *forme fruste* of a psychiatric illness (see text). However, patients with CSD do have an increased prevalence of psychiatric disorders, typically anxiety or depressive disorders, compared with individuals with other neuro-otologic illnesses.

* The first three features may be used to make a diagnosis of CSD. The clinical history would include 3 months or more of persistent, posture-related unsteadiness or dizziness provoked by motion of self and one or both of the visual stimuli.

experience prolonged exacerbations of unsteadiness and dizziness lasting for hours or days even after minimizing or eliminating exposure to provocative stimuli. In fact, the severity of CSD symptoms at a given time reflects the accumulated exposure to motion stimuli over the preceding hours or days, which makes patients continually vulnerable to further exacerbations.

- Severity of CSD symptoms at a given time reflects the accumulated exposure to motion stimuli over the preceding hours or days.
- The prevalence of difficulties with visual stimuli was significantly greater in patients with CSD (80%) than in patients with other disorders (20%).
- Vestibular neuritis, BPPV, and other peripheral vestibular disorders triggered more cases of CSD than central vestibular conditions.
- In most cases, CSD has a sudden onset, starting after a single event.
- The diagnosis of CSD should not be confused with, or attributed to, psychiatric morbidity.

Patients with psychiatric disorders, such as panic disorder, agoraphobia, and specific phobia of dizziness, also experience increased symptoms with provocative stimuli. Here, too, there are important dif ferences between patients with CSD and individuals with these psychiatric disorders. Fear is not a prominent part of CSD. Patients with anxiety disorders avoid provocative situations, because they fear adverse consequences, such as becoming incapacitated or causing a scene in public. In contrast, individuals with CSD avoid provocative situations to limit exposure to noxious physical sensations. Preliminary results from the Mayo Clinic validation study of CSD[19] are beginning to clarify these dif ferences. Most (75% to 98%) patients with active neuro-otologic conditions reported sensitivity to self-motion as part of their illnesses, although patients with CSD had prolonged unsteadiness or dizziness rather than time-limited vertigo experienced by those with the other disorders. Furthermore, the prevalence of difficulties with visual stimuli was significantly greater in patients with CSD (80%) than in patients with other disorders (20%)

and these visual symptoms listed in Table 19-1 appeared to be quite sensitive and specific for CSD.

The first 3 rows of Table 19-1 may be used clinically to identify patients with CSD. An individual with 3 months or more of nonvertiginous dizziness or unsteadiness that is postural in nature and exacerbated by his or her own movements, exposure to large-field visual stimuli, or performance of small-field precision visual tasks may be given the diagnosis.

Precipitating Events

In two large studies of CSD,[8,19] the most common triggering events, accounting for 25% of all cases, were previous acute vestibular disorders. Vestibular neuritis, BPPV, and other peripheral vestibular disorders triggered more cases of CSD than central vestibular conditions, most likely because they are more prevalent disorders. Other precipitants included panic attacks, especially in young adults (15% to 20%); vestibular migraine, more likely in women (15% to 20%); generalized anxiety (15%); mild traumatic brain injury (concussion or whiplash), especially in young men (10% to 15%); and dysautonomias (7%). Other medical problems, such as dysrhythmias and adverse drug reactions, accounted for about 2% of cases. These problems seemed to trigger CSD because they caused acute bouts of vertigo, dizziness, or disruption of gait and stance.

In most cases CSD has a sudden onset, starting after a single event. It also may begin with recurrent bouts of self-limited symptoms that consolidate into a persistent pattern. Most patients are able to identify one or more precipitating events, such as a single bout of vestibular neuritis or recurrent episodes of vestibular migraine or panic disorder. CSD rarely has an insidious onset. Although such cases occur, this presentation warrants reevaluation of the clinical history with particular attention to progressive vestibular or degenerative neurological conditions.

Medical Comorbidity

The original definition conceptualized CSD as a sequela of a triggering medical or psychological event, not as a condition that might coexist with other illnesses.[8] This was an error. Clinical experience and initial data from the Mayo Clinic validation study of CSD [19] indicate that the syndrome coexists with a wide variety of other disorders, including episodic and chronic neuro-otologic disorders, ranging from Ménière's disease to stroke-related, central vestibular deficits. As a result, the original diagnostic criterion about normal or insignificant physical examination and laboratory testing has been revised as shown in the fifth row of Table 19-1. Evidence of active neuro-otologic deficits on physical examination or vestibular laboratory testing no longer excludes a diagnosis of CSD, but identifies coexisting conditions.

Behavioral Predisposition and Comorbidity

In addition to their investigations of the core symptoms and precipitants of CSD, Staab and Ruckenstein studied the role of behavioral factors in its development and clinical course. This research coincided with investigations by other clinical scientists[6,21-23] to identify three ways the behavioral factors contribute to CSD.

- First, individuals with anxious, introverted temperaments or preexisting anxiety disorders may be particularly predisposed to developing CSD.
- Second, CSD may be more likely to occur in individuals who have high levels of anxiety, hypervigilance about vestibular symptoms, and excessive worry about potentially adverse outcomes during an acute bout of a neuro-otologic illness or other triggering event.
- Third, comorbid anxiety and depression occur in many, but not all, patients with CSD, adding considerable morbidity.

This last point is important in making accurate clinical diagnoses. Studies from Germany,[3] Sweden,[24] and the United States,[17] showed that 60% of patients with CSD had clinically significant anxiety and 45% had clinically significant depression. These rates were higher than those encountered in similar studies of patients with Ménière's disease, vestibular migraine, or BPPV, which found psychiatric comorbidity rates of 35% for anxiety and 20% for depression.[19,25,26] However, 25% of patients with CSD do not have significant psychiatric symptoms, and the core elements of CSD do not differ in patients with or without anxiety or depression. Therefore, the diagnosis of CSD should not be confused with, or attributed to, psychiatric morbidity.

Pathophysiological Mechanisms of CSD—Old and New Hypotheses

The Old Idea

Both Brandt and Dieterich, in defining PPV,[3] and Staab and Ruckenstein, in defining CSD, [7] hypothesized that classical and operant conditioning were the principal pathophysiological mechanisms for this disorder. Acute vestibular events were thought to be particularly potent unconditioned stimuli, because they produce strong physiological responses that sensitize (condition) postural and ocular motor reflexes to subsequent exposures to internal

or external motion stimuli. This was thought to trigger a heightened awareness of postural control, thereby reinforcing (classically conditioning) hypersensitive postural reflexes. At the same time, operant conditioning reinforces safety behaviors such as avoidance of provocative situations and use of safety aids (e.g., going out with a trustworthy companion). Anxiety that accompanies the triggering events may augment these conditioning processes. Catastrophic thoughts and dysphoric ruminations about the potential consequences of vestibular symptoms (e.g., crashing the car or becoming handicapped) were thought to perpetuate CSD and increase the likelihood of developing comorbid anxiety or depression.

A New Concept

The hypothesis that CSD is caused by classical and operant conditioning is based on well-established behavioral and cognitive theories, but no data specifically support it. Rather, several lines of evidence suggest a different mechanism, albeit one that still involves threat systems in the brain. CSD almost always starts with an acute process, such as an episode of a neuro-otologic disorder, other medical event, or panic attacks with prominent dizziness. Occasionally, patients will describe insidious onset, but that is uncommon. This means that nearly all triggers for CSD require patients to adapt quickly to an acute disruption of safe and secure mobility. During the early phases of acute events, such as the sudden onset of vestibular neuritis, patients must suppress input from damaged sensory systems and shift preferences to intact sensory systems to maximize detection of accurate spatial orientation cues. Patients must also shift into high-risk postural control strategies (e.g., moving cautiously, using supports) and maintain a greater degree of vigilance about the environment. These changes are adaptive in the short run, but should return to normal when no longer needed (e.g., as vestibular compensation develops). A similar pattern of shifting into high-risk postural control strategies followed by return to normal would be expected in response to episodic vestibular illnesses such as Ménière's disease or vestibular migraine. In these cases, the cycle of response would occur repeatedly. CSD may be caused by panic attacks or other intense stress reactions. These situations also may disrupt mobility, necessitating a similar shift out of and back into normal postural control strategies. Regardless of the nature of triggering events, CSD is now conceptualized as either persistent failure to readapt after the initial impairment to posture, balance, and oculomotor control resolves or persistent use of high-risk balance control strategies that are out of proportion to what is necessary to compensate for chronic neuro-otologic deficits. High anxiety during the early phase of balance disruption seems to impair the readaptation process, making it more likely that CSD will develop, possibly compounded by anxiety or depressive disorders.

CSD is now conceptualized as either persistent failure to readapt after the initial impairment to posture, balance, and oculomotor control resolves or persistent use of high-risk balance control strategies that are out of proportion to what is necessary to compensate for chronic neuro-otologic deficits.

The effect of anxiety on balance control strategies has been investigated in normal individuals and patients with CSD. Anxious individuals and patients with CSD do not tolerate postural disturbances as well as normal individuals. For example, Carpenter and colleagues,[27] found that anxious but otherwise normal subjects responded more quickly to rotational perturbations while standing on a posture platform than less-anxious subjects. They employed a stiffer and more reactive postural control strategy in which they made more postural corrections resulting in narrower postural displacements than their more relaxed counterparts. In a separate study,[28] patients with PPV had a similar stiff, highly reactive postural response to visual vection stimuli while standing on a posture platform that produced smaller displacements than normal subjects.

Anxious patients employ several features of high-risk postural control strategies in situations in which they are not needed.[29] One of these is co-contraction of antigravity muscle groups, which changes the natural sway pattern during steady stance to one with a higher frequency, but lower amplitude of movement. This causes anxious individuals to perform worse than normal subjects on simple postural tasks that do not require high-risk postural control strategies. In situations in which higher risk strategies are needed, anxious and non-anxious individuals perform similarly. For example, healthy college students with high anxiety swayed more than their less-anxious classmates while standing on a posture platform with eyes open but not with eyes closed. Both groups swayed more with eyes closed, but between-group differences were more noticeable during the easier task.[29] These studies show that threat systems in the brain instinctively alter balance performance, depending on the context of the posture control task and anxious state of the individual. Patients with CSD appear to have a reduced tolerance for postural disturbances, including motion of their bodies and movement in the visual surround. They make more high-frequency, low-amplitude postural corrections (a high-risk postural control strategy) in routine balance situations when that is not necessary.

The longitudinal development of CSD-type symptoms has been observed in two prospective studies of patients enrolled within a few days of acute vestibular events (vestibular neuritis or BPPV).[22,23] Thirty percent of subjects reported persistent dizziness or unsteadiness at 3[23] and 12[22] months after their acute vestibular illnesses. Importantly, chronic dizziness persisted despite the fact that 90% of symptomatic patients demonstrated full compensation or recovery from their peripheral vestibular deficits. Acute behavioral responses distinguished patients who developed chronic dizziness from those most likely to recover. In particular, high levels of anxiety at the onset of vertigo, excessive vigilance about vestibular symptoms, and catastrophic thinking about possible adverse outcomes were the best predictors of persistent dizziness and unsteadiness.[22] Two smaller investigations took these studies a step further by measuring postural sway in addition to anxiety and physical symptoms during recovery from acute vestibular neuritis.[30,31] Patients who had comorbid generalized anxiety swayed more than those without anxiety when standing with eyes closed or when watching an optokinetic stimulus moving toward their damaged ear.[31] The postural sway pattern during the acute period of illness had frequency content primarily in the vestibular range (i.e., dominated by the abnormal vestibular signals). Symptomatic recovery coincided with the appearance of a sway pattern with a broader range of frequencies, indicative of a return to a more flexible postural control strategy making use of multiple sensory inputs. Patients who retained the acute vestibular sway pattern reported chronic dizziness throughout the 9-month study. These longitudinal studies suggest that high anxiety magnifies postural instability and reactivity to motion stimuli during acute vestibular events and then inhibits readaptation to postural control strategies that are associated with recovery.

Figure 19.1[32] is a schematic of this new hypothesis. A triggering event induces a necessary switch to a high-risk postural control strategy. Under normal circumstances, vestibular, medical, and behavioral recovery (i.e., readaptation) take place over a time course determined by the nature of the triggering event. In the presence of predisposing factors such as an anxious, introverted temperament or preexisting anxiety disorders, the re-adaptation process is disrupted. Instead of recovery, the system enters a perpetual loop in which continued use of high-risk postural control strategies maintains a heightened reactivity to motion stimuli, making previously benign situations highly provocative. Predisposing factors also increase the risk of behavioral comorbidity. In this integrated model of CSD, behavioral and neuro-otologic factors are equally important elements of the pathophysiological mechanisms. Without their interaction, CSD would not exist. Thus, CSD is a condition that lies at the interface between postural control and threat systems in the brain. Neuroanatomical links between these systems have been identified from cortex to brainstem.[33] In health, they support safe and efficient postural control. In illness, they seem to be the neural substrate for CSD.

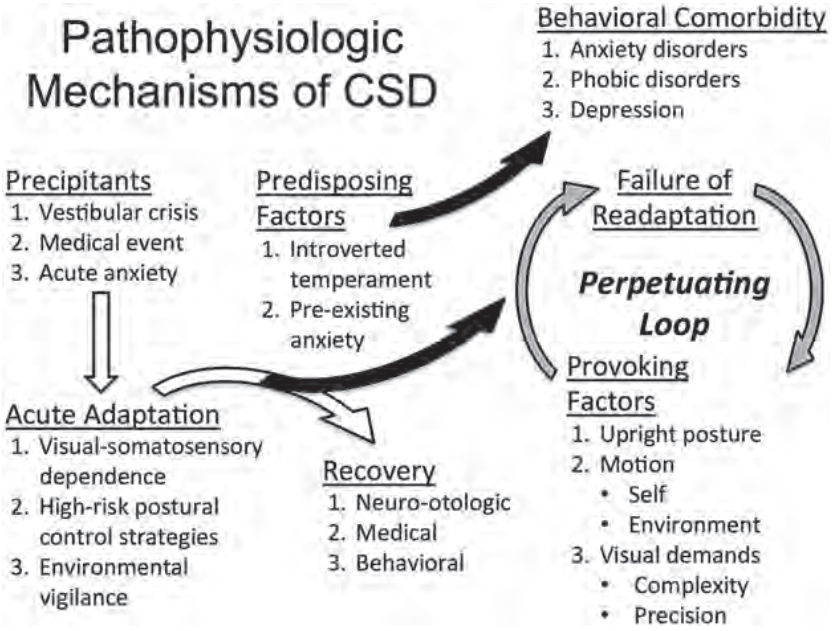

Figure 19.1 Figure 19.1. A new model of pathophysiological processes underlying chronic subjective dizziness (CSD). At the time of a precipitating event, postural control mechanisms react to maintain balance function to the greatest extent possible. When the acute event has passed, full recovery of neuro-otologic, medical, and behavioral function is expected. However, when predisposing factors are present, postural control systems may fail to readapt, instead entering a cycle of sustained hypersensitivity to provocative motion stimuli. Predisposing factors also increase the risk of behavioral comorbidity. (From Staab JP. Behavioral neurotology. In: Bronstein AM, ed. *Oxford Textbook of Vertigo and Imbalance*, Oxford University Press, Oxford, UK, 2013, used with permission.)

Differential Diagnosis

The diagnosis of CSD is made by inquiry into its key elements (Table 19-1). The role of physical examinations, vestibular laboratory tests, and neuroimaging are not to diagnose CSD itself, but to identify possible coexisting conditions and distinguish it from other disorders in its differential diagnosis. The most important aspects in taking the clinical history in a patient with suspected CSD are to elicit the core symptoms of CSD and separate past from present (i.e., to distinguish what a patient HAD previously from what he or she HAS currently). In Case Study 19-1, the patient's vestibular neuritis had been resolved for 2 years. Physical examination and vestibular laboratory testing confirmed excellent compensation, indicating that it was not the patient' s active illness. Diagnostic impressions and treatment plans have to account for chronic or recurrent neuro-otologic conditions (e.g., Ménière's disease), because these require ongoing care, but interventions for long-resolved problems will not improve present symptoms.

Several neuro-otologic and psychiatric disorders are included in the differential diagnosis of CSD. Neuro-otologic diagnoses include vestibular migraine, bilateral vestibulopathy, peripheral neuropathy, vestibular paroxysmia, orthostatic tremor, perilymph fistula, episodic ataxias, neurodegenerative disorders (spinocerebellar ataxias, multisystem atrophy), central vestibular syndromes, mild traumatic brain injury, autonomic dysregulation, and mal de débarquement. Specific clinical features distinguish each of these from CSD. CSD may be present in 30% of patients with vestibular migraine.[12] Patients with migraine headaches and distinct episodes of vestibular symptoms plus chronic unsteadiness and dizziness are likely to have coexisting vestibular migraine and CSD. Patients with orthostatic tremor report unsteadiness or dizziness on standing, but that usually improves with walking. They do not complain of difficulties with complex or demanding visual stimuli. Electromyographic or posturographic recordings should identify the pathognomonic 13- to 18-Hz tremor of the lower legs when patients are standing still. [34] Individuals with profound bilateral vestibular loss (BVL) have postural unsteadiness or dizziness exacerbated by head movements. They may have difficulties with motion-rich environments but do better if they can minimize their own movements. Patients with CSD are sensitive to environmental motion stimuli, even when they are still.Additionally, oscillopsia, common in patients with BVL, is not a feature of CSD. BVL may be identified by a bilaterally positive head impulse test or bilaterally reduced caloric responses. Patients with peripheral neuropathies, cerebellar degeneration, and movement disorders may report postural unsteadiness or dizziness, but they do not have much trouble with complex visual stimuli. Furthermore, their symptoms are usually progressive, unlike the waxing and waning course of CSD. Physical examinations demonstrate expected sensory-motor signs. Chronic dizziness and motion sensitivity may develop as part of the post-concussive syndrome, starting within days of concussive or whiplash injuries. CSD is not a late development following head injuries. Type 1 neurally mediated reflex syncope (aka vasovagal or neurocardiogenic syncope) and postural orthostatic tachycardia syndrome may mimic or coexist with CSD.[8] Symptoms and signs are predominantly orthostatic in autonomic dysregulation versus predominantly postural in CSD (see Relationship to Posture). Exertional symptoms are more prominent in autonomic dysregulation. Orthostatic blood pressure or tilt table abnormalities are not found in CSD.[35] Persistent sensations of rocking or swaying occur in mal de débarquement, but that develops after boat, train, airplane, or automobile trips, typically lasting at least several hours. Patients with this disorder usually report that they feel better when moving (e.g., riding in a car) and then feel bad again after stopping, which is opposite the typical pattern of motion sensitivity in CSD.

Primary psychiatric disorders in the differential diagnosis of CSD include panic disorder , generalized anxiety disorder, functional gait disorders, and somatic symptom disorders. Dizziness and lightheadedness are the second most common symptoms of panic attacks after chest pain and shortness of breath. Chronic dizziness may be one of several physical symptoms associated with generalized anxiety. Patients with functional gait disorders (aka conversion disorder) may complain of unsteadiness or dizziness, but their primary problems are trouble with walking. Unusual gait patterns on examination reveal this diagnosis.[11] Patients with somatic symptom disorders (previously somatization disorders) frequently report nonspecific dizziness, constant spinning vertigo, or swirling movements of the visual surround in multiple directions. These vestibular symptoms almost invariably coexist with other somatic symptoms and do not coalesce into the distinct pattern of CSD.

Treatment

Treatment options for CSD include the habituation form of vestibular and balance rehabilitation therapy (vestibular habituation), two classes of serotonergic medications (selective serotonin reuptake inhibitors [SSRIs] and serotonin norepinephrine reuptake inhibitors [SNRIs)]), and cognitive behavior therapy (CBT). No large, randomized, controlled trials of therapeutic interventions for CSD have been completed, so these recommendations are based on

smaller studies and extensive clinical experience. Most of the initial research on physical therapy interventions for vestibular disorders focused on subjects with chronic dizziness.[14] These early studies predated publication of the definition of CSD. Nonetheless, they have been informative for developing vestibular habituation protocols for CSD, because descriptions of their subjects suggest that many had CSD. Seven uncontrolled trials of SSRIs and SNRIs for patients with chronic dizziness, most of them targeting CSD, have been completed in the United States and Japan.[35-42] Five CBT trials have been done, including one with 6 and one with 12 months of follow-up.[15-18,43] Vestibular habituation, medication, and CBT may be used alone, or more frequently, in combination for CSD.

Vestibular Habituation Exercises

Vestibular and balance rehabilitation therapy is postulated to work by promoting compensation for peripheral or central vestibular deficits (see Chapters 9 and 22). However, a careful review of its formative research suggests that it may exert its therapeutic benefits through an additional (perhaps entirely different) mechanism, one that is well suited to treating CSD.[14] The pivotal trials of vestibular rehabilitation were conducted in the early 1990s on patients with chronic, nonspecific dizziness. The authors of those studies usually did not report detailed neuro-otologic assessments for their subjects. Study inclusion criteria were symptomatic in nature, targeting deficits in function rather than specific diagnoses. Nevertheless, these investigations certainly included patients with CSD, most with symptoms lasting for months to years, well beyond the time period for compensation after an acute vestibular injury. This suggests that the first vestibular rehabilitation protocols were primarily habituation strategies that reversed patients' classically conditioned hypersensitivity to motion stimuli and operantly conditioned alterations in gait and stance. Some of the early investigators used the term *habituation therapy* for their treatment.

To date, there have been no studies of vestibular habituation designed specifically for patients with CSD using modern clinical trial methods. Therefore, the benefits of vestibular habituation for patients with CSD must be extrapolated from clinical experience and the early studies of mixed groups of patients with chronic nonspecific dizziness. These suggest that vestibular habituation may reduce the severity of vestibular symptoms, increase mobility, and enhance daily functioning in 60% to 80% of patients.[14] Vestibular habituation may also reduce anxiety and depression in patients with chronic dizziness.[14] Successful treatment of patients with CSD using vestibular habituation requires a gentler approach than what is typically used to treat individuals who have just sustained an acute vestibular injury. Habituation exercises must be less intense at the beginning of therapy and must be increased more gradually or they will exacerbate rather than lessen symptoms. Patients whose vestibular habituation exercises are too vigorous at the beginning are likely to stop therapy prematurely and consider it to be a failure. Consistent benefits are usually obtained after 8 to 12 weeks of daily, home-based exercises, but maximum improvement may require 3 to 6 months of diligent therapy. Table 19-2 lists several keys to developing successful vestibular habituation strategies for CSD.

■ Table 19-2 **CHARACTERISTICS OF A SUCCESSFUL VESTIBULAR HABITUATION PROGRAM FOR CHRONIC SUBJECTIVE DIZZINESS**

Pacing	• Habituation exercises for CSD begin more gently and increase more slowly than the compensation exercises for acute vestibular deficits. • A daily exercise plan overcomes instinctive avoidance of provocative stimuli even if it starts with just a few minutes of habituation activities that the patient performs at home. • Scheduled breaks during exercises improve adherence and limit the potential to exacerbate symptoms.
Persistence	• The habituation process for CSD may take more time than the compensation process for acute vestibular deficits. • Full benefits of vestibular habituation may not be realized for 3–6 months.
Visual flow and visual complexity	• Exercises that include visual flow and complex visual stimuli address the visual symptoms of CSD.
Real world settings	• Use of indoor and outdoor settings that patients typically encounter promotes reintegration into daily activities.

Medication

Four open label, prospective trials and one lar ge retrospective study of SSRI treatment for patients with chronic dizziness have been conducted in the United States and Japan.[37-40] The studies were not all designed to tar get CSD specifically, but they used inclusion criteria that ensured enrollment of patients with CSD. Collectively , these studies included 190 patients who had CSD symptoms with or without additional neuro-otologic or psychiatric problems. All six commercially available SSRIs were tested in at least one study . A sixth investigation examined the efficacy of the SNRI milnacipran in 40 patients with CSD symptoms using an open label design similar to one of the previous SSRI trials. [40] A seventh study investigated the benefits of the SNRI venlafaxine in 32 patients formally diagnosed with both CSD and vestibular migraine.[42] Chronic non-vertiginous dizziness and unsteadiness were the primary target symptoms in all seven trials. Clinically meaningful reductions in these primary symptoms were observed in 60% to 70% of all patients who entered the trials (intent to treat analysis) and more than 80% of patients who completed at least 8 to 12 weeks of treatment (observed cases). About 20% of patients could not tolerate an SSRI or SNRI because of typical adverse effects of these medications (e.g., gastrointestinal upset, sleep disturbance, sexual dysfunction). No major problems were encountered with increased dizziness, even though dizziness is listed as a side effect of all 10 SSRIs and SNRIs that are available commer - cially in the United States. In patients with comorbid anxiety and depression, psychiatric symptoms improved along with vestibular symptoms. Taken together, these seven studies indicate that SSRIs and SNRIs can be used safely and effectively to treat patients with CSD. Table 19-3 provides dosing guidelines for medication treatment. None of the medications is superior to the others in benefits or side effects. Eight to 12 weeks of treatment may be required to obtain a clinically significant response, so a treatment trial should be continued for at least 12 weeks with the medication dose reaching at least the midpoint of the therapeutic range before considering it to be ineffective, except in cases of medication intolerance.[7] Maintenance treatment is recommended for a minimum of 1 year , longer for patients with persistent or recurrent neuro-otologic or psychiatric disorders, in keeping with the results of one of the SSRI trials which included a successful 1-year maintenance period. Patients who do not tolerate or respond to one medication have a good chance of responding to a second one. [7] In clinical care, treatment is switched from SSRIs to SNRIs or vice versa after two failed trials of medications in either class.

The clinical trials of SSRIs and SNRIs have made these medications the mainstay of pharmacological treatment for CSD. Mirtazapine and tricyclic antidepressants are used clinically but have not been the subjects of studies. Benzodiazepines and other vestibular suppressants are not effective as a primary treatment for CSD, though patients with high levels of comorbid anxiety may benefit from a short course of a benzodiazepine while initiating more definitive treatment with a serotoner gic antidepressant and vestibular habituation.

■ Table 19-3 **DOSING STRATEGIES FOR SEROTONERGIC ANTIDEPRESSANTS FOR CHRONIC SUBJECTIVE DIZZINESS**

Medication	Initial Therapy[a] Daily Dose (mg)	Titration (2 weeks) Daily Dose (mg)	Titration (4–6 weeks) Daily Dose (mg)	Therapeutic Range Daily Dose (mg)
Selective serotonin reuptake inhibitors				
Fluoxetine	5 to 10	10 to 20	20 to 40	20 to 60
Sertraline	12.5 to 25	25 to 50	50 to 100	50 to 150
Paroxetine	5 to 10	10 to 20	20 to 40	20 to 60
Citalopram	5 to 10	10 to 20	20 to 40	20 to 40
Escitalopram	2.5 to 5	5 to 10	10 to 20	10 to 20
Fluvoxamine[b]	25	25 to 50 twice daily	50 to 100 twice daily	50 to 100 twice daily

■ Table 19-3 **DOSING STRATEGIES FOR SEROTONERGIC ANTIDEPRESSANTS FOR CHRONIC SUBJECTIVE DIZZINESS—cont'd**

Medication	Initial Therapy[a] Daily Dose (mg)	Titration (2 weeks) Daily Dose (mg)	Titration (4–6 weeks) Daily Dose (mg)	Therapeutic Range Daily Dose (mg)
Serotonin and norepinephrine reuptake inhibitors				
Venlafaxine	25 to 37.5	37.5–50	75 to 150	75–225
Milnacipran[c]	12.5 to 25 twice daily	25 to 50 twice daily	50 twice daily	50 to 75 twice daily
Duloxetine[d]	20 to 30	40 to 60	40 to 60	40 to 60

[a] Most patients can be prescribed the higher initial dose to start treatment. Those who do not tolerate that dose and those who prefer to start treatment more cautiously may be prescribed the lower initial dose.
[b] Fluvoxamine is typically started at 25 mg once daily for 1 to 2 weeks and then increased to twice-daily dosing thereafter.
[c] Milnacipran is typically started at 12.5 mg once daily for 2 to 3 days and then increased to twice-daily dosing thereafter.
[d] Duloxetine has not been studied in clinical trials for chronic subjective dizziness (CSD), but it is used clinically.
Desvenlafaxine has not been studied in clinical trials for CSD. It is available only in 50-mg and 100-mg capsules, which does not permit gradual titration; therefore, it is not used commonly in clinical practice for patients with CSD.

Psychotherapy

Five controlled trials of CBT have been conducted for patients with chronic dizziness. Three trials specifically selected patients with CSD,[16,17,18] whereas the other two enrolled subjects with chronic nonspecific dizziness.[15] The three oldest studies included 39 patients and 40 control subjects. A systematic review of their results found that CBT had a medium effect for reducing dizziness (effect size = 0.46). The fourth trial included 41 patients with CSD randomly assigned to active treatment or wait-list control.[16] The authors reported large reductions in dizziness and dizziness-related avoidance symptoms after just three sessions of CBT (effect sizes = 0.98 to 1.15). All of these results were measured at the end of active psychotherapy treatment. Two studies examined long-term outcomes. One that included patients with long-standing CSD found that short-term benefits of CBT were not sustained at 1 year,[17] whereas another that enrolled patients within the first two months of development of persistent dizziness showed that benefits were largely maintained a 6-month follow-up. These studies give a mixed picture of the efficacy of CBT for CSD, but suggest that it may be more effective if employed as early as possible in the course of illness. CBT may also be used to treat comorbid anxiety and depressive disorders in patients with CSD.

CASE STUDY 19-1

Diagnosing CSD

A 34-year-old man was referred for neuro-otologic consultation because of sensations of unsteadiness and non-vertiginous dizziness that had persisted for 2 years after a bout of acute vertigo, nausea, vomiting, and ataxia. His initial vertiginous symptoms had resolved gradually over 6 to 8 weeks, but he was left with persistent sensations of unsteadiness when standing and veering to either side when walking that were not visible to others. He felt that he had to concentrate on walking to maintain his desired direction. He had no auditory symptoms or other neurological complaints. Head movements in any direction aggravated his symptoms. He worked stocking shelves in a large discount store, but often left work early because walking up and down the aisles increased his symptoms. The movement of customers in the store bothered him, even if he stood still. He previously enjoyed playing computer games with friends but could no longer use his laptop computer for more than 10 minutes without a break. The patient had no psychiatric history but said his friends thought of him as a worrywart. Neuro-otologic examination was normal. Vestibular laboratory tests revealed a fully compensated 45% left peripheral vestibular weakness, but no other abnormalities.

Continued

CASE STUDY 19-1

Comment

This case is typical of CSD triggered by vestibular neuritis. The patient's peripheral vestibular deficit was acquired during the acute phase of illness but was well compensated at the time of his consultation. Therefore, it could not have been the cause of his persistent unsteadiness or dizziness. Note that his symptoms changed in character from vertigo and ataxia to non-vertiginous dizziness and unsteadiness *without* gait disturbance at about the time that compensation for an acute peripheral insult would be expected (i.e., 4 to 8 weeks).The patient's motion-provoked symptoms were quite common for CSD. He had difficulty with his own head movement that was not direction or position specific. He could not tolerate full field visual flow, regardless of whether it resulted from his own movements (e.g., walking in the aisles) or motion of objects around him (e.g., customers milling about). He also had difficulty with small-field, precision visual tasks (e.g., using his computer). His worrywart temperament may have been a risk factor for developing CSD, but he had no psychiatric morbidity.

CASE STUDY 19-2

A 33-year-old woman was evaluated for a 3-year history of chronic daily dizziness punctuated by recurrent episodes of vertigo that lasted for 1 to 2 hours, usually associated with bilateral retro-orbital pain and headache. Her symptoms began with an acute attack of vertigo that occurred while driving across an unfamiliar bridge in rush-hour traffic. She feared that she would lose control of her car, so she pulled to the side of the road until the worst of the vertigo subsided, which took about 20 minutes. Following that incident, she developed persistent sensations of swaying or rocking whenever she stood or sat upright. This symptom increased with her own movements, motion of objects around her, and watching television or reading. She continued to have acute attacks of vertigo 2 to 3 times a month. About 75% of those attacks were accompanied by eye pain or headache with light and sound sensitivity and nausea that lasted for hours. She had panic attacks on days when her dizziness flared. She stopped driving and depended on friends or family members to accompany her on shopping trips. Otherwise, she stayed home where she felt most confident about her balance. Neuro-otologic evaluation and vestibular laboratory testing were entirely normal. Psychiatric examination revealed that she was shy as a child. She had several panic attacks shortly after graduating from high school, but those resolved spontaneously.

Comment

This patient had CSD, vestibular migraine, panic disorder, and agoraphobia. Vestibular migraine caused her recurrent attacks of vertigo, even though every vertigo attack was not associated with a migraine headache. The first episode of vestibular migraine on the bridge also triggered a recurrence of panic attacks and agoraphobic symptoms to the extent that she became nearly housebound. CSD was responsible for her persistent postural unsteadiness and dizziness as well as her day-in-and-day-out motion sensitivity. This case should not be confused with mal de débarquement (MdD). Even though the initial event occurred in a vehicle, it was a spontaneous attack of rotary vertigo, which is not a symptom of MdD. Rocking sensations may occur in both CSD and MdD, but patients with these two conditions respond differently to motion stimuli as discussed in the section on the differential diagnosis of CSD.

CASE STUDY 19-3

Treating CSD

The patient of Case Study 19-1 was diagnosed with vestibular neuritis, referred to a physical therapist for vestibular rehabilitation, and given a prescription for meclizine, which was not helpful and sedated him too much. He attended two physical therapy sessions, but he could not tolerate the intensity of the exercises, so he did not return. His wife became concerned that he was more anxious than usual, so his primary care physician prescribed the SSRI paroxetine at a dose of 20 mg dailyHe took it for 2 days, but experienced nausea and increased dizziness, so he stopped it. At that point, his primary care physician referred him to a tertiary neuro-otology center where his clinical history, examination, and laboratory findings were confirmed. A psychiatric consultant identified his anxious temperament but concluded that his anxiety was not at the level of a major anxiety disorder. The patient was diagnosed with (1) vestibular neuritis, resolved; (2) fully compensated left peripheral vestibular hypofunction; and (3) CSD. He was instructed to perform a series of vestibular habituation exercises starting with modest head movements, followed by twirling a striped umbrella in front of him, and finally walking in a hallway while looking from side to side at pictures on the walls. The duration of habituation exercise was increased gradually from 2 to 15 minutes twice daily. The patient was also prescribed sertraline (another SSRI) at 12.5 mg daily for 1 week, then 25 mg daily for 1 week, then 50 mg daily. He returned to work after 8 weeks of combined treatment.

Comment

This case demonstrates three pitfalls in treating patients with CSD. First, the patient had already compensated for his bout of vestibular neuritis. Therefore, vestibular suppressants were not indicated. Second, vestibular habituation exercises are a first-line intervention for CSD, but they must be introduced gradually to avoid symptom exacerbation and premature withdrawal from therapy. Third, SSRIs and SNRIs are the principal pharmacological treatments for CSD, but they also have to be started at lower doses and increased more slowly than usual for depression (please see Table 19-3). No specific interventions were needed for the patient's lifelong anxious temperament, though sertraline had the potential to dampen it.

Summary

A condition of persistent non-vertiginous dizziness and unsteadiness was first described as phobic postural vertigo 27 years ago. Its definition was updated 7 years ago to focus more clearly on its core physical symptoms (Table 19-1). The streamlined condition was called chronic subjective dizziness. It is the second most common diagnosis made in tertiary neuro-otology centers that recognize it, just behind BPPV and ahead of vestibular migraine. CSD is defined by persistent (>3 months) symptoms of non-vertiginous dizziness and unsteadiness that are worse in an upright position and can be exacerbated by exposure to provocative motion stimuli in the environment or by performance of precision tasks. The clinical course of CSD is typically chronic, but fluctuating in severity over many months or years. CSD is almost always triggered by an acute neuro-otologic, medical, or psychiatric illness. Two large studies have established its differential diagnosis and identified common comorbidities. New hypotheses about its pathophysiological mechanisms have been developed. Perhaps most importantly, studies conducted on three continents suggest that it may be treated successfully with vestibular habituation exercises, SSRI and SNRI antidepressants, and CBT.

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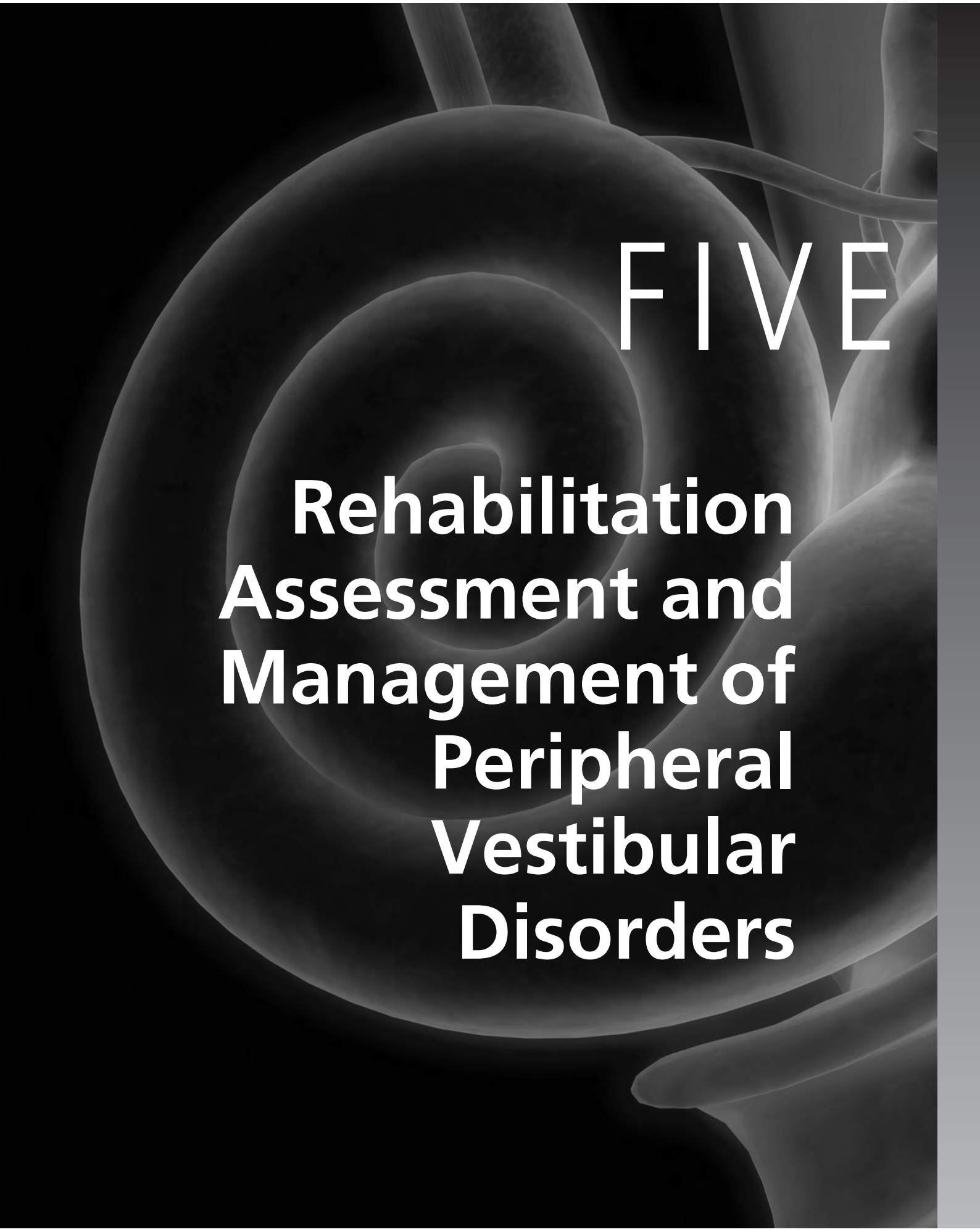

FIVE

Rehabilitation Assessment and Management of Peripheral Vestibular Disorders

CHAPTER 20

Physical Therapy Management of Benign Paroxysmal Positional Vertigo

Susan J. Herdman, PT, PhD, FAPTA ■ *Jeffrey M. Hoder, PT, DPT, NCS*

Benign paroxysmal positional vertigo (BPPV) is a mechanical problem of the peripheral vestibular system resulting from otoconia being displaced into the semicircular canals. It is the most common cause of recurrent and episodic vertigo from a peripheral vestibular disorder. In the general population, BPPV has an incidence of 64 new cases per 100,000 people per year. With such a high incidence in the general population, an understanding of the disease and its proper management becomes imperative. Treatment of posterior semicircular canal BPPV has been studied extensively, and there is considerable research that supports "best practice" for its assessment and management. We first describe the proposed mechanisms for BPPV and then explore the assessments and various treatment alternatives for this disorder.

Characteristics and History

BPPV is characterized by brief episodes of vertigo, with the perception of either the environment or one's self spinning when the head is moved into certain positions. It has been reported in adults of all ages, although it is relatively uncommon in children.[1,2] Although BPPV can occur spontaneously in many patients, it has been known to follow head trauma, labyrinthitis, or ischemia in the distribution of the anterior vestibular artery.[1] Spontaneous remission of this condition is common. For those patients in whom the episodic vertigo persists, this disorder can be annoying, disruptive, and often results in significant changes in normal activities.

Patients with BPPV commonly report vertigo triggered by lying down, rolling over in bed, bending over, and looking up. Common situations in which vertigo is provoked include getting out of bed, gardening, washing hair in the shower, and going to the dentist or beauty parlor. Other complaints associated with BPPV include balance problems that may last for hours or days after the episodic vertigo has stopped and more vague sensations such as lightheadedness or a feeling of floating (Table 20-1).

Historical Mention of BPPV-like Vertigo

Adler first described the clinical presentation of BPPV in 1897 in his paper on "unilateral vertigo" and recognized that the posterior and anterior semicircular canals (SCC) were most likely the affected structures.[3] He reported, "Active or passive movements of the head toward the diseased side . . . elicit severe vertigo, which can be so intense that patients become pale, diaphoretic, and strongly resist any further head turning . . . illusory movements of the environment occurred . . . in the respective *planes of the anterior and posterior semicircular canals* of the affected ear."[3]

The clinical syndrome of recurrent positional nystagmus was described in further detail by Barany, a Nobel laureate otologist from Sweden, in 1921.[4] Barany detailed the case of a 27-year-old woman who experienced recurrent attacks of vertigo for 2 weeks. ". . . these attacks only occurred when the patient lay on her right side. When she did this, there occurred a strong rotatory nystagmus to the

■ Table 20-1 **FREQUENCY OF COMPLAINTS IN 100 CONSECUTIVE PATIENTS WITH BPPV**

Complaints	Frequency (%)
Poor balance	57
Sense of rotation (vertigo)	53
Trouble walking	48
Lightheaded	42
Nausea	35
Queasy	29
Spinning inside head	29
Sense of tilt	24
Sweating	22
Sense of floating	22
Blurred vision	15
Jumping vision	13

Tusa RJ, Herdman SJ. Adapted from Canalith Repositioning for Benign Paroxysmal Positional Vertigo. *American Academy of Neurology*. 3B5.002.[5]

right with a vertical component upwards, when looking to the right was purely rotatory, and when looking to the left was purely vertical."[4] Barany noted that hearing, caloric reactions on both sides, and neurological examination were completely normal.[4]

Barany described several of the characteristics that are now known to describe BPPV:

1. Nystagmus is provoked by positional changes against gravity, resulting in complaints of vertigo.
2. The nystagmus and complaints of vertigo are brief in duration, despite maintaining the provocative position.
3. The nystagmus, as well as the complaints of vertigo, is fatigable with repeated exposure to the provocative position.

It was not until 1952 that two British otologists, Charles Hallpike and Margaret Dix, described a technique to elicit the positional nystagmus, as well as adopting the term "positional vertigo of the benign paroxysmal type."[6]

Proposed Mechanisms for BPPV

Two mechanisms have been proposed to explain the characteristic presentation of BPPV. In 1962, Schuknecht attributed the disorder to otoconia crystals adhered to the cupula, which he termed "cupulolithiasis" (Fig. 20.1) after noting basophilic staining masses of granular material attached to the cupula of the posterior canal in two patients with a history of BPPV.[7] Presumably, the presence of the debris adhering to the cupula significantly increases the density of the cupula and, therefore, produces an inappropriate deflection of the cupula of the posterior canal when the head is positioned with the affected ear below the horizon. The result is vertigo, nystagmus, and nausea.

Thus, cupulolithiasis is characterized by (1) immediate onset of vertigo when the patient is moved into the provoking position, (2) the presence of a nystagmus, which appears with the same latency as the complaints of vertigo, and (3) persistence of the vertigo and nystagmus as long as the person's head is maintained in the provoking position. Because the cupula remains deflected as long as the patient is in the provoking position, the nystagmus and vertigo will persist, although the intensity may decrease slightly because of central adaptation or because the patient may also have canalithiasis.[4] We have since learned that cupulolithiasis as a cause of BPPV is relatively uncommon.

Although the presence of debris on a cupula accounts for the clinical presentation of cupulolithiasis, debris adhering to the cupula has a greater prevalence than does BPPV, raising questions as to the validity of this theory.[8,9]

A second theory, canalithiasis, was proposed in 1979 by Hall et al[10] (Fig. 20.2). The canalithiasis theory suggests that degenerative debris from the utricle (possibly fragments of otoconia) are free-floating in the long-arm of the semicircular canal. Otoconia are more than twice the

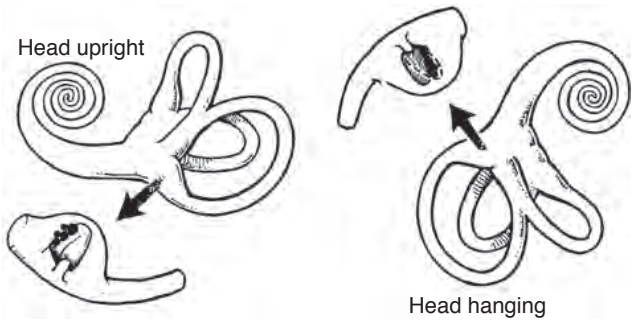

Figure 20.1 In cupulolithiasis, the debris is adhering to the cupula of the semicircular canal (shown here for posterior canal). Movement into the head-hanging position, gravity displaces the weighted cupula resulting in an abnormal signal from that canal. (Modified from Herdman, SJ, et al, 1993, with permission.[11])

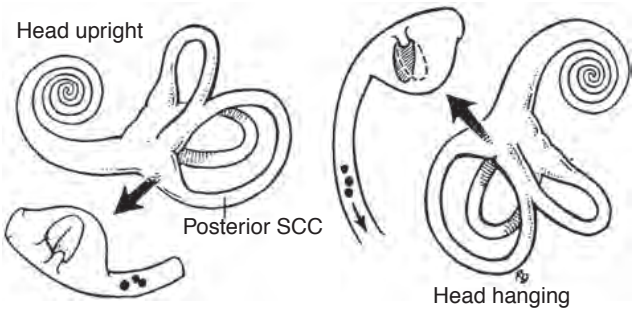

Figure 20.2 In canalithiasis, the calcium carbonate crystals are floating freely within the long arm of the canal (shown here for posterior canal). When the head is moved into the head-hanging position (Hallpike-Dix Test), the debris migrates to the most dependent portion of the canal. The movement of the debris subsequently causes the endolymph to move, which in turn overcomes the inertia of the cupula and an abnormal signal is sent to the central nervous system. (Modified from Herdman, SJ, et al, 1993, with permission.[11])

density of the endolymph within the semicircular canals. Therefore, a positional change of the canals with respect to gravity would result in movement of the otoconia, which in turn would overcome the inertia of the endolymph. The movement of the endolymph would deflect the cupula, causing nystagmus and complaints of vertigo. A latency of the response is related to the time needed for the cupula to be deflected by the pull of the endolymph. The intensity of vertigo and nystagmus are related to the degree of deflection of the cupula. Vertigo and nystagmus stop as the position is maintained as a result of cessation of endolymph movement as the otoconia settle in the most dependent portion of the canal.

Thus, BPPV from canalithiasis is characterized by (1) a delay in the onset of the vertigo by 1 to 40 seconds after the patient has been placed in the provoking position; (2) the presence of a nystagmus, which appears with the same latency as the complaints of vertigo; and (3) a fluctuation in the intensity of the vertigo and nystagmus that increases and then decreases, disappearing within 60 seconds. BPPV from canalithiasis is the most common form of this disorder.

Visualization of cloudy debris within the posterior SCC was made during a canal plugging surgery and lends support for this mechanism.[12] The theory of canalithiasis is consistent with all the clinical features of typical BPPV; however, it is important to note that there is no direct evidence that any intervention actually places the debris back into the utricle.[13]

Etiology

The exact cause of BPPV is mostly unknown. Over 50% of patients with BPPV are considered to have the idiopathic form (Table 20-2).[14] Under normal circumstances, it is recognized that otoconia are dislodged, absorbed, and renewed constantly. Thus, displacement of otoconia into one of the semicircular canals is always a possibility. Interestingly, the side on which a person sleeps may be an etiological factor in BPPV. Shigeno et al found that patients with recurring BPPV were more likely to sleep on the affected side in BPPV than were those patients who did not have recurrence.[15]

The age of the patient may be a contributing factor in idiopathic BPPV. Among patients with dizziness, the incidence of BPPV is known to increase with increasing age (Table 20-3). This may be, in part, due to fragmentation of otoconia, which has been noted in older adults.[16] It is extremely unusual for a child to have BPPV. The youngest documented case to date was a child age 5 years.[17] Children with episodic vertigo most commonly have a migraine disorder called Benign Paroxysmal Vertigo of Childhood, unrelated to the displacement of otoconia.[18,19]

The second most common cause of BPPV is head trauma (see Table 20-2). Head trauma–induced BPPV may include relatively minor injuries such as those associated with flexion-extension injuries of the neck (whiplash) as well as more severe head trauma. Dispenza et al (2010) reported that BPPV was the cause of vertigo in 33.9% of their total caseload of patients with whiplash.[20] When BPPV is associated with head trauma, it typically appears within a few days of the trauma, although it may go unrecognized depending on the severity of the patient's other injuries. BPPV from traumatic brain injury may be more recalcitrant to treatment than is the idiopathic form of BPPV. However, remission rates are equivalent.[21]

BPPV also occurs in combination with viral neuronitis. Most cases of unilateral vestibular neuronitis affect the superior branch of the vestibular nerve. This branch supplies the utricle and the superior and horizontal SCCs.

■ Table 20-2 **ETIOLOGY OF BPPV**

Diagnosis	# of Patients	% of Patients
Idiopathic	287	58.0
Post-traumatic	90	18.2
Vestibular neuritis	42	8.6
VBI	13	2.6
Other	63	12.7

495 patients (adapted from Baloh et al, 1987[1]; Katsarkas and Kirkham, 1978[14])

■ Table 20-3 **PERCENT OF BPPV BY AGE (DATA FROM A TERTIARY, SPECIALTY CLINIC)**

Age (yrs)	# of Dizzy Patients	# Dizzy Patients with BPPV	% Dizzy Patients with BPPV
0–9	22	0	0.0
10–19	52	1	1.9
20–29	123	3	2.4
30–39	360	45	12.5
40–49	485	77	15.9
50–59	411	91	22.1
60–69	539	136	25.2
70–79	752	188	25.0
80–89	392	108	27.6
90–99	33	11	33.3

Emory University, Dizziness and Balance Center

Theoretically, with the utricular degeneration that occurs during acute vestibular neuronitis, otoconia break free and float into the still functioning posterior SCC (which is innervated by the inferior branch of the vestibular nerve). The patient presents with vestibular hypofunction *and* posterior SCC BPPV.

Current Diagnosis and Management

Three different maneuvers can be used to evaluate an individual for the presence of BPPV. For all the maneuvers, observation of the direction and duration of the nystagmus by the clinician is critical to determining canal involvement and to developing a treatment plan. Therefore, it is important that the patient understands what to expect. During testing, patients should keep their eyes open and remain in the provoking position. To discourage the patient from attempting to move out of the provoking position once symptoms begin, prior to testing, the clinician should explain that the vertigo will stop or decrease if the patient remains in the position. If the patient's history suggests which side is affected, it is best to test the presumed unaffected side first to minimize nausea. For patients with severe nausea and a history of emesis associated with vertigo, the testing maneuvers should be performed more slowly, although this decreases the likelihood of provoking the nystagmus.

> Testing and treating patients with BPPV who may also have cervical spine and cervical–medullary junction disorders must be done with caution.[22,23] The positions used to diagnose and manage BPPV require orientating of the SCC with respect to gravity to provoke migration of the otoconia. The positioning of the patient can be modified to limit or omit neck extension and rotation while maintaining proper orientation of the canals within the plane of movement.
>
> Testing for vertebrobasilar artery insufficiency (VBI) remains controversial. On the one hand, assessments for VBI are not sensitive and have 0% positive predictive value for identifying VBI.[24,25] On the other hand, the consequences of misidentifying BPPV for VBI and vice versa are untenable. Fortunately, testing for BPPV can be performed without extending or rotating the neck by using a tilt table. If the clinician decides that testing for VBI should be included as part of the examination, testing should consist of cervical spine active range of motion testing with the patient in a seated position. Cervical positions should be sustained for 10 seconds each, before testing for BPPV, as recommended by Humphriss.[23]

A history of episodic vertigo with position changes and a positive Dix-Hallpike Test are the clinical practice guidelines for diagnosis of BPPV.[26,27] All tests for BPPV can be performed in room light, but the use of Frenzel lenses or an infrared camera system to prevent fixation suppression of horizontal and vertical nystagmus may increase the likelihood of a correct diagnosis. Torsional nystagmus is not suppressed by fixation.

Tests that Identify BPPV Affecting the Vertical Canals

Dix-Hallpike Test

The Dix-Hallpike Test, sometimes called the Barany maneuver or the Nylen-Barany maneuver, is considered the Gold Standard test for the diagnosis of BPPV (Fig. 20.3)[6,26-28] The combination of a history of episodic vertigo with position changes and a positive Dix-Hallpike Test are the suggested clinical practice guidelines for diagnosis of BPPV.[26,27]

This maneuver places the posterior SCC of the downside ear in the plane of the pull of gravity. Debris adhering to the cupula or free-floating in the long arm of the canal will shift away from the cupula, resulting in vertigo and

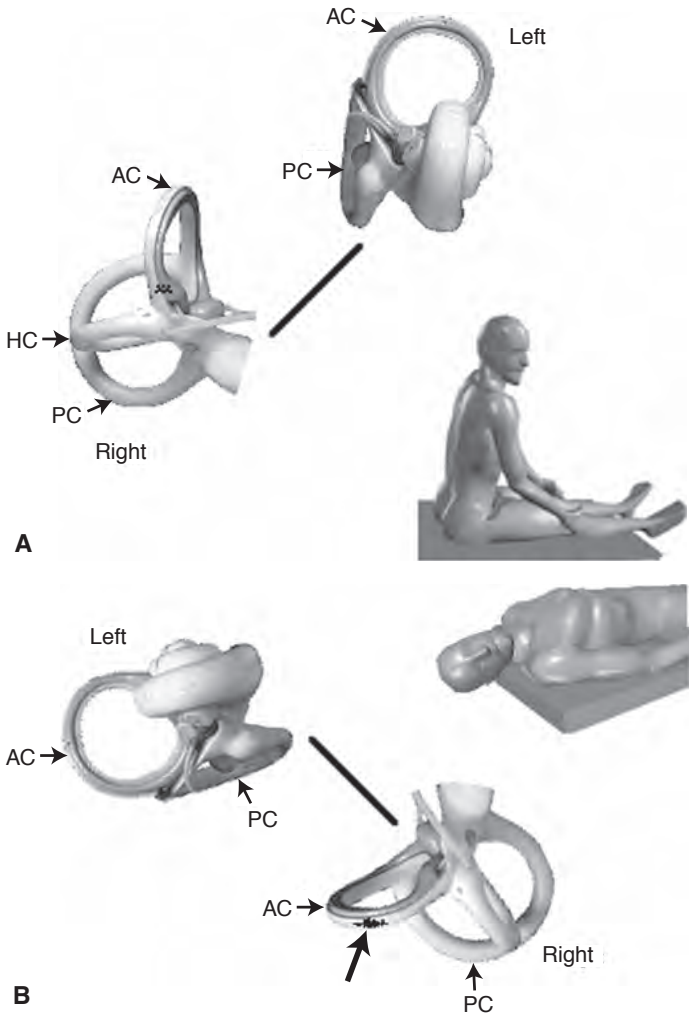

Figure 20.3 Position testing for anterior or posterior canals BPPV: Dix-Hallpike Test. The patient sits on the examination table and actively rotates his/her head 45 deg horizontally toward the labyrinth to be tested **(A).** The patient then quickly lies straight back "en bloc" with the therapist guiding the movement so that the head is hanging over the edge of the examination table by 20 deg to 30 deg (position **B**). The patient is asked if he/she has vertigo and is observed for nystagmus. The patient then slowly returns to a sitting position, assisted by the therapist, if needed, with the head still turned 45 deg and nystagmus is observed again (not shown). This test is then repeated with the head turned 45 deg in the other direction. The orientation of the vestibular labyrinths are shown for each position.

nystagmus. In most cases of BPPV, nystagmus and vertigo occur within a few seconds of the position change, but occasionally the signs and symptoms will have a longer delay until onset, even as much as 30 seconds. If the patient has BPPV, vertigo and nystagmus will be provoked when the affected ear is inferior. The patient may also experience vertigo upon returning to the sitting position. The test can then be repeated with the patient's head turned to the other side. If the debris is within the posterior semicircular canal, the resultant nystagmus during testing will be upbeating and torsional towards the side being tested.

Sidelying Test

The Sidelying Test (Fig. 20.4) is particularly useful in patients who cannot tolerate the Dix-Hallpike position because of neck or back problems.

Halker et al (2008) were able to calculate the sensitivity and specificity of both the Dix-Hallpike and the Sidelying Tests using data presented in a Cohen (2004) article.[28,29] For the Dix-Hallpike test, the estimated sensitivity was 79% [95% confidence interval (CI) 65-94], specificity was 75% (33-100), positive likelihood ratio (LR) was 3.17, and the negative LR was 0.28. For the Sidelying Test, they reported an estimated sensitivity of 90% [with a 95% CI of 79-100] and a specificity of 75% [95% CI of 33-100]. The positive likelihood ratio was 3.59 and the negative likelihood ratio was 0.14.[28] In a more recent article, unfortunately with only the abstract published in English, the sidelying test demonstrated concurrent validity with Dix-Hallpike for identifying both posterior and anterior SCC BPPV.[30]

Other Tests

Several other tests have been described as being useful for identifying anterior SCC BPPV. However, the evidence is limited to descriptions of case series. None of these tests report sensitivity or specificity. Only one test is mentioned in several articles[31-34] in which the observation of downbeating and torsional nystagmus is performed when the patient lays straight back and the head is hanging between

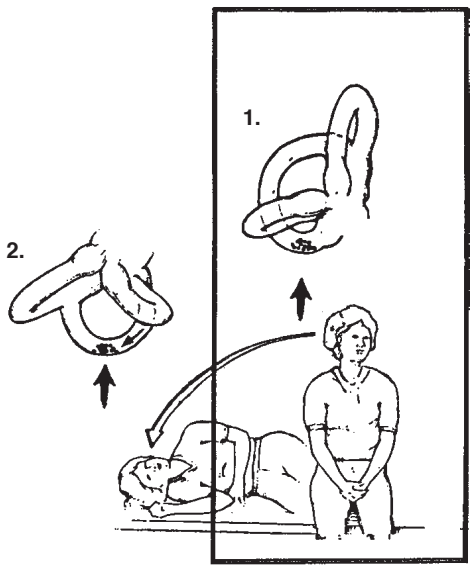

Figure 20.4 Shown here for testing the right labyrinth, the Sidelying Test is performed by asking the patient to turn the head 45 deg away from the side to be tested (1). The patient then lies down *rapidly* to the side opposite the direction of head turn (2). After waiting until any provoked nystagmus stops, or 30 sec if there is no nystagmus, the patient *slowly* returns to sitting. After waiting to be sure that the patient does not experience any vertigo in sitting, the test is repeated to the opposite side.

30 and 60 degrees. The four articles reported on a total of only 22 cases.

Tests for Identifying BPPV Affecting the Horizontal Canals

Roll Test

In patients with horizontal SCC BPPV, the Hallpike-Dix and Sidelying Tests may not provoke vertigo and nystagmus, because the final position does not place the horizontal SCC (HSC) in the plane of the pull of gravity. The most commonly used test for the horizontal SCC is the Roll Test (Fig. 20.5), in which the patient is supine and turns (rolls) his/her head in the plane of the horizontal SCC.[35-37] The elicited nystagmus is horizontal and may have a torsional component. In the canalithiasis form of Horizontal SCC BPPV, the nystagmus is geotropic; that is, the fast phase beats toward the Earth and is brief in duration. In the cupulolithiasis form of Horizontal SCC BPPV, the nystagmus is apogeotropic (the fast phase beats away from the Earth) and is prolonged in duration. The affected side is considered to be the more symptomatic side in canalithiasis and the less symptomatic side in cupulolithiasis.

The Roll Test for Horizontal SCC BPPV has only face validity, and its sensitivity and specificity have not been reported. However, the efficacy of the Roll Test has been assessed indirectly in a study of the Bow and Lean Test.[38]

Bow and Lean Test: A Diagnostic Test for Horizontal SCC BPPV

Because identifying the side of involvement using the degree of symptom severity may be challenging, the "Bow and Lean Test" has been proposed as an addition to the Roll Test to identify the affected side in HSC-BPPV.[39] First, the Roll Test is performed to determine whether the HSC-BPPV is the canalithiasis or cupulolithiasis form, based on the duration of the nystagmus. Then, the Bow and Lean Test (BLT) is used to determine which horizontal SCC is affected by observing the direction of the nystagmus (Fig. 20.6).

A single randomized controlled study reports that identification of the affected side in HSCC canalithiasis form using BLT significantly increases treatment efficacy compared with the Roll Test alone.[39] In this prospective single blinded study, the affected side of horizontal SCC was determined either by the Roll Test alone (n = 61) or by the Roll and Bow and Lean Tests performed together (n = 164). In both groups, they used the duration of nystagmus during the Roll Test to identify whether the patient had canalithiasis or cupulolithiasis. In the group diagnosed using the Roll Test only, the side to be treated was based on the nystagmus being stronger on the affected side for canalithiasis and the nystagmus being stronger on the unaffected side for cupulolithiasis. For the group diagnosed

Figure 20.5 In the Roll Test, the patient lies supine with the head elevated 15–20 deg **(A)**. This places the horizontal canals in the plane of the pull of gravity. The head is then quickly rolled to the left **(B)** to see if that will provoke the vertigo and nystagmus. Careful observation of the nystagmus is essential to determine whether the patient has the canalithiasis or the cupulolithiasis form of HC BPPV. The head can then be brought back to the neutral position **(C)**. After waiting until nystagmus ceases, the head is rapidly turned to the right **(D)**. Vertigo and nystagmus will be elicited in both the head left and head right positions. When the patient's vertigo and nystagmus stop, the patient's head is returned to neutral **(E)**.

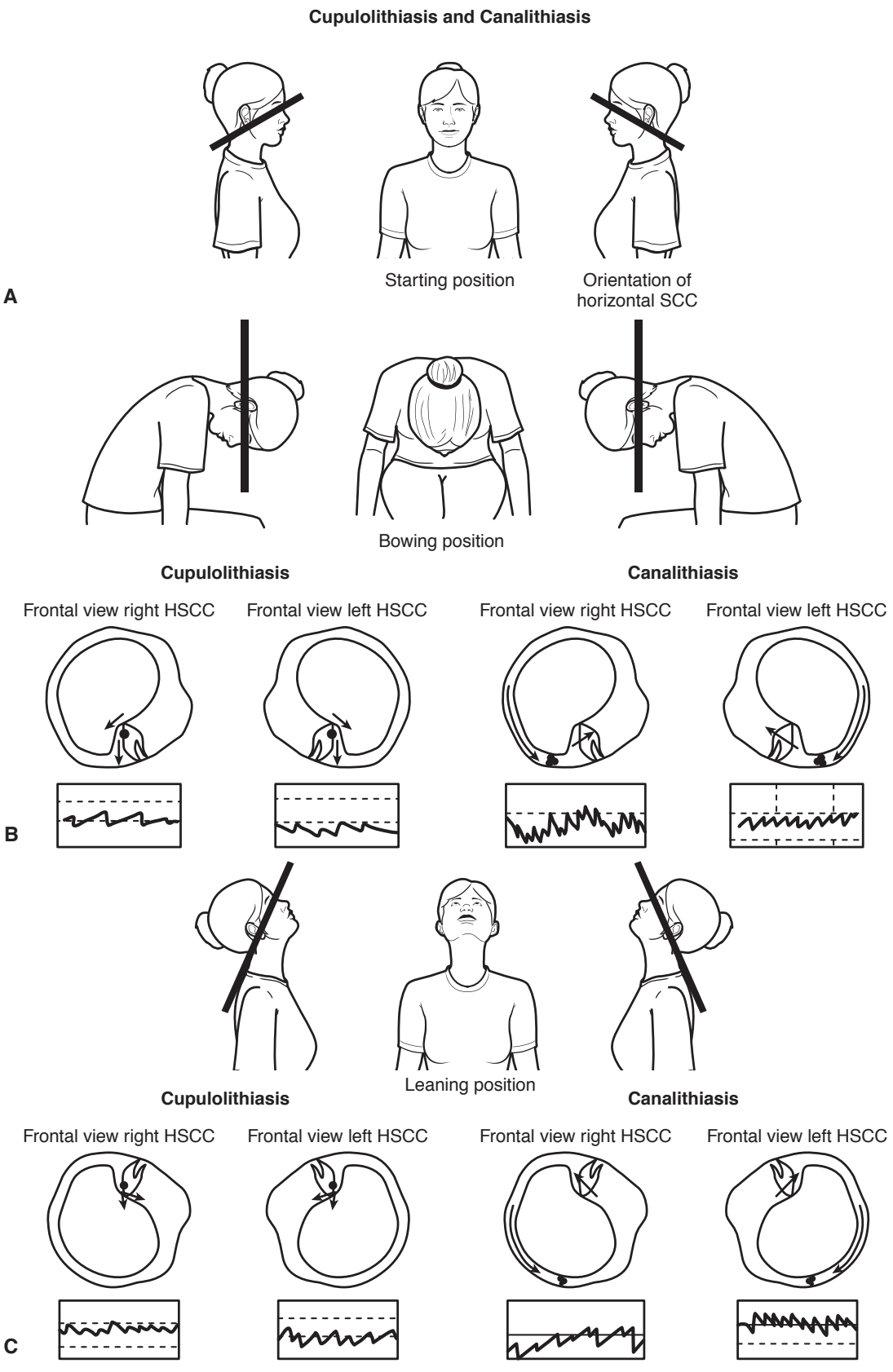
Cupulolithiasis and Canalithiasis
A
Starting position
Orientation of horizontal SCC
Bowing position
Cupulolithiasis
Canalithiasis
B
Frontal view right HSCC
Frontal view left HSCC
Frontal view right HSCC
Frontal view left HSCC
Leaning position
Cupulolithiasis
Canalithiasis
C
Frontal view right HSCC
Frontal view left HSCC
Frontal view right HSCC
Frontal view left HSCC

Figure 20.6 After the Roll Test has been used to determine if the patient has canalithiasis or cupulolithiasis, the Bow and Lean Test is used to determine which horizontal canal is involved. The starting position is shown in **(A)**. In the "bow" position **(B)**, the nystagmus beats away from the affected side if the patient has the cupulolithiasis form of BPPV and toward the affected ear in the canalithiasis form of BPPV. In the "lean" position **(C)**, in cupulolithiasis the nystagmus beats toward the affected ear but in canalithiasis, the nystagmus beats away from the affected ear.

using both the Roll Test and the BLT, the *direction* of nystagmus during the BLT was used to determine which side was affected. The additional information provided by the BLT to determine side of involvement improved the efficacy of treatment of those with HSC-BPPV canalithiasis from 67.4% to 83.1% after two sessions of canalith repositioning, and those with cupulolithiasis from 61.1% to 74.7%. This suggests that the BLT may be a useful adjunct to perform following the Roll Test, especially when the affected side is difficult to determine.[38]

Testing Sequence

One can easily assess all three canals for BPPV efficiently with the following testing sequence. If at any time during this series of tests, you find nystagmus and vertigo appropriate for BPPV, you should stop the testing and treat the patient. For sake of discussion, assume the patient tells you they experience vertigo when they lie on the right side. First, perform the Dix-Hallpike test on the left side. Then do the test on the right side. Let's assume the patient still has no vertigo. Before you sit the patient up from the right side, perform the Roll Test by having the patient turn their head quickly from center to the left. After 30 sec, have them quickly turn the head from center to the right. After 30 sec, have the patient sit up. If the patient had vertigo and nystagmus during the Roll Test, perform the Bow and Lean Test next. If the patient did not have nystagmus and vertigo during the Roll Test, perform the Sidelying Test. If you are still unable to provoke the nystagmus and vertigo, ask the patient to perform whatever movement/position they believe will produce their symptoms.

Nystagmus

The characteristics and direction of the nystagmus noted during testing will determine which of the semicircular canals is involved (Table 20-4). In general, BPPV associated with semicircular canals that are oriented in the vertical plane will present with a combination of vertical and torsional nystagmus, and BPPV associated with the horizontal semicircular canals will present with horizontal and torsional nystagmus. Most cases of BPPV involve the posterior canal. BPPV involving the posterior semicircular canal accounts for up to 85% of the total cases, with cases of horizontal SCC being from 5% to 13.6% and anterior SCC from 1.2% to 13%.[5,40]

■ Table 20-4 **NYSTAGMUS FEATURES BY CANAL AFFECTED IN BPPV**

Canal Affected	Initial Response in Dix-Hallpike
Posterior	Upbeating and torsional (torsional toward affected ear)
Horizontal: canalithiasis	Geotropic (right-beating in head right position, left-beating in head left position)
Horizontal: cupulolithiasis	Apogeotropic (left-beating in right head position, right-beating in head left position)
Anterior	Downbeating and torsional (torsional toward affected ear)

Both the Dix-Hallpike and the Sidelying Test have been used to identify Anterior SCC BPPV. In both tests, the contralateral anterior SCC is in the plane of the pull of gravity (see Figs. 20.3 and 20.4). The anterior canal of the ipsilateral (downside) ear is also in a dependent position, and if the head is sufficiently below the horizontal, the maneuver may trigger vertigo because of anterior SCC involvement of the ipsilateral ear. Thus, with both the Dix-Hallpike Test and the Sidelying Test, identification of the involved side in Anterior SCC should be based on the direction of the torsional component of the nystagmus rather than which ear is dependent during testing. The nystagmus in anterior canal BPPV is *downbeating* and torsional towards the affected side.

Algorithm for Treatment of BPPV

Figure 20.7 is an algorithm for arriving at the appropriate treatment for BPPV. Identification of the affected

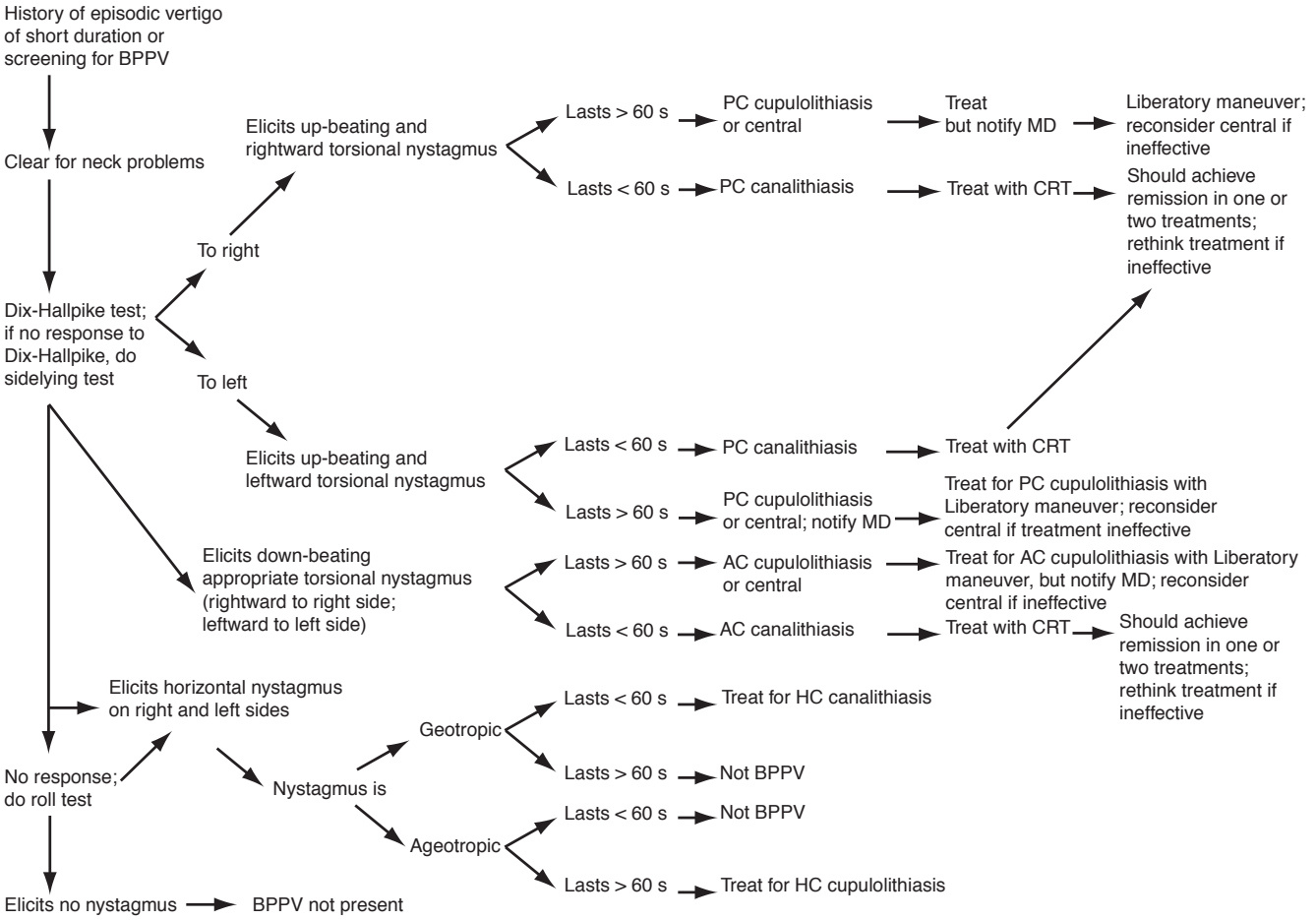

Figure 20.7 Algorithm for assessment leading to treatment of benign paroxysmal positional vertigo (BPPV). Identification of the direction and duration of the nystagmus leads to the determination of the canal involved and whether the BPPV is from canalithiasis or cupulolithiasis. This information directs the appropriate choice of treatment. AC = anterior canal; BPP = benign paroxysmal positional vertigo; CRT = canalith repositioning treatment; MD = medical doctor; PC = posterior canal.

side, which canal is involved (based on direction of nystagmus), and whether the problem is caused by canalithiasis or cupulolithiasis leads to specific treatments or to consideration that the nystagmus may be of central origin.

Evidence-based Treatments

Successful treatment is dependent on identifying which canal is involved and whether the debris is free-floating or adhering to the cupula. There are three basic bedside treatments for BPPV, each with its own indications for use: Canalith Repositioning, and Liberatory and Brandt-Daroff Habituation Exercises. Variations of these treatments have been developed depending on which semicircular canal is involved. Studies on the efficacy of these treatments that provide strong evidence that these treatments result in remission exist primarily for the Canalith Repositioning Treatment for Posterior SCC BPPV.[41-44] Some evidence now supports the treatment of Horizontal SCC BPPV.[45] The results of these studies must be interpreted cautiously, however, because of the high incidence of spontaneous remission that occurs in patients with BPPV. Several authors report that spontaneous recovery occurs within 3 to 4 weeks of onset,[46,47] but others suggest that spontaneous recovery may still occur even several months from onset.[48] Imai et al reported in 2011 that the natural course of remission in the apogeotropic form of horizontal SCC BPPV is an average of 13 days, and the geotropic form is an average of 16 days.[49]

For a given patient, the choice of which exercise is most appropriate depends on which canal is involved and whether the patient has the canalithiasis or cupulolithiasis form of BPPV.

Repositioning Treatment: Treatment of Posterior SCC Canalithiasis Form of BPPV

CASE: Your patient is a 74-year-old woman who experienced vertigo with nausea when she rolled over in bed in the morning about 12 days ago. She thinks the vertigo lasted less than a minute, although she continued to feel off-balance for the rest of the day. She now avoids lying down and has been sleeping on three pillows. She was seen by a physician three days ago, who diagnosed BPPV based on the patient's history and her complaints of vertigo when she was tested using the Dix-Hallpike maneuver. However, he did not have Frenzel or infrared goggles and was unable to observe nystagmus. He treated her with the Canalith Repositioning Treatment and referred her to you for follow-up care. On examination today, she has an upbeating and rightward torsional nystagmus with vertigo that lasts for 8 seconds when moved into the right Dix-Hallpike position. She states, however, that the vertigo is 25% better than it was when the physician treated her three days ago. You are considering repeating the treatment for the canalithiasis form of right posterior SCC BPPV.

The canalith repositioning treatment (or maneuver or procedure), initially proposed by Epley, was developed based on the theory that debris free-floating in the long arm of the SCC could be moved through the canal and back into the utricle.[50,51] This treatment approach has been studied extensively and is now commonly used for the management of posterior SCC positional vertigo.

The original Canalith Repositioning Procedure (CRP) (Fig. 20.8) was developed for treatment of posterior SCC BPPV and consisted of the following five key elements[50]:

1. Premedication of the patient
2. Specific positions used in the maneuver
3. The timing of shifts from one position to another
4. Use of vibration during the maneuver
5. Post-maneuver instructions

As more and more clinicians have used the CRP, numerous modifications of the procedure have been proposed. These modifications encompass elimination of premedication regimens, positions used, timing, use of vibration and post-treatment instruction. Some of the modifications resulted in less optimal outcome, while others simplified the CRP as proposed by Epley without affecting outcome. We will use the term "canalith repositioning treatment (CRT)" to distinguish the modified treatment approaches from the originally proposed procedure (Epley Maneuver).

1. Premedication of the patient: In the original treatment, patients were premedicated approximately 1 hour before treatment using either transdermal scopolamine or diazepam.[50] The purpose of premedicating a patient with a history suggesting BPPV is to reduce nausea and prevent vomiting during testing and treatment. In contrast to the original procedure as proposed by Epley, most reports do not mention medicating patients before treatment.[52-55] We recommend that although most patients do not need premedicating, an exception would be the patient with emesis associated with BPPV. In that situation, the use of an antiemetic medication such as promethazine HCL or prochlorperazine (Phenergan® and Compazine® respectively) would be appropriate and humane. As an alternative or supplement to medication, we suggest that the patient with a history of significant nausea and vomiting be moved *slowly* into the provoking position. If no vertigo or nystagmus is evoked, the positioning maneuver can be repeated more rapidly. Furthermore, if a vertical and torsional nystagmus and vertigo occur during positional testing, the patient should immediately be taken through the CRT rather than subjecting the patient to repeated provoking tests.
2. Specific positions used in the maneuver: the specific sequence of head positions recommended involves moving the head in 90-degree increments so the "debris" will migrate through the long-arm of the posterior canal and into the utricle (see Fig. 20.8). Note that with each change in head position, the debris should move away from the cupula of the posterior semicircular canal. Therefore, *the direction of the observed nystagmus should be the same in each position*. The original series of position changes suggested by Epley has remained unchanged since his original paper. Herdman et al (1993) compared the use of the five positions suggested by Epley with use of only four positions, omitting the position in which the patient's head is turned so that the nose is down toward the floor, and found a significant difference in the rate of remission.[11] When only four positions were used (sitting to Dix-Hallpike position to opposite Dix-Hallpike position to sitting), a 57% rate of remission was achieved. In contrast, when all five positions were used

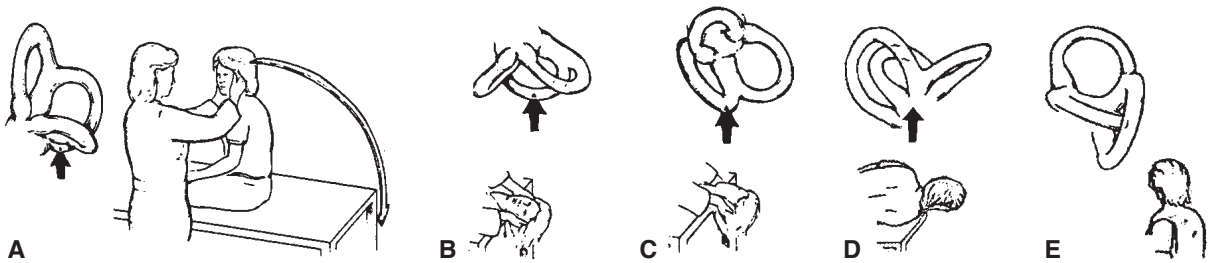

Figure 20.8 Treatment used for the canalithiasis form of both posterior and anterior SCC BPPV (arrows point to location of debris in the left posterior SCC). In the Canalith Repositioning Treatment, the patient is first taken into the Dix-Hallpike position (**A** to **B**) and kept there until the nystagmus stops or for twice the duration of the initial nystagmus. The patient then rolls his/her head, with assistance from the clinician, slowly through moderate extension toward the unaffected side (**B** to **C**) and kept in the new position until the nystagmus stops or for twice the duration of the initial nystagmus. The patient is then rolled onto the unaffected side with the head turned 45 deg down (toward the floor) and kept there until the nystagmus stops or for twice the duration of the initial nystagmus (**C** to **D**). In each of these positions, the patient may experience a short spell of vertigo as the otoconia move away from the cupula through the posterior canal. Finally, keeping the head deviated toward the unaffected side, the patient then slowly sits up (**D** to **E**) and can then straighten the head. The same maneuver would be used for the canalithiasis form of anterior SCC BPPV on the left side. (Modified from Tusa RJ and Herdman SJ. Canalith Repositioning for Benign Positional Vertigo. Education Program Syllabus 3BS.002, *Amer Acad Neuro*, Minnesota, 1998, p 13.[5])

or when the four-position maneuver was repeated several times, remission was 83%. Several studies have shown that when patients are moved through four positions multiple times, a 93% remission rate can be achieved.[11,55] Wolf et al found a 93.4% remission rate in patients treated with only the first four positions when multiple repositioning treatments are performed during the same clinical visit.[55] Therefore, it seems that either approach—using five positions or using four positions but repeating the maneuver multiple times—is effective. However, for patient comfort, using the five-position sequence is recommended rather than repeating the four-positions maneuver multiple times.

3. The timing between position changes within the maneuver: the total timing between the position changes in the CRT is based on the combination of the latency until nystagmus begins plus the time until the nystagmus stops. Once the patient's head has been moved into a new position, that position is maintained until the nystagmus slows down. At that point, the head is moved into the next position in the sequence. If nystagmus is not observed in any given position, which happens frequently, then the timing is based on the duration of nystagmus established during the initial Dix-Hallpike maneuver. Several studies have used a longer period between each change in position[11,55,56] with essentially the same results as found by Epley.[50] It is useful to know that it is not necessary to move the patient through the different positions quickly. Waiting longer between changes in positions allows the nausea to decrease and helps to prevent actual emesis.
4. Use of vibration during the maneuver: in the original procedure, Epley suggested that two different applications of mechanical vibration to the mastoid of the affected side be used during treatment. During one cycle, a standard electromagnetic bone conductor vibrator is used, and then in another cycle of the maneuver, a handheld vibrator (80 Hz) is used.[50] The theoretical purpose of the vibration was to prevent the debris from adhering to the walls of the canal and to aid in the movement of the debris through the canal. Several studies have failed to identify any difference in outcome whether a vibrator is used or not used,[57-59] and a Cochrane review concluded that there was no evidence that mastoid oscillation was beneficial.[60] Epley also advocated the use of mastoid vibration in the case of blockage of the SCC by the debris, or "canal jam," such as when a patient has no nystagmus on testing.[61] However, it is important to recognize that in a case of a complete "canal jam," there would be no nystagmus or complaints of vertigo with change in head position. The debris would essentially block movement of the endolymph, and therefore, the cupula would not be displaced.

5. Post-treatment instruction: The last component of the original CRT was the post-treatment instruction. The patient was advised to keep the head upright for 48 hours following the treatment, including sleeping with the head elevated 45 degrees. The follow-up visit for reassessment, and if necessary another treatment, would be scheduled for 1 week later. Asking patients to keep the head upright for 48 hours, presumably preventing the loose otoconia in the utricle from moving back into the posterior semicircular canal, is difficult for many patients. Patients would report not sleeping well or a sore neck. Fortunately, these post-treatment instructions do not appear to be necessary to the success of the treatment[60,62,63] Massoud and Ireland[62] examined the need for post-treatment instructions of staying upright for 48 hours. Two groups of patients were given the CRT or another treatment (the Liberatory maneuver) and were asked to sleep upright for 2 nights and then on the normal side for an additional 5 nights. Two other groups were given the same maneuvers but were not given any post-maneuver instructions other than to avoid brisk head movements for 1 week. All patients had a follow-up visit at 1 week after treatment. They found no statistical difference in the four groups (analysis of variance, P greater than 0.2). The success rate of the maneuvers was 88% to 96%. Similar results were reported by Nuti et al,[63] who examined 52 patients at 20 minutes, 24 hours, and 7 days after the Liberatory maneuver for posterior canal BPPV. A Cochrane Review found a statistically significant difference in remission rates between patients with and without post-treatment instructions but concluded that the clinical impact was insignificant. In summary, there does not appear to be a strong benefit to requiring patients to remain upright overnight after the canalith repositioning maneuver. Having patients stay upright for as little as 20 minutes after the maneuver may be sufficient for remission of BPPV.

There is strong evidence to support the use of the canalith repositioning treatments as safe and effective treatments compared with no treatment for posterior canal BPPV (Table 20-5). Cochrane reviews yielded a statistically significant effect in favor of the Epley Maneuver over control, in terms of complete resolution of symptoms and conversion from a positive to a negative Dix-Hallpike Test without serious adverse effects.[41,42] The reviews support the use of the Epley Maneuver as a safe, effective treatment for posterior canal BPPV, based on the results of 292 patients in five mostly randomized controlled trials with the importance of spontaneous remission of BPPV involving the posterior SCC, citing 20% to 38% of control patients were found to have a negative Dix-Hallpike maneuver at follow-up.[41,42] Helminski et al (2010) and Teixeira et al (2006) performed systematic reviews of the literature with similar results related to treatment of posterior canal BPPV through the use of canalith repositioning procedures, including the Epley Maneuver.[43,44] Both conclude that canalith repositioning procedures are more effective than controls in managing PC BPPV. Complications of nausea, vomiting, fainting, or conversion to horizontal canal BPPV occurred in 6% to 12% of patients.[53] Most controlled randomized treatments have used repeated repositioning maneuvers within one treatment session. Gordon and Gadoth (2004), however, found no significant difference in outcome between those patients treated with repeated maneuvers within a single session (92% remission) and those patients treated with only one maneuver (80%).[64]

CASE: Your patient is a 49-year-old man who experienced vertigo when he was hanging wallpaper about 24 days ago. He thinks the vertigo is brief but, when asked, states that he always moves his head when the vertigo starts. He was seen by a physician 2 weeks ago and was given a prescription for Valium® but has taken it only twice, because it makes him sleepy. He has not returned to work. The physician has referred him to you for treatment of his vertigo. On examination today, oculomotor tests are normal except for an upbeating and rightward torsional nystagmus with vertigo that lasts for 60 seconds when moved into the right Dix-Hallpike position, at which time you moved him out of the Dix-Hallpike position. You decide that the patient has the cupulolithiasis form of right posterior SCC BPPV because of the persistence of the nystagmus and vertigo.

In 1988, Semont et al proposed a single "liberatory" maneuver designed to move the debris from the posterior SCC and reposition it back into the utricle (Fig. 20.9).[65] The technique uses inertia and position changes against gravity to move the debris mechanically. The treatment has several names including Semont maneuver, Liberatory treatment, or brisk treatment.[11,65,66] Presumably, the rapid acceleration and deceleration of the movement from the initial sidelying position to the second (opposite side) sidelying position (Fig. 20.9) can dislodge debris adhering to the cupula of the SCC and thus is considered as a treatment for posterior SCC cupulolithiasis as well as a treatment for posterior SCC canalithiasis.

■ Table 20-5 TREATMENT EFFICACY FOR POSTERIOR SCC CANALITHIASIS

Treatment Maneuver	References	# of Subjects	Outcome
Systematic review modified Epley vs. Sham	Helminski et al 2010[44]	Of 10 studies identified via literature review, 2 RCTs double-blinded, 2 Quasi-RCTs met inclusion.	Odds in favor of resolution of BPPV were 22 times (95%, CI 3.41–141.73) and 37 times higher (95%, CI 8.75–159.22) in persons receiving CRP vs. sham.
Systematic review Epley vs. Sham	Teixeira and Machado 2006[43]	5 RCT trials found.	Positive evidence of efficacy for Epley's maneuver at 1 week and 1 month.
Systematic review modified Epley vs. Sham	Hilton and Pinter 2004[42]	Of 22 studies that met inclusion criteria, 5 RCTs were included; others were generally poor methodological quality.	Epley maneuver is safe and effective treatment of Posterior canal BPPV vs. sham. Patients who received Epley completely resolved symptoms (95%, CI 1.84–13.16) and conversion to negative Dix-Hallpike (95%, CI 2.21–14.56)
RCT, prospective, single-blinded; modified Epley vs. Sham	Lynn 1995[47]	n = 36 Conversion to negative Dix-Hallpike	89% cure rate for CRP group vs. 27% cured in Sham group
RCT, prospective, single-blinded	Froehling et al 2000[67]	n = 50 Conversion to negative Dix-Hallpike	67% cure rate for CRP vs. 38% cure rate for Sham
RCT, modified Epley vs. Sham	Munoz 2007[68]	n = 81 Conversion to negative Dix-Hallpike	Single treatment: 34.2% cure rate for CRP group vs. 14.6% in Sham group; Resolution of dizziness: 31.6% of CRP group and 24.4% of Sham group. One week later, CRP group and Sham group both received the CRP, and 61.8% and 57.1%, respectively, had negative DH test results.
RCT, modified Epley vs. Sham	Von Brevern 2006[69]	n = 67 Conversion to negative Dix-Hallpike	80% cure rate for CRP, 10% cure rate for Sham
RCT, modified Epley vs. Sham	Yimtae 2003[70]	n = 58 Conversion to negative Dix-Hallpike	75.9% cure rate for CRP, 48.2% cure rate for Sham

Semont and colleagues reported on a large cohort of patients with BPPV (n = 711) treated over an 8-year period.[65] They found a remission rate of 84% after a single treatment and of 93% after two treatments. Recurrence of the symptoms was infrequent (4%). In a prospective, randomized study that included 30 subjects treated with the Semont maneuver, Herdman et al (1993) reported remission of symptoms and/or signs of significant improvement in 90% of the cases after a single treatment. [11] In a later study, Ireland (1994) examined the effectiveness of the Liberatory maneuver in a series of patients using the patients as their own controls.[71] The patients were first treated with the Liberatory maneuver but on the *unaffected* side; none of the patients had relief from their symptoms. The patients were then "treated" with just the post-maneuver instructions to keep the head upright for 48 hours. Again, at the end of 1 week, all patients were symptomatic. Finally, the patients were treated with the Liberatory maneuver on the *affected* side. At the end of 1 week, all patients were symptom-free. Although the

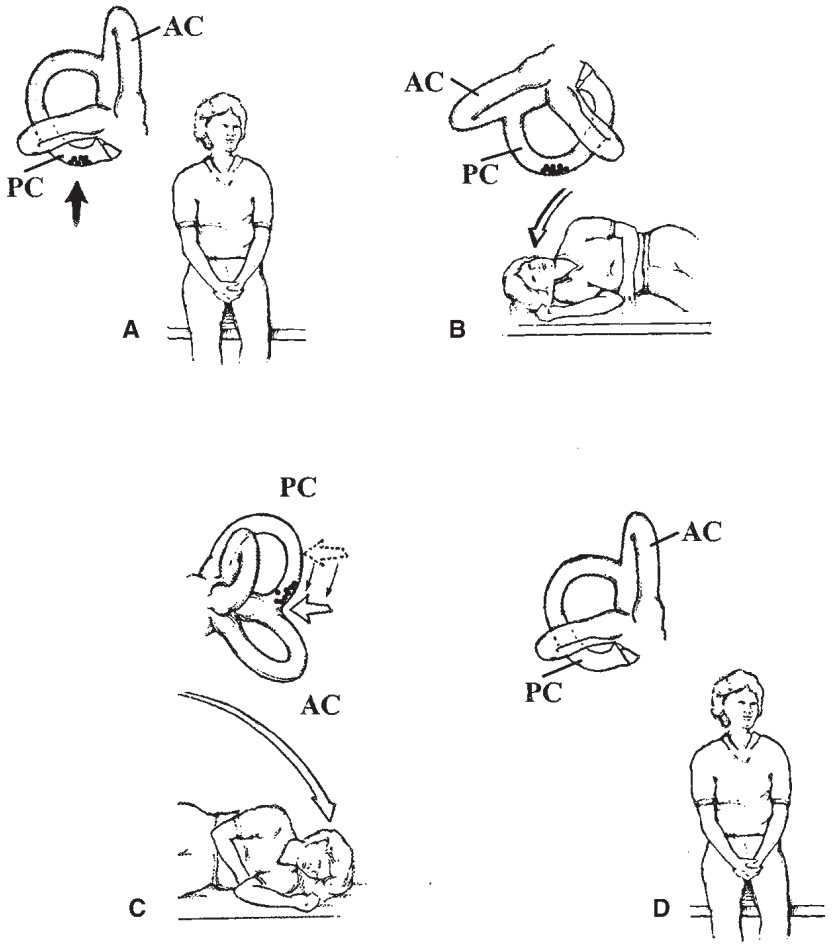

Figure 20.9 In the Liberatory maneuver for posterior SCC BPPV (12-5.2), the patient sits on the examination table sideways and the head is rotated 45 deg toward the unaffected side **(A)**. The patient is then moved quickly onto the affected side (parallel to the plane of the affected posterior SCC) **(B)**. After 1 minute, the patient moves rapidly, with assistance, through the initial sitting position to the opposite side while the head is still positioned 45 deg toward the unaffected side **(C)**; nose will now be 45 deg down toward the floor). The patient holds this position for 1 minute and then, keeping the head turned toward the unaffected side, moves slowly to a sitting position **(A)**.

number of subjects in this study is small (n = 10), the results support the effectiveness of the Liberatory maneuver as treatment for BPPV. Other studies have reported somewhat lower remission rates. Hausler and Pampurik (1989) and Levrat et al (2003) reported remission of symptoms in 51% and 62.6% of their subjects treated, respectively.[72,73]

The Liberatory maneuver can also be used in patients with canalithiasis, in which the success rate is as high as 86.8% after one treatment.[74] In a placebo-controlled, double-blinded study, there was a significantly greater rate of remission in patients treated using the Liberatory maneuver (86.8%) compared with a sham treatment (0%) at 1 and 24 hours after treatment.A second study showed similar findings at 4 days post-treatment with 84% remission in the treated group compared with 14% in a sham treated group of patients with PC SCC BPPV.[75]

Because the Liberatory treatment may dislodge material adhering to the cupula and move the debris toward the utricle, we believe this is the optimal treatment for the cupulolithiasis form of BPPV of the posterior SCC. However, Semont did not describe the nystagmus in his large series of patients, and the incidence of cupulolithiasis is relatively infrequent. Therefore, it remains unclear what the treatment efficacy is for the Liberatory or Semont maneuver with the cupulolithiasis form of BPPV. Evidence suggests that it is also appropriate for patients with the canalithiasis form of posterior SCC BPPV. It is important to note that the Liberatory maneuver may be difficult to perform with elderly patients, because of the quickness of movement required of the procedure.

Treatment of Anterior SCC BPPV

CASE: Your patient is a 36-year-old female who needed to be seen urgently in your clinic because of complaints of severe vertigo. The patient has not gone to a physician at the time of your initial evaluation. She reports that she woke up with severe complaints of vertigo and vomiting

yesterday, and although she is no longer vertiginous, she does not feel well and is somewhat off-balance. Past medical history is not significant; the only medication she is taking is for birth control. She rates her vertigo as of yesterday as a 9/10 on a visual analogue scale and her dizziness and imbalance today as a 6/10. Oculomotor exam in sitting is normal. You perform the Dix-Hallpike Test slowly because of the vomiting she had yesterday; she develops a rapid downbeating and leftward torsional nystagmus in the left Dix-Hallpike position, and she describes vertigo concurrent with the nystagmus. The nystagmus and vertigo last for approximately 20 seconds. She has some nausea but no vomiting, and you proceed to treat her with the CRT for left anterior SCC BPPV.

The evidence for treatment efficacy for Anterior SCC BPPV canalithiasis is limited to descriptive studies (Table 20-6). The majority of patients treated seem to respond to the CRT starting on the affected side (see Fig. 20.8).[76,77] Complicating the issue of which might be the most effective treatment is the fact that there are at least seven proposed treatments for Anterior SCC BPPV canalithiasis, some with only a few subjects.[76-85] Additionally, some studies used the same treatment for both the canalithiasis and cupulolithiasis forms of anterior SCC BPPV.[77]

■ Table 20-6 TREATMENT EFFICACY FOR ANTERIOR SCC BBPV CANALITHIASIS

Treatment Maneuver	References	# of Subjects	Outcome
Canalith repositioning maneuver starting on affected side	Lopez-Escamez et al 2006[31]	14	71% at day 30
	Jackson et al 2007[77]	55	100% in 1.32 maneuvers
Canalith repositioning maneuver starting on unaffected side	Korres et al 2008[78]	2	100%
Sitting to supine with head hanging to head flexed 30 degrees in 30-second intervals	Yacovino et al 2009[31]	13	84.6% in one maneuver; 100% in two
Reverse CRM – starting with nose down toward affected side; roll to back, head still, toward affected side; turn head 45 degrees opposite side, sit up	Zapula 2008[80]	1	100%
	Honrubia et al 1999[81]	41	50%
Partial roll – start on unaffected side with nose down, then nose horizontal, then nose up 45 degrees all in 30-second increments; then sit up for 3 minutes; repeated 2 to 4 × daily	Rahko 2002[82]	57	92.98% at 1 week
Dix-Hallpike to unaffected side for 2 minutes; then elevate head to level 1 minute and then return to sitting with chin tucked (30 degrees)	Kim et al 2005[83]	30	40.7% after one and
		5	92.3% after three maneuvers
	Korres et al 2008[84]		60% in one treatment; 100% by two treatments
Forced Prolonged Position – supine head hang 60 degrees for 30 minutes then in head vertical for 24 hours	Crevits 2004[85]	2	100%

Treatment of Horizontal SCC Canalithiasis BPPV

CASE: Your patient is a 21-year-old woman who came to your department with severe vertigo and imbalance. She was accompanied by her father and was dependent on him for support while walking. Her exam was unremarkable except for vertigo and a geotropic nystagmus during the Roll Test that was worse in the right roll position. Because the geotropic nystagmus was brief, less than 20 seconds in duration, in the right and left roll positions, you conclude she has the canalithiasis form of right horizontal SCC BPPV.

Numerous treatments have been proposed for the treatment of Horizontal SCC canalithiasis (Bar-B-Que Roll, 270 degree roll, the modified Liberatory maneuver for horizontal SCC canalithiasis, and Forced Prolonged Positioning) and for Horizontal SCC cupulolithiasis (Gufoni for cupulolithiasis, Forced Prolonged Positioning for cupulolithiasis).

Bar-B-Que Roll Treatment for Horizontal Canalithiasis

The CRP as proposed by Epley for the treatment of posterior SCC canalithiasis has been modified for horizontal SCC BPPV into a treatment referred to as the Bar-B-Que or Roll treatment.[86,87] This treatment is performed as a full 360-degree roll or as a 270-degree roll. In both treatments, the patient should be asymptomatic by the time they are in the prone position (Fig. 20.10).

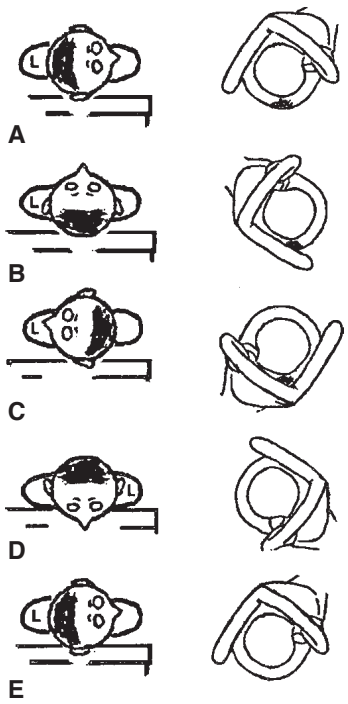

Figure 20.10 Bar-B-Que roll or Canalith repositioning treatment (CRT) for horizontal SCC BPPV. **(A)** The patient lies supine on the examination table or bed with the affected ear down (shown here for right horizontal SCC BPPV). **(B)** The patient's head is then slowly rolled away from the affected ear until the face is pointed up; this position is maintained for about 15 sec or until any vertigo stops. **(C)** The patient then continues to roll the head in the same direction until the affected ear is up. This position is also maintained for 15 sec or until the dizziness stops. **(D)** The patient then rolls the head and body in the same direction until the face is down and stays in that position for 15 sec. At this point in the treatment, the patient should be asymptomatic if the treatment has been effective. The patient can either sit up by moving first to a hands and knees position and then sitting sideways or can get off the treatment table by sliding one leg to the floor, keeping the head straight ahead. Alternatively, the head and body are rolled in the same direction to the original position with the affected ear down, and then the patient slowly sits up, keeping the head level or pitched down 30 deg. These two variations of the CRT for horizontal SCC BPPV are referred to as the 270-degree roll and the 360-degree roll, respectively. Patients can be taught to perform this treatment at home. The follow-up visit is usually scheduled for within a few days of the treatment. (Modified from Tusa and Herdman, 1998.[5])

The Liberatory Maneuver for Horizontal Canal Canalithiasis or Appiani Maneuver

The theory underlying the use of the Liberatory maneuver for treatment of horizontal SCC canalithiasis is to move the head in such a way as to cause debris floating freely in the horizontal SCC to move into the utricle (Fig. 20.11). Although called the "Liberatory maneuver" by Appiani and colleagues (2001), it is a treatment for canalithiasis and not, as with Semont's 'Liberatory' maneuver for posterior canal BPPV, a treatment that would be effective for cupulolithiasis.[88]

Forced Prolonged Position (FFP) for Horizontal Canal Canalithiasis

Another treatment option for horizontal SCC canalithiasis BPPV is Forced Prolonged Position (Fig. 20.12) described by Vannucchi and colleagues.[89] The patient goes to bed and lies in sidelying on the affected side (more symptomatic side) for 30 to 60 seconds and then slowly rolls over toward the healthy ear until he or she is in sidelying with the healthy ear down. He or she then remains lying on the unaffected side all night. The forced prolonged position can be combined with the roll treatment by having the patient perform a roll treatment followed by utilizing the FFP at night. Should the client need to get up at night, the maneuver is repeated.

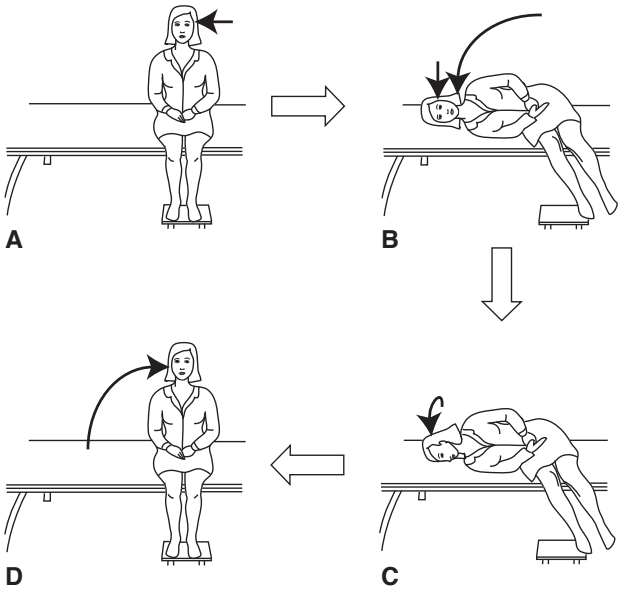

Figure 20.11 In the Appiani treatment (shown here for left-sided HC canalithiasis), the patient starts sitting on the treatment table **(A)**. The patient is quickly brought to a side-lying position on the *unaffected* side and stays in this position for 2 min **(B)**. The patient's head is rapidly rotated 45 deg down toward the table, again staying in this position maintained for 2 min **(C)**. The patient is slowly brought back to sitting and the cervical spine returned to a neutral position. This treatment has also been referred to as the Gufoni treatment for canalithiasis **(D)**. (From Appiani GC, et al, 2001;).[88]

Treatment of Horizontal SCC Cupulolithiasis

The Casani (or Gufoni) maneuver for cupulolithiasis is also known as the modified Semont maneuver for horizontal Canal Cupulolithiasis and is the only treatment for horizontal SCC cupulolithiasis for which there is evidence of treatment efficacy (Fig. 20.13).[90] Casani et al (2002) report remission of symptoms in 75% of patients with one maneuver. The proposed mechanism for this treatment is a rapid movement designed to dislodge otoconia from the cupula of the horizontal SCC. If necessary, it can then be followed by the roll treatment to move the now liberated debris out of the horizontal SCC.[91]

Treatment Efficacy Horizontal SCC BPPV

Until recently, the evidence for treatment efficacy for horizontal SCC BPPV has been limited to descriptive and case studies (Tables 20-7 to 20-9). Complicating the issue of which treatment might be the most effective is the fact that the same treatment is sometimes advocated for both canalithiasis and cupulolithiasis. Another complicating issue is the interchangeable use of several authors' names

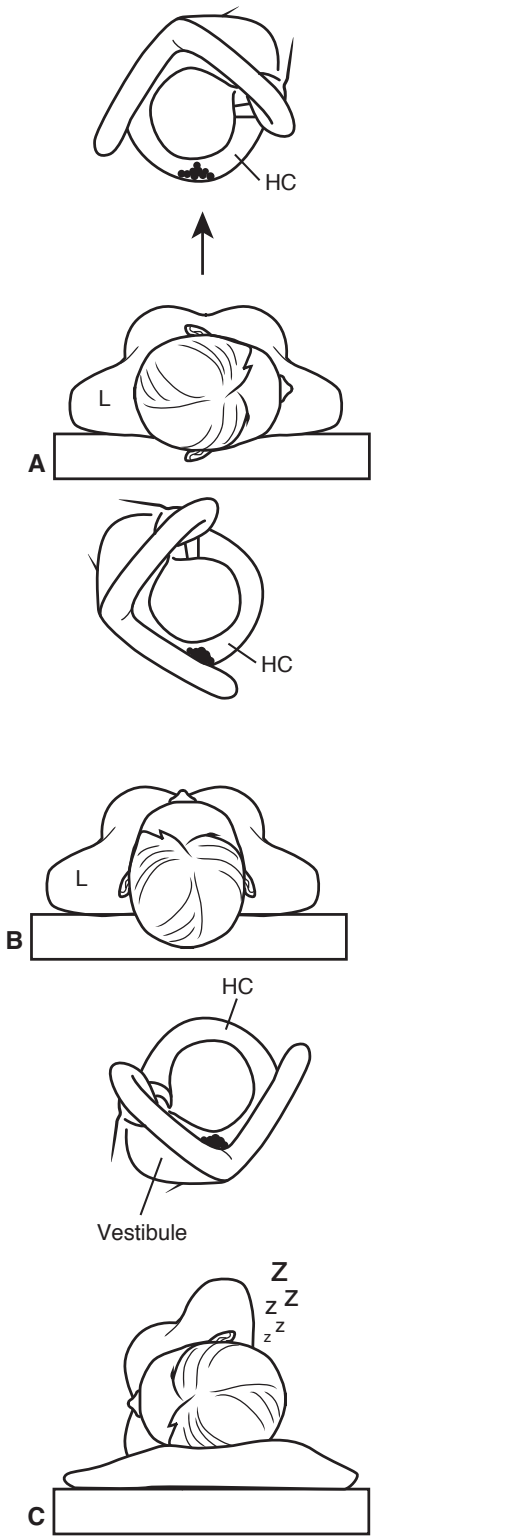

Figure 20.12 Shown here for the right-side canalithiasis of the HSCC, in the Forced Prolonged Position treatment, the patient lies on the affected ear **(A)** for 20 sec or more, and then slowly rolls toward the healthy ear until the healthy ear is down **(B** and **C)**. The patient remains in this position all night.

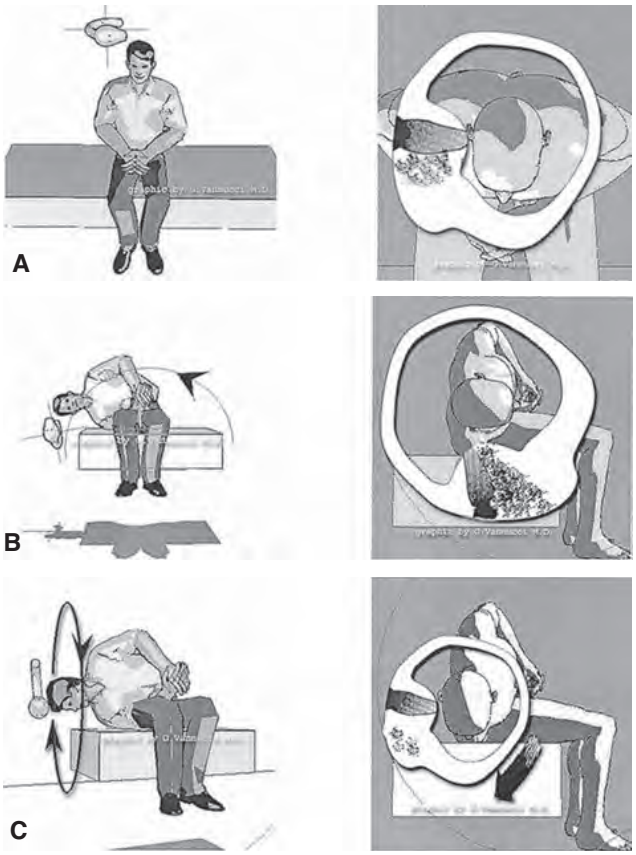

Figure 20.13 The primary treatment for HSCC cupulolithiasis is the Casani maneuver. In the Casani maneuver (also known as the Gufoni maneuver), the patient moves quickly from sitting **(A)** to side-lying position on the *affected* side **(B)**. The patient then quickly turns the head so the nose is down 45 deg **(C)** and remains in that position for 2–3 min before sitting up again. This treatment has also been referred to as the Gufoni treatment for cupulolithiasis. (From Casani et al, 2002.[90])

for the same treatments. A recent randomized study compared the Roll and Appiani (called Gufoni in article) treatments to a sham treatment in 170 consecutive patients with the canalithiasis form of horizontal SCC BPPV.[92] Both treatments resulted in a higher remission rate (69.1% and 60.9%, respectively) than a sham treatment (35.4%) on the day of treatment. At 1 month later, patients treated with either the Roll or the Appiani treatments were still significantly better than those receiving the sham treatment.

Another study compared the Appiani and Casani maneuvers (referred to as Gufoni maneuvers for horizontal SCC canalithiasis and horizontal SCC cupulolithiasis, respectively) to a modification of the maneuvers in which patients moved to the sidelying position in stages rather than in a single rapid movement.[93] Both geotropic and apogeotropic forms of BPPV were treated. For patients with the geotropic form, patients lay toward their unaffected side, and for patients with the apogeotropic form of BPPV, patients lay toward the affected side. Treatment using the modified form resulted in a similar remission rate (93%) as treatment with the original form of these maneuvers (88%). However, the modified form resulted in only a 2% conversion to PC-BPPV compared with a 16% rate for the original maneuvers.[93]

Nonspecific Treatment: Brandt-Daroff Exercises

Brandt and Daroff were the first to offer a treatment of BPPV.[48] The treatment consisted of repeated movements into and out of provoking head positions on a serial basis (Fig. 20.14).

In 1980, Brandt and Daroff studied the efficacy of a treatment on 67 patients with BPPV. The patients were supervised performing the exercise every 3 hours. Within 3 to 14 days, 66 of the patients experienced complete resolution of symptoms. Although they named the treatment "habituation exercises," they suggested that recovery occurred too quickly for there to be a habituation effect (some in 24 hours). Instead, Brandt and Daroff proposed a mechanical means to promote loosening and ultimate dispersion of the otolithic debris from the cupula.[48] Based on studies in guinea pigs, individual otoconia particles dissolve in endolymph within 100 hours.[46] Therefore, the Brandt-Daroff treatment may partially work by breaking up otoconia to allow them to dissolve. In contrast to Brandt and Daroff's early findings, when these Brandt-Daroff exercises were compared with CRT, patients treated using Brandt-Daroff had poorer outcome (25% remission) at 7 days post-treatment compared with patients treated with CRT (80.5% remission).[94]

Comment on Testing and Treatment

Clinicians often express concern that the rotation and extension of the neck during testing and treatment may be contraindicated in some patients. However, careful screening can alleviate these concerns, or if the clinician is still unsure about proceeding with testing or treatment, the positions can be modified (consider the use of a tilt table) so the *labyrinth* is in the correct position *without rotation or extension of the neck*. Humphriss et al (2003) recommend that the following parameters be met in every patient before the Dix-Hallpike test is performed.[95]

1. With the patient seated, can the head be rotated 45 degrees to the right for 30 seconds and then to the left for 30 seconds without pain?
2. With the patient seated, can the head be rotated 45 degrees to either side and extended back for 30 seconds without pain?

■ Table 20-7 TREATMENT EFFICACY FOR HORIZONTAL SCC CANALITHIASIS

Treatment Maneuver	References	# of Subjects	Outcome
Supine with head straight ahead, then three rapid 90-deg rolls toward unaffected ear, then sit (Lempert maneuver)	Lempert and Tiel-Wilck 1996[86]	2	100%
Lempert maneuver: Supine with head straight ahead, then three rapid 90-deg rolls toward unaffected ear, then sit (n = 29) Control: untreated (n = 34; consented but all at different hospital from treated group)	Sekine et al 2006[96]	63	At 1 week: treated 78.6%; untreated control group 69.1% (no significant difference)
Forced prolonged positioning: lie supine, rotate head and body to unaffected side, lie in that position for 8–12 hours	Vannucchi et al 1997[89]	74	90% remission for FPP in 3 days Pseudo randomized; control group untreated sample of convenience
Vannuchi maneuver followed by FFP	Nuti et al 2005[97]	18	88.8% symptom and nystagmus free with one treatment series; 100% in two
Patient sits w/ head straight ahead, quickly lies on *unaffected* side for nystagmus duration plus 1 minute, quickly turns head 45 degrees down for 2 minutes, slowly sits up	Appiani et al 2001[88]	32	78.12% with one maneuver; 100% with two maneuvers
After converting HSCC BPPV from cupulolithiasis to canalithiasis, treated for canalithiasis form using Gufoni for canalithiasis (aka Appiani) or FPP	Appiani et al 2005[91]	16	100% remission from canalithiasis
Gufoni maneuver: Patient sits w/ head straight ahead, quickly lies on *unaffected* side 2 minutes, quickly turns head 45 degrees down for 2 minutes, slowly sits up; then repeated	Francesco et al 2009[98]	58	79.3% complete resolution with one session; 13.8% converted to PSCC BPPV; 6.9% no benefit
Randomly assigned to 360 Bar-B-Que roll plus FPP for 12 hr (n = 54) or to Gufoni (aka Appiani) maneuver for canalithiasis (n = 58)	Casani et al 2011[99]	132	Bar-B-Que + FPP: 61% remission with one treatment; 79.6% with three treatments
Untreated control group (refused treatment or could not be treated) (n = 20)			Gufoni (aka Appiani) 86% remission with one maneuver; 93.1% with three treatments

■ Table 20-8 TREATMENT EFFICACY FOR HORIZONTAL SCC CUPULOLITHIASIS

Treatment Maneuver	References	# of Subjects	Outcome
Cupulolithiasis (n = 9) Seated, quickly lie on *affected* side, turn head down 45 degrees and stay in that position for 2–3 minutes, rapidly sit up (called Casani or Gufoni for cupulolithiasis)	Casani et al 2002[90]	9	Resolution in 44.4% in one, 55.5% in two, and 66.6% in three maneuvers
FFP: lie on side of *weaker* nystagmus for 12 hr	Chiou et al 2005[100]	29	93.1% cases resolved with FFP
1. Lie on side of *weaker* nystagmus all night for two weeks (FPP-one); 2. For those who converted from cupulolithiasis to canalithiasis with FFP-one; Lempert maneuver 3. For those with no responses to FFP-one; lie on side of *greatest* nystagmus all night for 2 weeks 4. If converted to canalithiasis, treated with Lempert maneuver (n = 1)	Boleas-Aguirre et al 2009[101]	22	1. 68% (n = 15) free of vertigo and nystagmus 2. Converted to canalithiasis (n = 1) treat with Lempert or to vertical canal (n = 3) treat with Epley; 100% resolution 3. No response to FFP-one (n = 3), then treated with FPP-two with 66% resolution

■ Table 20-9 STUDIES TREATING BOTH CANALITHIASIS AND CUPULOLITHIASIS FOR HORIZONTAL SCC

Form of HSCC BPPV	Treatment Maneuver	References	# of Subjects	Outcome
Canalithiasis (n = 55)	Bar-B-Que roll followed by forced prolonged positioning	Casani et al 2002[90]	82	Resolution in 67.4% with one, 80% in two, and 89.1% in three maneuvers
Cupulolithiasis (n = 9)	Seated, quickly lie on *affected* side, turn head down 45 degrees and stay in that position for 2–3 minutes, rapidly sit up			Resolution in 44.4% in one, 55.5% in two, and 66.6% in three maneuvers
Control (n = 18)	Vestibular suppressant medication; sample of convenience			Resolution in 16.6% in 1 week; 44.5% in 2 weeks
Cupulolithiasis Utricular-side	FFP: lie on side of weaker nystagmus for 12 hr	Chiou et al 2005[100]	29	93.1% cases resolved with FFP
Canalithiasis (n = 36) and Cupulolithiasis (n = 10)	Bar-B-Que roll for 360 degrees in 90-degree steps; each position held for 30 sec	Escher et al 2007[102]	46	74% resolution after one maneuver; 80% after two; 85% after maximum of three; form of BPPV had no affect on outcome
Canalithiasis (n = 18) Cupulolithiasis (n = 13)	BBQ Roll Modified Brandt-Daroff (not described)	Jackson et al 2007[77]	31	BBQ Roll: Average number of treatments to remission = 1.34 Modified Brandt-Daroff: Not reported

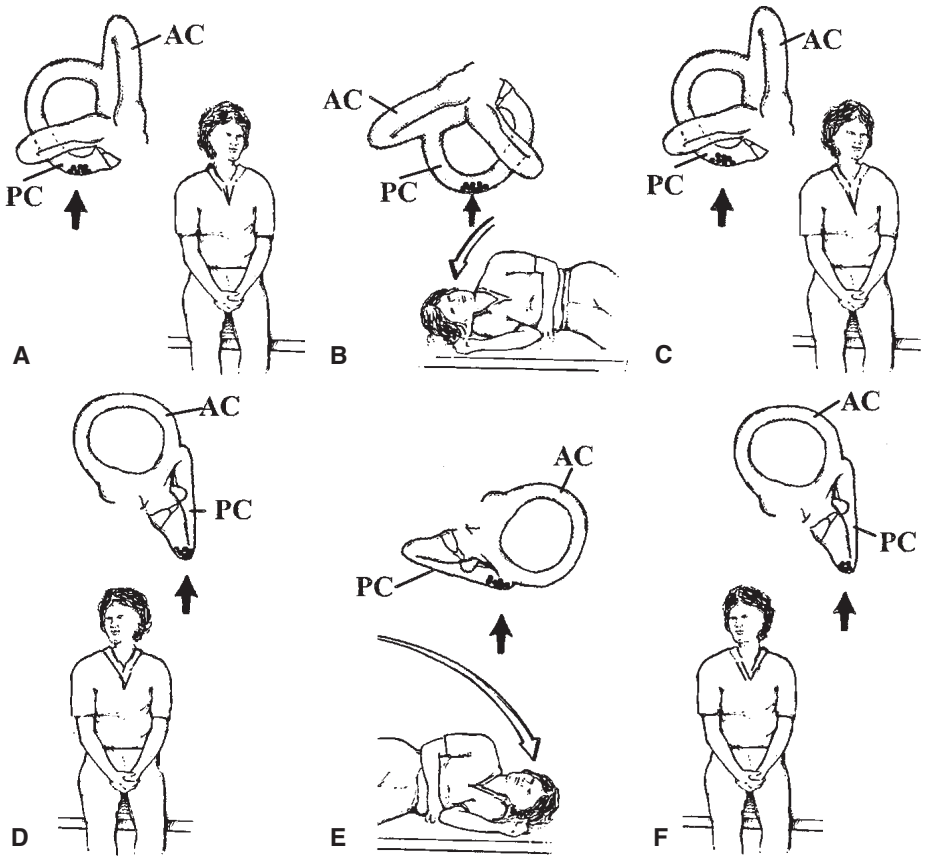

Figure 20.14 Brandt-Daroff treatment for treatment of posterior semicircular canal benign paroxysmal positional vertigo. **(A** and **B)** The patient is moved quickly into the side-lying position on the affected side (shown here as right side) and stays in that position until 30 sec after the vertigo has stopped. **(C)** The patient then sits up and again waits for the vertigo to stop. The patient then repeats the movement to the opposite side **(D)**, stays there for 30 sec after vertigo stops **(E)**, and sits up **(F)**. The entire treatment is repeated 10 to 20 times, three times a day, until the patient has no vertigo for 2 days in a row. AC = anterior canal; PC = posterior canal. *Black arrows* indicate position and movement of debris. (Modified from Tusa and Herdman, 1998.[38])

Humphriss et al (2003) consider a number of disorders to be absolute contraindications to performing the Dix-Hallpike test, and presumably also to performing some of the treatments (Table 20-10).[95] We would suggest that most individuals with these disorders can be tested and treated, but certain precautions must be taken to ensure patient safety, especially avoiding extension and rotation of the patient's neck.

Managing Persistent Imbalance in Patients with BPPV

CASE: Your patient is a 76-year-old woman with the canalithiasis form of posterior SCC BPPV. On her initial visit, you appropriately treated her with a CRT, as that was her primary problem. She has returned for her second visit 7 days later. She reports that she has had no vertigo since the treatment but feels very off-balance and staggers at times when walking. On her assessment today, she has no nystagmus or vertigo in the right or left Dix-Hallpike positions, and you conclude that her BPPV is in remission. She rates her disequilibrium while walking as a 4.1/10. Her balance confidence (ABC score) is a 67%. She can perform the Romberg test for 30 seconds with eyes open and closed, but her single leg stance time is only 3 seconds. Her fall risk score (DGI) is an 18/24, and her gait speed is slow for age and gender. One possibility is that her balance will probably improve over the next 2 weeks, but you are concerned that with her age, gender, single leg stance time, and DGI score, she is at a greater risk for falling than usual. Therefore, you decide to give her exercises to help improve her balance.

Over 50% of patients with posterior SCC BPPV subjectively report imbalance.[103,104] Imbalance relative to age-matched controls has been quantified using computerized static and dynamic sway platform testing.[105-111] The CRT and Liberatory maneuver may completely resolve positional nystagmus and vertigo based on repeat Dix-Hallpike Test and improved balance based on repeat computerized platform testing,[105,106,108,111] but 8% to 14% of patients may still complain of residual imbalance.[11,112,113] One possibility is that this imbalance is caused by BPPV that is not sufficient to deflect the cupula during testing. Another possibility is an underlying vestibular hypofunction that was not detected during the initial evaluation.[114] Several studies have suggested that a concurrent vestibular

■ Table 20-10 PRECAUTIONS TO TESTING AND TREATMENT

Precautions	Signs	Symptom Presentation
• Cervical spine instability e.g., atlantoaxial instability • Occipitoatlantal instability e.g., RA, Down's syndrome	Unsteadiness with walking	Neck pain, facial pain, headaches, paresthesia in hands, feet, face, or tongue; head and neck feeling unstable. Dizziness and vertigo.
• Prolapsed intervertebral disk with radiculopathy	Restricted neck movements. Muscle weakness in the arm and/or hand	Severe neck pain with referred pain in arm, hand, and fingers. Paresthesia and numbness in the arm, hand, and fingers, down the line of nerve distribution.
• Cervical myelopathy	Unsteadiness on feet	• Pain in neck and/or arms. Loss of power in arms, hands, legs. Numbness or paresthesia in hands and/or feet. Increased tone in limbs.
• Arnold Chiari malformation	Downbeating nystagmus with neck extension	• Occipital headaches aggravated by coughing or sneezing; in severe forms – ataxic gait, progressing to spastic quadriparesis, severe bulbar problems, and respiratory difficulties.
• Vascular dissection syndromes	Horner's syndrome	• Facial and neck pain, acute retro-orbital pain.
• Previous cervical spine surgery		• Patient may be symptom-free or have residual neck and/or arm pain.
• Acute trauma to neck ("whiplash") – Contraindicated if insufficient ROM of cervical spine		• Neck, arm, facial, trunk pain with restricted neck movement. Vertigo, dizziness symptoms. Symptom severity will vary.
• Rheumatoid arthritis	Arthritic deformities, especially feet/hands, knees/elbows.	• Neck pain and restricted movement. May have signs of cervical spine instability (see above).
• Carotid sinus syncope	• Fainting/blackouts on turning head or buttoning up shirt, or pressure on neck.	
• Aplasia of the odontoid process	Cervical spine instability	• Neck pain, transient neurological symptoms

Modified from Humphriss RL, Baguley DM, Sparkes V, et al. Contraindications to the Dix-Hallpike manoeuvre: a multidisciplinary review. *International J Audiology*. 2003;42:166-173.[21]

hypofunction is not the underlying cause of this residual imbalance. Blatt et al[106] and Di Girolamo et al[105] quantified the degree of imbalance in patients with posterior canal BPPV who did not have evidence of vestibular hypofunction (based on bithermal caloric testing) using computerized dynamic posturography before and after CRT. Approximately 66% of the patients did not improve to normal values by age after CRT, yet none of the patients had signs or symptoms of BPPV.

So what is the cause of persistent imbalance in patients in remission from BPPV? One possibility is the age of the patient. In the study by Blatt et al, those patients

who improved to within normal values by age were younger ($p < 0.05$).[106] No difference was found between groups for time from onset of BPPV, history of falls, or the patients' rating of the intensity of vertigo, although the patients' subjective rating of the intensity of disequilibrium approached significance. Another possibility is that the increased postural sway by some individuals on these tests may be a result of a separate, preexisting problem with balance that occurs in the elderly, such as disuse disequilibrium, which would not improve after CRT. Alternatively, it is possible that it takes time for the otoliths to fully recover following CRT. This latter explanation is supported by the study of Giacomini et al, who found that balance on a static platform did not fully recover in patients with BPPV until 12 weeks after CRT.[105] This has significant implications for the rehabilitation of patients with BPPV. If balance does not improve with treatment of the BPPV, then patients should be reexamined to rule out vestibular hypofunction or some other cause for imbalance. If appropriate, patients should then be referred for specific exercises to improve balance.

Unraveling Complicated Cases

Unfortunately, not all patients with BPPV have obvious signs and symptoms, nor do they necessarily have involvement of only one semicircular canal. The cases below illustrate some of the more confusing presentations. All patients in the cases below were tested using Frenzel or IR goggles. Additionally, Figure 20.15 presents an algorithm that may provide some assistance in making decisions when confronted by difficult cases.

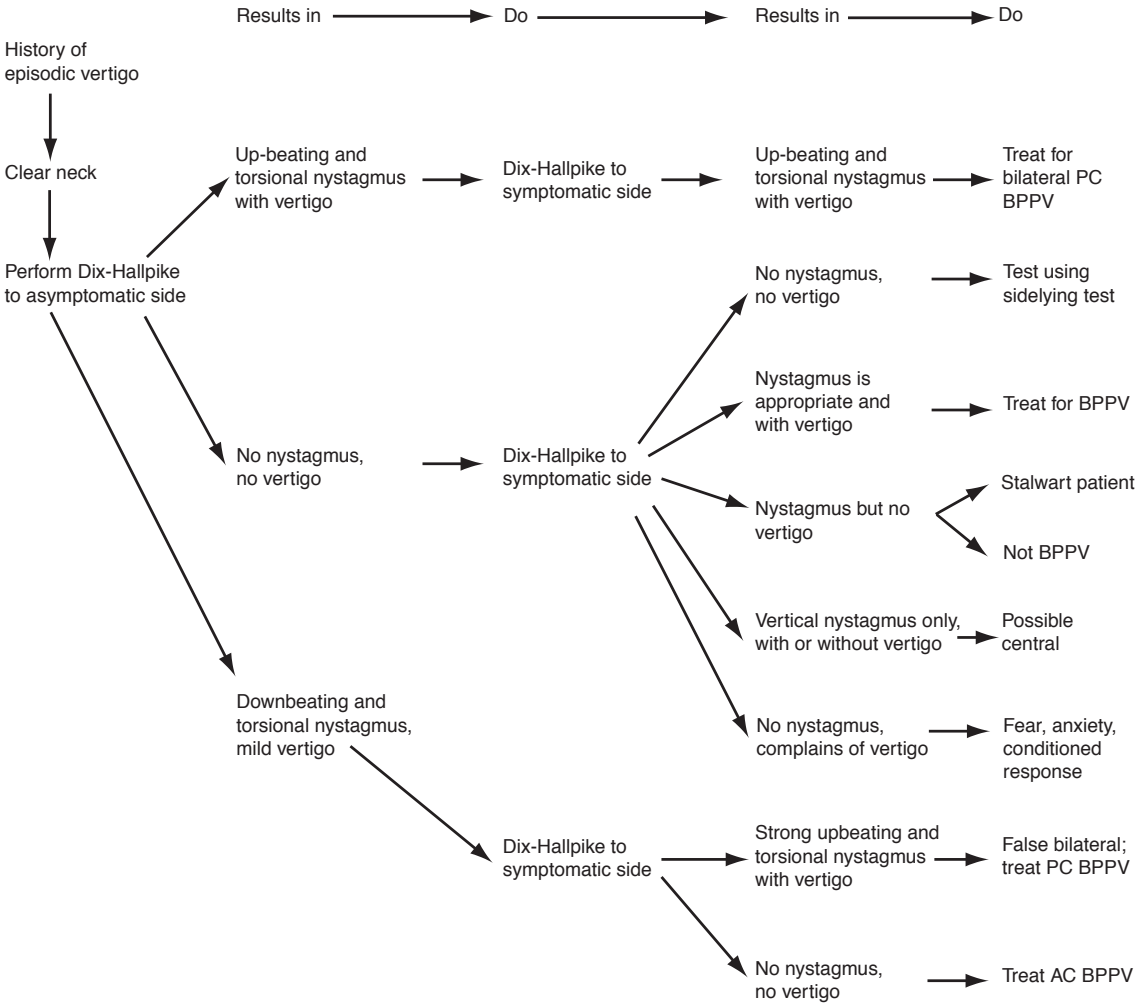

Figure 20.15 Algorithm for assessing, interpreting, and treating complicated cases of benign paroxysmal positional vertigo.

CASE STUDY 20-1

Your patient has a history of vertigo that lasted for days and is now positional. On examination, the patient has normal pursuit and saccadic eye movements. There is no nystagmus in room light. There is a positive head thrust test during head movement to the left. Using infrared goggles, you observe an upbeating and leftward torsional nystagmus in the left Dix-Hallpike position and the patient concomitantly complains of vertigo. The initial nystagmus resolves within 15 seconds, and then a persistent right-beating nystagmus without complaints of vertigo is observed. In the right Dix-Hallpike position, there is a persistent right-beating nystagmus again without vertigo.

Explanation

The patient may have a left vestibular neuritis affecting the superior branch of the vestibular nerve and resulting in a left horizontal SCC hypofunction. The left UVH would explain the direction-fixed, right-beating nystagmus without vertigo in both the right and left Dix-Hallpike positions. The superior branch of the vestibular nerve also supplies the utricle and anterior canal but not the saccule or posterior canal. Thus, the patient could have degeneration of the utricle with release of otoconia, which could then move into the still functioning posterior SCC causing posterior SCC canalithiasis BPPV. It is estimated that approximately 10% of all patients with vestibular neuritis will also develop BPPV.[115]

Resolution

Treat the left-sided BPPV first because that is the cause of the patient's primary complaint of vertigo. When the patient is in remission from BPPV, ask the patient if that has resolved all the symptoms. If it has not, and they report imbalance or vertigo with quick head movements, ask them if there has ever been an episode of vertigo that lasted for one day or more (consistent with vestibular neuritis). Then perform head thrust test to confirm horizontal canal hypofunction and refer the patient for a caloric test. If the patient has unilateral vestibular hypofunction, begin vestibular rehabilitation as necessary.

CASE STUDY 20-2

The patient has no complaints of vertigo when moved into the left Dix-Hallpike position but does have a persistent downbeating nystagmus upon observation. When moved into the right Dix-Hallpike position, the patient develops an upbeating and rightward torsional nystagmus (with concurrent complaints of vertigo) that decreases over 15 seconds but then becomes a persistent downbeating nystagmus.

Explanation

The upbeating and torsional nystagmus with vertigo that lasted only 15 seconds is consistent with the canalithiasis form of posterior SCC BPPV. The downbeating nystagmus observed after the upbeating and torsional nystagmus resolves is not simply a reversal of the primary nystagmus of BPPV. First, it is persistent. If it were a reversal of the BPPV nystagmus, it would last only seconds. Second, it was observed when the patient was in both the right and the left Dix-Hallpike positions. The persistent downbeating nystagmus in this case suggests a central problem in addition to the BPPV (Appendix 20A).

CASE STUDY 20-3

When you test the patient in the left Dix-Hallpike position, there is a vigorous upbeating and leftward torsional nystagmus associated with intense complaints of vertigo lasting 25 seconds. In the right Dix-Hallpike position, there is a downbeating and rightward torsional nystagmus with only mild complaints of vertigo lasting 15 seconds.

Explanation

There are two possibilities: either the patient has the canalithiasis form of posterior SCC BPPV on the left side and the canalithiasis form of anterior SCC BPPV on the right side, or this is an example of false bilateral BPPV. In the latter situation, the patient actually has posterior SCC BPPV on the left side; the mild observation of downbeating and torsional nystagmus appearing concurrently with mild complaints of vertigo that occurs when the patient is in the right Dix-Hallpike position is actually produced by the posterior SCC BPPV on the left side. The orientation of the labyrinth when the person was in the right Dix-Hallpike position was such that it caused the otoconia in the left posterior canal to drift *toward* the cupula. False bilateral BPPV is more common than true bilateral BPPV.

Resolution

Treat the patient for left posterior SCC canalithiasis BPPV. If the nystagmus and vertigo resolve on both sides, then it was a false bilateral BPPV. If, after you achieve resolution of the posterior SCC BPPV on the left, the patient still has downbeating and rightward torsional nystagmus when in the right Dix-Hallpike position, then it was a true case of bilateral BPPV—go ahead and treat for the anterior SCC canalithiasis BPPV.

CASE STUDY 20-4

Your patient has a history of vertigo when he lies down and rolls to the right. He has avoided lying on his right side since the first episode three weeks ago. You explain BPPV to him and what the testing entails before performing testing. In the left Dix-Hallpike position, he has no nystagmus or vertigo but starts sweating slightly. In the right Dix-Hallpike position, he has no nystagmus but complains of dizziness and becomes markedly diaphoretic. These symptoms resolve in a several minutes once the patient returns to a sitting position.

Explanation

It is quite possible that your patient had BPPV, but it has already resolved. The patient, however, has become fearful, and there is a conditioned autonomic nervous system response when the patient is moved into a position that previously provoked the vertigo.

Resolution

First, explain to the patient that he probably had BPPV. Reassure the patient that it is not unusual to have a response to the position that had provoked the initial episode. However, the patient still needs to be able to tolerate lying down. This is a situation in which the habituation approach would be appropriate (see Chapters 22 and 30). The patient can begin by moving into a semi-supine position—one that elicits very mild symptoms and gradually works toward becoming more comfortable lying down (see Chapter 30 for management suggestions).

CASE STUDY 20-5

You have been treating the patient for BPPV for 3 weeks but have not yet achieved resolution of the signs and symptoms. You started with CRT and did that treatment on two visits during the first week and even had the patient performing the treatment at home. When you asked the patient to demonstrate how they were doing the treatment, you noticed that the patient lifted her head when she rolled from the initial Dix-Hallpike position to the opposite side. You corrected that problem but after a week of performing the CRT at home, the patient still was not better. You then tried the Liberatory maneuver and taught the patient's spouse how to do the treatment at home. This was tried daily for another week without success. The spouse was able to demonstrate the treatment correctly, so that was not the problem. You then had the patient perform the Brandt-Daroff exercise at home for another week, still without resolution.

CASE STUDY 20-5

Explanation

Failure to respond to treatment for BPPV should be a "red flag" that something else may underlie the patient's signs and symptoms. The literature shows that 90% or more of patients with BPPV have complete resolution in one or two treatments.

Resolution

In the scenario presented above, the patient should be reevaluated by the referring physician for another disorder such as central positional vertigo.

CASE STUDY 20-6

Your patient is a 46-year-old male who was in a motorcycle accident 6 weeks ago. Although he does not remember what happened, his wife says that he suffered a concussion, several broken ribs, and considerable "road rash." He has been referred to you because of vertigo that occurs whenever he lies down. On testing, his oculomotor examination is normal except for the observation of brief downbeating and rightward torsional nystagmus, with complaints of vertigo, when he is in the right Dix-Hallpike position. You treat him for the canalithiasis form of right anterior SCC BPPV with the CRT. One week later, upon reassessment, there are no signs or symptoms suggesting BPPV. Three weeks later, the patient returns with a reoccurrence of his BPPV and again is successfully treated using the CRT, although two treatments are required. Five weeks later the patient again returns with complaints of positional vertigo, and testing shows he has right anterior SCC canalithiasis BPPV.

Explanation

Head trauma is the second most common cause of BPPV.[1] There is conflicting evidence as to whether or not reoccurrence of BPPV is more common following head trauma compared with the idiopathic form of BPPV. Two studies suggest that reoccurrence is more common in BPPV from head trauma than the idiopathic form of BPPV.[116,117] Other factors associated with reoccurrence are older age and vestibular neuritis.[116] However, another study, of 192 patients with BPPV from head trauma and 112 patients with idiopathic BPPV, failed to find those relationships.[118] There also is conflicting evidence as to whether anterior SCC BPPV is more common following head trauma than in the idiopathic form of BPPV. Dlugaiczyk et al reported that anterior SCC and multiple SCC involvement are more common in head trauma, but another study by Suarez et al failed to find any difference in which SCC was involved.[113,119]

CASE STUDY 20-7

Your patient is a 56-year-old male with complaints of vertigo and imbalance. His oculomotor examination is normal except for positional testing. In the right Dix-Hallpike position, you observe him to have a persistent left-beating nystagmus with complaints of vertigo, so you switch to the Roll Test. During the Roll Test, he has a persistent left-beating nystagmus with his head rolled to the right and a persistent right-beating nystagmus when his head is rolled to the left, with complaints of vertigo in both head positions. You follow this by performing the Bow and Lean Test, which is positive for left side involvement.

Explanation

The nystagmus exhibited during positional testing is apogeotropic, and this patient has the cupulolithiasis form of horizontal SCC BPPV.

Resolution

One treatment approach would be to perform the Casani maneuver (also known as Gufoni for cupulolithiasis, Fig. 20.13) for horizontal SCC cupulolithiasis and then recheck the patient on a subsequent visit. The other approach would be to treat for left horizontal SCC cupulolithiasis and then, on the assumption that the debris dislodged from the cupula is now floating freely in the long arm of the horizontal SCC, follow with a treatment for left horizontal SCC canalithiasis such as the Appiani or the 270-degree maneuvers.

CASE STUDY 20-8

Your patient is a 58-year-old woman with complaints of vertigo whenever she bends over or gets into or out of bed. Upon testing, she has brief right-beating nystagmus with vertigo in the right Dix-Hallpike position and brief left-beating nystagmus with vertigo in the left Dix-Hallpike position. Her vertigo is more intense when she is in the right Dix-Hallpike position. Additionally, when the left-beating nystagmus resolves, she develops a second brief bout of nystagmus and vertigo, but this time the nystagmus is upbeating and leftward torsional and reverses when she sits up.

Explanation

It would appear that this patient has multiple canal involvement with a combination of the canalithiasis form of right horizontal SCC BPPV (indicated by the geotropic nystagmus and her complaints of vertigo) and the canalithiasis form of left posterior SCC BPPV.

Resolution

In this case, start by treating the more symptomatic form of BPPV. When that has resolved, treat the other SCC involved.

Summary

The use of the CRT in the treatment of BPPV involving the posterior SCC is supported by a significant number of randomized controlled trials. Other forms of BPPV appear to be successfully treated, but the evidence is much weaker and more controlled trials are needed. Certainly, one key to successful treatment is identifying which canal is involved, what side the problem is on, and whether it is the canalithiasis or cupulolithiasis form of BPPV and then using the appropriate treatment, summarized in Table 20-11. The second key to successful treatment is focusing on the patient's understanding of the procedures you will use and their symptoms, for example, nausea, imbalance, and his or her level of anxiety.

■ Table 20-11 **MANAGEMENT GUIDE**

Semicircular Canal	Posterior or Inferior SCC	Anterior or Superior SCC	Horizontal or Lateral SCC	Unknown SCC
Provoking maneuvers	Dix-Hallpike or Sidelying tests	Dix-Hallpike or Sidelying tests	Roll Test, Bow and Lean test; may occur with Dix-Hallpike	All tests negative, may have occurred before coming to clinic
Nystagmus	Upbeating and torsional toward affected side	Downbeating and torsional toward affected side	Geotropic and brief if canalithiasis; apogeotropic and persistent if cupulolithiasis	Not observable
Affected side	Torsional toward affected side	Torsional toward affected side	Canalithiasis – beats toward affected side during 'bow'; cupulolithiasis – beats toward affected side in 'lean'	Could not be determined
Optimal treatment	Canalith repositioning treatment or Epley maneuver	Canalith repositioning treatment	Canalithiasis – 270-degree roll or the Appiani maneuver Cupulolithiasis – Casani maneuver	Brandt-Daroff
Precautions	May convert to AC or HC BPPV during treatment	Can be confused with central nystagmus, which is often downbeating, especially if persistent	Causes greater nausea, vomiting, and imbalance	May not be BPPV

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Appendix 20-A Differential Diagnosis: BPPV versus Central Postional Nystagmus and Vertigo

Richard A. Clendaniel, PT, PhD

Although benign paroxysmal positioning vertigo (BPPV) is a common finding that is relatively easy to diagnose and treat, there are causes of positional nystagmus and positional vertigo that are a result of either abnormalities within the central nervous system or other peripheral vestibular conditions. These conditions will not respond to the conservative measures described for the treatment of BPPV. The goal of this appendix is to help the clinician identify the signs and symptoms of positional nystagmus and positional vertigo that are not consistent with BPPV and are suggestive of other disorders.

Central Positional Nystagmus

Two types of central positional nystagmus have been identified: central positional nystagmus without vertigo (CPN) and central positional nystagmus with vertigo (CPV).[1,2] Central positional nystagmus without vertigo is characterized by nystagmus that persists as long as the head is held in the provoking position.[1] The nystagmus is typically in one direction (vertical, horizontal, or torsional), unlike the mixed vertical torsional nystagmus seen in posterior and anterior semicircular canal BPPV. Central positional nystagmus may be seen in elderly patients when they are in supine. The elicited nystagmus is typically vertical. In the absence of other findings on the examination, the CPN is thought to be benign. In other individuals, the CPN may be seen in conjunction with either upbeating or downbeating spontaneous nystagmus while the patient is seated. The CPN in these cases is typically greater than the spontaneous nystagmus observed in sitting.

The downbeating spontaneous nystagmus and CPN have been associated with a variety of central disorders including Chiari malformation, multiple sclerosis, olivopontocerebellar atrophy, and brainstem infarction.[3] These patients had other oculomotor signs such as impaired smooth pursuit and impaired VOR cancellation. The actual pathophysiology causing the downbeating nystagmus is not well understood at this point, but it is thought that the downbeating nystagmus results from an imbalance between the anterior and posterior semicircular canal pathways. Recall that the semicircular canal inputs are separated at the level of the vestibular nuclei into vertical (pitch), horizontal (yaw), and roll pathways. A lesion that causes either an increase in the central anterior semicircular canal pathways or a decrease in the central posterior semicircular canal pathways would lead to downbeating nystagmus.

The upbeating spontaneous nystagmus and CPN have been associated with central disorders such as tumor, stroke, and multiple sclerosis affecting brachium conjunctivum or the ventral tegmental tract.[4] Many of these patients also had findings of abnormal smooth pursuit. The presumed pathophysiology for the upbeating nystagmus is thought to be the opposite of that for the downbeating nystagmus. The upbeating nystagmus is caused by a higher level of neural activity in the central posterior semicircular canal pathways relative to the central anterior semicircular canal pathways.

Given the lack of symptoms of vertigo with the positional tests, the unidirectionality of the positional nystagmus, and the other oculomotor findings, it should not be difficult for the clinician to differentiate between CPN and BPPV.

Central positional nystagmus with vertigo can present with persistent, positional-induced nystagmus and

vertigo. In this type of CPV, the nystagmus is downbeating without a torsional component, and the nystagmus and vertigo persist as long as the head is in the provoking position. The nystagmus and vertigo do not fatigue, nor do they habituate with repeated testing. This pattern of CPV has been attributed to cerebellar tumors or to hemorrhage dorsolateral to the fourth ventricle.[5,6]

Unlike CPN, CPV may present in a manner remarkably similar to BPPV. Central positioning nystagmus with vertigo is often characterized by brief episodes of positioning vertigo and nystagmus, which has been called pseudo-BPPN.[2,7] Based on clinical findings and experimental animal studies, there is a large degree of overlap in the signs (latency, duration, and fatigability) as well as the symptoms associated with BPPV and CPV. For example, the latency to the onset of nystagmus in cases of CPV has been reported to be between 0 and 5 seconds, and in the case of experimental lesions to the cerebellar nodulus, up to 50 seconds.[8-12] Likewise, the duration of the elicited nystagmus and symptoms is typically relatively brief (5–60 sec) and mimics that seen in BPPV canalithiasis.[8,13] Unlike the duration of BPPV canalithiasis, the duration of the nystagmus in the animals with experimental lesions to the nodulus could persist up to 3 minutes, similar to that seen in BPPV cupulolithiasis.[12] Fatigability of the response to repeated testing within a short time frame, although not always tested in the clinic, is another hallmark of BPPV and may also be seen in individuals with CPV.[8,11,14] However, fatigability of the response is not seen in all patients with CPV, just as it is not seen in all patients with BPPV.[2,8]

Although there are similarities in the latency, duration, and fatigability of BPPV and CPV, there do appear to be differences in the patterns of the elicited nystagmus. The pattern of nystagmus in CPV can take a variety of forms. Findings from clinical studies have reported pure vertical (downbeating or upbeating), pure torsional, pure horizontal (either apogeotropic, or geotropic), and mixed vertical and horizontal patterns of nystagmus.[2,8,13-15] In animals with experimental lesions to the cerebellar nodulus, downbeat, downbeat with torsion, and horizontal patterns of nystagmus have been observed.[11,12] Although most of these patterns of nystagmus do not fit with BPPV, downbeating torsional nystagmus could be caused by anterior semicircular canal BPPV, and horizontal nystagmus could be caused by horizontal semicircular canal BPPV, making accurate diagnosis difficult. In addition, there are instances when the nystagmus seen in individuals with BPPV may not appear to fit the mixed vertical and torsional nystagmus seen in posterior and anterior semicircular canal BPPV. For example, the horizontal position of the eye in the orbit will accentuate either the vertical or torsional component of the nystagmus, which may make it difficult to determine if the nystagmus is mixed vertical and torsional, pure vertical, or pure torsional. Also, lid closure or rapid blinking may make it difficult to assess the pattern of nystagmus. Either scenario would increase the difficulty in making an accurate diagnosis.

Another differentiating feature between CPV and BPPV is the presence of other neurological findings and symptoms, which would not be expected in cases of BPPV. Associated symptoms, such as sudden hearing loss, tinnitus, fainting sensations, progressive imbalance, and vomiting have been reported in individuals with paroxysmal, positioning vertigo of central origin (CPV).[16,17] An abnormal oculomotor exam is often seen in individuals with CPV.[7] The presence of other neurological signs and symptoms, however, is not a requirement for CPV. There are numerous reports of individuals with CPV and no other signs or symptoms.[8,10,13,15] The similarities and differences between BPPV, CPV, and CPN are summarized in Table 20-A.

■ Table 20-A CHARACTERISTICS OF BPPV COMPARED WITH TWO COMMON FORMS OF CENTRAL VERTIGO AND NYSTAGMUS

Features	BPPV	CPV	CPN
Latency	1–15	0–5	0
Duration	5–60 seconds (longer in horizontal canal and in cupulolithiasis)	5–60 seconds	Persistent as long as the head is in the provoking position
Direction of nystagmus	In the plane of the stimulated canal. Most commonly mixed upbeating and torsion, may be downbeating and torsion, or horizontal	Pure vertical, pure torsional, or horizontal	Typically pure downbeat, may be pure upbeat or horizontal

■ Table 20-A CHARACTERISTICS OF BPPV COMPARED WITH TWO COMMON FORMS OF CENTRAL VERTIGO AND NYSTAGMUS—cont'd

Features	BPPV	CPV	CPN
Symptoms	Vertigo	Vertigo	No vertigo
Fatigability	Typical	Possible	No
Nausea and vomiting	Unusual with a single test; may develop with repeat testing	More frequently seen with a single test	No
Associated neurological signs and symptoms	None	None, cerebellar signs, or oculomotor signs	None, spontaneous nystagmus, or oculomotor abnormalities

Various causes of CPV have been reported in the literature, including cerebellopontine angle tumors, lesions affecting the cerebellar vermis or the fourth ventricle, and an infarction of the cerebellar nodulus.[15-17] A more common form of CPV is migrainous vertigo and positional nystagmus. Spontaneous, positional, or a combination of spontaneous and positional nystagmus can be seen during an acute migraine with associated symptoms of dizziness.[18] In these cases the nystagmus can be vertical, horizontal, or torsional and may change with different positional tests. In patients with a history of migrainous vertigo that were assessed when they were asymptomatic, 12% to 28% (depending on the time of assessment) demonstrated significant positional nystagmus.[19]

Other Causes of Positional Nystagmus/Vertigo

Vertebrobasilar insufficiency (VBI) has been reported to produce episodic bouts of vertigo and nystagmus.[20,21] These findings have been seen in both positional testing as well as in sitting with cervical rotation. Symptoms associated with VBI would be dependent of the position of the head on the trunk (neck position), rather than the position of the head in space (positional testing). Consequently, nystagmus and vertigo seen in the Dix-Hallpike Test should be reproducible in sitting with cervical extension and rotation if the cause of the signs and symptoms were a result of VBI. Vertiginous symptoms are common in cases of VBI, but isolated episodes of vertigo in VBI is controversial, with conflicting reports as to the incidence of this finding.[22,23]

Perilymphatic fistula and superior canal dehiscence can make the membranous labyrinth susceptible to pressure changes such as Valsalva maneuvers, or the Dix-Hallpike Test, where the head is hanging below the horizontal plane. These conditions are typically accompanied by hearing loss. The production of nystagmus and symptoms are not dependent on the position of the head in space and can often be reproduced in an upright (seated) position with the Valsalva maneuver.

Lastly, positional nystagmus may be seen following a unilateral vestibular loss. The nystagmus is horizontal (geotropic or apogeotropic), and the pattern of nystagmus will remain constant (geotropic or apogeotropic) in each Dix-Hallpike or Roll Test. Thus the nystagmus may appear similar to horizontal semicircular canal BPPV (canalithiasis or cupulolithiasis). Whereas individuals with horizontal semicircular canal BPPV are very symptomatic, individuals with positional nystagmus as a result of unilateral vestibular loss are either asymptomatic or have mild symptoms.

Summary

There are causes of positional vertigo and nystagmus not caused by BPPV. The challenge to the clinician is to identify the cause of the positional vertigo, such that appropriate treatment can be initiated. There are differences in the clinical presentation of the various causes of positional vertigo, but at times it may be difficult to determine whether the positional vertigo is a result of BPPV or another cause, such as CPV. The latency, duration, and symptoms may be very similar in cases of BPPV and CPV. There are generally differences in the pattern of elicited nystagmus in BPPV and CPV. The one consistent difference between BPPV and CPV is that BPPV will respond to maneuvers such as the canalith repositioning maneuver

or Semont maneuver, but CPV will not respond to these treatments. If the presumed BPPV is not responding to the therapeutic maneuvers, the clinician should be suspicious of possible CPV.

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CHAPTER 21

Physical Therapy Assessment of Vestibular Hypofunction

Susan L. Whitney, DPT, PhD, NCS, ATC, FAPTA
Susan J. Herdman, PT, PhD, FAPTA

Patients with peripheral vestibular hypofunction differ with respect to the onset and clinical course of their disability and to the final level of recovery, depending on the type and extent of vestibular deficit. Despite these differences, such patients have many of the same symptoms—dizziness, lightheadedness, vertigo, nystagmus, blurred vision, postural instability, fear of movement, gait disturbances, and occasional falling.[1] In addition, these patients may experience anxiety, depression, and fear related to their disability.[2-6] In fact, people with vestibular dysfunction report that they are significantly impaired by their disability.[7-9] As a result of one or more of these symptoms, patients with peripheral vestibular hypofunction often cope with their disability by avoiding certain movements and decreasing their activity level.[10] This habit, if not treated, will lead to the unfortunate results of physical deconditioning and an alteration of the patient's lifestyle.[10,11]

The purpose of this chapter is to provide an overview of patient problems and the key components of the clinical examination as well as the more comprehensive examination. We use the International Classification of Functioning, Disability and Health (ICF) scheme for the organization of this chapter.[12] The ICF was developed by the World Health Organization specifically to provide a framework for the "description of health-related states" that includes both positive experiences and negative consequences of disease.[12] This scheme consists of three domains that can be used to describe the effects of different disorders or diseases on a person's health, with a number of environmental and personal factors that affect each of those domains (Box 21-1).

The ICF model differs from other models of disablement in that it provides a more comprehensive depiction of the health of an individual. The model shifts the emphasis away from impairment and disability to a more balanced perspective that includes "health." Table 21-1 provides a description of how one can choose a self-report tool for use with persons with vestibular dysfunction based on which areas of the ICF are included in the measure (Fig. 21.1). Use of the ICF enables comparison of similar constructs across cultures to better describe rehabilitation outcomes.

Normal Structure and Function versus Impairment

Patients with vestibular hypofunction may express a multitude of symptoms. These symptoms emerge from functional deficits in vestibulo-ocular and vestibulospinal systems (Box 21-2) and from the results of sensory mismatch and physical deconditioning.[13]

Box 21-1

HEALTH CONDITION

Normal Function and Structure versus Impairment (body level)	Activities versus Limitations (individual level)	Participation versus Restriction (societal level)

Contextual Factors

Environmental Factors:		Personal Factors:	
e.g.	Natural environment	e.g.	Gender, age
	Support and relationships		Comorbidities
	Attitude of family		Social background
	Attitude of society		Education and profession
	Services, systems, policies		Past experience
	Products and technology		Coping and character style

■ Table 21-1 **MEASUREMENT TOOLS USED ACCORDING TO ICF DOMAIN**

	ICF Domain		
Tool	**Normal/Abnormal Structure and Function**	**Activity/ Limitation**	**Participation/ Restriction**
Head-thrust test	X		
Dix-Hallpike test	X		
Strength	X		
Range of motion	X		
Endurance	X		
Millon Behavioral Medicine Diagnostic	X		
Vestibular Coping Questionnaire	X		
Balance confidence (ABC scale)	X	X	
Gait speed		X	
Fall risk (Dynamic Gait Index)		X	
Dynamic Visual Acuity		X	
Physical Activities Scale for the Elderly		X	X
Medical Outcomes Study 36-Item Short-Form Health Survey			X
Sickness Impact Profile			X

■ Table 21-1 **MEASUREMENT TOOLS USED ACCORDING TO ICF DOMAIN—cont'd**

		ICF Domain	
Tool	**Normal/Abnormal Structure and Function**	**Activity/ Limitation**	**Participation/ Restriction**
Dizziness Handicap Inventory			X
Vestibular Disorders Activities of Daily Living Scale			X
Disability Score			X
Vestibular Activities and Participation Scale		X	X

Vestibulo-ocular Function and Dysfunction

The vestibulo-ocular reflex (VOR) is the primary mechanism for gaze stability during head movement. During movements of the head, the VOR stabilizes gaze (eye position in space) by producing an eye movement of equal velocity and opposite direction to the head movement. The ratio of eye velocity to head velocity is referred to as the *gain of the VOR*. The ideal gain in a normal subject would equal 1. VOR gain has been shown to be reduced to 25% in human beings immediately after unilateral labyrinthine lesions for head movements toward the affected side.[14,15] During the acute stage, VOR gain is also reduced to 50% for head movements toward the unaffected side.

Other eye movements, such as saccades and pursuit, are not affected by vestibular loss. Saccadic eye movements are rapid movements that allow refoveation of stationary targets. Pursuit eye movements enable the individual to visually follow a moving object across the visual field (smooth pursuit) without making compensatory head movements. Normally, the vestibulo-ocular, pursuit, and saccade systems work cooperatively to stabilize gaze during head movements.[16] If impaired smooth pursuit or hypermetric saccades are identified during the examination, the clinician should strongly consider that the person has central dysfunction.

Perception of Head Movement and Position

Normally, signals from the labyrinth provide accurate information concerning head movement and position. These vestibular signals are synchronized with visual and somatosensory inputs, and the nervous system is able to appropriately interpret the combination of signals. Whenever there is an acute or sudden asymmetry of vestibular function, of course, the brain interprets this abnormal signal as continuous movement of the head. The patient then experiences a spinning sensation even when he or she is not moving at all. Within a week, this asymmetry resolves and the spontaneous spinning sensation ceases. However, in patients with chronic vestibular dysfunction, the loss of a vestibular response to head movement becomes a persistent problem with actual movement of the head.

According to Norré,[17] the disturbed vestibular function produces a sensory input different from the one expected under normal conditions. This abnormal vestibular signal is in conflict with normal signals provided by the visual and somatosensory systems, and the resultant "sensory conflict" is thought to produce the symptoms associated with motion misperception.[17] Clinically, patients complain of lightheadedness or dizziness associated with particular head or body movements. Norré[17] has referred to this condition as "provoked vertigo," which is attributable to the asymmetry in the dynamic vestibular responses following a unilateral vestibular lesion.

Postural Instability

Independent and safe ambulation depends on the ability to successfully perceive the relevant features of one's environment while maintaining appropriate orientation of the body with respect to the support surface and gravity[18] Information necessary for postural control is derived from an integration of sensory inputs from the visual, somatosensory, and vestibular systems.[19]

Impairment in the function of vestibulospinal reflex (VSR) itself is believed to contribute to postural disturbances in patients with peripheral vestibular disorders. Lacour and associates[20] showed that unilateral vestibular neurotomy induces asymmetrical excitability in ipsilateral and contralateral spinal reflexes in baboons. Similarly ,

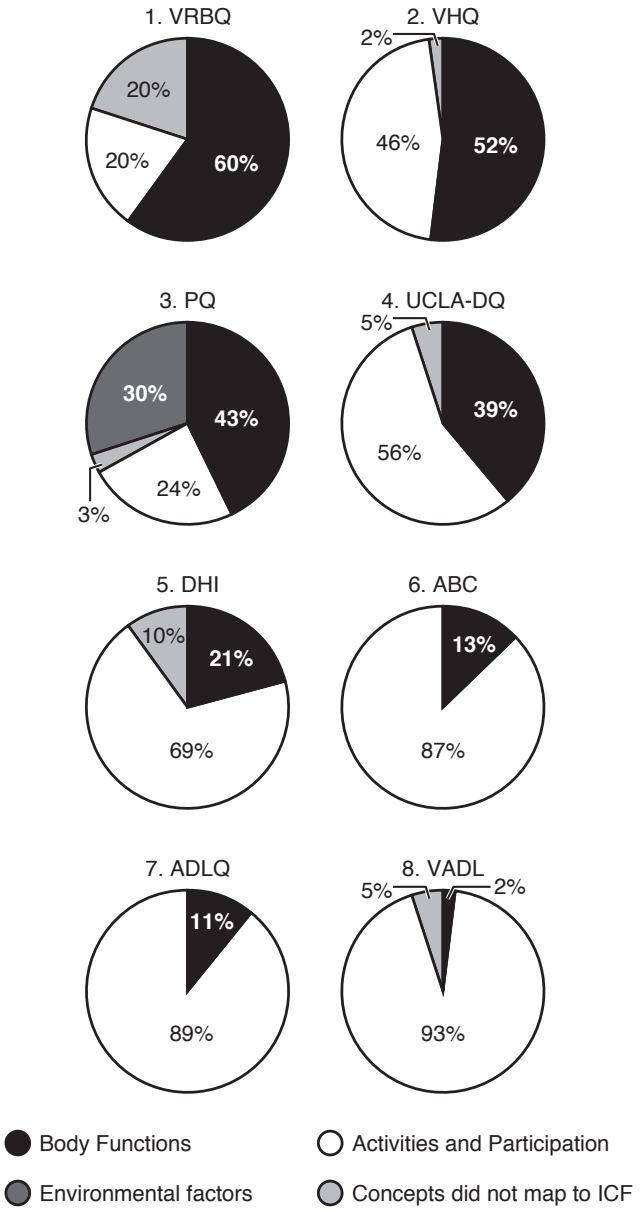

Figure 21.1 The WHO classification of several self-report instruments used to quantify change in vestibular rehabilation according to the ICF. 1. The Vestibular Rehabilitation Benefit Questionnaire (VRBQ), 2. The Vertigo Handicap Questionnaire (VHQ), 3. The Prototype Questionnaire (PQ), 4. The UCLA Dizziness Questionnaire (UCLA-DQ) 5. Dizziness Handicap Inventory (DHI), 6. The Activities-specific Balance Confidence Scale (ABC),[21] 7. The Activity of Daily Living Questionnaire (ADLQ), and 8. The Vestibular Disorders Activities of Daily Living Scale (VADL). Adapted with permission from the Journal of Physical Therapy, 2011.[22]

Allum and Pfaltz[23] reported that tibialis anterior responses in patients with unilateral peripheral vestibular deficits are enhanced contralateral to, and reduced ipsilateral to, the side of the lesion during support surface rotations. These patients also had reduced neck muscle activity and greater-than-normal head angular accelerations during response to the support surface rotations.

Cervical Range of Motion

Limitations in cervical range of motion can be a common clinical finding in patients with vestibular hypofunction, perhaps because of the head movement–induced symptoms and instability.[24,25] People self-restrict head movement to minimize the symptoms that occur with a change of head position and with head movement.

Physical Deconditioning

Changes in a patient's overall general physical condition (deconditioning) can be considered the most potentially disabling consequence of vestibular dysfunction. This finding may be associated with a patient's tendency to restrict movements that potentially provoke symptoms.

Activities versus Limitation

The loss or reduction of the VOR and vestibulospinal function translates into changes in function at the level of the individual. Poor VOR gain means that the person will have difficulty seeing clearly during head movements, especially unpredictable ones. Poor VSR results in changes in the person's confidence in balance, diminished gait speed, and an increased risk for falling. Patients with unilateral peripheral vestibular hypofunction frequently experience gait instability in situations that require them to move their heads while walking, to turn, or to stop quickly.[26-29] Walking with head movement appears to be particularly complex for persons with vestibular dysfunction,[30] and limited activity is often the result. In addition, clinical observation of the gait of a person with vestibular hypofunction commonly reveals numerous problems including:

- deviations such as veering left or right
- widened base of support
- decreased gait speed
- shortened step lengths
- decreased arm swing
- diminished ability to perform multiple tasks while walking
- occasional head or trunk tilting
- impaired perception of vertical
- decreased head and trunk motion[31-33]

For some patients, the use of an assistive device such as a cane reduces gait instability by acting as an additional proprioceptive cue [34-36] or by slowing gait speed.

Box 21-2

QUESTIONS TO ASK A PATIENT WITH A VESTIBULAR DISORDER

1. Do you experience spells of vertigo (a sense of spinning)? If yes, how long do these spells last?
2. When was the last time the vertigo occurred?
3. Is the vertigo spontaneous, induced by motion, induced by position changes?
4. Do you experience a sense of being off-balance (disequilibrium)? If yes, is the feeling of being off-balance constant, spontaneous, induced by motion, induced by position changes, worse with fatigue, worse in the dark, worse outside, worse on uneven surfaces?
5. Does the feeling of being off-balance occur when you are lying down, sitting, standing, or walking?
6. Do you stumble, stagger, or side-step while walking?
7. Do you drift to one side while you walk? If yes, to which side do you drift?
8. At what time of day do you feel best? _____ worst? _____
9. How many times per day do you experience symptoms?
10. Do you have hearing problems?
11. Do you have visual problems?
12. Have you been in an accident (e.g., motor vehicle)?
13. What medications do you take?
14. Do you live alone?
15. Do you have stairs in your home?
16. Do you smoke? If yes, please indicate how much per day.
17. Do you drink alcohol? If yes, please indicate how much.

Participation versus Restriction

As a result of many factors, including fear of falling, embarrassment about staggering while walking, pre-morbid personality, and discomfort with head movement–induced symptoms, patients may adopt a more sedentary lifestyle, frequently abandoning exercise routines or recreational activities they had pre-vestibular disorder.[10,37] If untreated, such changes could lead to more serious physical and psychosocial consequences, with significant restrictions in the person's participation in activities at the level of the society.

Physical Therapy Evaluation

The inclusion of different measurement tools to assess outcome in the ICF domains Normal/Abnormal Structure and Function (organ/body level), Activity/Limitation (person level), and Participation/Restriction (societal level) ensures that all possible problems associated with a patient's vestibular dysfunction are considered in planning the treatment program and in determining whether the treatment has been effective. Table 21-1 lists a variety of measurement tools according to ICF domain. These tools are discussed in the context of the full examination.

History

Medical History

When treating via direct access, the clinician may not have access to any information other than patient report of the present condition plus past medical history. Treatment via direct access is becoming more common for persons with vestibular disorders because access to a therapist is often quicker than access to a specialized physician. Referral from a local internist or general practice physician may provide only a diagnosis of "dizziness" or "vertigo," which is why it is crucial to develop differential diagnosis skills. If the patient has been seen by a specialist, physical therapy is often initiated after vestibular laboratory testing is complete and the physician has determined the patient's diagnosis.

The patient's current and past medical histories are important pieces of information that should be obtained by the therapist at the initiation of the physical therapy examination. Such information may assist in the identification of problems that could ultimately affect the patient's rehabilitation prognosis and outcome. For example, concurrent disease processes, such as peripheral vascular disease and peripheral neuropathy, will affect and prolong the patient's functional recovery. Diabetes, heart disease, old neck and back injuries, neurological dysfunction,

psychiatric comorbidities, a history of migraines, and preexisting or long-term visual dysfunction (e.g., strabismus) are examples of disorders that affect the ability of the person to compensate for the vestibular loss. Obtaining a complete medication history from the patient also is vital because many medications can produce or enhance dizziness (see Chapters 10 and 14). Some people may be taking certain medications that act to reduce the patient's symptoms by depressing the vestibular system. These medications may also delay vestibular adaptation and therefore may prolong the recovery period. The therapist should consult with the physician to determine the possibility of reducing the dose of such a medication or even eliminating it completely. Other people need medication to proceed with rehabilitation, especially those with anxiety and persons with central vestibular dysfunction. Persons with central vestibular disorders who have constant dizziness complaints may benefit from a central vestibular suppressant so that their symptoms, especially severe nausea, are better under control.[38,39] They may not be able to tolerate rehabilitation efforts without such medication to control their symptoms.

Subjective History

The subjective history of the patient's condition is critical in the evaluation of the patient with a peripheral vestibular problem. Questions that go beyond those usually asked by a physical therapist should be considered (see Box 21-2). The use of a questionnaire that the patient can complete before the initial visit is also helpful and saves time (see Appendix 21-A). A complete description of the patient's symptoms should be documented, so that functional progress can be later assessed. Think about documenting which symptom(s) the patient is experiencing, the duration of the symptoms(s), and the circumstance(s) in which the symptoms occur (e.g., spontaneous versus position-specific). Knowing what positions, movements, or situations aggravate the patient's symptoms is important in treatment planning. In addition, the patient should be asked questions about the type, frequency, duration, and intensity of symptoms and whether symptoms are of a fluctuating nature. Knowing the type of onset and frequency of the symptoms is very helpful in determining the physical therapy diagnosis and prognosis.

Intensity of symptoms like vertigo and disequilibrium can be measured by means of a visual or verbal analog scale similar to that used in the assessment of pain.[40-42] Some therapists use a 0 to 10 scale, and others use a 0 to 100 scale.[41,43] One commonly used technique is to ask the patient to rate symptom intensity using a visual analogue scale. For instance, disequilibrium (the patient's sense of being off-balance) is rated while sitting quietly and while walking. The scale is anchored at one end as "no imbalance at all" and at the other end as "the worst it can be." Another scale can be used for the intensity of head movement-induced dizziness. In that assessment, the patient rates the intensity of his/her dizziness while sitting quietly and then again after 1 minute of horizontal head movement at 1 Hz (see Appendix 21-A). These visual analogue scales have only moderate reliability.[42] It is very helpful to have patients rate their symptoms, yet not all patients are able to provide a number. Those who are confused have great difficulty rating their symptoms, and family members often attempt to "help" the patient, which is not useful. When it is impossible for the patient to provide a numerical rating, a "little, medium, or lot of dizziness" rating scale can help guide intervention. Finally, the patient can be asked the percentage of time in the last week that his/her dizziness has interfered with daily activities.

Questions related to the patient's perceived disability and psychosocial status should also be included in the initial assessment. Many different tools can be used to measure perceived disability, and most of them include items that assess function at the activity/limitation and participation/restriction levels. Using the Medical Outcomes Study 36-Item Short-Form Health Survey (SF-36) is one method that could be used to determine whether the patient is doing more in the home or community after therapy.[43-49] Other therapists may use the Sickness Impact Profile or other health status measures.[50] These tools help the therapist determine whether the patient is feeling better and is more active. Use of health status inventories is an excellent way to determine whether the patient has actually improved.[51] Shepard and colleagues[52] have suggested the use of a disability scale to objectively document a patient's perceived level of disability (Table 21-2). The Disability Scale is quick and easy to use and a change in the score of one point is significant. Test-retest reliability is high (ICC 1.0).[42] This 6-point Disability Scale has descriptors that range from having no disability (negligible symptoms) to having long-term disability. *Long-term disability* is defined as the inability to work for more than 1 year.[52] The Disability Scale can be incorporated into the initial and discharge physical therapy evaluations to document treatment outcome. As an alternative, the Post-Therapy Symptoms Scale, also developed by Shepard and colleagues,[52] can be used at discharge to determine whether the patient perceives a change in disability (Table 21-3). The Disability Scale is also useful in predicting treatment outcome; Shepard and colleagues[52] found that patients who rated their disability as a 4 or 5 were less likely to show significant improvement with rehabilitation.

■ Table 21-2 DISABILITY SCALE

Criterion	Score
Negligible symptoms	0
Bothersome symptoms	1
Performs usual work duties but symptoms interfere with outside activities	2
Symptoms disrupt performance of both usual work duties and outside activities	3
Currently on medical leave or had to change jobs because of symptoms	4
Unable to work for over 1 year or established permanent disability with compensation payments	5

Adapted from Shepard NT, et al: Habituation and balance retraining: A retrospective review. *Neurol Clin*. 1990;8:459.

■ Table 21-3 POST-THERAPY SYMPTOM SCALE

Criterion	Score
No symptoms remaining at the end of therapy	0
Marked improvement remaining at the end of therapy	1
Mild improvement, definite persistent symptoms	2
No change in symptoms relative to therapy	3
Symptoms worsened with therapy activities on a persistent basis relative to pre-therapy period	4

Adapted from Shepard NT, et al: Habituation and balance retraining: a retrospective review. *Neurol Clin*. 1990;8:459.

Many patients believe they have a psychological problem rather than a physical one. The patient's condition is one that cannot be seen by family and friends. Often, the condition is not well understood and may have been misdiagnosed or not addressed by the medical community.[53,54] When interacting with such a patient, the therapist must reassure him or her that others share the disorder . This reassurance is essential, because in some cases, stress or emotional trauma magnifies symptoms (see Chapter 18). It often helps to have written information to give the patient and the patient's family. One source for this information is the Vestibular Disorders Association (VEDA) a support group that has a variety of brochures about living with a vestibular disorder (http://vestibular.org/).

Several tools have been developed to define the patient's subjective symptoms of dizziness in an objective manner.[52,55] The Dizziness Handicap Inventory (DHI) is a useful clinical tool that can clarify the patient's symptomatic complaints and perceptions of his or her functional abilities (embedded in Appendix 21-A; see also Appendix 21-B)[9,21,55-58] Items relate to functional, emotional, and physical problems that the patient may have, and these items reflect primarily activities and participation plus body functions domains of the ICF. This inventory can be administered quickly during the initial and discharge visits, to quantify whether or not the patient thinks he or she has improved. In addition, the DHI may also help the therapist identify persons who may be at risk for falling. [9] Scores higher than 60 have been related to reported falls in persons with vestibular dysfunction.[9] The DHI has high test-retest reliability ($r = 0.97$)[55] and therefore can be used to track changes with treatment. In addition, the DHI has been shown to be correlated to the Activities-specific Balance Confidence (ABC) Scale in persons with vestibular disorders.[5] The ABC scale is a 16-item tool that quantifies balance confidence across a continuum of progressively more challenging situations from mobility in the home to walking on icy sidewalks.[59] The ABC scale demonstrates good test-retest reliability ($r = 0.92$) and convergent and criterion validity. Total scores range from 0% to 100%, with baseline scores of less than 80% considered abnormal. A significant change in the ABC score was either a return to a score of at least 80% or a change of more than 10 points.[60]

Fall History

Some patients with vestibular dysfunction fall.[1,27,60-63] Interviewing the patient about their history of falls is very important, because individuals often misinterpret the question "Have you fallen?" When asked, the patient answers "no," but with probing, it is common to find out that the patient has fallen. A *fall* is defined as involuntarily moving to the ground or lower surface. Carefully defining what a fall is with the patient provides the therapist better data to interpret the patient's condition. People who have fallen two or more times in the last 6 months are at high risk for falling again.[64] The patient can also be asked about having "near falls." *Near falls* are defined as (1) taking several steps to hold onto a wall, table, or top of a chair or (2) being caught or supported by another person.[64,65] Other

important issues include: (1) whether the patient has been injured during a fall, (2) the conditions under which the fall(s) occur, (3) how the patient has modified his or her lifestyle after a fall or fall-related injury, and (4) whether the patient sought medical intervention as a result of the fall. Unexplained falls are always of concern.The treating physician should be notified immediately if the person is falling with no known provocative cause. Fortunately, certain exercise programs can reduce fall risk, even in the frail older adult.[66,67] Others[68-71] have demonstrated that fall risk is reduced after vestibular rehabilitation in persons with unilateral hypofunction.

Functional History

To obtain a complete picture of functional status, the therapist should question the patient about previous and current activity levels (Box 21-3). A history of the patient's activity level is an important component of the assessment, which often characterizes the extent of the patient's disability. The Vestibular Activities of Daily Living Scale is an excellent example of a tool that will assist the therapist in identifying limitations in activity and participation (Box 21-3).[10,72]

Some patients avoid leaving their homes because exposure to highly textured visual stimulation, such as light flickering through trees or walking in stores, increases their disequilibrium.[73-76] This common experience is referred to as the "shopping aisle syndrome."These patients may have a limited ability to interact with their environment and, over time, tend to adopt a more sedentary lifestyle. Occasionally, patients develop phobias associated with their symptoms, including fears of elevators and of heights. Whitney and colleagues[77] have reported that 50% of the people referred to the investigators' tertiary care physical therapy clinic stated that they were always wary of heights.

Box 21-3

CURRENT FUNCTIONAL STATUS

Are you independent in self-care activities?

Can you drive:
- In the daytime?
- In the nighttime?

Are you working? If yes, occupation:

_____

Are you on medical disability?

Can you perform all your normal parenting activities?

Do you have difficulty:
- Watching TV?
- Reading?
- Being in stores or malls?
- Being in traffic?

Do you have difficulty walking up and down ramps, stairs, walking on grass?

Patient Goals

At the beginning of the assessment, the patient should be asked about expectations of physical therapy and functional goals.[78] After the assessment is complete, the therapist and the patient should discuss whether these goals are realistic and attainable. The patient's goals may have to be mutually modified by the therapist and the patient. The final level of recovery for most patients with unilateral vestibular hypofunction (UVH), without other complications, should be a return to full activities. Conditions that may make recovery more difficult, and therefore need to be recognized in the setting of goals, include the patient's premorbid physical condition and personality profile.[6] Occasionally, patients with significant vestibular dysfunction make remarkable functional gains but still experience significant symptoms at the conclusion of rehabilitation.

Clinical Examination

The clinical examination of a patient with vertigo and disequilibrium is usually comprehensive (Box 21-4) and therefore is time-consuming. Discretion should be used as to which portions of the examination must be performed on each patient. The full examination is described here but, where possible, conditions are described for which different portions of the examination would be unnecessary. Many of the elements of the therapist' s assessment are also discussed in Chapter 10. Key elements of the clinical examination are listed in Box 21-5.

Oculomotor and Vestibulo-ocular Testing

The oculomotor examination is one part of the overall assessment of the "dizzy" patient that may have been performed by a neurologist or otolaryngologist before referral for physical therapy. It is therefore not always included in the physical therapy assessment. If the patient is self-referred, it is an *essential* aspect of the physical examination.

First, the patient is observed for the presence of spontaneous nystagmus in room light. In patients with unilateral peripheral vestibular hypofunction, spontaneous

Box 21-4

SUMMARY OF THE CLINICAL EXAMINATION OF THE "DIZZY" PATIENT

Oculomotor Examination (in room light)

Non-vestibular—extraocular movements, pursuit, saccades, VORc, diplopia.

Vestibular—skew, spontaneous and gaze-evoked nystagmus, VOR to slow and rapid head thrusts, visual acuity test with head stationary and during gentle oscillations of the head.

With Frenzel lenses—spontaneous and gaze-evoked nystagmus, head shaking–induced nystagmus, tragal pressure–induced nystagmus, nystagmus with and without fixation, hyperventilation-induced nystagmus, and positional nystagmus.

Sensation

Somatosensation—proprioception, light touch, vibration; quantified tests: vibration threshold, tuning fork test

Vision—visual acuity and field

Coordination

Optic ataxia/past pointing, rebound, diadokokinesia, heel to shin, and postural fixation

Range of Motion (active and passive)

Upper and lower extremity, neck (rotation, extension, flexion, lateral flexion)

Strength (gross)

Grip, upper extremity, lower extremity, trunk

Postural Deviations

Scoliosis, kyphosis, lordosis

Positional Testing

Dix-Hallpike test, side-lying test, roll test

Motion Sensitivity

Motion- and position-induced dizziness

Sitting Balance (active or passive, anterior-posterior, and lateral)

Weight shift, head righting, equilibrium reactions, upper and lower extremity, ability to recover trunk to vertical

Static Balance (performed with eyes open and closed)

Romberg test, sharpened Romberg test, single leg stance, stand on rail, force platform

Balance with Altered Sensory Cues

Eyes open and closed, foam.

Dynamic Balance (self-initiated movements)

Standing reach (Duncan), functional (Gabell and Simons), Fukuda's stepping test

Ambulation

Normal gait, tandem walk, walk while turning head, singleton to right and left, Dynamic Gait Index, Timed "Up & Go"

Functional Gait Assessment

Obstacle course, double-task activities, stairs, ramps, grass, sand

nystagmus will be observable in room light during the acute stage after onset of the lesion and is the result of the imbalance in the tonic or resting firing rate of the vestibular neurons. Within a few days of onset, the patient should suppress the nystagmus with visual fixation. Patients in this acute stage often complain of having difficulty reading and watching television. In later stages, with infrared goggles in place, the therapist may still be able to visualize the spontaneous nystagmus that is absent in room light.

Next, the patient is examined for gaze-holding nystagmus, the inability to keep the eyes steady when looking about 15 to 20 degrees eccentrically. Direction-fixed gaze-holding nystagmus (the direction of the nystagmus is the same regardless of eye-in-orbit position) is consistent with acute unilateral vestibular hypofunction. Direction-changing gaze-holding nystagmus is consistent with a central lesion, usually in the posterior fossa.

Patients can also be tested for ocular alignment, particularly for skew deviations that can occur during the acute stage of a unilateral vestibular loss (UVL). Skew deviations, in which the eye opposite the side of the lesion is elevated, occur because of the loss of the tonic otolith input from one side. Normally, the tonic input holds the eyes level within the orbit; when there is a unilateral vestibular loss, the eye on the side of the lesion drops in the orbit and the patient complains of vertical diplopia. As

Box 21-5

KEY ELEMENTS OF THE CLINICAL EXAMINATION

Oculomotor

In room light—skew deviation, spontaneous and gaze-evoked nystagmus, VOR head-thrust test, visual acuity test with head stationary and during gentle oscillations of the head

With Frenzel lenses—spontaneous nystagmus, positional nystagmus, and nystagmus with and without fixation

Range of Motion

Neck (rotation, extension, flexion, lateral flexion)

Positional Testing

Dix-Hallpike test, side-lying test, roll test

Motion Sensitivity

Motion- and position-induced dizziness (motion sensitivity test)

Static Balance (performed with eyes open and closed)

Romberg test, sharpened Romberg test, single-leg stance

Balance with Altered Sensory Cues

Eyes open and closed, foam

Ambulation

Normal gait, tandem walk, walk while turning head, Timed "Up & Go"

Functional Gait Assessment

Obstacle course, double-task activities, stairs, ramps, grass, sand

Fall Risk

Dynamic Gait Index

Activity Level

Physical Activities Scale for the Elderly

Quality of Life

Medical Outcomes Study 36-Item Short-Form Health Survey, Dizziness Handicap Inventory, Vestibular Disorders Activities of Daily Living Scale, Disability Score, Vestibular Rehabilitation Benefits Questionnaire,[79,80] Vestibular Activities and Participation Scale[22]

with spontaneous nystagmus from UVL, skew deviations from UVL should resolve within 3 to 7 days after onset.

Smooth pursuit is tested by asking the patient to track, with the eyes, an object moving in the central 20 degrees of the visual field while the head is stationary Typically, this test assesses central pathways involved in smooth pursuit and the motor function of cranial nerves III, IV, and VI. Smooth pursuit is abnormal if, during tracking, the eye movements are disrupted by saccades. One important thing to remember is that abnormal smooth pursuit is never a sign of a peripheral vestibular deficit.

For the patient with nystagmus, determining the quality of pursuit eye movements may be difficult. In addition, care must be taken to distinguish gaze-evoked nystagmus from end-point nystagmus. End-point nystagmus, which is normal, occurs when the eyes are at the extreme end of their range of motion. It is also important to determine, in the initial patient history, whether the patient has any premorbid eye disorder, including asking about a history of strabismus. Patients with "lazy eye" disorders may have disconjugant smooth pursuit, which can confound the results of the testing.

Saccadic eye movements are tested by simply asking the patient to look back and forth between two horizontal or two vertical targets. In healthy individuals, the target can be reached with a single eye movement or with one small corrective saccade. It is important to have discrete targets to observe so that the clinician can definitively determine if they consistently overshoot the target position (see Chapters 10 and 11 for more details).

The patient can then be asked to voluntarily fixate on a moving target while the head is moved in the same direction. This procedure tests the ability to suppress or "cancel" the vestibulo-ocular reflex (VORc) and is a function of the parietal lobe. Results should agree with the observations made during the smooth-pursuit test. If VORc is impaired, one should suspect a central nervous system disorder.

Next, the VOR itself is tested. The head-thrust or head impulse test is one test that an experienced physical therapist can perform to assess the function of the

vestibular system itself. The test traditionally involves an unpredictable, high-acceleration, small-amplitude head thrust in the horizontal plane,[81,82] although the head thrust test can be performed in the planes of the each of the semicircular canals.[83] The patient sits with the head pitched in 30 degrees of cervical flexion (using an imaginary line from the inferior rim of the ocular orbit to the external acoustic meatus). The patient is instructed to maintain visual fixation on the examiner's nose. The patient's head is then gently grasped, and a small-amplitude (5 to 10 deg) but high-acceleration (3,000 to 4,000 deg/sec ^{2}) thrust is applied horizontally. When the head impulse stops, the eyes are observed to see if they are still directed toward the target (the nose). If there is a decrease in vestibular function, a corrective "overt" saccade, a rapid eye movement that returns the eyes to the target, will be observed. If the head impulse test at near distance is abnormal (corrective saccade is present), the test is repeated with the participant looking at a target place more than 2 m away. Repeating the test with a distant tar get helps reduce the false-positive results seen in some older individuals because of poor ability to accommodate to a near tar get. Testing should be performed with appropriate visual correction (glasses) for the distance tested although sometimes it is difficult to visualize the eyes. The sensitivity of the test has been reported to be 54%, and specificity to be 100%.[84] During the acute stage, corrective saccades occur even with slow head rotations. In the chronic stage, people with vestibular deficits often are able to maintain fixation during slow head movements using the pursuit eye movement system but make corrective saccades to regain the target with rapid head movements. In addition, the presence of corrective saccades is dependent on the severity of the deficit, and in general the head impulse test is most likely to be positive with deficits greater than 75%. It is imperative that the examiner carefully view the eyes during the first head thrust maneuver, because they may be able to generate "covert" saccades on subsequent repetitions of the test that will be missed by the examiner.

Another evaluation of VOR function is to measure the degradation of visual acuity that occurs with head movement.[40,85] We use a modified ETDRS chart with SLOAN letters (Lighthouse Distance Visual Acuity Tests, Long Island City, New York) for the clinical version of this test. A handheld Snellen card is not as appropriate, because it measures the patient's vision at a distance of only 18 inches, a distance that older patients in particular have difficulty accommodating. In addition, the letters on the Snellen card are not balanced for difficulty nor is there an equal decrease in letter size from line to line. In the clinical Dynamic Visual Acuity (DVA) test, the patient is first asked to read a wall eye chart with the head stationary. Then the patient is asked to read the chart while the head is oscillated at 2 Hz by the examiner. Using a metronome helps standardize the test.[86,87] In normal individuals, visual acuity changes by one line in younger individuals or by two lines in older individuals. In patients with uncompensated, unilateral vestibular loss, visual acuity degrades by three or four lines. Computerized systems for measuring visual acuity during head movement are now available (see Chapter 11) and have the added benefit of being able to determine dynamic visual acuity for rightward and leftward head movements separately. The NIH Toolbox has a DVA test and has established test-retest reliability.

Brandt and colleagues[88] suggest that distance acuity poorer than 20/50 has a significant effect on postural stability. Additionally, balance can also be affected by visual field loss[89]; patients with monocular vision may have particular difficulty with depth perception, which would affect their ability to walk up and down stairs. The test becomes much more difficult to perform with progressive or bifocal lenses, especially in the pitch plane.

Eye movements can also be observed with the use of Frenzel lenses or video oculography (VOG). Frenzel lenses magnify the eyes, with light inside them to help with visualization, enabling the clinician to observe eye movements and greatly decreasing the patient's ability to stabilize the eyes with visual fixation. The VOG systems permit the examiner to visualize the eyes in all positions because infrared cameras record eye movements and transmit the image to either a computer or television monitor. Either one or both eyes may be visualized, depending on the system used. Clinical assessment of oculomotor function using Frenzel lenses should include spontaneous and gaze-evoked nystagmus, head shaking–induced nystagmus,[90] tragal pressure–induced nystagmus, hyperventilation-induced nystagmus, and positional nystagmus (see Chapter 10). The VOG systems allow the playback of the eye movements, permitting the patient to visualize their eye movements, and also permitting the clinician to permanently record what was seen. There are significant advantages to being able to record eye movements with complex eye movements and conditions. In addition, the ability to determine if the nystagmus changes with fixation can significantly help determine if you are dealing with a peripherally or centrally generated nystagmus.

During the assessment of eye movement function, the patient is asked to report any symptoms of blurred vision or dizziness. Tests that involve repeated head movements (VOR, head shaking–induced nystagmus, and dynamic visual acuity testing) may exacerbate the patient's symptoms. If there is a significant increase in symptoms, the patient may be unable or may refuse to continue with the testing. Explaining the importance of the information

to be gained from the test and acknowledging the patient's symptoms often helps the patient to accept the discomfort caused by the test. If the patient is experiencing significant symptoms, some of the testing can be deferred, because if the patient becomes too sick, he or she may not return for a follow-up visit.

Sensory Evaluation

Sensation of the extremities can be tested to rule out concurrent pathology and assist in treatment planning. Perhaps the most important of these is the assessment of kinesthesia and proprioception, although profound sensory loss affecting touch and pressure sensitivity would also affect postural stability and raise the risk of a fall.[91,92] These tests may not be performed in all subjects but should be considered, especially in older individuals and in patients with diabetes or peripheral neuropathy.

Proprioception can be assessed by having the patient close the eyes and then moving the patient's great toes either up or down and asking the patient to identify the position of the toe. Care must be taken to make these relatively small movements, or the test becomes too easy. The patient must also be instructed not to guess at the answer. This traditional test of proprioception does not appear to be very sensitive, and patients are quite accurate in perceiving whether the toe is up or down even when other tests indicate sensory deficiencies. Kinesthesia can be tested by slowly moving the toe either up or down and asking the patient to state the direction of the movement as soon as he or she first perceives movement. Again, the patient should be instructed not to guess. Perception of the direction of the movement should occur before the toe is moved more than 10 to 15 degrees, although each clinician must develop his or her own internal standard for what is normal. Vibration can be tested with application of a tuning fork to a bony prominence. One method is to ask the patient to identify when the vibratory sensation stops, and then to dampen the tuning fork unexpectedly. Another method is to let the vibration diminish naturally and to time the difference between when the patient and the clinician stop feeling the vibration. Again, each clinician must develop his or her own sense of normal. Devices are also available that quantify vibration thresholds. These devices enable the clinician to compare the patient with age-matched normal subjects and to follow changes over time. Diminished sensation in the toes may not affect postural stability; if a patient appears to have no sensation in the toes, then the clinician should perform the same tests on the ankles.

The visual, vestibular, and somatosensory systems all show decrements with age, and the clinician should be familiar with these normal changes to differentiate them from pathological changes.[93] Furthermore, certain disorders that can affect perception, such as cataract formation, are more likely to occur in the older person. There is some evidence that a decrease in VOR gain occurs with aging, at least at higher frequencies, and there is a more limited adaptive capability with increased age.[14,15]

Multisystem involvement can impede the patient's functional progress. A patient with reduced vestibular function and a deficit in somatosensory cues is likely to compensate by using visual mechanisms. This patient is limited functionally if placed in a situation devoid of visual information. For example, many patients with reduced somatosensory cues and a peripheral vestibular deficit have difficulty walking in the dark. A treatment plan that does not consider the patient's sensory loss may be ineffectual, and the patient's functional independence could suffer.

Coordination

Vestibular deficits per se do not result in poor coordination or in limb ataxia. Assessment of coordination is especially important, however, as part of the preoperative and postoperative examination of patients with cerebellar angle tumors. Finger-to-nose and heel-to-shin movements and the ability to perform rapid alternating movements of fingers or feet are gross tests that may be used to subjectively assess the patient's coordination. Other tests of cerebellar function include truncal stability and tests of tone, such as postural fixation and the rebound phenomenon; however, surgery for vestibular schwannoma usually does not cause these deficits.

Range of Motion and Strength

Range of motion and strength is not assessed in every patient. However, in some patients it is an important part of the assessment, such as in patients with neck pain. Although the neck, trunk, and extremities can be included in this assessment, special attention should be paid to the neck's range of motion. Patients in whom head movement exacerbates symptoms may voluntarily restrict active neck motion and may eventually lose that range of motion. Furthermore, many of the other assessments involve passive movement of the neck (e.g., Head Impulse Test, head shaking–induced nystagmus, dynamic visual acuity, positional testing), and any limitations in movement or pain associated with neck movement should be identified before those tests are attempted. In some patients, when neck range of motion increases, there appears to be an associated decrease in dizziness symptoms.[25] A detailed examination of extremity strength and range of motion is often unnecessary; however, a quick screen indicates whether more-detailed testing would be appropriate. It is common

in the elderly patient to find weakness in the distal extremities, which can be recognized in this quick screening and dealt with through use of a home exercise program to "tune up" the system and reduce the patient's risk of falling.

Postural Examination

In addition to assessment of range of motion, the patient's posture should be evaluated. Predisposing orthopedic conditions or postural deviations may complicate the rehabilitation prognosis. Anterior-posterior and medial-lateral views of both the patient's sitting and standing postures should be assessed. Typically, posture is not affected in people with peripheral vestibular dysfunction.

Positional and Movement Testing

Clinically assessing the positions and movements that provoke the patient's symptoms is important. In this portion of the evaluation, attempts are made to replicate the various positions and movements experienced by the patient throughout the day (Table 21-4). The activities are rated by the patient as to whether they provoke no symptoms or mild, moderate, or severe symptoms. In some situations, the patient may be unable, or unwilling, to perform the task. This reluctance is especially apparent in the patient who experiences severe dizziness early in the evaluation. In addition, a patient may refuse to move into or out of a specific posture because of the fear of eliciting symptoms.

The most important positional test is the Dix-Hallpike test and its variants (Fig. 21.2).[94] This test is most commonly used in patients who complain of vertigo only when they move into certain positions, but it should be included in almost all assessments of people with complaints of dizziness.[95] Vertigo and nystagmus occurring when the patient is moved into the Dix-Hallpike position is used to diagnose benign paroxysmal positional vertigo (BPPV) (see Chapter 20 for details). The patient should be cautioned in advance that the maneuver can cause dizziness or vertigo but nonetheless should be performed. Two other positional tests can be performed: the sidelying test, which is a variation of the Dix-Hallpike test, and the roll test for the horizontal semicircular canals. It is also important to perform the Dix-Hallpike and other positional tests, because some patients may be referred to therapy with a "diagnosis" of dizziness or vertigo, both of which are symptoms, not diagnoses. After the Dix-Hallpike test is performed, it may become obvious that these patients have BPPV or another vestibular disorder that causes positional nystagmus and vertigo such as fistula. There are also people who have "subjective" BPPV, as reported by Haynes and associates,[96] who describe symptoms typical of BPPV yet do not exhibit the typical

■ Table 21-4 **MOTION SENSITIVITY QUOTIENT***

Baseline Symptoms	Intensity	Duration	Score
1. Sitting to supine			
2. Supine to left side			
3. Supine to right side			
4. Supine to sitting			
5. Left Dix-Hallpike position			
6. Return to sit from left Dix-Hallpike position			
7. Right Dix-Hallpike position			
8. Return to sit from right Dix-Hallpike position			
9. Sitting, head tipped to left knee			
10. Head up from left knee			
11. Sitting, head tipped to right knee			
12. Head up from right knee			
13. Sitting, turn head horizontally 5 times			
14. Sitting, move head vertically 5 times (pitch)			
15. Standing, turn 180 degrees to the right			
16. Standing, turn 180 degrees to the left			

*See Chapter 22 for further information.

nystagmus during the Dix-Hallpike maneuver. Haynes and associates reported that 86% of those with "subjective" BPPV reported significant improvement in symptoms after treatment with the Semont maneuver.[96] Recognizing BPPV and providing the proper intervention can significantly enhance the patient's quality of life (Fig. 21.2).[97]

BPPV is a common cause of vertigo.[98] This peripheral vestibular deficit is easily and effectively treated with physical therapy.[99,100] In contrast, vertebral artery compression is relatively rare, and unfortunately, clinical tests for vertebral artery compression are not reliable (see Chapter 31). Vertebral artery compression often causes some symptoms that are like those of BPPV. Occlusion of blood flow with compression of the vertebral artery produces neurological symptoms, such as numbness, weakness, slurred speech, and mental confusion, as well as vertigo and nystagmus. Persons with BPPV do not experience neurological symptoms such as slurred speech, mental confusion, weakness, or numbness. If there is any level of suspicion that the person has vertebrobasilar compromise, they should be referred out for blood flow studies.

Figure 21.2 Dix-Hallpike test used primarily to evaluate for benign paroxysmal positional vertigo. The head is turned to one side and the patient is moved from sitting into a supine position with the head hanging over the end of the table. The patient is then observed for nystagmus, and complaints of vertigo are noted. The patient is then returned to the upright position. (From Herdman, 1990.[95])

Examples of other movements that can be tested are rolling, supine to sit, reaching in sitting toward the floor, and sit to stand. All these movements can be tested at various speeds and with the patient's eyes open and closed. The therapist should exercise care when having patients perform these maneuvers while standing with the eyes closed, because this is often very difficult for patients and may cause them to lose their balance, especially if the patient experiences severe vertigo. A more formalized assessment of what positions and movements induce symptoms in patient is the Motion Sensitivity Quotient (MSQ). The MSQ is typically used to determine what positions result in an increase in symptom intensity and to quantify the patient's symptom intensity and duration. The test consists of 16 changes in position, and the results are used to develop a treatment program to reduce symptom intensity (see Chapter 22).

The speed at which movement is performed can affect the patient's symptoms. For example, a quickly performed movement could increase the patient's symptoms, whereas the same movement performed at a slower speed may not. Varying the speed and the conditions under which the patient performs the task may affect the patient's functional ability. Positional and movement testing is limited only by the imagination of the therapist. For one patient seen in our clinic, the only position that increased her symptoms was the "all-fours" position, which she would assume for looking under the bed (Fig. 21.3).

Head Movement Induced Symptoms: In addition to testing positions and movements that incorporate multiple body segments, the patient is asked to perform head

Figure 21.3 Patients may experience dizziness or vertigo in positions other than those normally tested. Shown here is the provoking position for one patient who experienced vertigo only when bending over and turning her head.

movements. The head movements are typically tested with the patient sitting. These movements are performed at various speeds and with the eyes open and closed.The patient is asked to report whether these movements provoke symptoms and whether the symptoms are of a mild, moderate, or severe intensity. Verbal and visual analog scales have been used in an attempt to better quantify the patient's symptom intensity.[40,41] The same movements are tested in the standing position or during gait, if the patient can tolerate further testing.

Balance Assessment

Sitting Balance

Few patients with chronic vestibular disorders have difficulty with balance in sitting, but for some patients, including an assessment of sitting balance may be appropriate. Acutely, people will have deficits in sitting balance in the first few days post-acute peripheral vestibular insult. Patients should be tested while they are leaning anteriorly and posteriorly as well as right and left; tests should be performed both actively and passively. The patient can be observed for weight-shifting ability, head righting, equilibrium reactions in the upper and lower extremities, and the ability to recover to a trunk vertical position. Having the patient reach in sitting in all directions without support is also valuable information that can be gained in the clinical examination. The Ottawa Sitting Scale may be helpful in the acute care setting to quantify sitting abilities.[101]

Static Balance

Static balance tasks have been used clinically to objectively document balance function.[102-106] Single-leg stance (SLS), Romberg, and Sharpened or tandem Romberg tests are often included in a static balance test battery and can be performed with the patient's eyes open or closed.[106-108] Traditionally, the variable of interest in this testing has been the time that the patient maintains the position. Normative data for different ages have been established for SLS, Romberg, and sharpened Romberg tests.[106] The Romberg test has been shown to have low intrasubject variability when measures are repeated over a 5-day period.[107]

Patients with vestibular deficits may have normal performance on these tests.[109] Tests of static balance, such as the Romberg, are fairly easy. Patients may have difficulty with this test only during the acute stage after onset of the vestibular deficit or if they have a coexisting peripheral neuropathy. It is also important to remember that patients with balance disorders other than from vestibular dysfunction may have difficulty with these tests. Having difficulty maintaining stance with the Romberg test does not necessarily mean the subject has vestibular dysfunction. Table 21-5 lists the expected results for static and

■ Table 21-5 **EXPECTED TEST RESULTS IN PATIENTS WITH UNILATERAL VESTIBULAR LOSS (UVL)**

Test	Acute UVL	Compensated UVL
Nystagmus	Spontaneous and gaze-evoked in light and dark; head shaking–induced nystagmus	Spontaneous in dark; may have head-shaking–induced nystagmus
Vestibulo-ocular reflex (VOR)	Abnormal with both slow and rapid head thrusts	Abnormal with rapid head thrusts toward side of lesion
Romberg	Often, but not always positive	Negative
Sharpened Romberg	Cannot perform	Normal with eyes open; cannot perform with eyes closed
Single-leg stance	Cannot perform	Normal
Modified Clinical Test for Sensory Interaction in Balance (CSTIB)—foam, eyes closed	Most cannot perform	Normal
Gait	Wide-based, slow cadence, decreased rotation, may need help for a few days	Normal
Turn head while walking	Cannot keep balance	Normal; some may slow cadence

dynamic balance tests in patients with acute and compensated UVL.

Measures of Sway

During performance of static balance tasks, medial-lateral or anterior-posterior stability can be objectively documented with the use of "high-tech" tools such as force platforms, accelerometers,[110-112] or of simple tools such as a sway grid.[112,113] When one is assessing and attempting to replicate standing sway measures, the distance the subject stands from a stable visual target, upper extremity positioning, type or lack of footwear, and foot position of the patient should be standardized. Brandt[88] hypothesizes that one explanation for the variability often found on the Romberg test is the inconsistent positioning of the patient with respect to a target used for visual fixation. The distance that the patient stands from the target should be standardized and should be within 1.5 meters. Kirby and associates[114] employed five different foot positions to determine the effect of foot position on sway. They determined that subjects standing in double-limb stance were most stable in the 25-degree toe-out position. In addition, they observed that subjects had the greatest medial-lateral sway when their feet closely approximated each other. The standard Romberg position is with heel and toes together.

Postural sway correlates well with measures of the DHI.[21,56] Of course, both postural sway and the responses to the DHI questionnaire are under "voluntary control," and what is measured is subject to errors according to what the patient wants to convey. Sway patterns of patients with central disorders differ from that of patients with other vestibular diagnoses. Yoneda and Tokumasu[113] reported that sway patterns seem to be different among patients with Ménière's disease, BPPV, and vestibular neuronitis, and that these patterns differed from those of a normal comparison group.

Altering Sensory Cues

The modified Clinical Test for Sensory Interaction in Balance (CTSIB) should be included in the rehabilitation assessment.[28,115-117] In some ways this test is an extension of the Romberg test, which assesses the effect of removing visual cues on postural stability. Referred to formerly as the "foam and dome" test (Fig. 21.4), the CTSIB assesses the influence of vestibular, somatosensory, and visual inputs on postural control. In the modified version, the "dome" portion of the test is no longer used.

In the modified CSTIB (mCTSIB), standing on the foam surface and closing the eyes alters somatosensory input and eliminates visual input. In this situation, vestibular input is the most accurate information about postural stability. Patients with uncompensated unilateral peripheral vestibular loss may have difficulty maintaining an upright posture when both visual and support-surface information are altered.[19] Subjects with unilateral peripheral vestibular dysfunction often lose their balance when standing on foam with the eyes closed (using the mCTSIB protocol).

According to Nashner,[19] symmetry and constancy of vestibular information are critical in providing an absolute reference for reorganization of senses in conflicting conditions. The inability of the vestibular system to provide

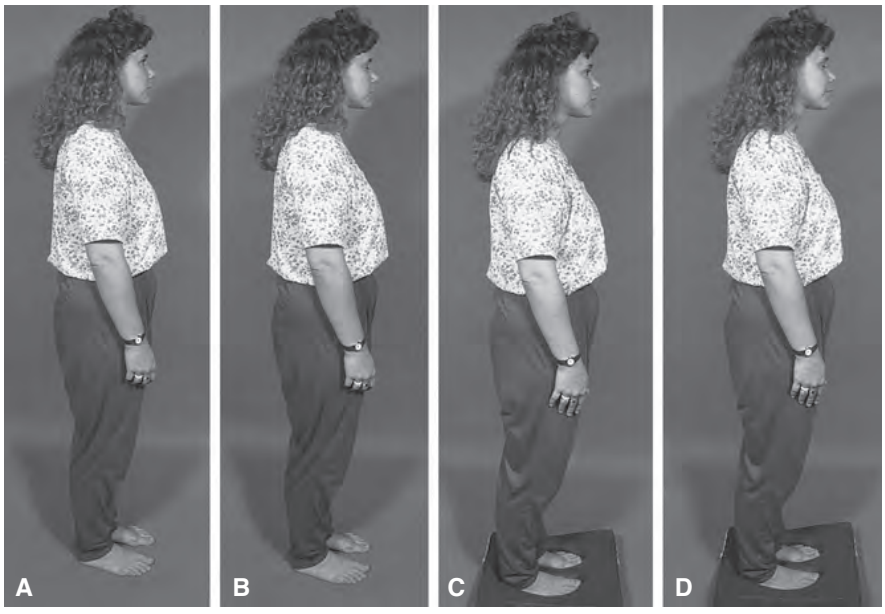

Figure 21.4 The modified Clinical Test of Sensory Integration of Balance: The subject is standing on a level support surface with eyes open **(A)** and with eyes closed **(B)**, and then on a compliant surface with eyes open **(C)** and eyes closed **(D)**. The time the patient takes to perform each task is recorded, and the amount of sway and the patient's movement strategy are documented qualitatively. The results of this test help determine whether the patient is dependent on certain sensory cues.

this information may explain why patients with unilateral vestibular lesions often report postural instability when riding on an escalator or when walking on thick carpet across a dimly lit room. If a patient is unstable when both visual and somatosensory cues are altered, a treatment plan might be designed to improve the function of the remaining vestibular system. Depending on the patient, an alternative treatment strategy may focus on altering the patient's environment so that visual and somatosensory cues are maximized, in an effort to overcome the vestibular loss.[118-120]

Self-Initiated Movements

In addition to static tests of balance function, self-initiated movements and dynamic tests of balance should be examined. Self-initiated weight shifts performed in different directions can be assessed to determine whether the patient moves freely and symmetrically.[121] Self-initiated movements should also be tested in functionally relevant and rich contexts—for example, having the patient reach to pick an object from the floor or place an object on a high shelf. Altering the environment so that there are many visual distractions may also be a way to make the task more difficult. Reaching for an object on the floor among many objects on the floor might make the task more difficult for the patient.

The functional reach test has been developed as a way to assess the subject's balance and willingness to reach outside the base of support.[56,122-127] This test functionally and reliably documents performance on a self-initiated task. The subject is asked to reach as far forward as possible. The extent of movement is measured with a simple yardstick (Fig. 21.5). Duncan and coworkers[124] have shown that functional reach, as a measure of a subject's margin of stability, correlates well with center of pressure measures obtained from a force platform. This test can be modified so that reaches in different directions are documented (Fig. 21.6).[121] Functional reach has also been shown to be related to falls in older male veterans[124] Generally, scores of 6 inches or less indicate that the person is at high risk for falling.[110] This test has been shown to have concurrent validity with certain items of the Functional Independence Measure and is useful in determining the risk of falling in older adults. It has also been shown to demonstrate change over the course of rehabilitation,[126] although Wernick-Robinson and colleagues[127] suggest that functional reach measures have less value in patients with vestibular dysfunction.

The standing reach test is administered easily and requires very little equipment. Such a functional measure of balance can easily be used in the clinic or home care setting to document whether the patient has made gains in physical therapy. The mini-BESTest might be considered to assess balance in persons with vestibular disorders, because the test is promising but has not yet been validated in persons with vestibular disorders.[128]

Movement Strategy

During the balance assessment, the therapist should observe and document the patient's movement strategy.

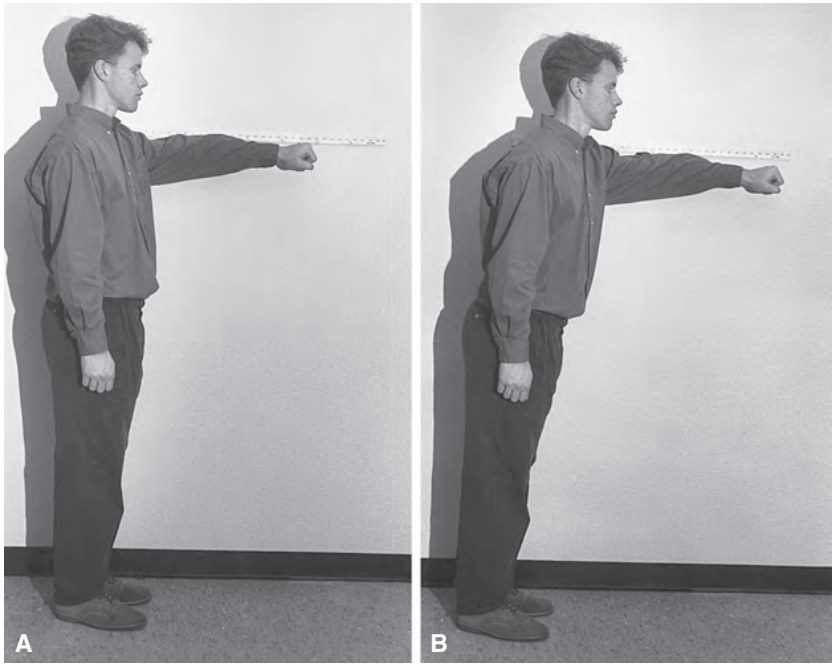

Figure 21.5 The distance a patient can reach is one measure of functional balance.[119] **(A)** The patient's acromion is lined up with a yardstick while the patient's arm is held parallel to the yardstick. **(B)** The patient reaches forward as far as possible while keeping both feet flat on the ground.

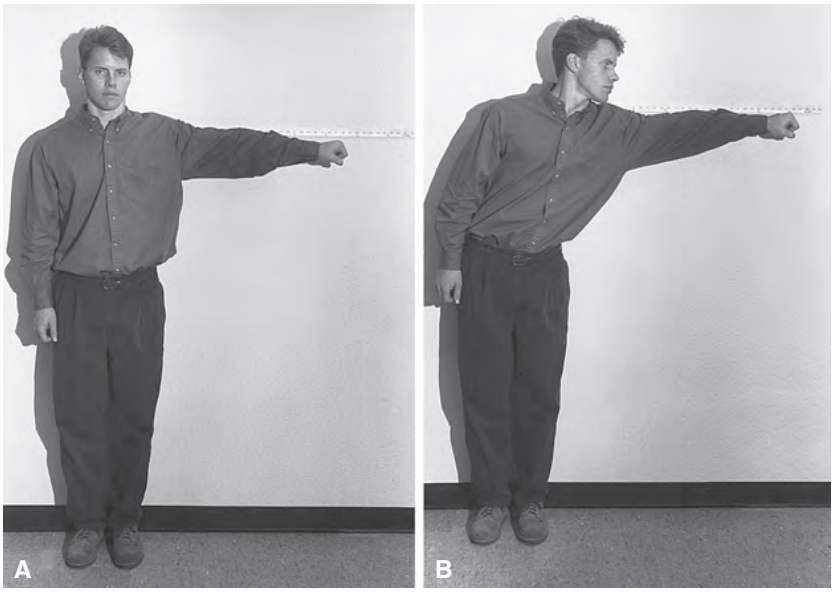

Figure 21.6 The distance a patient can reach to the side is another possible functional measure of balance. **(A)** The patient's acromion is lined up with the edge of a yardstick. **(B)** The patient reaches to the side as far as possible while keeping both feet flat on the floor.

Three types of movement strategies have been described for controlling anterior-posterior displacements of the center of mass.[129] The ankle strategy produces shifts in the center of mass via rotation of the body about the ankle joints. According to Horak and Nashner,[130] the ankle strategy elicits a distal-to-proximal firing pattern of ankle, hip, and trunk musculature. This activation pattern exerts compensatory ankle torques, which are believed to correct for small postural perturbations. The hip strategy controls movement of the center of mass by flexing and extending the hips. Unlike the ankle strategy, the muscle activation sequence associated with the hip strategy occurs in a proximal-to-distal fashion. This strategy produces a compensatory horizontal shear force against the support surface. The hip strategy occurs in situations in which the ankle is unable to exert the appropriate torque necessary to restore balance. This situation arises when the task of maintaining balance is more dif ficult, such as when an individual stands on a small, narrow support surface. Finally, a stepping strategy is used when the center of mass is displaced outside the base of support. This strategy is employed in response to fast, large postural perturbations.

To function safely and independently throughout the life span, humans are required to respond, through their movements, to a variety of task situations and environmental contexts. Postural strategies may be task-specific, and therefore categorizing postural strategies can prove to be difficult. In addition, individuals vary greatly with respect to body size, proportion, and weight. These considerations, taken together with age-related changes, make the task of categorizing postural strategies dif ficult. Instead, the physical therapist may be more successful in documenting whether the patient's individual strategy is *efficient, safe, and successful* with respect to achieving the task goal. With this approach, the clinician's expectations of the patient's responses are not as biased. This notion also has implications for treatment. Current motor learning theory suggests that the learner will be more successful in the task when the learner, not the teacher or therapist, selects the appropriate movement strategy.[131]

Gait Evaluation

Evaluation of the patient's gait provides a dynamic and functional assessment of the patient's postural control mechanism. The gait assessment can be obtained through clinical observation, videotape analysis, computerized gait mats, or computerized motion analysis (there is evidence of reliability for each).[132-134] A videotape record of the patient's gait is easily obtained clinically and can be extremely useful for documentation and patient education.

The patient's gait should be assessed in as many situations as are realistically accessible to the therapist. Analysis of the patient walking down a crowded versus a non-crowded hallway may yield very dif ferent information about the patient's gait function. Similarly, the patient should be instructed to walk at a normal speed, slowly, and quickly. Common clinical observations indicate that patients with vestibular disorders, like other patients, have greater gait instability when asked to walk at a non-preferred speed. They also often have difficulty changing gait speed while they are walking.

During gait assessment, the physical therapist documents the patient's movement strategy, the presence of

gait deviations, and whether the patient reports any abnormal sensation of movement. The patient with a peripheral vestibular disorder may select an overall strategy for gait that limits movement of the head and trunk. Clinically, this gait is characterized as being stiff or robot-like, and the patient may use excessive visual fixation while walking. Some of the typical gait deviations demonstrated by these patients include inconsistent step lengths, veering to the right or left, a widened base of support, a listing of the head and/or trunk to one side, decreased rotation through the trunk and neck (and decreased arm swing), and "en bloc"[135] movements and slow turns. In addition to observable gait deviations, patients with a unilateral peripheral vestibulopathy may associate dizziness with their gait instability.

A complete assessment of the patient's gait function should include having the patient perform a variety of tasks while walking. Many of these tasks cause the patient to lose balance; therefore, during this portion of the gait assessment, the physical therapist may need to guard the patient but without becoming a part of the patient's postural control system. The goal of this assessment is to learn how the patient, not the therapist, solves the problem of postural control.

One of the gait tasks performed by the patient is to walk while moving the head, either to the left and right or up and down. Head movements up/down and right/left are components of the Dynamic Gait Index (DGI), [136] a very helpful tool for use in the quantification of gait dysfunction. The therapist documents whether the patient experiences symptoms (dizziness, lightheadedness, etc.), loss of balance, or an exaggeration of gait deviations. Such a detailed assessment facilitates documentation of the patient's progress. The functional gait assessment is often used for younger people rather than the DGI, as the test was designed to negate a possible ceiling effect of the DGI.[28] The HiMat has also been used recently for patients with higher level gait disorders, although none of the tests appear optimal for those persons with blast injuries and vestibular complaints.[137-140]

Many patients with unilateral peripheral vestibular loss experience difficulty when asked to perform gait tasks that require an anticipatory mode of motor control. One such task requires the patient to walk quickly and then to stop immediately on the therapist's command. To maintain a level of uncertainty, the patient should not be informed ahead of time of the required pivot direction. The speed of the pivot is another factor that must be considered.The patient may also slow gait speed as a strategy to avoid disequilibrium during the task. The patient may also slow gait speed to avoid the anticipatory requirements of the task. In such instances, the patient should perform the task at a faster speed so that the therapist obtains a more complete picture of the patient's gait function.

Observing the ability of the patient to negotiate an obstacle course may also provide valuable information about his or her functional balance (Fig. 21.7) and the patient's ability to function in an anticipatory mode of control. To assess the patient's functional balance, the patient self-selects the path to negotiate the obstacle course. In addition, the patient decides whether or not to pick up or step over an object in the course. The patient also decides to negotiate the course quickly or slowly. If, on the other hand, the patient's anticipatory control is of interest, the therapist directs which path the patient should follow. Task variability and uncertainty are manipulated by the

Figure 21.7 Patients with vestibular disorders may have difficulty negotiating an obstacle course. The patient selects the path of the course to follow and whether to step over or pick up objects.

therapist. For example, the therapist may decide at which point in the course to throw a ball toward the patient (Fig. 21.8). A temporal constraint may also be added to the task. Asking the patient to negotiate the course as quickly as possible enhances task difficulty and provides a means to subjectively document the patient's progress.

The ability of the patient to perform gait tasks while manipulating an object with the hands should also be assessed. Patients with unilateral vestibular loss frequently complain of increased gait instability when carrying a basket of laundry up a flight of stairs or when carrying a bag of groceries. Clinically, the patient's ability to monitor postural control while manipulating an object can be tested in a variety of ways. The patient can be asked to walk, pick up one or more objects off the floor, and continue walking. Documenting the strategy used by the patient to perform this task is important. Many patients with vestibular loss bend at the knees and avoid flexing the head or bending at the hips. This strategy may be selected in an attempt to minimize provocation of symptoms or loss of balance.A patient may also be asked to negotiate a flight of stairs while carrying objects of varying weight or size. This task is important to include in the gait assessment, because a patient who demonstrates little difficulty with other gait tasks may express extreme postural instability with this task.

Other gait tasks that the therapist can assess are sidestepping, backward walking, tandem walking, walking in a figure of eight, and marching and/or jogging in place. If feasible, the patient should be asked to perform these tasks at various speeds with the eyes open and closed.

Additional tests can be used to assess functional balance in subjects with peripheral vestibular dysfunction. The DGI, as developed by Shumway-Cook and associates[27,30,136,141-143] is very helpful in quantifying gait dysfunction in people with vestibular disease (Box 21-6). It is currently undergoing a revision that is not available at the time of publication (personal communication, A Shumway-Cook). This eight-item test takes less than 10 minutes to perform and requires little equipment (a shoebox, two cones, and stairs). Scoring is based on the concept of no, minimal, moderate, or severe gait dysfunction when performing the eight gait tasks. It has been related to scores on the Berg Balance Scale,[141,142] a tool to assess balance. Scores of 19 or less on the DGI have been related to falls in community-living elderly adults. [138] In patients with severe vestibular disability, we have seen scores as low as 3 out of 24! The DGI is very useful for quantifying gait dysfunction in people with vestibular dysfunction and can be easily used to determine whether the therapy intervention is effective.

The Berg Balance Scale (BBS) is also a useful tool to use if the patient has balance dysfunction![143-146] Not all patients with peripheral vestibular disorders have balance disorders.[97] Scores of 36 and less on this test indicate a 100% risk of falling in older adults![141,143,147,148] This scale consists of 14 items that include sitting, standing, and reaching activities with a total point value of 56. It has been shown to be a reliable and valid test with many different patient populations and is very useful in assessing change in a patient's balance over time. Concurrent validity of the BBS and the DGI has been established in a mixed group of patients with vestibular dysfunction.[143]

Another tool that might be helpful to assess gait over time is the Timed "Up and Go" (TUG) test. [149] This test consists of having the subject rise from a standard height chair with armrests, walk 3 m, turn, and return to sit in the

Figure 21.8 The obstacle course can also be used to determine how well the patient responds to external perturbations. In this situation, the patient maintains postural control while simultaneously stepping over a box and catching a ball.

Box 21-6

DYNAMIC GAIT INDEX

1. Gait Level Surface

Instructions: Walk at your normal speed from here to next mark (20 ft).

Grading: Mark the lowest category that applies:

- Normal: Walks 20 ft, no assistive devices, good speed, no evidence of imbalance, normal gait pattern.
- Mild impairment: Walks 20 ft, uses assistive devices, slower speed, mild gait deviations.
- Moderate impairment: Walks 20 ft, slow speed, abnormal gait pattern, evidence of imbalance.
- Severe impairment: Cannot walk 20 ft without assistance, severe gait deviations, or imbalance.

2. Change in Gait Speed

Instructions: Begin walking at your normal pace (for 5 ft). When I tell you "Go," walk as fast as you can (for 5 ft). When I tell you "Slow," walk as slowly as you can (for 5 ft).

Grading: Mark the lowest category that applies:

- Normal: Able to smoothly change walking speed without loss of balance or gait deviation. Shows a significant difference in walking speeds between normal, fast, and slow speeds.
- Mild impairment: Is able to change speed but demonstrates mild gait deviations; *or* has no gait deviations but unable to achieve a significant change in velocity; o*r* uses assistive device.
- Moderate impairment: Makes only minor adjustments to walking speed; *or* accomplishes a change in speed with significant gait deviations; *or* changes speed but has significant gait deviations; *or* changes speed but loses balance, but is able to recover and continue walking.
- Severe impairment: Cannot change speeds; *or* loses balance and has to reach for wall or be caught.

3. Gait with Horizontal Head Turns

Instructions: Begin walking at your normal pace. When I tell you to "Look right," keep walking straight, but turn your head to the right. Keep looking to the right until I tell you to "Look left"; then keep walking straight and turn your head to the left. Keep your head to the left until I tell you to "Look straight"; then keep walking straight but return your head to the center.

Grading: Mark the lowest category that applies:

- Normal: Performs head turns smoothly with no change in gait.
- Mild impairment: Performs head turns smoothly with slight change in gait velocity (i.e., minor disruption of smooth gait path or uses walking aid).
- Moderate impairment: Performs head turns with moderate change in gait velocity, slows down, staggers but recovers, can continue to walk.
- Severe impairment: Performs task with severe disruptions of gait (i.e., staggers outside 15-degree path, loses balance, stops, reaches for wall).

4. Gait with Vertical Head Turns

Instructions: Begin walking at your normal pace. When I tell you to "Look up," keep walking straight, but tip your head and look up. Keep looking up until I tell you "Look down." Then keep walking straight and turn your head down. Keep looking down until I tell you to "Look straight"; then keep walking straight, but return your head to the center.

Grading: Mark the lowest category that applies:

- Normal: Performs head turns with no change in gait.
- Mild impairment: Performs task with slight change in gait velocity (i.e., minor disruption to smooth gait path or uses walking aid).
- Moderate impairment: Performs tasks with moderate change in gait velocity, slows down, staggers but recovers, can continue to walk.
- Severe impairment: Performs task with severe disruption or gait (i.e., staggers outside 15-degree path, loses balance, stops, reaches for wall).

5. Gait and Pivot Turn

Instructions: Begin walking at your normal pace. When I tell you "Turn and stop," turn as quickly as you can to face the opposite direction, and stop.

Grading: Mark the lowest category that applies:

- Normal: Pivot turns safely within 3 seconds and stops quickly with no loss of balance.
- Mild impairment: Pivot turns safely in greater than 3 seconds and stops with no loss of balance.
- Moderate impairment: Turns slowly, requires verbal cuing, requires several small steps to catch balance after turn and stop.

Continued

Box 21-6

DYNAMIC GAIT INDEX—cont'd

- Severe impairment: Cannot turn safely, requires assistance to turn and stop.

6. Step Over Obstacle

Instructions: Begin walking at your normal speed. When you come to the shoe box, step over it, not around it, and keep walking.

Grading: Mark the lowest category that applies:
- Normal: Is able to step over box without changing gait speed; no evidence of imbalance.
- Mild impairment: Is able to step over box, but must slow down and adjust steps to clear box safely.
- Moderate impairment: Is able to step over box but must stop, then step over. May require verbal cuing.
- Severe impairment: Cannot perform without assistance.

7. Step Around Obstacles

Instructions: Begin walking at your normal speed. When you come to the first cone (about 6 ft away), walk around the right side of it. When you come to the second cone (6 ft past first cone), walk around it to the left.

Grading: Mark the lowest category that applies:
- Normal: Is able to walk around cones safely without changing gait speed; no evidence of imbalance.
- Mild impairment: Is able to step around both cones, but must slow down and adjust steps to clear cones.
- Moderate impairment: Is able to clear cones but must significantly slow speed to acomplish task, or requires verbal cuing.
- Severe impairment: Unable to clear cones, walks into one or both cones, or requires physical assistance.

8. Steps

Instructions: Walk up these stairs as you would at home (i.e., using the rail if necessary). At the top, turn around and walk down.

Grading: Mark the lowest category that applies:
- Normal: Alternating feet, no rail.
- Mild impairment: Alternating feet, must use rail.
- Moderate impairment: Two feet to stair, must use rail.
- Severe impairment: Cannot do safely.

From Shumway-Cook and Woollacott, 1995.[136]

chair. It is closely related to speed of gait and is a quick method to assess gait performance over time in any physical setting. In people with vestibular dysfunction, it is often helpful to have the subject turn to the right and also to the left to detect any asymmetry. The test takes less than 1 minute to complete and requires only a chair and a stopwatch. In a mixed group of patients with vestibular disorders, scores of 11.1 seconds or greater were 5 times more likely to have reported a fall in the previous 6 months.[30] At 11.1 seconds, the sensitivity of the test was 80% and the specificity was 56%, suggesting that the tool may be helpful for use in persons with vestibular dysfunction.[30] The TUG has been modified to include components that assess an individual's ability to allocate attention.[148,150] Although the modified TUG has not yet been validated in patients with vestibular deficits, it adds an important component to balance testing—that of the effect of cognitive impairments on balance.

The four-square-step test has been validated with persons with vestibular disorders.[151,152] Straight canes are aligned at 90-degree angles to each other, similar to a plus sign. The subject is asked to step into each block twice, going forwards and then repeating the pattern backwards as they are timed and instructed to perform the task as quickly as possible. The test was originally designed to assess risk of falling in older adults. It functionally provides information about the ability to make quick changes of direction and tests the ability to step backwards.

Red Flags

As the assessment is being performed, certain "red flags" may appear. Signs and symptoms of central nervous system pathology must be recognized and reported to the referring physician (Box 21-7). It is possible for individuals to be referred to a balance and vestibular clinic with undiagnosed central nervous system disease. Multiple sclerosis, brainstem transient ischemic attacks (TIAs), Parkinson's disease, cerebellar disorders, and migraines are but a few of the disorders that have been diagnosed in patients

Box 21-7

"RED FLAGS" DURING ASSESSMENT THAT INDICATE A NEED TO ASK FURTHER QUESTIONS

Numbness

Tingling

Weakness

Unilateral hearing loss

Slurred speech

Progressive hearing loss

Tremors

Poor coordination

Upper motor neuron signs and symptoms:
- Presence of Babinski sign
- Spasticity
- Clonus

Loss of consciousness

Rigidity

Visual field loss

Memory loss

Cranial nerve dysfunction

Spontaneous nystagmus in room light after 2 weeks

Vertical nystagmus without torsional component (not benign paroxysmal positional vertigo) in room light

presenting to our clinics with the diagnosis of dizziness. It is very important that individuals who treat people with vestibular dysfunction become very good at making a physical therapy diagnosis on the basis of the patient's symptoms and physical findings. The patient history gathered early in the assessment often guides the experienced clinician in deciding which path to follow in further evaluating the patient. Red flags are often identified in the history if the correct questions are asked of the patient. For example, patients who describe their "dizziness" as a sensation of being pushed or having sensory changes associated with their "dizziness" may actually be experiencing TIAs (Box 21-7).

Transition from Assessment to Treatment

The following points are offered as guidelines for the development of a treatment program based on the assessment.

Is There a Documented Vestibular Deficit?

The results of any formal vestibular function tests should be reviewed. If vestibular testing has been performed, it is very helpful to obtain the results before initiating treatment. If the vestibular function test results are normal, the therapist may or may not be dealing with a vestibular deficit. The results of the typical vestibular function tests confirm the presence of horizontal semicircular canal deficits. Of course, a patient may have a vertical canal lesion without a horizontal canal problem, but that is most likely to occur in BPPV, which is usually easily recognized. Otolith and central vestibular lesions are more difficult to identify, and the therapist must rely on patient history or on the presence of other deficits that localize the problem to the central nervous system ifVEMP or subjective vertical testing is not available. The "bucket test" might be helpful as a clinical test of the subjective visual vertical, because it discriminates fairly well between normal results and persons with vestibular dysfunction (both peripheral and central).[153] Persons with normal vestibular test results may still have a vestibular disorder . Only one-fifth of the vestibular apparatus (the horizontal canal) is typically tested by laboratory vestibular function testing, although utricular and saccular function tests now are available (see Chapters 11 and 12.

What Type of Vestibular Problem Does This Patient Have?

The vestibular function test results indicate whether the patient has a peripheral, unilateral, central, or mixed disorder (both peripheral *and* central) or a bilateral vestibular deficit as well as the severity of the deficit. Some of the exercises for patients with vestibular deficits are designed to improve the remaining vestibular function or to induce compensatory responses from the central nervous system and therefore are not appropriate for patients with complete vestibular loss. Similarly, exercises for BPPV (see Chapter 20) will not help the patient with a UVL (see Chapter 22).

Not All Dizzy Patients Have a Vestibular Lesion

Although nystagmus and complaints of dizziness and vertigo are common in patients with vestibular deficits, these problems can occur in other , non-vestibular disorders. Nystagmus may be caused by medications or brainstem or cerebral hemisphere lesions, or may be congenital. Dizziness may occur in patients with peripheral somatosensory deficits (like patients with peripheral vestibular deficits, such patients feel dizzy when they are standing

but not when they are sitting), with central nervous system lesions, or with other medical problems such as low blood pressure, or as a side effect of medications. Vertigo, most frequently associated with peripheral vestibular deficits, can occur with central lesions (often, patients with central lesions may not complain of vertigo) and with other medical problems, such as presyncope. These patients may have balance problems and may need an individualized exercise program but not necessarily exercises designed to improve vestibular function. It is not known what interventions are optimal for persons with central vestibular dysfunction, although there is some evidence that they tend to improve with rehabilitation.[154]

Assess and Reassess

The initial assessment is directed at identifying problems associated with the vestibular deficit, such as increased dizziness or decreased visual acuity with head movements, and with the functional limitations of the patient. The exercise plan, therefore, may include vestibular exercises but must also address the specific problems and level of function of the individual patient. Developing a problem list enables the therapist to set goals and devise specific exercises for each of those goals. As the patient responds to the exercises, reassessment is necessary to allow modification of the treatment program. The participation of the patient is *key* in developing a plan of care that is mutually agreed on and feasible for the patient's lifestyle.

Quantify the Assessment

Although not all components of the assessment yield quantifiable data, many tests do, such as the patient's subjective complaints, DVA, measures of postural stability, and most of the gait and balance measures. Taking the time to objectively measure the patient's performance is important for the therapist to determine the outcome of the patient's intervention, to modify the intervention, to justify further intervention, and to determine when to terminate intervention.

Determining Whether There Has Been Improvement

One of the most difficult parts of the assessment is deciding what criteria to use in determining whether the patient's problem has actually improved. Many studies that examine changes in patients with performance of vestibular exercises use a statistically significant change in test score as the criteria for "improvement."[33,43,155-157] However, statistically significant change does not necessarily indicate a *clinically meaningful change*, especially from the patient's perspective. Some studies have begun to define change according to criteria that may be more clinically relevant, but the criteria can vary considerably among studies. Brown and colleagues[118] defined clinically significant change for the DHI as a change greater than 18 points, but the investigators did not describe how they arrived at these criteria.

A second method of examining improvement has been to determine whether the patient's performance has returned to within normal values for age. This information exists for only some of the outcome measures we use, such as the computerized DVA test and gait speed.[40,157] A third method would be to use values based on comparisons with other information. For example, the cutoff value for low risk for falls using the DGI was established by comparing test scores against the person's history of falling.[142,143] A fourth approach has been to combine the scores for several tests and then define outcome on the basis of the total or weighted score. Whitney and colleagues[61] used a composite score based on the ABC, DHI, and DGI scores. Finally, criteria for improvement can be based on change in score that is beyond the within-subject variability of the measures.[68] Clearly, more research is needed in this important area.

Summary

The physical therapy assessment is multifaceted and aimed toward identifying the patient's specific functional deficits and to quantitatively establish the effects of the vestibular deficit on the patient's vestibulo-ocular and vestibulospinal systems and subjective complaints of disequilibrium and vertigo. The results of the assessment are used to identify specific patient problems and to develop treatment goals for the patient. The results of the assessment also provide the basis for determining whether the interventions used are successful. The aim of exercise in the rehabilitation of patients with vestibular disorders is to promote vestibular compensation and functional recovery.

Acknowledgments

The authors wish to acknowledge the contribution of Diane F. Borello-France, PhD, PT, who cowrote this chapter in the first edition of *Vestibular Rehabilitation*. Some of the work described here was supported by NIH grant DC05384 (SLW) and NIH grant DC03196 (SJH).

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Appendix 21-A
Initial Evaluation Form

Evaluation

Initial: Yes No Follow-up: Yes No
Date: _____
Patient: _____ Medical Record #: _____
D.O.B. _____ Age: ____
Referring physicians and physicians to whom we should send report (please give addresses):
Describe the major problem or reason you are seeing us:
When did this problem begin? _____
Specifically, do you experience spells of vertigo (a sense of spinning)? Yes No
If yes, how long do these spells last? _____
When was the last time the vertigo occurred? _____
Is the vertigo:

spontaneous:	Yes	No
induced by motion:	Yes	No
induced by position changes:	Yes	No

Do you experience a sense of being off-balance (disequilibrium)? Yes No
If yes, is the feeling of being off-balance:

constant:	Yes	No
spontaneous:	Yes	No
induced by motion:	Yes	No
induced by position changes:	Yes	No
worse with fatigue:	Yes	No
worse in the dark:	Yes	No
worse outside:	Yes	No
worse on uneven surfaces:	Yes	No

Does the feeling of being off-balance occur when you are:

lying down	Yes	No
sitting	Yes	No
standing	Yes	No
walking	Yes	No

Do you or have you fallen (to the ground)? Yes No
If yes, please describe _____
How often do you fall? _____
Have you injured yourself? Yes No
If yes, please describe: _____
Do you or have you had "near falls"? Yes No
Do you stumble, stagger, or side-step while walking? Yes No
Do you drift to one side while you walk? Yes No
If yes, to which side do you drift? Right Left

Pertinent Past Medical History

Do you have:

Diabetes:	Yes	No
Heart disease:	Yes	No
Hypertension:	Yes	No
Headaches:	Yes	No
Arthritis:	Yes	No
Migraines	Yes	No
Neck problems:	Yes	No
Back problems:	Yes	No
Pulmonary problems:	Yes	No
Weakness or paralysis:	Yes	No
Hearing problems:	Yes	No
If yes, describe _____		
Visual problems:	Yes	No
If yes, describe _____		
Have you been in an accident?	Yes	No
If yes, please describe _____		
When did it occur? _____		

What medications do you take?
_____
_____
_____

Social History

Do you live alone? Yes No
If No, who lives with you?_____
Do you have stairs in your home? Yes No If yes, how many? _____
Do you smoke? Yes No If yes, please indicate how much per day_____
Do you drink alcohol? Yes No If yes, please indicate how much _____
Do you have trouble sleeping? Yes No

PANAS (Positive and Negative Affective Scale)[1]

The scale below consists of a number of words that describe different feelings and emotions. Read each item and then mark the appropriate answer in the space next to that

word. Indicate to what extent you generally feel this way—that is, how you feel on the average. Use the following scale to record your answers:

1 2 3 4 5
very slightly or not at all | a little | moderately | quite a bit | extremely

_____ interested _____ irritable _____ jittery _____ strong _____ nervous
_____ enthusiastic _____ distressed _____ alert _____ active _____ excited
_____ ashamed _____ afraid _____ upset _____ inspired _____ hostile
_____ guilty _____ determined _____ proud _____ scared _____ attentive

How would you describe your functional level of activities before this problem developed?

_____

Current Functional Status

Are you independent in self-care activities: Yes No
Can you drive: In the daytime? Yes No In the nighttime? Yes No
Have you modified your driving habits? If yes, in what way? _____
Are you working? Yes No Not applicable If yes, occupation: _____
Are you on Medical Disability? Yes No
Can you perform all your normal parenting activities?Yes No Not applicable
Are you able to:

Watch TV comfortably?	Yes	No
Read?	Yes	No
Go shopping?	Yes	No
Be in traffic?	Yes	No
Use a computer?	Yes	No

Initial Visit: Disability Scale[2]

Please pick the one statement from the following list that best describes how you feel:[2]

___ Have negligible symptoms
___ Have bothersome symptoms
___ Perform usual work duties but symptoms interfere with outside activities
___ Symptoms disrupt performance of both usual work duties and outside activities
___ Am currently on medical leave or had to change jobs because of symptoms
___ Have been unable to work for more than 1 year or have established permanent disability with compensation payments

Final Visit

Please pick the one statement from the following list that best describes how you feel:[2]

_____ No symptoms remaining at the end of therapy
_____ Marked improvement remaining at the end of therapy
_____ Mild improvement, definite persistent symptoms at the end of therapy
_____ No change in symptoms in relation to therapy
_____ Symptoms worsened with therapy activities on a persistent basis compared with pretherapy period

Assessment

Subjective Complaints

PANAS[1] Score/significance: _____
Rate the following symptoms from 0 (none) to 10 (as bad as it can be):
Vertigo: (0–10) _____ Disequilibrium: (0–10) _____ Oscillopsia (0–10) _____
What time of day do you feel best: _____ worst: _____
How many times per day do you experience symptoms? _____
Major problems: _____
Disability score: _____ (pre-therapy)
_____ (post-therapy)

Motion Sensitivity Quotient:[2]

_____

MSQ = {(Total score) × (# of positions with symptoms)} ÷ 20.48.
(Don't forget to adjust for baseline intensity so MSQ is based on change in intensity.)
MSQ: 0–10 = mild; 11–30 = moderate; 31–100 = severe.

Baseline Symptoms	Intensity (0–5)	Duration*	Score (Intensity + Duration in Points)
1. Sitting to supine			
2. Supine to left side			
3. Supine to right side			
4. Supine to sitting			
5. Left Dix-Hallpike			
6. Return to sit from left Dix-Hallpike			
7. Right Dix-Hallpike			
8. Return to sit from right Dix-Hallpike			
9. Sitting, head tipped to left knee			
10. Head up from left knee			
11. Sitting, head tipped to right knee			
12. Head up from right knee			
13. Sitting, turn head horizontally 5 times			
14. Sitting, move head vertically 5 times			
15. Standing, turn 180° to the right			
16. Standing, turn 180° to the left			

*Duration: 5–10 sec = 1 point; 11–30 sec = 2 points; >30 sec = 3 points.

Oculomotor Examination

Room light:

A. Spontaneous nystagmus Y N
B. Gaze-holding nystagmus Y N
C. Smooth pursuit _____
D. Saccadic eye movements _____
E. VORc _____
F. VOR slow _____
G. VOR rapid head thrusts _____
H. Visual acuity stationary: _____ Dynamic _____

Frenzel/IR (recorded Y or N):

A. Spontaneous nystagmus? Y N
B. Gaze-holding nystagmus? Y N
C. Horizontal head-shaking–induced nystagmus? _____
D. Vertical head-shaking–induced nystagmus? _____
E. R Dix-Hallpike maneuver: nystagmus _____ vertigo _____
F. L Dix-Hallpike maneuver: nystagmus _____ vertigo _____
G. Supine roll head right: nystagmus _____ vertigo _____
H. Supine roll head left: nystagmus _____ vertigo _____
I. Pressure test _____

Quantitative Dynamic Visual Acuity (DVA): static _____right _____left _____

Caloric Test: _____

Stance Postural Control

A. Alignment eyes open: _____
(erect, head still, base of support)
B. Self-initiated weight-shifting: _____
(strategy)
C. Reactive responses (sternal shove): _____
(strategy)

Balance Tests

A. Romberg: eo _____ ec _____ Normal/abnormal for age? _____
B. Sharpened Romberg: eo _____ ec _____ Normal/abnormal for age? _____
C. Single leg stance: eo _____ ec _____ Normal/abnormal for age? _____
D. Functional reach: _____(normal ≥12 inches)

Gait

Assistive devices: _____ Orthoses? _____
A. At self-initiated pace:
Speed: _____ Normal for age? _____
Cadence: _____ Base of support: _____
Step length: _____ (equal, each foot passes the other)
Arm swing: _____
Head and trunk rotation: _____
Path: Straight? Y N Swerves: R L
Staggers: Y N Side-steps: Y N
B. As fast as possible: Can perform Cannot perform
Speed: _____ Normal for age? _____
Cadence: _____ Base of support: _____
Step length: _____ (equal, each foot passes the other)
Arm swing: _____
Head and trunk rotation: _____
Path: Straight? Y N Drifts: R L
Staggers: Y N Side-steps: Y N
C. Gait deviations: (20 foot path, 12 inches wide)
_____
D. Walk, turn head: Can perform Cannot perform
Cadence: _____ Base of support: _____
Path: Straight? Y N Drifts: R L
Staggers: Y N Side-steps: Y N
E. Walk, move head vertically
Cadence: _____ Base of support: _____
Path: Straight? Y N Drifts: R L
Staggers: Y N Side-steps: Y N
F. Walk, turn head and count backwards out loud by _____:
Cadence: _____ Base of support: _____
Path: Straight? Y N Drifts: R L
Staggers: Y N Side-steps: Y N
G. Walk, turn around rapidly:
Normal: _____
Has difficulty/loses balance turning right _____ turning left _____
H. Fall Risk (Dynamic Gait Index or Functional Gait Assessment): _____

Functional Gait

Independent: min/mod/max assist
Dependent: slow, cautious
Stairs _____ Inclines _____
Uneven surfaces _____ Carpet _____

Subsystems

A. ROM:
Cervical_____
R- UE _____ L- UE _____ R- LE _____ L- LE _____
B. Strength: R- UE _____ L- UE _____ R- LE _____ L-LE _____
C. Soft tissue problems: Y N Location: _____
Nature: (spasm?) _____
D. Sensation: (vibration, proprioception, kinesthesia) LEs _____
E. Muscle tone: UEs _____ LEs _____
F. Cerebellar: Finger to nose R _____ L _____
Rapid alternating movement: R _____ L _____
Optic ataxia R _____ L _____
Tremor R _____ L _____
Heel to shin—looking R _____ L _____
Heel to shin—not looking R _____ L _____
Rapid alternating movement R _____ L _____
G. Pain: Y N Location _____

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Appendix 21-B Dizziness Handicap Inventory[1]

Name: _____ Date: _____

The purpose of this scale is to identify dif ficulties that you may be experiencing because of your dizziness or unsteadiness. Please check Yes, No, or Sometimes for each question. Answer each question as it pertains to your dizziness or unsteadiness only.

	Yes	No	Sometimes
P1. Does looking up increase your problem?	_____	_____	_____
E2. Because of your problem, do you feel frustrated?	_____	_____	_____
F3. Because of your problem, do you restrict your travel for business or recreation?	_____	_____	_____
P4. Does walking down the aisle of a supermarket increase your problem?	_____	_____	_____
F5. Because of your problem, do you have difficulty getting into or out of bed?	_____	_____	_____
F6. Does your problem significantly restrict your participation in social activities such as going out to dinner, the movies, dancing, or to parties?	_____	_____	_____
F7. Because of your problem, do you have difficulty reading?	_____	_____	_____
P8. Does performing more ambitious activities like sports or dancing or household chores such as sweeping or putting dishes away increase your problem?	_____	_____	_____
E9. Because of your problem, are you afraid to leave your home without having someone accompany you?	_____	_____	_____
E10. Because of your problem, are you embarrassed in front of others?	_____	_____	_____
P11. Do quick movements of your head increase your problem?	_____	_____	_____
F12. Because of your problem, do you avoid heights?	_____	_____	_____
P13. Does turning over in bed increase your problem?	_____	_____	_____

	Yes	No	Sometimes
F14. Because of your problem, is it difficult for you to do strenuous housework or yardwork?	_____	_____	_____
E15. Because of your problem, are you afraid people may think you are intoxicated?	_____	_____	_____
F16. Because of your problem, is it difficult for you to walk by yourself?	_____	_____	_____
P17. Does walking down a sidewalk increase your problem?	_____	_____	_____
E18. Because of your problem, is it difficult for you to concentrate?	_____	_____	_____
F19. Because of your problem, is it difficult for you to walk around your house in the dark?	_____	_____	_____
E20. Because of your problem, are you afraid to stay home alone?	_____	_____	_____
E21. Because of your problem, do you feel handicapped?	_____	_____	_____
E22. Has your problem placed stress on your relationships with members of your family or friends?	_____	_____	_____
E23. Because of your problem, are you depressed?	_____	_____	_____
F24. Does your problem interfere with your job or household responsibilities?	_____	_____	_____
P25. Does bending over increase your problem?	_____	_____	_____
Total	_____	_____	_____
	(4)	(0)	(2)
Total: _____ F _____ E _____ P _____			
(36) (36) (28)			

Appendix Reference

1. Jacobson GP, Newman CW. The development of the dizziness handicap inventory. *Arch Otolaryngol Head Neck Surg*. 1990;116:424. Copyright © 1990 The American Medical Association.

CHAPTER 22

Physical Therapy Treatment of Vestibular Hypofunction

Susan J. Herdman, PT, PhD, FAPTA
Susan L. Whitney, DPT, PhD, NCS, ATC, FAPTA

Vestibular rehabilitation is considered an appropriate and valuable treatment approach for patients with v estibular hypofunction. This chapter provides the reader with the background necessary to treat patients with vestibular hypofunction. The chapter covers mechanisms of reco very as the basis for the exercise approaches used and similarities and differences among the v arious treatments and guidelines for the progression of the exercises. We provide a review of literature that supports the use of these e xercises. Finally, we provide case studies to illustrate the decision-making process in developing exercise programs.

> The previously used nomenclature to describe the exercises used in the treatment of v estibular hypofunction needs to be revised in light of recent findings on the mechanisms underlying improvement in physical function. The previously described "Adaptation" and "Substitution" e xercises are together called "Gaze Stabilization" exercises.

Goals of Treatment

The goals of physical therap y intervention are to (1) decrease the patient's disequilibrium (sense of being off-balance) and oscillopsia (visual blurring during head movement), (2) improve the patient's functional balance especially during ambulation, (3) improve the patient's ability to see clearly during head movement, (4) improve the patient's overall general physical condition, (5) enable the patient to return to a more normal level of activity and participation in society, and (6) reduce the patient's social isolation. Patients are usually seen by the physical therapist on an outpatient basis, although the initial interv entions often occur while the patient is in the hospital. Several centers within the United States and England employ physical therapists who screen for vestibular dysfunction in the emer gency room. An important part of the rehabilitation process is the establishment of a home e xercise program. The physical therapist must motivate the patient and ensure e xercise compliance. To do so, the physical therapist must identify the patient' s own goals and clarify to the patient the treatment goals as well as the potential effects of exercise.

Mechanisms of Recovery

Several different mechanisms are involved in the recovery of function following unilateral v estibular loss (UVL). These mechanisms include cellular recovery, spontaneous reestablishment of the tonic-firing rate centrally, adaptation of residual vestibular function, the substitution of alternative strategies for the loss of vestibular function, and habituation of unpleasant sensations. The following section relates the changes that are seen follo wing UVL to the mechanisms of recovery.

Cellular Recovery

Cellular recovery suggests that the receptors or neurons that were damaged and initially stopped functioning may recover. This has been demonstrated for vestibular hair cells in nonprimate mammals following aminoglycoside-induced loss.[1,2] There appears to be some functional recovery related to the anatomic recovery of hairs cells although there is a persistent deficit.[3] It is unclear at this time whether recovery of hair cells is a significant factor in recovery of vestibular function in human beings.

Spontaneous Nystagmus, Skew Deviation, and Postural Asymmetries in Stance

Nystagmus, skew deviation, and postural asymmetries in stance occur because of the asymmetric disturbances of static vestibular function, and all recover spontaneously as the static or tonic firing rate is rebalanced.[4,5] These symptoms and signs are caused by the disruption of tonic vestibulo-ocular and vestibulospinal responses. Unilateral loss of the input from the semicircular canals results in an asymmetry in those inputs, which is interpreted as head movement. The loss of the signal from the semicircular canals on one side results in spontaneous nystagmus. Unilateral loss of utricular inputs results in a skew deviation in which the eye on the side of the lesion drops in the orbit. Patients with skew deviations complain of a vertical diplopia.[6] Disruption of the tonic vestibulospinal responses produces an asymmetry in the muscle activity in the lower extremities, as measured electromyographically, while the patient is standing,[7] and in a postural asymmetry, which can be detected clinically.[8] These signs and symptoms resolve spontaneously within 3 to 14 days following onset of the unilateral vestibular deficit. The timing of the disappearance of these symptoms parallels the recovery of the resting firing rate of the vestibular nucleus neurons and is not dependent on other sensory inputs.[9,10]

Head Movement–Induced Disequilibrium, Imbalance, and Visual Blurring

These symptoms and signs are caused by the disruption or loss of the vestibular response to head movement, the dynamic portion of the vestibular system. Initially, the gain of the vestibulo-ocular reflex (VOR) is decreased by as much as 75% for head movements toward the side of the lesion resulting in visual blurring during head movement.[11] Presumably, the gain of the dynamic vestibulo-spinal response is also reduced and results in disequilibrium and gait ataxia.

There is a wealth of evidence that recovery from the dynamic disturbances of vestibular function requires both visual inputs and movement of the body and head.[5,12-15] The gain of the vestibulo-ocular response does not recover when cats or monkeys are kept in the dark following unilateral labyrinthectomy.[5,14] Recovery of vestibulo-ocular gain begins when the animals are returned to a lighted environment. Similarly, if animals are prevented from moving after unilateral vestibular nerve section, there is a delay in the onset of the recovery of postural stability. Thus, the recovery period is prolonged (Fig. 22.1).[15] The following

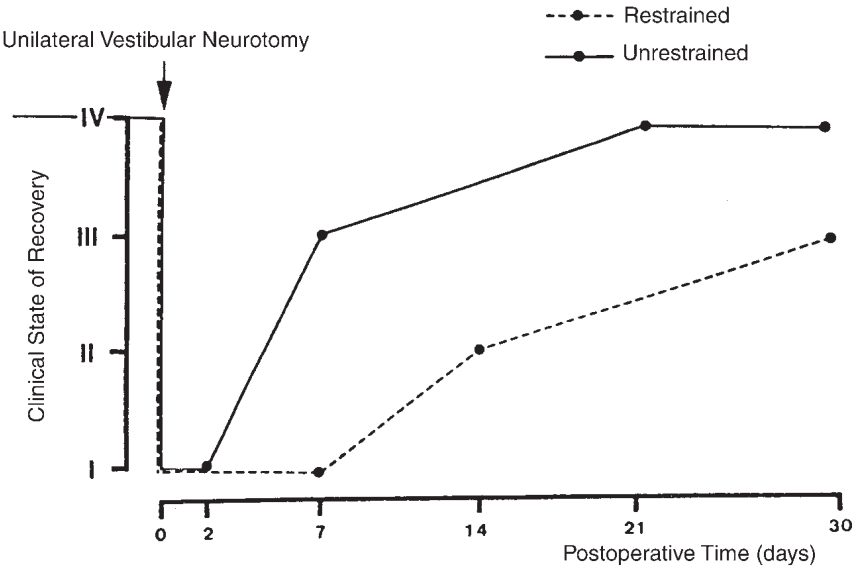

Figure 22.1 Effect of restricting mobility on rate of recovery following unilateral transection of vestibular nerve. Baboons that were restrained after unilateral vestibular nerve section had a delayed onset of recovery and a more prolonged recovery than those animals that were allowed free movement. (Reprinted with permission from Lacour M, Roll JP, Appaix M. Modifications and development of spinal reflexes in the alert baboon (papio papio) following an unilateral vestibular neurectomy. From *Brain Res*. 1976;113:255, with permission.[15])

section presents a brief overview of some of the mechanisms that contribute to the improvement in function in patients with vestibular hypofunction. For more details, see Chapters 2 and 9.

It was initially believed that the recovery of the signs and symptoms associated with unilateral vestibular hypofunction or loss was due solely to the adaptive capability of the vestibular system—that is, the ability of the vestibular system to make long-term changes in the neuronal response to input. Several studies have demonstrated that adaptation can be induced during the acute stage after UVL. Pfaltz[16] used optokinetic stimulations and increased VOR gain in patients with UVL compared with untreated patients. Szturm et al[17] reported increased VOR gain in patients following a course of vestibular adaptation exercises but not in a control group. The primary signal for inducing vestibular adaptation is retinal slip, or the movement of a visual image across the retina.[18] This slip results in an error signal that the brain attempts to minimize by increasing the gain of the vestibular responses (see Chapter 2). However, adaptation is probably not the only mechanism behind recovery in patients with vestibular hypofunction.[19] Recent research suggests that the substitution of alternative strategies may be the basis for functional recovery (see Chapter 9). Enhancement of gaze stability and dynamic visual acuity during predictable head movement is believed to be from central preprogramming, but during passive head movements, adaptation may be the primary mechanism of improvement.[19,20] For example, patients with vestibular hypofunction may make a saccade during the head thrust itself. These preprogrammed saccades occur at a latency that is too short to be voluntary or reflexive[21] and are not present in normal subjects.[22] There is evidence that they can be "uncovered" by making the amplitude of the head thrust unpredictable.[23] Another mechanism that has been described is *anticipatory* slow-phase eye movement. These slow-phase eye movements are initiated more than 5 milliseconds (ms) *before* the initiation of predictable head thrusts, are in the appropriate direction for the direction of the head movement, and are initiated centrally. Other slow-phase eye movements probably are initiated by the intact vestibular system, based on the latency to onset of these eye movements (less than 10 ms *after* the head thrust), which is within the range for the VOR.[24]

Recovery of postural stability may be caused by the use of visual and somatosensory cues instead of remaining vestibular cues. Although the substitution of visual or somatosensory cues as a strategy may provide sufficient information for postural stability in many situations, the patient will be at a disadvantage if trying to walk when those cues are inaccurate or not available, such as in the dark. At an extreme, some patients may modify their behavior to avoid situations in which visual or somatosensory cues are diminished, such as going out at night.

These mechanisms do not adequately substitute for the lost vestibulo-ocular function, which functions across frequencies of 8 Hz or more[25,26] for activities such as walking and running and gaze stabilization during unpredictable and predictable head movements. Preprogrammed saccades and anticipatory eye movements occur primarily with predictable head movements and not during unpredictable head movements. Other types of eye movements do not function across the necessary frequency range (Fig. 22.2). For example, pursuit eye movements are limited to less than 1 Hz and a maximum eye velocity of 60 degrees per second.[27] Saccadic eye movements would not be a particularly useful alternative for a poor VOR, because patients would not be able to see the target clearly during the actual eye movements.[28] The cervico-ocular reflex also does not operate in the appropriate frequency range, and well-controlled studies have failed to identify a cervico-ocular reflex in which the eye movements are in the compensatory direction.[29,30] Similar limitations exist for the substitution of somatosensory and visual cues for lost vestibular function (Fig. 22.3). Again, the vestibular system operates through a wider frequency and velocity range than do vision and somatosensation.[25,26,31-33]

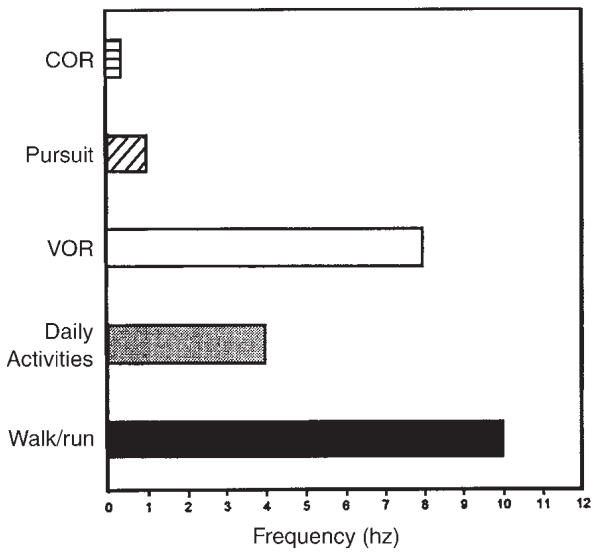

Figure 22.2 Frequency range over which the COR, smooth-pursuit eye movements, and the VOR can contribute to gaze stability compared with the frequency and velocity ranges for daily activities, walking, and running. Only the normal VOR operates over the frequency and velocity ranges of normal activities. (Modified from Herdman SJ. The role of adaptation in vestibular rehabilitation. From *Otolaryngol Head Neck Surg*. 1998;119:49-54, with permission.)

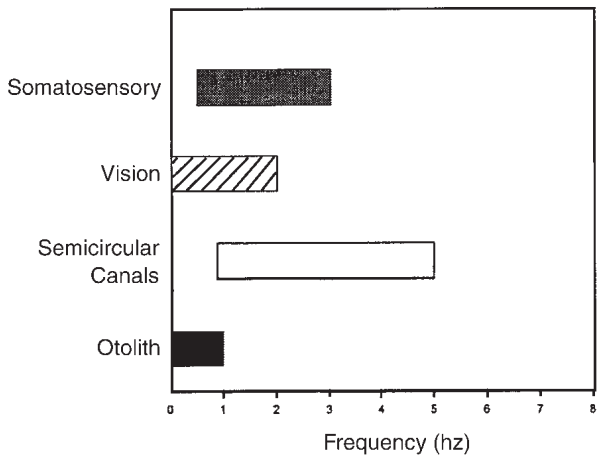

Figure 22.3 Frequency ranges over which somatosensory, visual, semicircular canal, and otolith inputs contribute to postural stability. This information has been determined only for stability during quiet stance and following sudden perturbations. (Modified from Herdman SJ. The role of adaptation in vestibular rehabilitation. From *Otolaryngol Head Neck Surg*. 1998;119:49-54, with permission.)

Patients may also restrict head movements as a means of seeing clearly or maintaining their balance. This strategy is not particularly desirable, because it would result in limited activity and would not provide a mechanism for seeing clearly or for maintaining balance during head movements. Restricting head movements can also lead to musculoskeletal impairments, which serve to compound the activity limitations from the vestibular deficit.

Reduction of Symptom Intensity

Habituation refers to a reduction in symptoms produced through repetitive exposure to the movement and presumably is a central process. The mechanism and neural circuitry are not well known.

Treatment Approaches

Several different approaches have been advocated in the management of patients with vestibular hypofunction. Three different approaches—gaze stabilization exercises, habituation exercises for the reduction of symptomatic complaints, and postural stabilization exercises—are presented. A summary and comparison of these different exercise approaches (Table 22-1) shows that there are elements common to all. Case studies are used to demonstrate the basis for specific exercises used in treatment and the progression of the patient's exercise program.

Gaze Stabilization Exercises

Three different exercises are used to enhance gaze stability in patients with vestibular hypofunction. In all, the challenge is for the eyes to stay on a target during a head movement. The mechanism by which the appropriate eye movement is generated varies from patient to patient. The first exercise is based on having the patient attempt to maintain fixation on a target while moving his or her head (Figure 22.4). These exercises were originally based on inducing adaptation of residual vestibular function by creating a small amount of retinal slip. The best stimuli

■ Table 22-1 **COMPARISON OF DIFFERENT EXERCISE APPROACHES FOR THE PATIENT WITH A PERIPHERAL VESTIBULAR DISORDER**

Movement	Gaze Stabilization	Postural Stabilization	Habituation
Incorporates head and neck exercises into the treatment approach	X	X	X
Uses a functional evaluation to assess the symptoms of the patient	X	X	X
Incorporates principles of motor control and learning into designing a treatment program	X	X	X
Practices mental exercises to increase concentration	X	X	X
Has the patient work in a variety of environments and task contexts	X	X	X

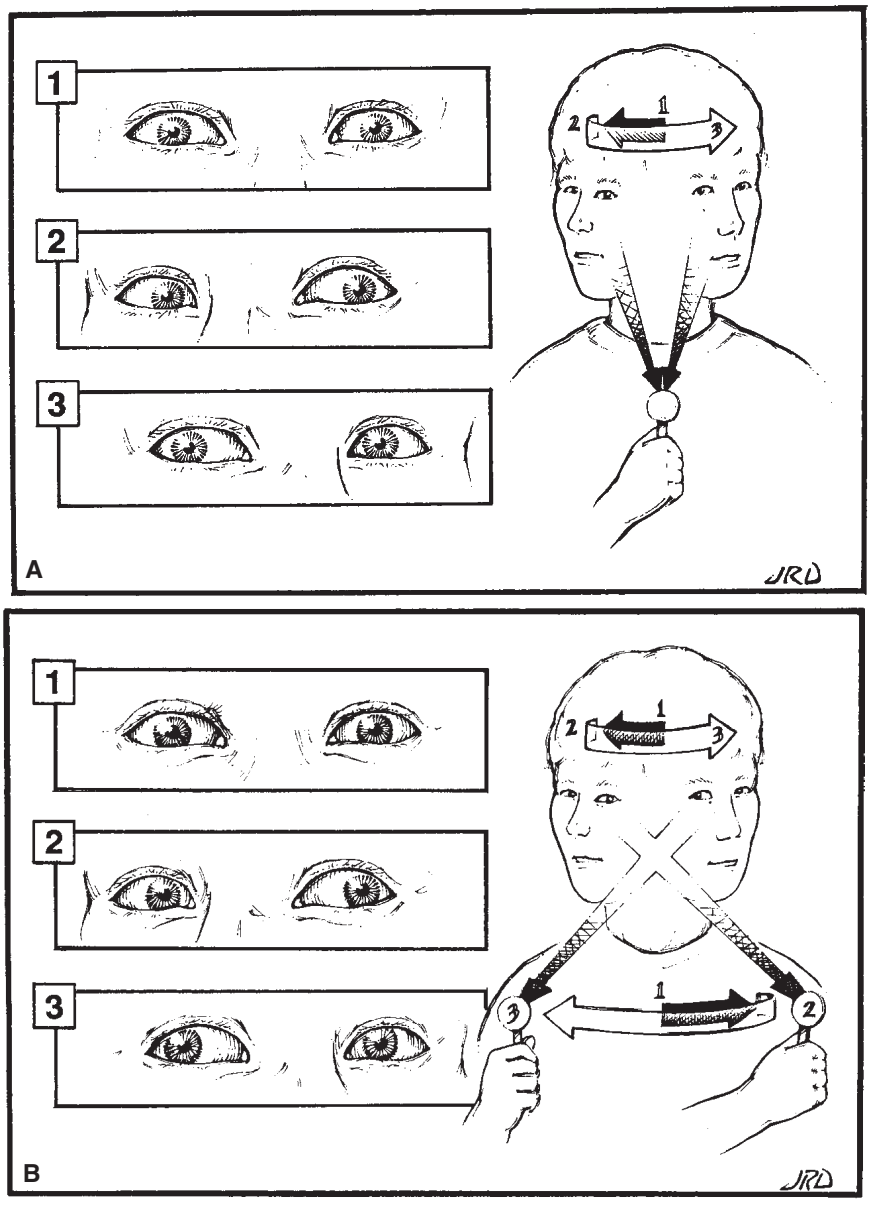

Figure 22.4 Exercises for gaze stabilization can include an X1 viewing paradigm **(A)** and an X2 viewing paradigm **(B)**. In the X1 viewing paradigm, the visual target is stationary, and the subject moves his head back and forth while trying to maintain visual fixation on the target. In the X2 viewing paradigm, the target and the head move in opposite directions, while the subject again keeps the target in focus. These exercises are performed using a small visual target (foveal stimulus) and a large visual target (full-field stimulus) with the head moving either horizontally or vertically. (Modified from Tusa RJ, Herdman SJ. Vertigo and disequilibrium. In: Johnson R, Griffin J, eds. *Current Therapy in Neurological Disease*. 4th ed. St. Louis: Mosby Year-Book; 1993:12, with permission.)

appear to be those that incorporate movement of the head and a visual input. Optokinetic stimulation (movement of visual world only) by itself also can increase the gain of the vestibular system, although perhaps not be as effectively as head movement combined with a visual stimulus.[18,34-36] Nevertheless, optokinetic stimuli have been used in the treatment of patients with vestibular hypofunction[16] or with chronic vestibular symptoms.[37]

Although there is evidence that these exercises do enhance recovery, mechanisms other than adaptation may underlie the improved gaze stability. The second exercise involves repeated eye and then head movements between two targets with the goal of maintaining fixation on the target during the head movement. Two letters (e.g., X and Z) are placed on a wall about 2 feet apart (the two targets are placed close enough together that when the patient is looking directly at one, he/she can see the other target out of the corner in the periphery). The patient is instructed to look directly at the X, being sure that the head is also lined up with the X (the nose should point at the X). Then the patient shifts gaze to the other target, keeping the head still. The patient turns his/her head toward the second target with the goal of keeping the target in focus. In patients with a poor or no VOR, other mechanisms must develop if the eyes are to remain on target during the head movement. Presumably, in this exercise, the underlying mechanism is central preprogramming.

The third exercise involves imagining fixation on a stationary target during a head movement performed with eyes closed. In this exercise, called the remembered target exercise, the patient looks at a target placed directly in front of him/her. Then the patient closes his/her eyes and, while trying to keep the eyes on the remembered target location, turns the head. The patient then opens the eyes to see if he/she is looking at the target. The exercise is repeated with the head starting in the center position. This exercise may enhance the use of cervical inputs to generate the eye movement that will keep the eye on the target, or it may enhance cortical co-activation, producing the head movement and the eye movement.

Habituation Exercises

These exercises are based on the concept that repeated exposure to a provocative stimulus will result in a reduction in the pathological response to that treatment. The first to develop exercises based on habituation were Cawthorne and Cooksey[38,39] in the 1940s. At the time, Cawthorne was treating patients with unilateral vestibular deficits and postconcussive disorders. In conjunction with Cooksey, Cawthorne developed a series of exercises that addressed their patients' complaints of vertigo and impaired balance. The Cawthorne-Cooksey exercises include pursuit and saccadic eye movements, movements of the head, tasks requiring coordination of eyes with the head, total body movements, and balance tasks (Box 22-1). Cawthorne and Cooksey recommended that the exercises be performed in various positions and at various speeds of movement. In addition, patients were required to perform the exercises with their eyes open and closed. According to Cawthorne and Cooksey,[38,39] performing the exercises with the eyes closed decreased the patient's reliance on visual information and possibly forced more effective compensation by vestibular and somatosensory mechanisms. Additionally, patients were trained to function in noisy and crowded environments. To encourage active participation, Cawthorne and Cooksey had patients exercise together in daily group sessions. Cawthorne and Cooksey

Box 22-1

EXAMPLES OF CAWTHORNE-COOKSEY EXERCISES FOR PATIENTS WITH VESTIBULAR HYPOFUNCTION OR POST-CONCUSSION

A. In bed
   1. Eye movements—at first slow, then quick
      a. up and down
      b. from side to side
      c. focusing on finger moving from 1 m to 0.3 m away from face
   2. Head movements at first slow, then quick; later with eyes closed
      a. bending forward and backward
      b. turning from side to side

B. Sitting (in class)
   1. as above
   2. as above
   3. Shoulder shrugging and circling
   4. Bending forward and picking up objects from the ground

C. Standing (in class)
   1. as A1 and A2 and B3
   2. Changing from sitting to standing position with eyes open and shut.
   3. Throwing a small ball from hand to hand (above eye level).
   4. Throwing ball from hand to hand under knee.
   5. Changing from sitting to standing and turning around in between.

D. Moving about (in class)
   1. Circle round center person who will throw a large ball and to whom it will be returned.
   2. Walk across room with eyes open and then closed.
   3. Walk up and down slope with eyes open and then closed.
   4. Walk up and down steps with eyes open and then closed.
   5. Any game involving stooping and stretching and aiming such as skittles, bowls, or basketball.

Diligence and perseverance are required, but the earlier and more regularly the exercise regimen is carried out, the faster and more complete will be the return to normal activity.

Source: Cawthorne-Cooksey exercises for patients with vestibular hypofunction. Reprinted with permission from Dix, MR. The rationale and technique of head exercises in the treatment of vertigo. *Acta Oto-rhino-laryng (Belg)*. 33;370:1979.

believed a group exercise format would be more economical and more fun for the patients, and would make it easier to identify a malingerer. Hecker et al[40] used the Cawthorne-Cooksey exercises to treat a group of patients with vestibular disorders and reported that 84% of the patients responded favorably. They also emphasized the importance of performing the exercises regularly.[40] In addition, they noted that emotional stress seemed to affect the patient's progress.

In the 1980s, Norre[41,42] proposed a different series of exercises for vestibular habituation training in the treatment of patients with unilateral peripheral vestibular loss. According to these investigators, an asymmetry in labyrinth function results in a "sensory mismatch." The disturbed vestibular signal produces an input to the brain that conflicts with information received from intact visual and somatosensory systems. This conflict, they believed, produced the symptoms experienced by patients with unilateral peripheral vestibular loss.

More recently, habituation exercises are based on the Motion Sensitivity Quotient (MSQ) as a means for evaluating initial problems and documenting outcome.[43] The MSQ, developed by Shepard and Telian, uses a series of movements and positions as the basis for establishing an individualized exercise program for patients with chronic unilateral vestibular hypofunction (Table 22-2).[43-45] Note that the Dix-Hallpike position is part of the MSQ; it is important to realize that the MSQ is *not* used in patients who have Benign Paroxysmal Positional Vertigo (BPPV). Calculation of the MSQ score involves:

- For each of the sixteen movements, the patient rates the intensity of symptoms on a scale of 0 to 5 after the movement is completed.
- The therapist calculates the number of seconds until the patient's symptoms return to baseline and applies the following criteria: less than 5 seconds = a score of 0; 5 to 10 seconds = a score of 1, 11 to 30 seconds is given a score of 2, and greater than 30 seconds is given a score of 3.
- The Intensity + Duration scores for all 16 movements is summed (added across Columns 1 and 2) and written down in Column 3.
- The numbers in Column 3 are added. That sum is multiplied by the number of positions that cause an increase of symptoms.
- The results of that multiplication are divided by 20.48 for the final score.

If the final score is 0 to 10, it indicates a mild motion sensitivity, 11 to 30 a moderate motion sensitivity, and 31 to 100 a severe motion sensitivity.

In the Habituation approach, up to four movements are chosen from the MSQ results to form the basis for the exercises. The patient performs these movements 2 or 3 times, twice a day. It is important that the patient perform the movements quickly enough and through sufficient range to produce mild to moderate symptoms.

■ Table 22-2 **MOTION SENSITIVITY QUOTIENT TEST FOR ASSESSING PATIENTS WITH DIZZINESS***

Baseline Symptoms	Intensity	Duration	Score
1. Sitting to supine			
2. Supine to left side			
3. Supine to right side			
4. Supine to sitting			
5. Left Dix-Hallpike			
6. Return to sit from left Dix-Hallpike			
7. Right Dix-Hallpike			
8. Return to sit from right Dix-Hallpike			
9. Sitting, head tipped to left knee			
10. Head up from left knee			

■ Table 22-2 **MOTION SENSITIVITY QUOTIENT TEST FOR ASSESSING PATIENTS WITH DIZZINESS*—cont'd**

Baseline Symptoms	Intensity	Duration	Score
11. Sitting, head tipped to right knee			
12. Head up from right knee			
13. Sitting, turn head horizontally 5 times			
14. Sitting, move head vertically 5 times (pitch)			
15. Standing, turn 180 deg to the right			
16. Standing, turn 180 deg to the left			

*MSQ = {(Total score) × (# of positions with symptoms)} + 20.48 MSQ score 0–10 + mild; 11–30 = moderate; 31–100 = severe. Duration: 5–10 sec + 1 point; 11–30 sec = 2 points; >30 sec = 3 points.

Source: From Shepard NT, Telian SA. Programmatic vestibular rehabilitation. *Otolaryngol Head Neck Surg*. 1995;112:173.[45]

As habituation occurs, the movements can be performed more rigorously. The patient should rest between each movement for the symptoms to stop. The symptoms should decrease within a minute after each exercise or within 15 to 30 minutes after all exercises have been performed. It may take 4 weeks for the symptoms to begin to decrease. The exercises may be performed for a week to 2 months before they are modified.

The habituation approach is not advocated for all patients. The elderly, especially, should not perform movements in which they rise quickly. Precautions include orthostatic hypotension and orthostatic intolerance. The habituation approach is often not tolerated well, because it takes time to decrease the symptoms, does not work on all patients, and compliance is difficult because of the symptoms that are exacerbated with the exercises. If treatment fails, counseling is advisable regarding changing activities or reorganizing the work area.

Postural Stabilization Exercises

Exercises should synthesize the use of visual and somatosensory cues with the use of vestibular cues as well as the possibility of central preprogramming to improve postural stability. For example, balance exercises should "stress" the system by having the patient work with and without visual cues or while altering somatosensory cues by having the patient stand on foam. Removing or altering cues forces the patient to use the remaining sensory cues. Thus, if the patient is asked to stand on foam with eyes closed, the use of vestibular cues will be fostered. The intensity of balance exercises while walking can be increased by having the person walk with a more narrow base of support, by adding head movement, or with unpredictable changes in direction of walking. The primary concerns when developing an exercise program to improve postural stability is how to challenge the patient's balance without causing the patient to fall. Fall risk should be determined before choosing the most appropriate exercises for a person with postural control deficits.

Treatment

General Considerations

1. Treatment should begin early. Most evidence suggests that patients will still improve even with long periods of time between the onset of the vestibular hypofunction to the initiation of exercises.[46,47] Logically, however, it is safer for the patient if fall risk can be decreased as quickly as possible. Furthermore, there is evidence that early intervention with vestibular exercises facilitates a decrease in symptoms and improves gait stability compared with no exercise in patients post resection of vestibular schwannoma.[48-50] In a prospective, randomized controlled trial (n = 53), an early customized vestibular rehabilitation exercise program in older adults (age greater than 50 years) post acoustic neuroma resection was more effective at enhancing postural control at 3 months and was maintained at 1 year compared with those who were provided general instructions.[49] In another

recent randomized trial in 40 persons with an acute vestibular disorder, Teggi et al reported that early vestibular exercises resulted in better Dizziness Handicap Inventory scores, less anxiety, less reliance on visual cues, and better walking, as assessed by the Dynamic Gait Index.[50] The exercise group means were all "better" than the control subjects group outcome means at 25 days post onset of the exercise program. Both studies suggest that the use of a supervised exercise program can enhance outcomes in persons with vestibular disorders.

2. Exercise can be for brief periods of time initially. Pfaltz's study[16] is important for showing that even brief periods of stimulation can produce VOR gain changes that would be particularly useful in the treatment of patients during the acute stage of recovery.
3. Patients with chronic peripheral vestibular deficits may have limited their movements, or at least their head movements, in an attempt to avoid precipitating the symptoms of dizziness and disequilibrium. Head movements must be encouraged in these patients both to induce improved function and to habituate the symptoms provoked by movement.
4. The goal for each gaze stabilization exercise is to maintain fixation on a target. In the X1 and X2 viewing paradigms, the patient is asked to move the head at a rate that is at the top of their ability to see clearly (Fig. 22.4). In fact, there should be a slight blurring of the target image indicating that there is some retinal slip. As mentioned before, the best stimulus to induce adaptation is one producing an error signal that the central nervous system attempts to reduce by modifying the gain of the remaining vestibular function. In the eye-head movement, the patient must be reminded that the goal is to see clearly as the head moves to the target. Finally, in the remembered target paradigm, the patient's goal is to see the target clearly when he/she open the eyes.
5. Exercises may cause an increase in the intensity of the patient's dizziness and disequilibrium. This is because the exercises involve head movement, which the patient has been avoiding, because moving the head provokes the patient's dizziness. This situation may be threatening to patients who are extremely fearful of experiencing their symptoms. Nevertheless, patients should be told that during physical therapy, there might be a period when they may feel worse before they feel better. To assist the patient through this period, the physical therapist should be accessible. For example, the patient should be instructed to telephone the therapist if the symptoms become severe or long lasting. In such instances, the physical therapist determines if the exercises can be modified or if the exercises should be discontinued until the patient is formally reevaluated. Excessive exacerbation of the patient's symptoms can also be avoided by careful exercise prescription. Initially, the patient may be provided with only a few key exercises. The patient is instructed to attempt the exercises 3 to 5 times per day; the number of repetitions is based on the therapist's assessment of the patient's exercise tolerance. It is a good idea to have the patient perform all exercises completely during the clinic visit in order to assess the response to the exercises. In some ways, the increase in symptoms is good, because there is some evidence that performing these exercises will result in a decrease in symptom intensity over the course of several weeks.[43-45,47,48,51] However, if the increase in intensity is great or if it persists for longer than 10 to 20 minutes after the exercise is finished, the patient may avoid performing the exercise or he/she may be less active for the rest of the day. Patients who become excessively "dizzy" while performing the exercises may even refuse to continue with rehabilitation.
6. For optimal recovery, exercises must stress the system in different ways.[46] For example, adaptation of the vestibular system is frequency dependent.[52,53] If the system is adapted at a specific frequency, gain will improve most at that frequency. Because normal movement occurs over a wide range of frequencies of head movement, the patient should perform the head movement exercises at many different frequencies for optimal effects. Different head positions can also be used to vary the exercise. Patients need to be instructed to gradually increase the speed of their head movements either from exercise session to exercise session or within a session. This creates a challenge to the central nervous system and may improve the effectiveness of the exercise.[53]

7. Response to exercise takes time. The early studies on vestibular adaptation used paradigms in which the stimulus was present for several hours or more.[54] This situation would not be appropriate for patients, especially during the acute stage. We now know that vestibular adaptation can be induced with periods of stimulation as brief as 1 to 2 minutes.[16] During the time in which the brain is trying to reduce the error signal, the patient may experience an increase in symptoms and must be encouraged to continue to perform the exercise without stopping. Each exercise shown in Figure 22.4, for instance, should be performed for 1 minute without stopping. The time for each exercise can then be gradually increased to 2 minutes.
8. Patients need to be instructed to gradually increase the speed of their head movements either from exercise session to exercise session or within a session. This creates a challenge to the central nervous system and may improve the effectiveness of the exercise.
9. Patients should always work at the top limit of their ability. Although the patient's morale can be lifted through activities that he/she can perform relatively easily, most exercises should stress the patient's ability. For example, with the eye-head exercises, the speed of the head movement should be increased as long as the patient can keep the visual target in focus. Or, the eye-head exercises can be modified by adding more targets (e.g., simulating the placement of mirrors in a car) and can also be integrated into the postural stabilization exercises.
10. Static balance exercises can be modified by decreasing the base of support, including going to the sharpened (tandem) Romberg position in stages and even to single leg stance (See Fig. 22.4 and Box 22-2), altering visual and somatosensory cues, adding head movement, or adding upper extremity movements. The X1 and X2 viewing paradigm exercises can be performed with the patient standing with a progressively more challenging base of support and/or with the patient standing on firm or foam surfaces. Eye-head movement exercises can be integrated into walking.
11. It is reasonable to expect improved function within 6 weeks in patients who are compliant about doing their exercises. Habituation of symptoms may take longer. Even in persons with chronic vestibular disorders, exercise improves function and gait.[55]

Figure 22.5 Modified single leg stance exercises: The patient places a hand on a counter for balance. Then the patient places the foot closest to the counter on the cup and tries to lift hand from the counter, at first intermittently and then for longer periods of time. The exercise is repeated, first turning to face the opposite direction so the foot to be placed on the cup is closest to the counter.

Problem-Oriented Approach

In a problem-oriented treatment approach, the physical therapy program is based on (1) the problem areas identified during the evaluation, (2) the patient's diagnosis, and (3) the patient's medical history. For example, the physical therapy program for a patient with Ménière's disease that has progressed to vestibular hypofunction would differ from that provided to a patient with hypofunction from acute vestibular neuronitis. Both patients will experience vertigo, and although vertigo resolves spontaneously (no exercises needed), the patient with Ménière's disease is more likely to have developed anxiety related to the episodes of vertigo or dizziness.[56] This anxiety may interfere with an exercise program directed toward the problems associated with the vestibular hypofunction, which will include head movements that induce dizziness.

A problem-oriented treatment approach is comprehensive and incorporates gaze stabilization, habituation, and postural stabilization exercises, according to the individualized needs of the person. In addition, the exercise program should incorporate functional activities and contemporary principles of motor learning and motor

control. The general treatment progression includes the following:

1. Increasing and alternating the speed of the exercises
2. Performing exercises in various positions and activities (i.e., head movements performed while sitting, then standing, during walking)
3. Performing exercises in situations of decreasing visual and/or somatosensory input (i.e., balance exercises with eyes open and eyes closed, walking with eyes closed)
4. Performing balance exercises while distracted (e.g., mental tasks)[57]
5. Exposing the patient to a variety of task and environmental situations and contexts (i.e., walking in the home to walking at a shopping mall)

Problem: Visual Blurring and Dizziness When Performing Tasks that Require Visual Tracking or Head Stabilization

Visual blurring and dizziness when performing tasks that require visual tracking or gaze stabilization during head movements is most likely a result of decreased VOR gain resulting in visual blurring during head movement and also a visual-vestibular sensory mismatch. Some caution should be taken when gaze stabilization exercises are used during the acute stage following unilateral vestibular loss. In this stage, the patient may complain of severe vertigo and may be nauseated and vomiting. Head movement will make these symptoms worse and the patient usually prefers to lie quietly, often in a darkened room or with eyes closed. At this stage, the patient may also be taking medications to suppress these vegetative responses and may be receiving intravenous fluid replacement. Slow, easy head movements, such as turning the head to look at someone, should be encouraged first. Good visual inputs (bright room lights, curtains open, television on) also should be encouraged during the first days after the acute onset of a vestibular deficit. Getting up out of bed may require assistance and sometimes an assistive device, because many people are unstable early after an acute vestibular event.

After 1 to 3 days, the symptoms of nausea and vertigo should have diminished or resolved, and the spontaneous nystagmus should be decreasing as the resting state of the vestibular neurons recovers. Patients can begin exercises to facilitate gaze stabilization as early as 1 to 3 days after the onset of the vestibular loss using gentle, active head movement. The X1 viewing paradigm exercise with horizontal head movement would be an appropriate starting point. The exercise would be performed while fixating a small (foveal), stationary target for 1 minute followed by a period of rest and would be done 5 times/day (see Fig. 22.4 and Box 22-3).

Box 22-2

EXERCISES TO IMPROVE POSTURAL STABILITY

There are many different balance exercises that can be used. These exercises are devised to incorporate head movement or to foster the use of different sensory cues for balance.

1. The patient stands on a firm surface with his feet shoulder width apart with eyes open, looking straight ahead at a target on the wall. He progressively narrows his base of support from feet apart to feet together to a semi–heel-to-toe position and even to the tandem (sharpened) Romberg position. The exercise is performed first with arms outstretched, then with arms close to the body, and then with arms folded across the chest. Each position is held for 15–30 seconds before the patient does the next most difficult exercise. The patient practices for a total of 2–10 minutes. The exercise can also be performed with eyes closed. For each position, eyes are closed intermittently at first and then for longer periods of time. The tandem Romberg position should not be tried.
2. The patient stands with his feet as close together as possible. He then turns his head to the right and to the left horizontally while looking straight ahead at the wall for 1 minute without stopping (X1 viewing exercise). The exercise can first be performed with both or one hand helping to maintain balance by touching a wall, if needed. The patient takes his hand or hands off the wall for longer and longer periods of time while maintaining balance. Progression involves having the patient move his feet even closer together.
3. A headlamp or a laser pointer can be attached to the patient's waist, shoulders, or head, and the patient can practice shifting his weight to place the light onto targets marked on the wall. This home "biofeedback" exercise can be used with the feet

Box 22-2

EXERCISES TO IMPROVE POSTURAL STABILITY—cont'd

in different positions and with the patient standing on surfaces of different densities.

4. The patient practices standing on a cushioned surface. Progressively more difficult tasks might be hard floor (linoleum, wood), thin carpet, shag carpet, thin pillow, sofa cushion. Graded density foam can also be purchased. The exercise is performed with eyes closed, but the patient should open his eyes when he moves his feet.
5. The patient stands on a firm surface, or a cushioned surface when ready, and pats a balloon back and forth to a partner.

Walking:

1. The patient walks, with someone to assist him if needed, as often as possible (acute disorders).
2. The patient begins to practice turning his head while walking. This will make the patient less stable, so the patient should stay near a wall as he walks.
3. Make walking more difficult by asking the patient to count backwards as he or she walks. This can be made progressively more difficult by having the patient perform the exercise while walking on different surfaces.
4. The patient practices walking with a more narrow base of support. The patient can do this first touching the wall for support or for tactile cues, and then gradually touching only intermittently, and then not at all.
5. The patient practices turning around while he walks, at first making a large circle but gradually making smaller and smaller turns. The patient must be sure to turn in both directions. This can also be tried with eyes closed by some patients.
6. The patient can practice standing and then walking on ramps, either with a firm surface or with a more cushioned surface.
7. The patient can toss and catch a ball over his shoulder while walking.
8. The patient can play kickball with a tennis ball against a wall.
9. Out in the community, the patient can practice walking in a mall before it is open and therefore quiet. They can practice walking in the mall while walking in the same direction as the flow of traffic, or walk against the flow of traffic.
10. The patient can practice maintaining balance while sitting and bouncing on a Swedish ball or while bouncing on a trampoline. This exercise can be incorporated with attempting to maintain visual fixation of a stationary target, thus facilitating adaptation of the otolith-ocular reflexes.
11. Walking and counting can be attempted but may be especially difficult for persons with bilateral vestibular loss.[58]

The next stage would be to add performing the exercise using vertical head movements. As the patient improves, he or she should try to perform the exercises for up to 2 minutes each. Although the patient may complain of increased dizziness or disequilibrium with head movements, neither is a reason to stop the exercises unless, as mentioned earlier, the symptoms persist for 10 to 20 minutes or more each time the patient performs the exercise. Vomiting or significant nausea, however, would be reasons for terminating or modifying the exercises.

VOR gain in the acute stage after unilateral vestibular loss is poor (0.25 to 0.5), and relatively slow head velocities and low frequencies should be used so that the patient can keep the visual target in focus at all times. As the patient improves (within a few days to a week or more), the exercises can be expanded to include use of a full-field stimulus (checkerboard) in addition to the small target he or she had been using. Within 1 or 2 weeks after onset, patients can begin the gaze stabilization exercises that require them to maintain fixation on a visual target that is moving in the opposite direction as their head movement (X2 viewing; see Fig. 22.4 and Box 22-3). This exercise should be performed with smaller head movements (and comparably small target movements), because the target cannot be kept in focus while viewing out of the corner of an eye. The head movements may have to be slower as well for the patient to maintain fixation. Both the X1 and X2 viewing paradigms should be performed at increasing head velocities as the patient improves. Within 2 or 3 days after onset, patients can begin to perform the

gaze stabilization exercises while standing, as a preparation for turning the head while walking. The eye-head exercise and the remembered target exercise are added to the patient's exercise program as he or she progresses.

Ultimately, the eye-head exercise should be incorporated into gait and other functional activities. Patients with vestibular deficits should also be instructed to perform functional tasks or games that require visual tracking or gaze fixation. For example, laser tag requires the patient to move the head while focusing the eyes on a moving target. The Wii, discussed in Chapter 28 has games that incorporate eye-head tracking activities that can be incorporated into the treatment program. Bouncing and catching a ball may be another appropriate task for some patients.The patient could be advised to bounce the ball off the floor, wall, and/or ceiling in an attempt to vary the task by changing direction of object motion and/or neck position. Alternatively, the patient can play kickball against a wall, which would require following the ball with eyes and head *and* rapid shifts in balance. Using multicolored or highly patterned balls may also increase task difficulty, because the moving pattern or high color contrast may greatly increase the patient's symptoms. Mental activities (counting backwards by three or seven) should also be incorporated during standing and gait as the patient improves.

Exposing the patient to highly textured visual environments may also assist in the remediation of visuovestibular interaction deficits. For example, the patient may be instructed to gradually perform tasks such as grocery shopping or walking through a shopping mall. Optokinetic stimulation can be incorporated into the treatment program to simulate complex visual environments.[37]

Problem: Exacerbation of Symptoms

If a patient experiences exacerbation of symptoms during the head movement assessment, habituation exercises can be incorporated into the exercise program. However, there is sufficient evidence that the gaze stabilization exercises also result in a decrease in symptom intensity. During the physical therapy assessment, positions or movements that provoke the patient's symptoms are identified based on the results of the Motion Sensitivity Test or during the history taking (see Chapter 21). These positions and movements are then incorporated into the patient's exercise program. Initially, if the patient has significant symptoms, habituation exercises may be modified so they can be performed in the supine or sitting position. Later, the patient can perform exercises while standing or even while walking. During the habituation exercises, the patient is instructed to hold the position for at least 10 seconds, or until the symptoms dissipate. In addition, the patient is instructed to perform each exercise 3 to 5 times. It is particularly important that the initial prescription of these exercises should avoid those movements/positions that produce severe symptoms. Many therapists have experienced that too aggressive an approach may lead to patient noncompliance. Instead, therapists should select movements and positions that produce a moderate level of symptoms.

Attempts are also made to include habituation exercises into the patient's daily routine. For example, the patient is instructed to incorporate neck diagonals into the task of loading and unloading the dishwasher. This task requires the patient to focus on the object and move the body, head, and arm synchronously to either pick up or place the object on a high shelf.

Problem: Static and Dynamic Postural Instability

The balance and gait exercises prescribed for a patient are based on the problems identified during assessment and will vary considerably depending on patient history and medical diagnosis. Recovery from unilateral vestibular neuronitis, labyrinthitis, or from a surgical procedure, such as vestibular nerve section, typically takes 6 weeks, although recovery may take up to 6 months in some patients. Patients with vestibular nerve section, for example, often return to work within 3 weeks. Recovery from resection of acoustic neuroma typically takes longer, although most of the recovery occurs within the first few weeks. After the first 2 or 3 weeks, the main complaints are fatigue, instability when turning quickly, and some increased difficulty walking in the dark. Patients may also complain of greater instability when walking on uneven surfaces or when there is a change in light intensity (opening a door to the outside or walking through intermittent shadows, such as trees).

Although many patients with peripheral vestibular hypofunction perform below the norm on static tests of balance during the acute stage, the most common complaint is imbalance during dynamic activities such as walking, especially with head turning. Rarely do these patients report an inability to stand on one leg or to walk with eyes closed. Instead, they may complain of difficulty walking on an uphill grade, through a cluttered room, or into a movie theater. Because of the nature of the patient's deficits, balance training should address the dynamic aspects of gait and be task directed. Balance and gait training are inseparable and therefore considered together in this discussion.

Based on the evaluation, the therapist identifies the patient's functional balance deficits. Therapeutic exercise prescription should address the patient's specific deficits. For example, a patient may experience disequilibrium

when the opportunity to use visual and/or somatosensory input for balance is minimized (e.g., walking at night). In this situation, emphasizing exercises and tasks that require the patient to focus on vestibular, instead of visual or somatosensory, input is important. Such exercises can include walking backward, sidestepping, and braiding performed with the eyes closed, marching in place on foam performed first with the eyes open and later with the eyes closed (Fig. 22.6 and Box 22-3), and walking across an exercise mat or mattress in the dark.

Another functional deficit is the instability experienced by patients when faced with situations that require movements of the head while walking. For instance, many patients indicate having difficulty shopping for groceries. To scan the grocery shelves for the desired item, the patient must walk while moving the head left, right, or diagonally. At the same time, the patient must continue to monitor the environment to prevent a collision with another shopper. As a result, this rather ordinary task creates an overwhelming challenge to the patient's postural control system. To overcome this challenge, the patient is first instructed to walk down a corridor while moving the head left and right, or up and down. Later, the patient performs the same task, while avoiding objects placed in the walking path. The most difficult condition would be to perform this exercise in an area with a complex visual environment (similar to a shopping aisle with people and carts).

Figure 22.6 Activities that promote use of vestibular information for balance are frequently included in the patient's home exercise program. In this example, the patient marches in place on a foam cushion, with the eyes open or closed. Closing the eyes maximizes the importance of vestibular information.

The last deficit to be discussed is the difficulty patients with vestibular deficits experience when their gait is unexpectedly disrupted. One seldom walks through a busy shopping mall without experiencing a sudden head-on encounter with another person. Such tasks require the postural control system to respond quickly. In some cases, the patient may need to improve the ability to anticipate forthcoming events. As indicated in the gait evaluation, an obstacle course can be devised to assess the patient's ability to anticipate or respond quickly to changes in task context. The obstacle course can also be used in treatment. The patient should be instructed to vary the course in as many ways as possible. Having a family member verbally direct the patient on the path to follow is helpful with verbal cueing for sudden changes of direction during ambulation. Commands are given at the moment the patient must encounter or avoid an obstacle, thereby maintaining a level of task uncertainty. Another task that requires the patient to respond quickly to externally imposed constraints is walking and pivoting to the left or right. Again, a family member directs the patient on when and in what direction to pivot. Family members can also quickly give the command to "stop."

Patients with vestibular disorders can experience difficulty with many different balance and gait tasks. This discussion considered only a few. With a thorough evaluation, the therapist may identify the patient's specific motor control problem. In such cases, the therapy program should not be limited to specific balance or gait exercises. Instead, the therapist should provide the patient with balance and gait activities that challenge the patient's postural control system in a variety of ways in different environments and on various surface areas to maximally challenge the patient's abilities.

Problem: Progression of Balance and Gait Exercises

Recovery of postural stability occurs more gradually than recovery of gaze stability. Following acute onset of vestibular loss, patients need assistance to get out of bed for 1 to 2 days after the onset, sometimes longer. They usually, although not always, need assistance with ambulation for the first few days. Patients with UVL usually can stand with feet together and their eyes closed within 4 to 5 days after the onset, although they still will have

Box 22-3

EXERCISES TO IMPROVE GAZE STABILITY

Acute stage (also used with chronic, uncompensated patients):

1. A business card, or other target with words on it (foveal target), is taped on the wall in front of the patient so he can read it. The patient moves his head gently back and forth horizontally for 1 minute while keeping the words in focus.

   This is repeated moving the head vertically for 1 minute.

   The patient should repeat each exercise at least three, preferably four or five times a day.

   The duration of each of the exercises is extended gradually from 1 to 2 minutes.

   Patients should be cautioned that the exercises may make them feel dizzy or even nauseated, but that they should try to persist for the full 1 to 2 minutes of the exercise, resting for 1 minute between exercises.

Subacute or chronic stage:

1. Eye-head exercises are added, at first with only two targets and performed only with horizontal head movements. The exercise is performed for up to 2 minutes, but the patient can take breaks at any time. Progression is to perform vertically or to add more targets (e.g., simulating mirror placement in a car).
2. The remembered target exercise can be added to the home exercise program. The exercise is performed for up to 2 minutes but the patient can take breaks at any time.
3. Depending on whether the X1 exercise with a small target induces any nausea or excessive dizziness, the X1 exercise is then repeated using a large pattern such as a checkerboard (full-field stimulus), moving the head horizontally. Progression would be to use the checkerboard but move the head vertically.
4. The patient holds a business card in front of him so that he can read it. He moves the card and his head back and forth horizontally in *opposite* directions, keeping the words in focus for 1 minute without stopping. This is repeated with vertical head movements and with a large, full-field stimulus. The duration is gradually extended from 1 to 2 minutes. The patient should repeat each exercise at least three, preferably five times each day.

Chronic stage:

1. The patient fixates on a visual target placed on the wall in front of him or her while gently bouncing up and down on an exercise ball or standing on a compliant surface.

increased sway. Gait will be grossly ataxic for the first week, but patients should be walking independently, albeit with a widened base of support, within 1 week. During this initial stage of recovery, several different balance and gait exercises are appropriate. Goals include increasing the patient's endurance while walking, improving stability while standing with a more narrow base of support (Romberg position) with eyes open and closed, and beginning to turn the head while walking. Exercises to improve balance in sitting or other positions are usually not necessary, but for postoperative patients, bending over must be avoided for several weeks.

Gait exercises can be more challenging for patients with chronic vestibular disorders, although, again, the starting point depends on the problems identified during the assessment (see Box 22-2). Patients can be taken through a series of exercises that stress their balance by gradually decreasing their base of support or by altering visual and somatosensory cues. Even if they are unable to maintain the position successfully for the required period of time, practicing will improve their balance. Patients with complete unilateral vestibular loss, however, rarely perform the sharpened Romberg with eyes closed at any age. More difficult dynamic balance exercises may include walking and turning suddenly or walking in a circle while gradually decreasing the circumference of the circle, first in one direction and then in the other. The patient needs the practice of walking in different environments, such as on grass, in malls (walking in an empty mall is easier than in a crowded mall, walking with the crowd is easier than against the crowd), and walking at night. Precautions to prevent falls should always be taken until the patient no

longer needs them, but it is important to avoid letting the patient become dependent on assistive devices for walking. Assistive devices may only be necessary the first few days after the vestibular event but are rarely used later in the rehabilitation process.

Problem: Physical Deconditioning

Physical deconditioning because of inactivity may be a significant problem for many patients with UVL. Many patients are advised to begin a regular walking program. The purpose of the walking program is twofold: first, to prevent deconditioning of the patient; second, to provide realistic balance challenges to the patient's central nervous system. Tasks such as walking on uneven terrain, walking through a shopping mall, or crossing the street challenge the patient in ways that cannot be simulated by a therapeutic exercise program (Fig. 22.7). When crossing the street, the patient must conform to the temporal constraints imposed by moving vehicles. Specifically, the patient must determine at what moment to step off the curb to avoid confronting a vehicle. When crossing the street at a busy intersection, this requirement becomes more difficult to fulfill. In addition, the patient's postural control system must make quick adjustments to offset perturbations caused by changes in terrain or motion of other people.

The initial program requires the patient to walk for 15 to 20 minutes four or more times per week. Over the subsequent weeks, the patient is instructed to increase to a 30-minute walk. Initially, the patient may be advised to walk in a familiar environment with few challenges. Later, the therapist encourages the patient to expand the walking program to other situations and contexts. Walking in a park and at a shopping mall are frequently recommended as the patient improves. When walking in these situations, patients are advised to experience as many challenges as possible. For example, riding an escalator in a shopping mall may provide an interesting challenge to the patient. The patient must remain balanced while standing on a moving support surface. In addition, the motion of other people toward or to the side of the patient may create a sense of dizziness or imbalance, enhancing the difficulty of the task. Such challenges are necessary to overcome if the patient is to manage safely in a variety of contexts without experiencing an exacerbation of symptoms.

Patients can also be encouraged to return to other activities, such as tennis or golf, which will help improve their overall fitness. These activities need to be added gradually. Activities such as using a stationary bike, although it should help improve fitness, will not help improve postural stability except indirectly by increasing strength or range of motion. Swimming can be safely performed by patients with UVL and is a good fitness exercise. If the patients want to return to ocean swimming, they should be advised that they might have some difficulty initially walking on the sand or standing on the sand in the water. (Patients with bilateral vestibular loss must be more cautious because, without visual cues, they will have difficulty knowing "which way is up" when underwater. Ocean swimming in particular may not be advisable. Swimming with goggles will help, but not in murky water.) For all patients, the precautions we all should follow need to be observed: never swim alone.

Figure 22.7 Patients may have difficulty with walking when they must conform to temporal constraints such as when crossing a street before the light changes.

Problem: Return to Driving

One of the more commonly asked questions is when the patient may begin to drive again. The legal ramifications may vary from state to state, but essentially patients should not drive if they cannot see clearly during head movements or if head movements result in significant dizziness (or disorientation). One guideline, that patients should be able to see clearly when making rapid and abrupt head movements, can be tested using the dynamic visual acuity test. If a patient decides to return to driving, he or she should be encouraged to begin on quiet, local streets or in a parking lot when it is empty. Driving at night and driving on high-speed roads should be delayed until the patient is comfortable with the quieter roads. Drivers' training may be beneficial but may or may not be covered through insurance. It is somewhat controversial as to whether persons with bilateral vestibular hypofunction should drive. A recent report suggests that persons with bilateral loss are safe to drive, yet patients have reported reductions in their perceived ability to drive.[59,60] See Chapter 7 for additional information related to driving and vestibular disorders.

Information the Physical Therapist Should Send the Physician

The physical therapy evaluation should be aimed at identifying the specific problems that patients with vestibular hypofunction often have (Table 22-3). Once the problems have been identified, the actual treatment plan is developed with an emphasis on the home exercise program. Reevaluation should occur mid-rehabilitation process and again at the end of the rehabilitation process. The referring physician therefore should be provided with objective assessment results that show whether or not progress has occurred. With the electronic medical record, sending reports to the physician hopefully becomes easier. Continued communication, especially if the patient is being managed with a pharmacological intervention, is very helpful to the referring physician. Adjustments in the medication can be

■ Table 22-3 **SPECIFIC PROBLEMS AND APPROPRIATE MEASUREMENT TOOL (SEE CHAPTER 21 FOR DETAILS)**

Potential Problem	Measurement Tool
Subjective Complaints	
Imbalance – especially walking	Visual analogue scale – while walking
Oscillopsia with head movement	Visual analogue scale – while walking
Dizziness with head movement	Visual analogue scale – 1 minute of head movement
Low confidence in balance	Activities Specific Balance Confidence Scale (ABC), Falls Efficacy Scale,[61] or the Falls Efficacy Scale International[62]
Anxiety and depression	Many different tools can be used
Limitations	
Decreased walking speed	Timed walking at preferred speed compared with age and gender norms
Increased risk for falling	Dynamic Gait Index Functional Gait Assessment
Poor vision during head movements	Dynamic Visual Acuity
Poor physical condition	6-minute walk test; Heart Rate, Respiratory Rate
Performance of basic ADLs and driving	Questionnaires
Restricted of participation in normal societal interactions	Disability Scale Dizziness Handicap Inventory Physical Activities Scale[63]

more easily made if symptom reports are objecti vely reported to the physician.

The goals of physical therap y intervention are to (1) decrease the patient's disequilibrium (sense of being off-balance) and oscillopsia (visual blurring during head movement), (2) improve the patient's functional balance especially during ambulation, (3) improve the patient's ability to see clearly during head movement, (4) improve the patient's overall general physical condition and activity level, (5) enable the patient to return to a more normal level of activity and participation in society, and (6) reduce the patient's social isolation. These goals should be directly linked to the exercises and the outcome measures used by the therapist.

Patients are usually seen by the physical therapist on an outpatient basis, although, in some cases, the initial treatments will occur while the patient is in the hospital. An important part of the rehabilitation process is the establishment of a home e xercise program. The physical therapist must motivate the patient and obtain compliance. To do so, the physical therapist must identify the patient's own goals and clarify to the patient the treatment goals as well as the potential ef fects of exercise.

The initial note should contain the following:

1. Quantification of the patient's subjective complaints and of the results of the examination. At a minimum, there should be one measure of subjective complaints, a measure of gait speed and of fall risk, and a measure of the patient's ability or inability to participate in societal activities. Normal values for age (and gender where appropriate) should be included for comparison with the patient's performance,
2. A list of problems specific for that patient, the goals for the patient, and a timeframe by which those goals should be met.
3. The specific exercises the patient will be performing related to the patient's goals.
4. An indication that the patient is expected to perform a home exercise program (HEP).
5. The number of clinical visits/week and the number of weeks that physical therapy will be required.
6. The expected ability of the patient to improve should also be included in the note.

A request for continuation of the therapy should:

1. Show evidence that the patient has shown improvement toward reaching those goals or gives an explanation as to why the patient has not improved. Again, quantified data should be presented.
2. The number of clinical visits/week and the number of weeks that physical therapy will be required.

Final note should:

1. Itemize what goals the patient has achieved.
2. Provide the data that demonstrates the change (baseline and discharge measures).
3. Explain why the patient is being discharged from physical therapy if the goals have not been achieved.

Evidence that Exercise Facilitates Recovery

Animal studies provided us with the first evidence that visuomotor experience facilitated the rate of recovery and improved the f inal level of reco very following vestibular dysfunction.[13-15] Furthermore, several studies suggested that exercise facilitated the process of vestibular compensation. For example, Igarashi et al[64] found locomotor equilibrium compensation occurred faster (7.3 days as compared with 13.7 days) in a group of squirrel monkeys exercising in a rotating cage compared with a none xercise group. Similar findings have been observed in cats follo wing unilateral labyrinthectomy.[12]

Fortunately, there is now considerable evidence in human patients that the use of specific exercises facilitates the level of recovery in patients with unilateral vestibular hypofunction.

> Availability of supporting evidence is important because supervised physical and occupational therapy adds to the health care costs of the patient. Current health care providers (insurance companies) are be ginning to require that treatments are supported by evidence and that continuation of patient visits is supported by documentation of progress through the use of v alid assessment tools. It is necessary, therefore, to demonstrate the effectiveness of these e xercise approaches in the rehabilitation of patients with v estibular hypofunction.

A Cochrane Review in 2011 on vestibular rehabilitation identified six additional studies that compared vestibular rehabilitation therapy for patients with unilateral peripheral vestibular hypofunction with sham treatments, no treatment, and other forms of v estibular rehabilitation therapy to the 21 identified in the 2007 review.[65,66] Exercises used in these articles include

habituation, adaptation, and substitution. The patients in all studies were either community dwelling or patients hospitalized for surgery who had been community dwelling before admission to the hospital. The authors concluded that:

1. moderate to strong evidence exists that vestibular rehabilitation is a safe and effective treatment for patients with unilateral vestibular hypofunction or loss.
2. moderate evidence exists that vestibular rehabilitation is an effective treatment of patients during the acute period onset of vestibular neuritis or after resection of vestibular schwannoma.
3. improvements have been reported in dizziness, gait and balance, vision during head movements, activities of daily living, and quality of life.
4. and moderate evidence exists that these improvements are sustained for months after the rehabilitation process ends.

Several key studies are summarized below under headings that represent the evidence provided. As with many investigations, these studies are limited in certain ways:

1. In many of these studies the exercise approaches were not described in sufficient detail to allow replication of the exercises used.
2. Although the study designs included analysis of group data, in most studies no information was provided on the individual level (e.g., the number of participants within a group who failed to improve).
3. With few exceptions, these randomized controlled trials did not examine the effect of specific factors, such as exercise approach, gender, time from onset, comorbidities, or environment on recovery.

I. Specific vestibular exercises result in a decrease in symptoms, improvement in postural stability, and improvement in gaze stability in patients with chronic unilateral vestibular hypofunction:[67]

This *prospective, randomized, controlled study* compared the effectiveness of vestibular exercises (customized exercise programs consisting of gaze stability, habituation, and balance exercises) to general conditioning exercises and to the use of vestibular suppressant medications in patients with chronic vestibular dysfunction. Patients were randomly assigned to one of the groups and were followed for 6 weeks. The investigators found a *significant decrease in dizziness only in the group of patients receiving vestibular rehabilitation*. Similarly, *only the vestibular exercise group had a significant improvement in postural stability* as indicated by decreased anterior-posterior sway and increased single leg stance time. The group receiving vestibular suppressant medications did not show improvement.[17]

This randomized study compared the effectiveness of unsupervised exercises and vestibular rehabilitation supervised by physical therapists. The patients in the unsupervised exercise group were given a handout and were instructed to "go home and do them." This group of patients was compared with a group of patients who performed a customized, supervised program of vestibular adaptation exercises. The authors found that a greater percentage of patients improved in the *vestibular adaptation exercise* group than in the unsupervised exercise group. Similarly, Topuz et al compared a supervised program of habituation exercises to home exercises only and reported a significant improvement in subjective complaints and self-perceived handicap in the supervised exercise group only.[68]

These studies provide some support for why it is not sufficient to simply provide patients with a list of what they could do to foster return to normal activities. For the most part, patients with vestibular hypofunction who end up in a neurologist's office are those patients who have not recovered on their own. Typically, they have avoided activities that involve head movement, because it increases their symptoms, they may be fearful of provoking another episode of vertigo, or they may be sedentary by habit or nature. *Giving these patients a handout of exercises to perform will not overcome these barriers to recovery.* Pavlou et al recently reported that their drop-out rate was 55% in a group of people provided a home DVD for exercise versus a 10% drop-out rate for a group of people with visual

vertigo symptoms who received a supervised exercise program.[69] Offering a patient a *customized home exercise program* appears to be more likely to result in reduced symptoms and improved independence in daily activities, however.[45,70]

In a study of patients with chronic vestibular deficits, Shepard and Telian compared the efficacy of customized vestibular exercise programs to a more generic exercise program. They used a delayed treatment paradigm. Subjects first were assessed to establish a baseline and reassessed at 1 month *before initiating any exercises*. This served as a control for spontaneous recovery. Subjects who had not shown spontaneous recovery were then stratified by age and by pretreatment disability levels to ensure that the two groups were similar. After 3 months of therapy, *only the vestibular rehabilitation group showed a significant reduction in dizziness during routine daily activities*. The *vestibular rehabilitation group also showed a significant improvement on both static and dynamic posturography, a reduction in motion sensitivity, and a decrease in asymmetry of vestibular function*. The generic exercise group improved only in their performance of static balance tests.[46]

The objective of this prospective, double-blinded, randomized, placebo-controlled study was to determine the effect of vestibular exercises on the recovery of visual acuity during head movement in patients with unilateral vestibular hypofunction. This question is of particular interest because patients with vestibular loss complain of visual blurring during head movement and report having difficulty driving, because they cannot see clearly while in a moving car. This limitation is not only a safety issue but also affects the ability of the patient to return to work. One group (13 patients) performed vestibular exercises designed to enhance the vestibulo-ocular reflex (VOR) and the other (8 patients) performed placebo exercises. Measurements of visual acuity during predictable and unpredictable head movements were made weekly using a computerized test. As a group, patients who performed vestibular exercises showed a significant improvement in visual acuity during predictable and unpredictable head movements, but those performing placebo exercises did not. Based on stepwise regression analysis, the leading factor contributing to improvement was vestibular exercises. Other factors examined included age, time from onset, initial DVA, oscillopsia, and duration of treatment. The investigators concluded that the use of vestibular adaptation exercises is the main factor involved in recovery of visual acuity during predictable and unpredictable head movements in patients with unilateral vestibular hypofunction. There is similar evidence of exercise-induced improvement in DVA in patients with bilateral vestibular deficits.[71]

II. Specific vestibular exercises result in a decrease in symptoms and in improvement in postural stability in patients with acute unilateral vestibular hypofunction:[48]

This prospective, double-blinded, randomized, placebo-controlled study examined whether patients with *acute unilateral vestibular loss* following resection of acoustic neuroma would benefit from vestibular rehabilitation. This study compared the effect of vestibular adaptation exercises with exercises designed to be "vestibular neutral" (head still). Both groups were instructed in safe ambulation every day. Exercises were initiated on postoperative day 3 in both groups. They found no difference in subjective complaints of vertigo between the groups over the course of the study. This was expected, because vertigo occurs as a result of the asymmetry in the tonic firing rate of the vestibular system and recovers spontaneously. There was, however, a significant difference in the complaints of disequilibrium by postoperative days 5 and 6. As a group, *patients performing the vestibular adaptation exercises had significantly less disequilibrium than did patients in the control group*. Differences between the groups were also noted for gait pattern, especially with horizontal head movement. *All the control subjects had increased ataxia or developed some ataxia when asked to turn their head while walking. In contrast, only 50% of the*

vestibular exercise group showed this gait disturbance. This resulted in the patients in the vestibular exercise group being safer while walking at the time they were discharged home than were the patients in the control group.[72]

This prospective, randomized study examined the effectiveness of gaze stabilization exercises performed beginning 3 days after surgery on reducing the patients' perception of dizziness/imbalance. They examined a cohort of 65 patients (30 exercise patients, 27 control patients, and -8 balance exercises only). The main finding was that there was less dizziness in those patients who performed vestibular exercises, based on the scores of the Dizziness Handicap Inventory.[49]

This *randomized, controlled* trial found that patients over the age of 50 years who performed a customized exercise program performed significantly better on standing balance, Timed Up and Go test, and Tandem Gait when compared with patients over 50 years of age who received only instructions to go about their normal activities. This effect persisted up to 12 weeks and also became apparent on the Dynamic Gait Index.

III. Exercises versus no exercises:[73]

Yardley et al reported on a comparison of customized home exercise programs based on habituation exercises with a control group who performed no exercises. In this *prospective randomized* study, patients with dizziness from a variety of causes (including non-vestibular) were assigned to one of the two groups. Outcome measures included scores of symptom severity, anxiety, provocative movements, and the sharpened Romberg test. The results of that prospective study showed that there was *significantly greater improvement in the patients who were on the vestibular habituation exercise program than on no exercises*. Vestibular exercises, even when unsupervised, thus appear to be beneficial.[74]

The purpose of this *prospective, randomized controlled* study was to assess the role of a computerized posturography-assisted training protocol combined with a home-based exercise program to spontaneous recovery in patients with subacute (2 weeks post onset) patients with UVH. Ten healthy volunteers were also studied. After 6 weeks of therapy, patients performing the combined computerized posturography and Cawthorne-Cooksey exercises improved significantly in most measures (modified clinical test of sensory organization and balance [mCTSIB] and limits of stability [LOS] tests), and there were no significant differences between these patients and a group of healthy volunteers. In contrast, the untreated group differed from healthy controls on several LOS parameters.

These studies are important, because they provide the evidence that vestibular exercises result in improved function when compared with no treatment control group.

IV. Are different treatment approaches equally effective for vestibular hypofunction?[75]

Only one study has compared the different vestibular exercises (habituation and gaze stabilization exercises) in the treatment of patients with UVH. The results should be considered as preliminary, because the study had a small number of subjects (n = 7). The study used disability, motion sensitivity, and visual acuity during head movement (computerized dynamic visual acuity, DVA) as the outcome measures but not gait and balance measures. Study patients were treated with either habituation exercises or gaze stabilization exercises (adaptation and substitution exercises), and both groups improved in all outcome measures. These finding were unanticipated, because although habituation exercises are designed to reduce symptoms, they theoretically should not result in an improved DVA. Similarly, gaze stabilization exercises are expected to improve DVA but not necessarily reduce symptoms, although this has been shown in at least one previous study.[46] The author suggests that the important factor for successful outcome for patients with UVH may be the inclusion of head movements in the exercises.

An important part of treatment is the education of the patient about the possible final level of recovery. This enables the patient (and the therapist) to set realistic goals.

It is possible that treatment efficacy is not only dependent on matching the exercise appropriate to the patient's problems but also is dependent on the characteristics of the individual patient.

V. *Are there factors that can be identified that are associated with who will and who will not show improvement?*[47]

Several studies on treatment of patients with vestibular hypofunction clearly state the reality that some patients do *not* improve.[75,76] For patients with UVH, between 12% and 25% of subjects do not improve, depending on what outcome measure is used (Table 22-4).[47] Of patients with BVH, overall outcomes are worse, with between 18% and 72% of patients failing to show improvement (see Chapter 23).[77] Identifying and assessing these non-responding patients is extremely important because, as clinicians, we are obligated to identify these patients, seek the reasons for poor outcome, and develop methods to reverse poor outcome.

Herdman et al (2012) examined data from 209 patients with UVH, all of whom had been treated with similar courses of vestibular adaptation, substitution, and balance and gait exercises.[47] As with several other studies, the results showed that although most patients improve with a course of vestibular exercises, some patients do not improve or improve in only certain areas (Table 22-5). This study examined the relationships between patient characteristics, pretreatment measures of subjective complaints and physical function, and treatment outcome. The primary findings of the study were:

- *Age and gender* did not affect which patients would improve.[47] However, the study showed that older patients walked more slowly and had poorer visual acuity during head movement (DVA) than did younger patients at discharge. This was expected, because there is a naturally occurring decrement in gait speed and in DVA with age. Older patients were also more likely to remain at risk for falls, based on the DGI score, than were younger patients. This simply may be a reflection of the slower gait speed as that is a component of scoring the DGI test, or it may indicate a lesser degree of recovery in older patients. Results from several other studies have identified the fact that *age is not a factor in achieving improvement* in subjective complaints, visual acuity during head movement, gait speed, and fall risk in patients following a course of vestibular exercises.[46,51,79-81] Less is known about the role of gender in recovery. Within

■ Table 22-4 **PRE-REHABILITATION EXPECTED TEST RESULTS IN PATIENTS WITH UNILATERAL VESTIBULAR LOSS**

Test	Acute UVL	Compensated UVL
Nystagmus (NOT affected by exercises)	Spontaneous and gaze-evoked in light and dark; head shaking induced increases nystagmus; with visual fixation, the spontaneous nystagmus decreases	Spontaneous in dark; may have head-shaking–induced nystagmus
Head Impulse Test of the VOR	Positive with a corrective saccade	May have covert saccades toward the side of the lesion; overt corrective saccade more likely when >75% asymmetry
Imbalance – especially while walking	Present with all walking and early on, during standing: slow, cautious gait	Essentially only with rapid turns toward affected side and sometimes in the pitch plane

Continued

■ Table 22-4 PRE-REHABILITATION EXPECTED TEST RESULTS IN PATIENTS WITH UNILATERAL VESTIBULAR LOSS—cont'd

Test	Acute UVL	Compensated UVL
Oscillopsia with head movement	May or may not be significant complaint	Little or no complaints, may be affected by depression[47]
Dizziness with head movement	Major complaint	Little or no complaints; affected by depression[47]
Confidence in Balance	ABC score <80%	ABC score >80%
Anxiety		Should decrease w/ return to activities
Depression		If situational, should decrease
Walking speed	~50% slow for age	Only 14.6% abnormally slow for age[47,78]
Risk for falling	DGI score ≤19 indicating risk for falling	67% no longer at risk; 22% improved but still at risk; 11% no improvement;[47]
Vision during head movements	Abnormal for age	78% improve significantly with majority to within normal for age[47]
Participation in normal societal interactions	Disability scores range from bothersome[1] – out of work or on long-term disability[5]	Disability scores shift to 0–2 except for those out of work for >1 year or are on long-term disability[51]
Performance of basic ADLs	Most can perform independently	Can perform independently
Driving	Unsafe	Safe although some may modify or limit driving. It remains somewhat controversial related to persons with bilateral vestibular disorders.
Romberg	Often, but not always positive	Negative
Sharpened Romberg	Cannot perform	Normal with eyes open; cannot perform with eyes closed
Single leg stance	Cannot perform	Normal, eyes open
CSTIB—foam, eyes closed	Most cannot maintain balance on foam, eyes open	Normal with eyes open; may be normal with eyes closed
Gait	Wide-based, slow cadence, decreased rotation, may need help for a few days	Normal except for very rapid turns toward affected side or in pitch plane
Turn head while walking	Cannot keep balance	Normal; some may slow cadence and veer

■ Table 22-5 MEAN (SD) IN OUTCOME MEASURES FOR PATIENTS WITH CHRONIC UNILATERAL VESTIBULAR HYPOFUNCTION BEFORE (BASELINE) AND AFTER A COURSE OF GAZE AND POSTURAL STABILIZATION EXERCISES (DISCHARGE). ADAPTED FROM HERDMAN ET AL, 2012.[47] MEAN AGE OF SUBJECTS WAS 59.3 + 14.9 YEARS.

Outcome Measures	oVAS	dVAS	ABC	% of Time Symptoms Interfere	Disability	DVA (Ipsilesional)	Gait Speed (meter/sec)	DGI
# of subjects tested	182	180	108	79	140	153	132	183
# of subjects abnormal at baseline	88	149	82	76	136	70	89	127
# of pre-post comparisons	65	113	69	20	68	67	59	119
Baseline	3.05 (2.66)	3.97 (2.54)	51.5 (16.9)	59 (29.8)	2.9 (0.95)	0.327 (0.134)	0.80 (0.15)	14.2 (4.0)
Discharge	0.79 (1.65)	1.58 (1.91)	78.5 (16.0)	24 (30.1)	1.39 (1.4)	0.210 (0.138)	1.01 (0.17)	19.8 (3.0)
Pre-post paired comparison: *p* value	<0.0001	<0.0001	<0.0001	<0.0001	<0.0001	<0.0001	<0.0001	<0.0001
95% Confidence Interval	1.58 - 2.94	0.11 - 0.15	31.7 - -22.3	*	1.1 - 1.8	0.095 - 0.139	-0.80 - -0.54	-6.15 - -4.71
% of subjects with significant improvement	78%	81%	78%	80%	75%	78%	85%	88%

* Confidence Interval not calculated because of small number of comparisons.
oVAS = subjective oscillopsia; dVAS = subjective disequilibrium; ABC = Activities specific Balance Confidence; DVA = Dynamic Visual Acuity; DGI = Dynamic Gait Index

patients who all had surgical removal of acoustic neuroma, gender was a significant factor in the patients' perception of handicap postoperatively, with male patients reporting a greater perception of handicap at 3 months postoperatively, but there was no gender effect at 1 year after surgery.[69] In patients with a variety of diagnoses, male patients reported worse disability scores than female.[44]

- *Time from onset*: Time from onset was not related to any outcome, supporting the use of these exercises in patients regardless of the chronicity of their vestibular dysfunction. Patients in this study had a mean time from onset of 14.5 months (range 1 week to 12 years; median 5.0 months). Findings from studies on patients with acute UVL are different and suggest that delaying the initiation of vestibular exercises results in a delay in the initiation of recovery/slows compensation.[1,72] In one relatively small study (n = 23), patients with acute loss of vestibular function following resection of acoustic neuroma started a vestibular rehabilitation program at 3 days postoperatively and had better postural stability and less disequilibrium than a control group who performed placebo exercises by 6 days postoperatively.[48] Another randomized controlled trial with a larger cohort of subjects (n = 65) confirmed these findings.[72] Even this result is somewhat controversial, because a third study found no difference in outcome between patients performing vestibular exercises and a placebo group during the acute stage post removal of acoustic neuroma.[82] There were differences, however, in the outcome measures and the exercises used, which may explain the different conclusions, but clearly more research is needed on the effect of time from onset on recovery.

A different issue is whether there is a critical period during which exercises *must* be initiated. The evidence seems to suggest that there is no critical period—that vestibular exercises are beneficial even for patients with chronic problems.[17,56] This has been shown directly through analysis of time from onset as a factor that might be involved in recovery.[46,51,77]

- *The vestibular deficit itself*: The only relationship this study identified between degree of deficit, as defined as percent asymmetry, was that patients with smaller asymmetries in vestibular function were more likely to return to normal DVA for age.[47] Degree of deficit was not related to improvement in other outcome measures. The extent to which the rate of development of the problem (e.g., sudden unilateral, slowly progressive loss as in vestibular schwannoma) affect the final level of recovery remains unclear. There is some evidence that points to the importance of the underlying etiology in outcome. For instance, within subjects who all had surgical removal of acoustic neuroma, tumor size and degree of preoperative canal paresis were significant factors in the patients' perception of handicap postoperatively.[80] In contrast, patients with surgically induced vestibular loss have a similar level of recovery as do patients with unilateral vestibular neuritis and labyrinthitis.[49]
- The *presence and number of different comorbidities* did not affect whether or not improvement occurred. This was in contrast to a study that showed that patients with migraine often improve physically but continue to perceive that they are disabled by their vestibular complaints.[83] In the Herdman et al study, the only comorbidity that affected outcome was the presence of anxiety and/or depression. Patients with anxiety and/or depression were more likely to have lower balance confidence and a higher percentage of time for which symptoms interfered with activities at discharge. As with many if not all chronic illnesses, the presence of depression and/or anxiety can impact quality of life. In some populations, depression is the variable most likely to influence quality of life.[84] However, Grunfeld et al noted that the presence of a vestibular lesion per se is not associated with depression.[85] Somewhat in contrast, Yardley et al found that negative mood, rather than true depression, was a factor contributing to self-perceived handicap in patients with dizziness.[86] Although there may be no direct association between vestibular deficits and depression, the presence of depression can have a serious impact on the potential for recovery. Krebs et al noted that some patients with vestibular hypofunction developed depression that prevented participation in vestibular rehabilitation.[77] This observation is supported by others who noted that treatments that appear to decrease quality of life have low adherence by patients,[87] while better outcomes and improved

quality of life are associated with increased treatment adherence.[88] Factors that have been identified as contributing to the self-perception of handicap or disability in patients with dizziness include somatization, negative mood, satisfaction with social support, self-esteem, and anxiety.[89,90] Psychiatric/psychological factors also help predict outcome. Symptoms of somatic anxiety predict an increase in handicap over a 7-month study of people with recurrent dizziness.[90] Another factor, maladaptive coping strategies, was not examined in the Herdman et al (2012) study but has been documented by others as being associated with decreased quality of life and with greater depression in patients with chronic medical illness.[91,92]

One study of patients with episodic vertigo identified four primary strategies for coping with vertigo: (1) problem-focused information seeking, (2) distraction, (3) denial, and (4) relinquishing responsibility.[91] The investigators found that "relinquishing responsibility" predicted handicap, even when the analysis controlled for other factors such as emotional distress. Because of the non-episodic nature of the symptoms in the patients with vestibular hypofunction, the relationship of coping strategies and psychological factors to outcome may be different than what has been already reported.

Several patterns were identified in which multiple factors accounted for a significant percentage of the recovery of some of the outcome measures.[47]

1. Patients who indicated that symptoms interfered with performance of their usual activities at a high level (greater percentage of the day) at discharge were those who had anxiety and/or depression and who indicated a high percentage of time symptoms interfered with activities (5TSI) at the time of the baseline assessment. These two factors together accounted for 83.7% of the outcome. Similarly, patients who rated their disability as high at discharge had higher baseline %TSI, worse baseline disability scores, and poor baseline DVA score. These three factors accounted for 47.8% of the disability score at discharge.
2. Patients who walked more slowly at discharge had slower baseline gait speed and were older. Approximately 55% of gait speed at discharge was accounted for by advanced age and slow walking at onset of symptoms.
3. Patients had lower (poorer) fall risk scores at discharge, as measured by the DGI test, if they had a history of falls, had poor baseline fall risk scores initially, and were older. These three factors accounted for 42.5% of DGI at discharge. Interestingly, initial DVA score, which has previously been shown to predict fall risk at discharge, was not a significant factor.[93]

These results provide some guidelines that therapists and other clinicians can use to develop expectations for recovery.

Several other factors must be taken into account when considering the potential for recovery of a patient.

- Medication: Several authors have suggested that vestibular-suppressant medications such as diazepam may delay or slow recovery,[44,94] but few studies actually have examined the influence of medication on the level of recovery. One randomized controlled study found that patients taking meclizine did not show an improvement in symptoms or in balance, whereas patients performing vestibular exercises did improve.[67] Patients treated with steroids for vestibular neuritis, on the other hand, appear to have less perception of handicap after treatment than those patients who were not treated with steroids.[95] The use of medication appears to be influenced by performing exercises. In a separate randomized trial, Venosa and Bittar suggested that persons who performed exercises acutely (symptoms that began less than 5 days before the start of the study) were significantly less likely to use medication at 3 weeks than those who were not provided vestibular exercises.[96] Those in the vestibular exercise group also had a greater number of normal Fukuda step tests and post head shake testing results recorded by a blinded tester. At 3 weeks post exercise in acute persons with vestibular disorders compared with a control group, they had similar symptom reports. However, only 13% continued to take medication in the exercise group, while 82% of subjects in the control group were taking medication daily.
- *Recovery may be delayed* or limited if the patient restricts head movement or if visual inputs are minimized.[10,14,15] Patients with vestibular lesions often prefer to keep their eyes closed and their heads still to minimize symptoms. Another factor that may delay recovery or limit the final level of recovery may be the use of medications that suppress vestibular function.[94]

Is recovery sustained after rehabilitation is stopped? Both anecdotal evidence and systematic studies suggest that recovery following vestibular loss may be "fragile." Symptomatic relapse may occur with extreme fatigue or stress, prolonged periods of inactivity, illness, or even a change in medication.[45] Patients need to be aware of this possibility and should understand that it does not indicate a worsening of the underlying pathology. Little is known as to whether patients who responded to vestibular rehabilitation with improved function need to continue with exercises to sustain that improvement. Krebs et al examined patients with UVH and BVH at 1 year after a course of vestibular rehabilitation.[77] They found that frequency of performing a home exercise program over the course of that year correlated with sustaining improvements in gait speed.[77]

More research is needed to determine other factors that may explain treatment outcome such as personality traits Many factors can affect the final level of recovery and should be kept in mind when talking to patients about their progress and anticipated recovery.

VI. Exploration of other treatment options

Several studies have examined whether training balance on force platforms has an added benefit to the more traditional vestibular exercise in patients with chronic unilateral vestibular hypofunction. Nardone et al (2010) compared postural control and subjective scores of stability in patients with chronic vestibular deficits performing Cawthorne-Cooksey habituation exercises with patients performing balance training on an oscillating force platform.[97] Both forms of exercise resulted in equally improved balance as measured on the posturography platform and improved subjective disequilibrium, but the maximal improvement occurred after patients had received both exercises.

Winkler and Esses (2011) noted that the use of exercises incorporating platform perturbation with changes in foot position and in visual cues plus balance exercises resulted in greater improvement than balance exercises alone in patients with chronic UVH.[98] Another study compared the effect of computerized dynamic posturography and treatment using optokinetic stimulation in patients with chronic UVH.[99] Both groups improved in the overall posturography score, but there were differences noted in the performance based on which sensory cues were altered. Finally, a study by Marioni et al (2012) showed that gaze stability and weight shifting exercises with biofeedback resulted in improved stability compared with no treatment.[74]

CASE STUDY 22-1

A 63-year-old woman developed vertigo with nausea and vomiting 4 months ago. The vertigo lasted for several days during which time she became dehydrated. She was hospitalized for 4 days during which time she was treated for dehydration and worked up for cardiac problems and stroke. Discharged from the hospital with a diagnosis of "vertigo," she was seen by a neuro-otologist for workup of her vertigo and diagnosed as having unilateral vestibular neuritis. A caloric test performed at that time showed right canal paresis (63% decrease on right). She had a course of vestibular rehabilitation, which she states was not beneficial. Presently, she complains of lightheadedness and dizziness with head movement. She is using a cane for ambulation, which she did not need before the onset of the vertigo. Review of records from 4 months ago show that at that time the patient was ambulating with a walker, and that her clinical visual acuity was 20/25 with her head stationary and increased to 20/100 during 2-Hz horizontal head oscillations (clinical DVA test). Additionally, she had restricted her participation in her normal social activities, no longer going to church functions or out to dinner with friends.

Past Medical and Social History

Significant weight gain in last 4 months. No history of trauma, exposure to ototoxic medications, no heart disease, diabetes, thyroid disorder, hypertension, arthritis, or treatment by psychiatrist. She lives alone in a one-story house with 4 steps to enter with a railing. She has no family living nearby.

Comment

This patient developed a unilateral vestibular hypofunction 4 months ago (based on the caloric). Vestibular neuritis typically affects the superior vestibular nerve

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resulting in horizontal canal hypofunction and utricular degeneration with sparing of the posterior canal. It is possible that the patient may develop BPPV as a result, and she should be screened for BPPV. It is not clear why the patient believes the previous course of vestibular rehabilitation was not beneficial. If the exercise program were too aggressive, it may have provoked excessive dizziness. Therefore, the patient may have not been compliant with the exercises. It is clear that she had made some improvement since the onset of the problem—she is walking now with a cane instead of a walker.

Subjective Complaints

She rates her disability as a 3/5 on the Disability Scale[43] indicating that her symptoms affect her work and leisure activities. The patient rates her disequilibrium as a 6.6 on a visual analogue scale from 0 (normal) to 10 (worst it could be). She rates her oscillopsia during ambulation as an 2.4 on a similar scale. Her balance confidence (ABC) is 56%. Her dizziness affects her activities 80% of the day.

Neurological Examination

Normal except as follows:

1. Patient has absent response to vibration (125 Hz) in her toes and poor kinesthesia in her toes and ankles. Ankle jerk responses are absent bilaterally.
2. Muscle strength in anterior tibialis muscle and in gastrocnemius muscles is 4-/5 bilaterally.
3. Unable to perform Romberg eyes open or closed for 30 seconds.
4. Walks slowly with widened base of support; turns "en bloc."

Comment

The peripheral neuropathy may affect her postural stability, especially in the dark, as well as the final level of functional recovery. It is unusual for the Romberg to be abnormal. This may be because of her peripheral neuropathy or the combination of peripheral sensory loss and vestibular hypofunction, and/or there may be an element of fear of falling. Her gait pattern is typical of a person with an uncompensated vestibular deficit.

Oculomotor Examination

The oculomotor examination in room light reveals:

1. Visual acuity is 20/25–4 OU (both eyes viewing) with head stationary and decreases to 20/63 - 3 during 2-Hz horizontal oscillations of the head.
2. Gaze stabilization is normal during slow head rotations but patient makes large corrective saccades with head thrusts to the right.
3. No spontaneous or gaze-holding nystagmus.
4. Dynamic Visual acuity testing using the computerized system could not be performed, because the patient developed a significant increase in dizziness with only 20 seconds of horizontal head movement.

Oculomotor examination using an infrared recording system to prevent fixation suppression of nystagmus reveals:

1. Horizontal head-shaking resulted in a left-beating nystagmus; there was no vertical head-shaking–induced nystagmus.
2. Movement into the Dix-Hallpike position to the left was negative, but to the right the patient developed a mild left-beating nystagmus without vertigo.

Comment

The decrease in visual acuity during head movement is consistent with an uncompensated vestibular deficit. The presence of corrective saccades with head thrusts to the right is consistent with a right defcit. The normal-appearing gaze stabilization with slow head movements may be a result of the function of the intact left side, residual function of the right vestibular system, or to substitution of the pursuit eye movement system for gaze stability. Her inability to tolerate/ perform repeated head movements for more than 20 seconds will limit her initial exercise program. Head-shaking–induced nystagmus is not always present in patients with unilateral vestibular deficits. The presence of a left-beating nystagmus following horizontal head shaking is consistent with her right-sided lesion. The left-beating nystagmus, without vertigo, when the patient was moved into the right Dix-Hallpike position, is from the asymmetry in the utricular signal produced by movement into that position and is not uncommon in patients with unilateral vestibular loss.

Balance and Gait

1. She has an abnormal Romberg with eyes open and closed. Her single leg stance time is less than 2 seconds eyes open. She walks with a slow cadence (0.63 m/sec; normal for age = 0.87 m/sec)[78] with and without cane, widened base of support, and decreased rotation through trunk and neck. She is unable to turn her head while walking and keep her balance.

Continued

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2. Fall risk: Her dynamic gait index is 14/24 indicating a risk for falling. She had difficulty with changes in gait speed, walking and turning her head horizontally and vertically, turning around quickly, stepping over an object, and going up and down stairs.

Plan for Vestibular Rehabilitation

Problems

1. Poor static balance and single leg stance time.
2. Disequilibrium and postural instability induced by head movement.
3. Poor balance in the dark.
4. Decrement in visual acuity with head movement.
5. Limited tolerance for head movement.
6. Ambulation with a cane.
7. Slow gait speed.
8. Is at risk for falling.
9. Decreased participation level.
10. Social isolation.

Plan

The patient was placed on gaze and postural stabilization exercises and some exercises based on functional activities. The exercises consisted of the X1 viewing paradigm, which the patient performed for 20 seconds each using a foveal target held in her hand. Initially, the patient performed these exercises sitting down, and she was instructed to wait for her symptoms of dizziness to decrease before starting the next exercise. She performed both horizontal and vertical head movements. This exercise was to be performed three times daily initially with an increase to five times daily in 1 week if tolerated and to perform standing when possible. The patient was also instructed to gradually increase the duration of each exercise with a goal of 1 minute each. The goals of these exercises were to improve gaze stability, decrease her symptomatic response to head movement, and encourage the patient to make head movements. Functionally, the exercises should improve her visual acuity during head movement and habituate the dizziness induced by head movement. She was also instructed in static balance exercises performed with eyes open and then while briefly closing her eyes. Unfortunately, she lived alone, so a family member was not available to help her with the exercises. For her safety, she was instructed to perform the balance exercises in a safe place in her home, such as in a corner, facing out, and to place a heavy chair in front of her, which she could grasp if needed. She was taught the step-over exercise, which she was to perform twice daily for 2 minutes (Fig. 22.8) with the goal of enabling her to safely step over small objects and up curbs. She was instructed to practice walking without her cane and without touching the furniture or walls. She was to perform this latter exercise in her house in the hallway for 3 to 5 minutes, twice daily. The goal of these exercises was to decrease her reliance on her cane and on any possible furniture or wall "crawling" while walking.

The patient was also asked to begin a walking exercise program, increasing her walks to 20 minutes daily. The patient performed all exercises at least once during this initial clinic visit to ensure that she was able to perform the exercises correctly and that the exercises did not make her so dizzy that she would stop performing them. Follow-up was scheduled for 1 week later, and the patient was told to call if she had questions or difficulty performing the exercises. After the initial follow-up visit, the patient was seen in clinic once a week.

One-Month Reevaluation

Subjective Complaints

The patient rated her disequilibrium as a 3.0 on a visual analogue scale from 0 (normal) to 10 (worst it could be). She rated her oscillopsia during ambulation as a 1.9 on a similar scale. This was a marked improvement over her initial visit. Her balance confidence was a 64% (meaningful change is greater than 10%), and she stated that her symptoms interfered with her activities 65% (meaningful change is 4.1%) of the time.

Balance and Gait

She could perform the Romberg with eyes open for 30 seconds but could not perform the Romberg with eyes closed for more than a few seconds. She no longer was using her cane. During ambulation, her cadence, base of support, step length, and rotation through trunk and neck were normal. Her gait speed had increased to 0.79 m/sec. When asked to turn her head from side to side while walking, there was a decrease in cadence, but the patient could perform the task with only a slight deviation from a straight path. She was able to walk safely on ramps, stairs, and on grass, although on grass she was very cautious and did not have much head movement. She had resumed local driving during

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Figure 22.8 Step-over exercise: The patient shifts his or her weight to **(A)** step over a rolled towel and then **(B)** step back. The patient can perform this first holding onto a counter and then without holding on.

the day but had not attempted to drive at night. She was able to go to church but had not yet resumed her evening social activities.

Comment

This patient is now showing good progress toward recovery following her unilateral vestibular loss. Her inability to perform the Romberg with eyes closed most likely reflects her peripheral neuropathy, because most people with UVH can perform the Romberg eyes open and eyes closed. A home program of exercises would be continued until the patient's recovery had plateaued. The use of a straight cane at night was recommended because of her combined peripheral neuropathy and vestibulopathy.

CASE STUDY 22-2

A 46-year-old male with complaints of dizziness and imbalance; scheduled for resection of left vestibular schwannoma next week. MRI with gadolinium showed a 1.5-cm tumor extending into the left cerebellar-pontine angle, with slight compression of brainstem or cerebellum. Facial nerve function appeared to be symmetrical and normal. He reports that his balance is worse in the dark and when going up stairs. He denies falls. Patient appeared quite concerned about the scheduled surgery. He expressed concerns about dying or being unable to return to work after surgery.

Past Medical and Social History

Headache (sinus or migraine), allergic to hay; no hearing left ear and a minimal high frequency loss right ear with a Speech Reception Threshold of 35 and no speech discrimination loss. Medications: none. Allergies: No known drug allergies. He lives with his wife and two children age 10 and 14 years. The bedrooms in his house are on the second floor; there is a railing on the stairs. There are 4 steps to enter his house from the front and 2 steps from the garage (but no railing in

Continued

CASE STUDY 22-2

the garage). He is independent in basic activities of daily living and drives day and night with no limitations. He has a busy practice as a trial lawyer.

Subjective Complaints

He rates his Disability as 0/5 indicating that he has negligible symptoms. His PANAS scale indicated both anxiety and depression. Head movement does not induce dizziness (VAS = 0/10) and only slightly increased his oscillopsia (VAS = 0.1/10) and disequilibrium (VAS = 2.0/10). His activities-specific balance confidence score (ABC) is 82%. Dizziness affects his activities 5% of the time.

Oculomotor Examination

Positive head thrust test to the left. With Frensel lenses, there was no spontaneous or gaze-evoked nystagmus. After head shaking, there was only a single drift of the eyes to the left and a single corrective saccade. No nystagmus was elicited with vertical head shaking. Visual acuity with the head stationary was 20/20 corrected. Computerized visual acuity during active head oscillations between 120 and 180 degrees per second was normal for rightward and leftward head movements.

Balance

His Romberg was normal and the patient could maintain the sharpened Romberg with eyes open for 30 seconds. He could not perform the sharpened Romberg with eyes closed. The patient ambulated with a normal stride length, base of support, trunk and neck rotation, and arm swing. His gait speed was slower than normal for his age (1.0 m/sec compared with a normal value of 1.13 m/sec).[77] He stated that he normally walks slowly. He did not appear to use excessive visual fixation to maintain his balance while ambulating. Fall Risk: DGI score = 22/24.

Comments

This patient is typical of many patients with vestibular schwannomas in that his initial complaint was decreased hearing. The absence of vertigo or even disequilibrium in these patients is because of the gradual vestibular loss occurring as the tumor grows rather than an abrupt onset as would occur with vestibular neuronitis. He has no central signs such as a direction changing gaze-holding nystagmus. The patient made corrective saccades with head movements toward the involved side, which indicates an inadequate vestibular response. The only abnormal finding found with assessment of his balance was an inability to perform the sharpened Romberg with eyes closed. We have found that the sharpened Romberg test with eyes closed is sensitive to unilateral vestibular loss but not specific for vestibular deficits in younger people; because many older community-living adults cannot maintain the sharpened Romberg position. His history of migraine headache may complicate his recovery.[100-102] Additionally, if he has migraines postoperatively, it may make it difficult for him to perform the gaze stabilization exercises.

The primary concerns expressed by the patient were that he might die during surgery or might not be able to work again after surgery. These concerns need to be addressed before surgery, if possible.

Intervention

Before surgery, the typical postoperative rehabilitation course was discussed with the patient and his wife. Emphasis was placed on the need to move his head postoperatively, even if it made him somewhat dizzy; the exercises he most likely would be performing, and his activity level when discharged home. Because of his concerns about the surgery, an appointment was set up with the neurologist. The neurologist prescribed an antianxiety and antidepressant medication for short-term use only. Surgical resection of the acoustic neuroma was performed 3 weeks later using a translabyrinthine approach, because the patient had no usable hearing in that ear (see Chapter 17). The patient did well postoperatively. He had moderate complaints of vertigo for a few days after surgery but no diplopia. He did have a complete facial paresis, but the surgical report noted that the facial nerve was intact. Initially, he had a spontaneous right-beating nystagmus that increased when he looked to the right. He expressed considerable relief that the surgery was over.

On postoperative day 3, he no longer had a right-beating nystagmus. His active neck range of movement was limited by 50% because of pain at the surgical site, which he described as a pulling sensation.

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Subjective Complaints

He rated his oscillopsia as a 7/10 with head stationary and as an 8.2 with his head moving. He noted that some of the visual blurring was caused by the ointment he was using in his eye because of the facial palsy. He rated his disequilibrium while walking (assisted to the bathroom) as a 6/10. He rated his neck pain as a 3/10.

Gait and Balance

He had a negative Romberg. Sharpened Romberg was not tested. He was able to ambulate independently but had a widened base of support and minimized rotation through his trunk. His gait was slow, and he occasionally side-stepped. He appeared to use excessive visual fixation while he walked for balance and slowed his gait when he turned his head while walking. Gait speed was 0.54 m/sec.

Treatment

Gaze stabilization exercises were initiated on postoperative day 3. The initial exercise used was the X1 viewing paradigm, which the patient was to perform both while sitting and while standing. While standing, he was to gradually decrease his base of support and bring his feet closer together. The exercise was to be performed using a foveal stimulus with horizontal head movements only. The patient was instructed to perform the exercises five times a day for 1 minute each. In addition, he was to walk, gradually increasing the distance walked.

Comment

This patient's postural instability and his performance on the various balance tests are typical of patients during the acute period following resection of vestibular schwannoma. Many patients experience vertigo immediately after the surgery because of the sudden asymmetry in vestibular function with removal of the tumor. This also accounts for the spontaneous nystagmus in this patient. His facial paresis could be a problem when he performs the gaze stabilization exercises, because he cannot blink to lubricate his eyes.

Early Postoperative Course

By postoperative day 6, the day of discharge, he was ambulating independently but still had an increased base of support and a slow cadence (0.7 m/sec). His gait was less ataxic, and he no longer used excessive visual fixation to maintain balance. He was independent on stairs with a railing. His exercise program consisted of the X1 viewing paradigms, using both a foveal and a full-field stimulus and moving his head horizontally for 1 minute and then vertically for 1 minute, that he was to perform standing five times a day. He also was instructed to begin practicing turning his head while walking, being careful because it would make him less stable.

Total exercise time each day was approximately 25 minutes. At this stage, patients are still not allowed to bend over or lift anything more than 5 pounds (risk of cerebrospinal fluid leak). Stair safety was discussed with the patient and his wife. Other topics discussed were the expectation that he would be very tired when he got home and even the next day because of the traveling; to resume his exercises either 1 or 2 days after he got home, depending on his fatigue; to try to establish a regular sleeping and meal schedule; and to remember that activity was beneficial.

Comment

Rapid recovery from the severe vertigo and imbalance for patients with unilateral vestibular loss, even after surgery, is typical. The effect of the vestibular loss, however, is still obvious (e.g., still has difficulty with walking while moving his head), and fatigue is a major problem after surgery. Most patients with unilateral vestibular loss are never able to perform the sharpened Romberg with eyes closed. We typically do not give these patients any kind of assistive device to use when walking. There are 2 or 3 days while they are in the hospital when walking with a cane might be helpful for some patients. Purchasing a cane for these patients is difficult to justify, because the cane will not be needed at discharge. There are exceptions to this rule, of course.

Outpatient Follow-up

At 2 weeks s/p surgery, his primary complaints are (1) his facial palsy; (2) dizziness with head movement; (3) imbalance when he turns his head, or walks on uneven surfaces (grass) or in the dark; (4) fatigue; and (5) sleeping poorly. He denies falls. He states that he has been compliant with the exercises and is walking

Continued

CASE STUDY 22-2

10 to 15 minutes daily but not by himself. He is not driving but wants to start as soon as possible. PMH: unchanged except for recent surgery and facial paresis with left eye irritation secondary to surgery. Medications: Puralube for eye.

Subjective complaints: He rates his disability as 4/5 indicating he is on medical leave but states that otherwise he would rate his disability as a 3/5. Head movement VAS = 2.1/10; oscillopsia VAS = 3.1/10; disequilibrium VAS = 4.2/10. Balance confidence (ABC) = 69%.

Balance: Fall Risk: DGI score = 18/24 (difficulty keeping his balance when walking with head movements.) Gait speed = 1.0 m/sec (nl ≥1.13 m/sec).

Visual Acuity: DVA = 0.236 for head movements to the left (normal < 0.162).

Facial movement: No appreciable movement

Treatment

The eye-head exercise was added to his exercise program, as well as walking with head movements horizontally and vertically and walking outside, increasing the time to 20 to 30 minutes daily. Protection of his left eye, because he had no blink reflex, was discussed including not rubbing the eye, wearing wraparound sunglasses to prevent dust or small particles from being blown into his eye and reinforcement of using a lubricant in his eye. Returning to driving was also discussed with the patient. It was suggested that he should not drive until he could see clearly and was not dizzy when he moved his head. The visual blurring in his left eye, because of the lubricant, was another reason to not resume driving yet.

At 4 weeks status post resection of left vestibular schwannoma, he still has complaints of dizziness and imbalance, which he notes is worse in the dark. He is independent in basic activities of daily living and had tried limited driving in the daytime but not at night primarily secondary to eye irritation. He has not returned to work secondary to fatigue, some imbalance with turning, and his facial palsy.

Subjective complaints: He rates his disability as 1/5 indicating his symptoms are bothersome.

Head movement VAS = 0.4/10; oscillopsia VAS = 0.3/10; disequilibrium VAS = 1/10. Balance confidence (ABC) = 82.5%.

Balance: Fall Risk: DGI score = 22/24 (still has difficulty keeping his balance when walking with head movements.) Gait speed = 1/10 m/sec.

Visual Acuity: DVA = 0.136 for head movements to the left (normal < 0.162).

Comment

He was doing very well after surgery. Primary problems are limited to walking on uneven surfaces, walking with head movement, and difficulties related to eye irritation. He had an appointment scheduled with an ophthalmologist for later in the week. The goal for this patient is a return to full activities, probably within 6 weeks of surgery. Other patients require a longer recovery period, and may not return to work for 3 months. The full recovery period following this surgery is 1 year with fatigue being the main problem. Within 6 months (and usually earlier), patients should be able to participate in sports such as tennis, racquetball, and golf, all of which are also good vestibular and balance exercises. They may have to change how they play and may have to shift to doubles tennis games. Patients will be aware of a sense of imbalance when they turn rapidly toward the side of the deficit, but usually do not have any loss of balance. Some patients complain that they have difficulty when balance is stressed, such as when walking in the dark on uneven surfaces or if they have to step backwards suddenly.

Patients who do not do well should be carefully screened for other problems that would complicate their recovery, such as the coexistence of visual changes, sensory changes in the feet, or central nervous system lesions that would prevent vestibular adaptation. Additionally, screening for psychological problems may be helpful. Some patients are fearful of moving the head. These patients may still benefit from vestibular exercises even several months after surgery, but will need to be on a more closely supervised program to ensure compliance.

This patient also had a facial palsy after surgery. The potential for recovery is good because the nerve was intact after surgery. We do not initiate facial exercises until the patient has more than faint voluntary movements, and then patients are cautioned to practice gentle facial movements rather than forceful movements. Another approach would be to send the patient for

CASE STUDY 22-2

EMG biofeedback instruction.[103] In patients with significant synkinesis, we use biofeedback training to improve the quality of the facial movements. Of main concern for patients with facial paresis or palsy is protection of the eye. If lid closure is absent or poor, patients may use either a cellophane moisture chamber or an eye patch to prevent drying of the eye and corneal damage. Patients using either type of patch should be advised to be careful when walking because they have only monocular depth perception cues. Surgical intervention, such as implanting a gold lid weight, may be necessary.

CASE STUDY 22-3

An 18-year-old woman is referred to physical therapy with a diagnosis of a right peripheral vestibulopathy. Current complaints include imbalance, especially with head movement, and a sense that her eyes "don't catch up with my head" during head movements. Patient no longer complains of vertigo and denies falls. She states that she still occasionally staggers, stumbles, and side steps when walking. PMH is non-contributory. Current meds include Prednisone and acyclovir. She is allergic to penicillin. She is independent in all activities of daily living. She has not returned to driving or to her part-time job as a housecleaner. She has returned to her college classes.

Neurological Examination

The general neurological examination was normal except for the vestibular system. Her MRI scan and audiogram were unremarkable. Vestibular laboratory testing included an oculomotor screening battery, static positional testing, caloric testing, rotational testing, and posturography. Test results showed a left gaze-evoked nystagmus, a left-beating nystagmus on positional testing, a right vestibular paresis on caloric testing, a left directional preponderance on rotational testing, and abnormal posturography (abnormal response for all six sensory organization conditions and abnormal adaptation to toes-up rotation on movement coordination).

Comments

The left-beating, gaze-evoked nystagmus, the left-beating nystagmus on positional testing, and the directional preponderance most likely reflect right vestibular paresis. The increased difficulty experienced by the patient on all six of the sensory organization tests is unusual, but not unheard of, during the chronic stage following unilateral vestibular deficits. In some patients, this finding may reflect a psychogenic component to the patient's complaints. Difficulty maintaining balance to sudden toes-up rotations of the support surface may signify a tendency toward retropulsion in some patients, but is a nonspecific finding in many patients with balance problems. It may indicate that the patient will have difficulty walking on uneven surfaces.

Physical Therapy Examination

Subjective complaints: She rates her disability as a 1 on a scale from 0 to 5 indicating her symptoms are bothersome.[43] She rates her dizziness with head movement as a 1.4/10, her oscillopsia while walking as a 1.4/10 and her disequilibrium while walking as a 1.9/10 (all scores represent decrement associated with movement; score of 10 is worse).

Visual Acuity: head stationary: LogMAR 0.018 (normal); with predictable head movement to the left: LogMAR = 0.076 (normal for age = 0.087); predictable head movements to the right: LogMAR = 0.125.

Static Balance: Negative Romberg; SR eyes open = normal, eyes closed = 4 sec (normal for age = 60 seconds)

Gait: decreased head movement, occasional stagger

Fall risk: Dynamic Gait Index Score = 23/24

Impression: subjective complaints of head movement induced dizziness, oscillopsia, and imbalance; abnormal visual acuity with 120 d/s head movements to right; abnormal gait pattern (decreased head movement); decreased activities (driving) all consistent with sub-acute UVL

Continued

CASE STUDY 22-3

Treatment Goals

1. 75% decrease in head movement induced symptoms (dizziness, oscillopsia, and disequilibrium) in 6 weeks
2. Normal visual acuity during head movement to right in 6 weeks
3. Normal static balance with eyes closed except for SR
4. Return to all normal activities in 6 weeks

Comment

Her complaints of disequilibrium with head movement, blurred vision, and a gait disturbance and the objective findings are all consistent with the disturbance of the dynamic vestibular responses.

Intervention

The patient was given a home exercise program that consisted of head movements in standing with eyes open, X1 and X2 exercises, and a walking program. She was told to perform the X1 and X2 exercises five times a day for 2 weeks. She was advised to walk daily. She was seen on two subsequent visits, each 2 weeks apart, upon which she was reevaluated and given a progression in her home exercise program. The following exercises were added to her home program over time: walking with head movements, walking with pivot turns, walking backwards, kickball, and circle walking. She was also given additional eye/head exercises (see substitution exercises).

After 4 weeks, the patient was retested in preparation for discharge. The patient no longer complained of symptoms with head movement. Her gait no longer revealed any abnormalities. She was able to tandem walk 15 steps with her eyes open and 3 steps with her eyes closed. In addition, she had no difficulty walking and moving the head left and right.

At discharge, her activity level had improved. She was walking for 45 minutes four times a week. She was able to drive and perform housework without difficulty. At discharge, the patient was instructed in a maintenance home program that consisted of head movements, eye/head movements, and a walking program.

SUMMARY

Patients with unilateral vestibular deficits can be expected to recover from the vertigo and/or disequilibrium they first experience. The final level of recovery should be to return to all or most activities. Other nervous system disorders can delay or limit the level of recovery. There is moderate to strong evidence in human beings that suggests that exercises facilitate recovery. Early intervention may not be important in optimizing recovery, because patients with chronic deficits still show improvement with a vestibular rehabilitation program, although some evidence exists that suggests that those who receive early intervention may require less pharmacological management and have better outcomes.

This chapter has presented treatment strategies for the physical therapy management of patients with unilateral peripheral vestibular hypofunction. Patients with unilateral peripheral vestibular hypofunction may present with a variety of functional deficits. The physical therapy treatment program should address all of the patient's functional deficits. The use of exercise in the rehabilitation of patients with vestibular disorders is aimed at promoting compensation for the vestibular loss. Several case studies are presented to illustrate the rehabilitation management of patients with unilateral peripheral vestibular hypofunction.

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CHAPTER 23

Physical Therapy Management of Bilateral Vestibular Hypofunction and Loss

Susan J. Herdman, PT, PhD, FAPTA
Richard A. Clendaniel, PT, PhD

Reduction or loss of vestibular function bilaterally results in difficulty maintaining balance, especially when walking in the dark or on uneven surfaces, and in a decrease in the patient's ability to see clearly during head movements. In addition, patients with bilateral vestibular hypofunction or loss (BVH or BVL) also complain of intense feelings of being off-balance and of strange but disturbing sensations in their heads with head movement. Because of these problems, patients with BVH may restrict their activities and can become socially isolated.

Vestibular rehabilitation can improve postural stability, decrease the sense of disequilibrium, and improve visual acuity during head movements enabling people with BVH to resume a more normal life. [1-5] Unfortunately, most patients have residual functional problems and subjective complaints.[5] The exercises used for patients with BVL are aimed at fostering the substitution of alternative strategies to compensate for the lost vestibular function and at improving any remaining vestibular function. This chapter presents the assessment and physical therapy treatment of these patients. Case studies are used to illustrate different points.

Etiology

Bilateral vestibular hypofunction and loss can occur secondary to a number of different problems including ototoxicity, meningitis, sequential vestibular neuritis, progressive disorders, autoimmune disorders, chronic inflammatory peripheral polyneuropathy, congenital loss, and neurofibromatosis. In most cases, however, BVH is considered to be "idiopathic," because the underlying cause cannot be identified (see Chapter 4). The incidence of the various forms of BVH is also not clear, because it varies depending on the type of clinical practice examined (Table 23-1).

Primary Complaints

Balance and Risk for Falling

Patients with bilateral vestibular loss are primarily concerned with their balance and gait problems. During the acute stage, they may feel off balance even when lying or sitting down. More typically, however, their balance problems become obvious only when they are standing or walking. Thus, patients who are bedridden, such as those who develop BVH after

■ Table 23-1 **REPORTED FREQUENCY OF TYPES OF BVH**

Study	Herdman et al[5] (n=69)	Zingler et al 2007[6] (n=255)	Rinne et al 1998[7] (n=53)	Brown et al 2001[8] (n=13)	Gillespie and Minor 1999[2] (n=35)
Mean age (years)	63.2 ± 14.4	62 ± 16	—	61.1 ± 18.2	56
Gender: % male	66.7%	62.0%	—	46%	51%
Etiology unknown	38.4%	51%	21%	38.5%	5.7%
Ototoxic	20.2%	13%	17%	61.5%	57.1%
Progressive	20.2%				
Sequential	23.9%	36%			14.3%
Autoimmune		7%	9%		5.7%
Ménière's/other otological		5%	13%		5.7%
Tumor			5.7%		
Plus cerebellar	excluded		13%		
Meningitis/other neurological	26%	11.4%			

receiving a vestibulotoxic medication such as gentamicin, often do not know they have a balance problem until they get out of bed. Typically, these patients have been treated with the ototoxic medication because of a serious infection. They are often debilitated, and their balance problems are initially attributed to weakness. Other patients, such as those with a progressive loss, may not notice their gradually worsening balance until it reaches a critical point and they start falling.

Even with full compensation, balance problems will persist. Although the other sensory and motor systems do help compensate for the vestibular loss, these systems cannot substitute completely for the loss of vestibular function (see Chapters 6 and 9). Normal postural stability while walking requires the combined use of at least two of three sensory cues (visual, vestibular, somatosensory). Patients who have no vestibular function, therefore, will have difficulty when either visual or somatosensory cues are also significantly decreased (e.g., walking in the dark). Although balance may be poor, it is not known what the actual frequency of falling is for patients with BVL. One study reported that 70% of patients with BVH under the age of 65 reported falls related to their bilateral hypofunction and 58% of those 65 to 74 years of age reported falls related to their BVH.[9] In both cases the incidence of falls exceeded the fall rate of healthy individuals of the same age. Interestingly, patients age 75 years and older had a lower fall rate than do healthy individuals; this was attributed to their use of assistive devices and limited activity level. Most patients are able to prevent falls even though they may side-step or stagger occasionally. However, even after a course of vestibular and balance rehabilitation, 20% to 30% of patients with BVH are still at risk for falling when they are discharged.[3,10]

Oscillopsia

Another problem for patients with bilateral vestibular loss is the visual blurring that occurs during head movements. Almost 70% of all patients with BVH complain of oscillopsia even after a course of gaze stabilization exercises.[10] Greater intensity of oscillopsia occurs in patients with absence of both

inferior and superior vestibular nerve function. In addition to the subjective complaint, patients also have a documented decrement in visual acuity during head movements. Interestingly, several studies have demonstrated that the subjective complaints of oscillopsia does not correlate with the actual decrement in visual acuity during head movement.[5,11,12]

Initially, loss of vestibular function results in a decrement in visual acuity even when the patient is stationary, if the head is not supported.[13] Even following the best compensation, some patients say that objects that are far away appear to be jumping or bouncing. This visual blurring, or oscillopsia, increases with irregular or unpredictable head movements such as would occur while walking. As a result, patients may not be able to read street signs or identify people's faces as they walk, or they may have difficulty seeing clearly while in a moving car. Severe oscillopsia will also impact postural stability because decreased visual acuity will affect the person's ability to use visual cues for stability.[14]

Sense of Disequilibrium, Imbalance, and Dizziness

Patients often complain of a sense of being "off-balance" that is separate from their actual postural instability. This feeling lessens or disappears when the person is lying down or sitting with the head supported. It increases dramatically when the person is moving. Although disequilibrium may decrease as a result of compensation, for up to 80% of patients it remains a serious and debilitating problem.[10] It can lead to decreased physical activity, social isolation, and depression. Another disturbing sensation that is more vaguely described by patients with terms such as "dizziness" and "spaciness" is also heightened by head movement and persists in as many as 60% of all patients. This head movement–induced dizziness is exacerbated by repeated head movement and may cause patients to avoid movement.

Physical Deconditioning

Poor physical condition can be a significant problem for patients with BVL. This can be caused directly by a decreased activity level because of the patient's fear of falling or because of the increased dizziness that occurs with movement. It is especially a problem for patients whose vestibular loss is secondary to ototoxic medications. These patients are already debilitated because of severe infection. Many patients on peritoneal dialysis, for example, develop infections that are treated with gentamicin, a vestibulotoxic aminoglycoside.

Assessment

The assessment of patients with BVL is similar to that for patients with unilateral vestibular deficits; therefore, only certain aspects of the assessment will be described here. Physical therapy assessment of patients with BVL must address subjective complaints, postural instability and oscillopsia, the patient's overall physical condition, and their ability to perform activities of daily living (ADLs). This assessment must also identify other factors that might affect recovery, especially visual and somatosensory deficits. A summary of the assessment and the usual findings for patients with chronic bilateral vestibular loss is presented in Box 23-1.

Box 23-1

TEST RESULTS ON PATIENTS WITH BVL

Oculomotor examination

- Abnormal findings in room light including: poor VOR to slow and rapid head thrusts; visual acuity with head stationary is usually normal but in the more acute stage, during gentle oscillation of the head, acuity changes to 20/100 or worse. Some patients will have normal dynamic visual acuity during active head movements in the compensated stage.
- With Frenzel lenses: No spontaneous, gaze-evoked, head-shaking–induced, tragal-pressure–induced, hyperventilation-induced, or positional nystagmus.

Sensation

- Somatosensory and visual information is critical to functional recovery and must be carefully evaluated.

Coordination

- Should be normal. The exception might be the patient with Neurofibromatosis 2 in whom other parts of the nervous system may be affected.

Range of motion

- Should be normal, but patients will voluntarily restrict head movements because head movement

Box 23-1

TEST RESULTS ON PATIENTS WITH BVL—cont'd

makes them less stable and also results in poor vision.

Strength (gross)

- Should be normal.

Postural deviations

- Should be normal.

Positional and movements testing

- Should not result in vertigo.

Sitting balance

- Patients may *have* difficulty maintaining their balance during weight-shifting in sitting during the acute stage, but should not have difficulty during the compensated stage.

Static balance

- Romberg: Abnormal during acute stage in many patients. Normal based on time in the compensated patient.
- Sharpened Romberg: Patients with complete or severe bilateral loss *may* not be able to perform this with eyes open and will not be able to perform this with eyes closed.
- Single leg stance: Difficult to perform even during compensated stage, with eyes open.
- Stand on rail: Usually not tested.
- CTSIB: Romberg will often be normal. Standing on foam surface: Difficult to perform with decreased base of support. Should not be attempted in many patients.
- Force platform: Normal or close to normal anterior-posterior sway with eyes open or closed during compensated stage on stable surface.

Balance with altered sensory cues

- Increased sway when visual *or* somatosensory cues are altered, loss of balance when both visual *and* somatosensory cues are altered.

Dynamic balance (self-initiated movements)

- Fukuda's Stepping Test: Normal with eyes open during compensated stage; cannot perform with eyes closed (rapid loss of balance).

Functional reach

- May be decreased with eyes closed.

Ambulation

- The patient's gait is usually at least slightly wide-based during compensated stage. There is a tendency to use visual fixation while walking and to turn "en bloc." Tandem walk cannot be performed with eyes closed.
- Walking backwards even with eyes open may be difficult and performed with caution
- Walk while turning head: Gait slows, base of support widens, step length decreases; may become ataxic.

History

For the most part, evidence on the relationship of etiology to the final level of recovery is conflicting. Extrapolation of the data from Brown et al (2001) shows that 40% of the subjects with an unknown etiology had improvement in balance confidence compared with 50% of patients with ototoxicity.[3] Gillespie and Minor (1999) also reported that the majority of patients with ototoxicity (n=20) improved.[2] Another study found that patients with an idiopathic etiology had lower disability scores and higher balance confidence scores at discharge, both indicating perceived improvement, compared with all other etiologies combined.[10] However, one advantage of knowing the underlying etiology of the BVH is in knowing the accompanying problems the patient may have. For example, the patient who has a spontaneous or sequential BVH is less likely to have other health problems that will affect recovery than the patient who had a severe infection and was treated with an ototoxic medication.

One common cause of BVH is the effect of ototoxic medications such as gentamicin. Once considered to be an idiosyncratic response, initial studies indicated that less than 3% of people who receive gentamicin develop a vestibular deficit.[15] Subsequent studies, however, indicate that the incidence of aminoglycoside ototoxicity ranges from 9% to 15%.[16-18] These are most likely conservative estimates based on relatively small studies (n less than 150 in each study) and the fact that vestibular toxicity was assessed using electronystagmography. The prevalence is as

great as 10% to 20%, however, in those patients with renal impairment, over the age of 65 years, taking loop diuretics, or with previous vestibular loss. This percentage increases to 40% among persons on renal dialysis who receive gentamicin. Furthermore, the patient who has a loss of vestibular function, with its resultant balance and visual problems, secondary to ototoxic medication may have to deal with significant anger and depression.

While taking the patients' histories, the clinician should identify the presence of specific comorbidities such as macular degeneration and cataracts. These disorders result in a gradual decrease in available visual cues and will have an adverse effect on balance in the future. Similarly, disorders that frequently result in peripheral neuropathies should be noted, such as diabetes or chronic alcohol abuse. Other comorbidities that influence outcome in patients with BVH had fair to strong correlations with specific outcomes.[10] Of particular interest was that a history of headaches correlates with a higher percentage of time that symptoms interfere with life, a greater intensity of oscillopsia and head movement–induced dizziness, and a lower balance confidence at discharge. Furthermore, the total number of comorbidities also influences outcome.[2,10] Gillespie and Minor (1999) found that poor outcome following a course of vestibular rehabilitation was related in part to having more than four comorbidities,[2] and Herdman et al reported that poor outcome, as indicated by higher disability scores at discharge, was correlated with greater numbers of comorbidities.[10]

Subjective Complaints

The patient's complaints of disequilibrium, oscillopsia, and head movement–induced dizziness can be assessed using a visual analogue scale.[19] The Dizziness Handicap Inventory[20] and the Vestibular Rehabilitation Benefits Questionnaire[21] are useful tools to assess the patient's perception of handicap and perception of quality of life. Balance confidence and a measure of activities of daily living should also be assessed.[22-25]

Vestibular Function

One important consideration in designing a treatment program is whether there is any remaining vestibular function. Vestibular function can be documented using tests such as the rotational chair and caloric tests. The presence of remaining vestibular function can be used as a guide in predicting the final level of recovery for patients.[2,3,10] Patients with incomplete bilateral vestibular loss are often able to return to activities such as driving at night and to some sports. Patients with severe bilateral loss may not be able to drive at night, and some patients will not be able to drive at all because of the gaze instability. Activities such as sports and dancing may be limited because of vision and balance problems.

In certain cases, vestibular function tests can also be used to follow the course of the vestibular loss and of any recovery of vestibular function that might occur.[26,27] Certain aminoglycoside antibiotics are selectively taken up by vestibular hair cells and result in a gradual loss of vestibular function. Typically, there is continued loss of vestibular function even after the medication is stopped. Some improvement in vestibular function may occur in hair cells that were affected by the ototoxic drug but did not die. Potentially, an increase in gain may also occur with the use of vestibular adaptation exercises. This has been demonstrated in patients with unilateral vestibular loss but not in patients with BVL.[28,29]

Visual System

Assessment of visual function should include a measure of visual acuity, because this can affect postural stability.[14] Measuring visual acuity during head movement is particularly important. The vestibulo-ocular system normally stabilizes the eyes during head movements; when there is no vestibulo-ocular reflex (VOR) to stabilize the eyes during head movement, small amounts of retinal slip (movement of image across the retina) will degrade vision. For instance, 3 deg/sec of retinal slip would cause visual acuity to change from 20/20 to 20/200.[30] As such, in patients with absent vestibular function, the head movement that occurs in a moving car can cause a degradation of visual acuity that would make driving unsafe.

Dynamic Visual Acuity

Assessment of visual acuity during head movement (dynamic visual acuity, or DVA) can be performed either clinically or using a computerized system. The advantage of the computerized system is that the test is standardized, and its reliablity has been established for some systems.[31] Clear differences in DVA scores occur among normal subjects, those with dizziness from nonvestibular causes, and patients with known vestibular loss (Fig. 23.1). Comparisons must be made, however, taking age into consideration (Table 23-2). The results of the computerized DVA test are both sensitive (90% for those over 65 years old and 97% for those under 65 years old) and specific (94%) for vestibular loss. The clinical DVA, however, is easy to perform and is sufficiently reliable to be useful as a guide to treatment and to treatment efficacy.[32] However, the test should be performed using an ETDRS™ vision chart rather than the more commonly used Snellen™ chart.[33]

Somatosensory System

Particular attention should be made to assess vibration, proprioception, and kinesthesia in the feet. Although mild deficits in sensation in the feet may have no effect

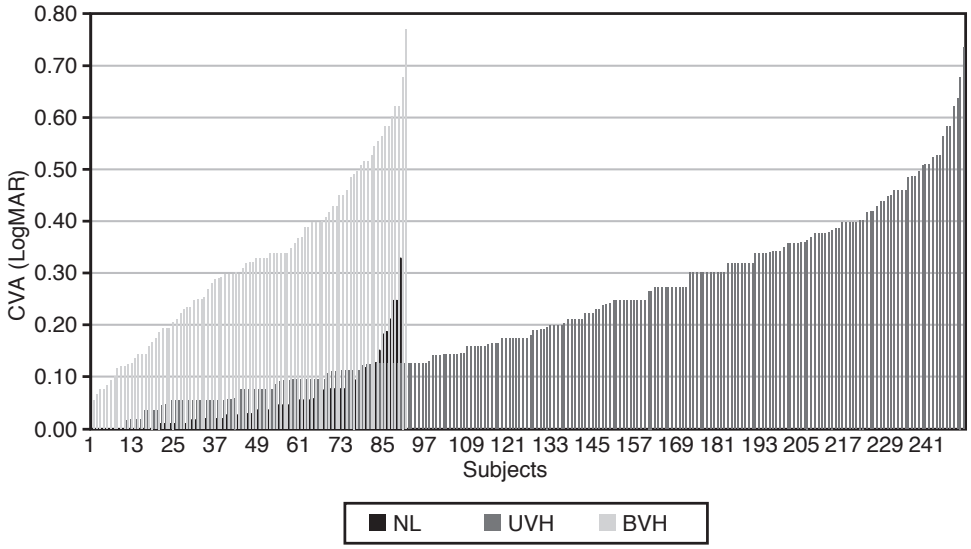

Figure 23.1 Distribution of dynamic visual acuity scores (LogMAR) using the computerized test in patients with unilateral and bilateral vestibular loss and in normal subjects. In the methodology used to determine these values, static visual acuity (SVA) had a minimal score of LogMAR 0.000, which is different from newer commercial versions of the test (see Herdman et al[31] for details on the methodology used). DVA is the expression of the decrement in visual acuity during head movement.

Table 23-2 DISTRIBUTION OF DVA SCORES IN NORMAL SUBJECTS BY AGE

Age (years)	20–29 (n=23)	30–39 (n=15)	40–49 (n=15)	50–59 (n=13)	60–69 (n=10)	70–79 (n=9)
Scores (LogMAR)						
Mean	0.021	0.024	0.032	0.059	0.078	0.187
1SD	0.024	0.027	0.032	0.037	0.058	0.089
2SD	0.048	0.053	0.064	0.073	0.115	0.178
Mean + 2SD	0.069	0.077	0.095	0.132	0.194	0.365

on postural stability in otherwise healthy individuals, in patients with vestibular loss, somatosensory deficits may have profound effects on balance and on the potential for functional recovery. As with visual system disorders, being aware of potentially progressive disorders affecting somatosensory information is important.

Balance and Gait

Patients with BVL must be given a detailed assessment of balance and gait. Obviously, static balance should be assessed first. In the acute stage, patients with bilateral vestibular deficits may have positive Romberg tests. In the compensated stage, the Romberg is usually normal in that the patient can hold the position for the normal time for their age. Although some patients will be able to perform the Sharpened Romberg with eyes open, they will not be able to perform the Sharpened Romberg with eyes closed. Patients with bilateral vestibular deficits will also have difficulty performing tests in which both visual and somatosensory cues are altered. An example of this would be Fukuda's Stepping Test in which the eyes are closed and the patient is marching in place. Patients may have normal tests with eyes open but will fall with eyes closed.

Determining whether patients use both visual and somatosensory cues to maintain balance or whether they depend on particular sensory cues is critical. It is important

to recognize that this may vary considerably from patient to patient and that it can change over the course of recovery. Bles et al[34] found that patients with BVL are initially more dependent on visual cues than on somatosensory cues for balance. With time, there is an improvement in the ability of patients to use somatosensory cues. This improvement varies from patient to patient, however, and needs to be carefully assessed (Fig. 23.2). If a patient is dependent on one sensory cue for balance (e.g., vision), then exercises that foster the ability to use the under-used sensory cues are appropriate (e.g., somatosensory).

The gait of many patients with bilateral vestibular deficits can be described as wide-based, slow, and ataxic. In other patients, gait may appear to be essentially normal unless challenged by the need to perform an unanticipated head or body movement. Patients decrease their trunk and neck rotation in an effort to improve stability by avoiding head movements. Arm swing is similarly decreased. Patients may use excessive visual fixation and therefore have increased difficulty if asked to look up or turn the head while walking. Patients typically turn "en bloc" and may even stop before they turn. Asking patients to turn their heads while walking results in increased ataxia and, often, loss of balance.

One of the main characteristics of gait of people with bilateral vestibulopathy it that is has sudden changes in trajectories compared with gait of healthy individuals.[35] That is, the patient may appear to walk normally at one moment and then staggers at the next moment.

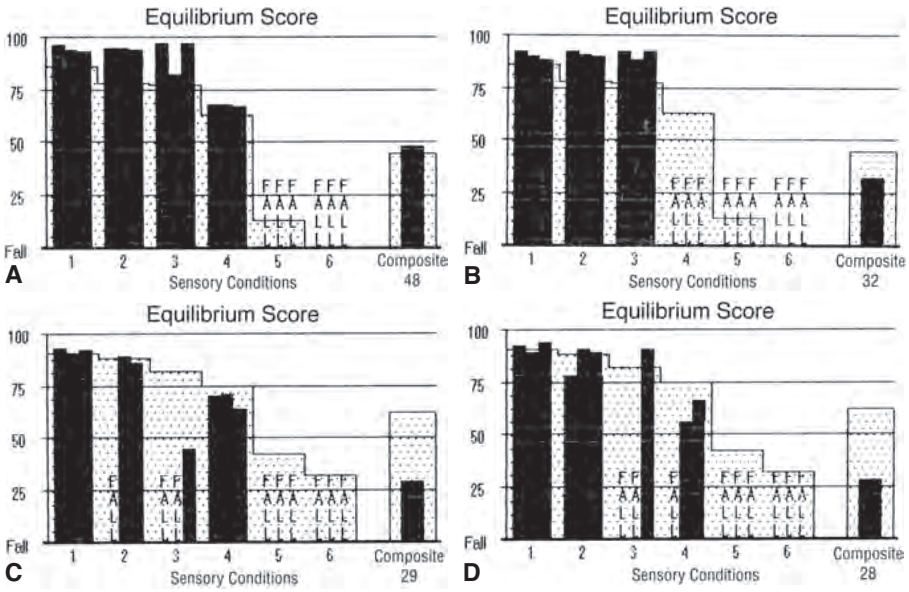

Figure 23.2 Posturography test results from patients with bilateral vestibular loss demonstrating the differences in ability to maintain postural stability when different sensory cues are altered or removed. Patients are tested using six different conditions:

	Available Cues	Unavailable or Altered Cues
Test 1	Vision, vestibular, somatosensory	—
Test 2	Vestibular, somatosensory	Vision absent
Test 3	Vestibular, somatosensory	Vision altered
Test 4	Vision, vestibular	Somatosensory altered
Test 5	Vestibular	Vision absent, somatosensory altered
Test 6	Vestibular	Vision altered, somatosensory altered

Results show patients who have difficulty when both visual and somatosensory cues are altered **(A)**, when somatosensory cues only are altered **(B)**, when visual cues only are altered **(C)**, and when either visual or somatosensory cues are altered **(D)**.

Mechanisms of Recovery

The mechanisms used to stabilize gaze in the absence of vestibular inputs have been well studied (Box 23-2). The mechanisms involved in maintaining postural stability are still somewhat less well understood, although research is being done in that area.

Gaze Stability

Subjects without vestibular function must develop different mechanisms to keep the image of the target on the fovea during head movements (see Box 23-2). Central preprogramming is probably the primary mechanism by which gaze stability is improved in patients with BVL. It is assessed by comparing the gain of compensatory eye movements during active and passive head movements.[36-40] The difficulty with central preprogramming as a substitute for the loss of vestibular function is that it would not be effective in situations in which head movements are unpredictable, such as perturbations while walking.

Other mechanisms used to improve gaze stability include modifications in saccadic and pursuit eye movements.[36,40-43] Patients with complete BVL may make hypometric saccades toward a visual target. Then, as the head moves toward the target, the eyes would be moved passively into alignment with the target. They may also make accurate saccadic eye movements during combined eye and head movements toward a target, and then make corrective saccades back to the target as the head movement pulls the eyes off the target. These strategies enable the patient to recapture a visual target following a head movement.[36] During a head movement in which the eyes should stay on the visual target, the eyes may be pulled off the target during the head movement (because of the absent VOR) and then the eyes make a compensatory saccade back to the target. Eventually, that compensatory saccade may occur during the head movement.[41,42] Pursuit eye movements can be used during low-frequency (and low-velocity) head movements to stabilize the eyes. The limits of smooth pursuit eye movements is dependent on the nature of the stimulus (predictable vs. unpredictable; sinusoidal vs. constant velocity). In general, smooth pursuit works well for sinusoidal stimuli at frequencies of up to 1 Hz (although this peak frequency decreases with age). For constant velocity stimuli, smooth pursuit can work well up to target velocities of 100 deg/sec. Recent evidence demonstrates that patients with bilateral vestibular deficits have higher smooth pursuit gains than normal controls, although their performance is still within the normal range.[43] They may also make high-velocity, slow-phase eye movements during the head movement.[40]

Potentiation of the cervico-ocular reflex (COR) was initially thought to contribute significantly to the recovery of gaze stability in patients with BVL.[36,44,45] In the COR, sensory inputs from neck muscles and facet joints act to produce a slow-phase eye movement that is opposite to the direction of the head movement during low-frequency, brief head movements. The COR, therefore, complements the VOR, although in normal subjects it is often absent and when present contributes, at most, 15% of the compensatory eye movement. In patients with complete BVL, the COR operates at frequencies of head movements of up to 0.3 Hz, well below the frequency range of head movements during normal activities.[40] Bronstein et al (1995) found that COR gain increased in patients during the early stages of recovery but then decreased, presumably as other compensatory mechanisms developed.[46] Therefore, although the COR may be increased in patients with BVL, in reality it does not operate at frequencies that would contribute significantly to gaze stability during the head movements that would occur during most activities.

Kasai and Zee[36] and Schubert et al[40] found that different patients with complete BVL use different sets of strategies to compensate for the loss of VOR. Therefore, exercises to improve gaze stability should not be designed to emphasize any particular strategy, but instead should provide situations in which patients can develop their own strategies to maintain gaze stability (see Box 23-2). No mechanism to improve gaze stability fully compensates for the loss of the VOR, however, and patients will continue to have difficulty seeing during rapid head movements.

Box 23-2

MECHANISMS USED TO STABILIZE GAZE

- Change amplitude of saccades
- Use corrective saccades
- Modify pursuit eye movements
- Central preprogramming of eye movements

Postural Stability

A study on the course of recovery of patients with complete bilateral vestibular deficits over a 2-year period has shown that patients switch the sensory cues on which they rely.[19] Initially, they rely on visual cues as a substitute for the loss of vestibular cues, but over time they become more reliant on somatosensory cues to maintain balance. In this study, patients were required to maintain balance when facing a moving visual surround. Over the 2 years

they were followed, they recovered the ability to maintain their balance to within normal limits in the testing paradigm except at high frequencies. The vestibular system functions at higher frequencies than do the visual or somatosensory systems, which would account for why neither visual nor somatosensory cues can substitute completely for loss of vestibular cues.

The contribution of somatosensory inputs from the cervical region to postural stability in patients with complete BVL is not clearly understood. Bles et al[47] found that changes in neck position did not affect postural stability in patients with complete BVL. They concluded that somatosensory signals from the neck do not contribute to postural stability. We do not know, however, if kinesthetic signals from the neck that occur during head movement would affect postural stability. Certainly patients with bilateral vestibular dysfunction become less stable when asked to turn their heads while walking. Another interpretation is that these patients are more reliant on visual cues to maintain postural stability, and thus, when the head moves and visual cues are degraded, their balance becomes worse. The contribution of somatosensory cues from the lower extremities to postural stability in patients with BVL is also not well understood. Certainly some patients are dependent on somatosensory cues rather than on visual cues. Perhaps more importantly, we do not yet know how the degree of somatosensory loss affects postural stability in these patients.

The loss of either visual or somatosensory cues in addition to vestibular cues has a devastating effect on postural control. Paulus et al[48] reported a case in which the patient had a complete bilateral vestibular loss plus a loss of lower extremity proprioception. This patient relied on visual cues to maintain his balance. When the effectiveness of the visual cues was degraded (i.e., by fixating on a visual target more than 1 meter away), his postural stability deteriorated significantly. Visual and somatosensory cues will not substitute fully for the lost vestibular contribution to postural stability.[49-51]

Compensatory Strategies

Patients can be taught, and often develop on their own, strategies to use when in situations in which their balance will be stressed. For example, they learn to turn on lights at night if they have to get out of bed. They may also wait, sitting at the edge of the bed, before getting up in the dark to allow themselves to awaken more fully and for their eyes to adjust to the darkened room. They should be advised to use lights that come on automatically and to have emergency lighting inside and outside the house in case of a power failure. Patients may need to learn how to plan to move around places with busy visual environments such as shopping malls and grocery stores. For some patients, moving in busy environments may require the use of some type of assistive device, such as a shopping cart or a cane, but for many patients with BVL, no assistive devices are needed after the patient becomes comfortable walking in that environment.

Evidence that Exercise Facilitates Recovery

Support for the use of exercises to improve physical function in patients with bilateral vestibulopathy was based originally on the result of studies of nonhuman primates (Igarashi et al).[52] Evidence now exists in people with BVH that supports the use of specific exercises (adaptation, substitution, balance and gait; see Chapter 22) to decrease subjective complaints, improve visual acuity during head movement, and improve postural stability during functional activities. Krebs et al,[1] in a double-blinded, placebo-controlled trial, found those patients performing customized vestibular and balance exercises had better stability while walking and during stair climbing than did patients performing isometric and conditioning exercises, such as using an exercise bicycle. Furthermore, the patients who had vestibular rehabilitation were able to walk faster. They used vestibular adaptation and eye-head exercises as well as balance and gait training. In a continuation of this study, Krebs et al[4] again demonstrated that, as a group, those individuals performing the vestibular rehabilitation exercises had increased gait velocity, improved stability while walking, and decreased vertical excursion of the center of mass while walking. They noted a moderate correlation between improved gait measures at 1 year and the frequency of performing the home exercise program over the preceding year. When the patients with BVL (n=51) were combined with those with a UVL (n=33), 61% of the patients demonstrated significant improvements in their gait. Another study of patients with BVH demonstrated improved visual acuity during head movement in patients with BVH.[5] In this double-blinded study, patients were randomly assigned to either perform gaze stabilization exercises (adaptation and substitution) or placebo exercises (saccades, without head movement, against a featureless background). As a group, patients who performed the vestibular exercises had a significant improvement in visual acuity during head movement, but the control group did not improve. Furthermore, 7 of 8 patients who performed the exercises improved compared with only 1 of 5 patients in the control group. Only type of exercise was significantly correlated with change

in DVA. Other factors examined, including age, time from onset, and initial DVA, were not significantly correlated with change in DVA.

The fact that not all patients with BVH improve with a course of vestibular rehabilitation is documented in several studies. Brown and colleagues noted that, as a group, the patients (n=13) had significant improvements in various measures (Dizziness Handicap Inventory, Activities-specific Balance Confidence Scale, Dynamic Gait Index, Timed "Up and Go" test, and Sensory Organization Test component of dynamic posturography).[3] However, not all patients benefited to the same degree. Brown and colleagues noted that 33% to 55% of the patients demonstrated what were considered clinically significant changes on the different measures.[3] In another retrospective study, Gillespie and Minor found that only 63% of the patients with BVL that received vestibular rehabilitation demonstrated improvements (defined as reported increased activity levels, reduced symptoms, and demonstrated normal gait velocity, normal Romberg test, or normal DVA).[2] More recently, a study examined the outcome of vestibular rehabilitation in 69 patients with BVH.[10] They found a significant improvement for the group as a whole for each outcome measure except oscillopsia and disability (Table 23-3). However, for balance confidence, percentage of time symptoms interfered with activities, visual acuity during head movement, gait speed, and fall risk, a majority of, but not all, patients improved. The percentage of patients who improved varied from 3.9% for head movement–induced dizziness to 81.3% for percentage of time symptoms interfered with activities, depending on the specific measurement (Table 23-3). The percentage of subjects who had worse scores at discharge varied from 0% for DVA, DGI, and dynamic posturography, to 28.6% for disequilibrium intensity (Table 23-3).

Not all exercise approaches are appropriate for patients with BVL. Telian et al[53] studied the effectiveness of a combination of balance exercises, vestibular habituation exercises, and general conditioning exercises for patients with bilateral vestibular deficits. They were unable to demonstrate a significant change in functional activity in these patients following treatment. Thus, vestibular habituation exercises do not appear to be appropriate for these patients. This makes sense, because habituation exercises are designed to decrease unwanted responses to vestibular signals rather than improve gaze or postural stability.

Treatment

The treatment approach for patients with complete loss of vestibular function involves the combined use of gaze stabilization exercises and exercises that foster the substitution of visual and somatosensory information to improve postural stability and the development of compensatory strategies that can be used in situations where balance is stressed maximally (Boxes 23-3 and 23-4). This approach is also used in the treatment of patients with unilateral vestibular hypofunction (see Chapter 22).

Once the patient's specific problems have been identified, the exercise program can be established. During the initial sessions, particular attention should be paid to the degree to which the exercises increase the patient's complaints of dizziness. The patient's perception of dizziness can be the major deterrent (limiting factor) to the patient's eventual return to normal activities. Head movement, a component of all exercises, may increase that dizziness. Also, the home exercise program typically requires that the patient perform exercises many times daily. Patients may find that they become increasingly dizzy with each performance of the exercises. It is important to explain to the patient that some increase in dizziness is expected when they begin the exercises and with any increase in the intensity of the exercises. One guideline to follow is that any increase in dizziness should decrease to pre-exercise levels within 10 to 20 minutes. Additionally, limit the exercises that involve head movement to only one exercise initially. Other exercises can be added and the frequency and duration of the exercises can be increased as the patient improves. Have the patient perform at least one complete set of all exercises at the time of the clinic visit. Patients should also be taught how to modify the exercises if the dizziness becomes overwhelming (Box 23-5). They should be strongly encouraged to contact the therapist if they continue to have difficulty. In those patients in whom dizziness continues to be a problem, we suggest medication and stress reduction techniques, such as walking, to try to reduce the effect of the dizziness on the patient's life.

Progression of Exercises

Changing the duration of any given exercise, the frequency of performance, and how many different exercises are given (Table 23-4) can modify the intensity of the exercise program to improve gaze stability. Patients will find the exercises more challenging if they have to perform them while standing as opposed to sitting. Exactly which exercises are given initially and the progression itself will depend on the individual patient. Concurrent with the exercises to improve gaze stability, of course, the patient should be instructed in exercises to improve postural stability. Again, the initial exercise program and the rate of progression should be customized for each patient.

■ Table 23-3 **MEAN (5D) IN OUTCOME MEASURES FOR THE GROUP AT BASELINE AND AT DISCHARGE**

Outcome Measures	oVAS	dysVAS	dzVAS	ABC	% of Time Symptoms Interfere	Disability Score	DVA (LogMAR)	Gait Speed (m/sec)	DGI	CDP
# of pre-post comparisons	28	35	26	32	16	23	23	31	47	15
Baseline	2.7 ± 2.8	4.2 ± 2.2	3.1 ± 2.1	49.4 ± 20.5	72.7 ± 27.0	2.9 ± 1.1	0.323 ± 0.107	0.69 ± 0.22	13.5 ± 3.6	42.1 ± 9.9
Discharge	1.1 ± 1.5	2.1 ± 2.2	1.7 ± 2.5	68.3 ± 18.7	31.4 ± 24.9	2.1 ± 1.4	0.195 ± 0.102	0.84 ± 0.18	18.1 ± 3.0	53.1 ± 8.8
Pre-post paired comparison: p value	0.006 (NS)	0.00002	0.0004	0.00004	0.0001	0.006 (NS)	0.00001	0.00001	0.00001	.00003
% of subjects with significant improvement	42.9% (n=12)	45.7% (n=16)	3.9% (n=1)	56.3% (n=18)	81.3% (n=13)	43.5% (n=10)	73.9% (n=17)	54.8% (n=17)	61.7% (n=29)	6.7% (n=1)
% subjects who returned to normal	32.1% (n=9) (score < 0.2)	17.1% (n=6) (score < 0.2)	34.6% (n=9) (score < 0.2)	28.1% (n=9)	6.3% (n=1)	13.0% (n=3)	52.2% (n=12)	25.8% (n=8)	36.2% (n=17)	6.7% (n=1)
Total subjects who improved or returned to normal	53.6% (n=15)	45.7% (n=16)	38.5% (n=10)	62.5% (n=20)	81.3% (n=13)	47.8% (n=11)	78.3% (n=18)	61.3% (n=19)	70.2% (n=33)	6.7% (n=1)
% of subjects who became worse	21.4%	28.6%	19.2%	6.3%	18.8%	8.7%	0%	3.2%	0%	0%

(Modified from Herdman et al, 2013.[10])

Box 23-3

EXERCISES TO IMPROVE GAZE STABILITY

1. To improve remaining vestibular function and central preprogramming
   - Tape a business card on the wall in front of you so that you can read it.
   - Move your head back and forth sideways, keeping the words in focus.
   - Move your head faster but keep the words in focus. Continue to do this for 1 to 2 minutes without stopping.
   - Repeat the exercise moving your head up and down.
   - Repeat the exercises using a large pattern such as a checkerboard (full-field stimulus).
     - *Note:* When training the patient to perform this exercise, watch the eyes closely. If the patient is making corrective saccades, the patient should slow the head movement down.
2. Active eye-head movements between two targets to foster the use of saccadic or pursuit strategies and central preprogramming.

   Horizontal targets:
   - Look directly at one target being sure that your head is also lined up with the target.
   - Look at the other target with your eyes and then turn your head to the target (saccades should precede head movement). Be sure to keep the target in focus during the head movement.
   - Repeat in the opposite direction.
   - Vary the speed of the head movement but always keep the targets in focus.
     - Note: Place the two targets close enough together so that when you are looking directly at one, you can see the other with your peripheral vision. Practice for 5 minutes, resting if necessary. This exercise can also be performed with two vertically placed targets.
3. Visualization of remembered targets to foster central preprogramming:
   - Look at a target directly in front of you.
   - Close your eyes and turn your head slightly, imagining that you are still looking directly at the target.
   - Open your eyes and check to see if you have been able to keep your eyes on the target.
   - Repeat in the opposite direction. Be as accurate as possible.
   - Vary the speed on the head movement.
   - Practice for up to 5 minutes, resting if necessary.

Box 23-4

EXERCISES TO IMPROVE POSTURAL STABILITY

The purpose of these exercises is to force you to develop strategies of performing daily activities even when deprived of vision, proprioception, or normal vestibular inputs. The activities are supposed to help you develop confidence and establish your functional limits. On all these exercises you should take extra precautions so you do not fall.

_____ 1. Stand with your feet as close together as possible with both hands helping you maintain your balance by touching a wall. Take your hand or hands off the wall for longer and longer periods of time while maintaining your balance. Try moving your feet even closer together. Repeat this for _____ minutes twice each day.

_____Repeat exercise #1 with eyes closed, at first intermittently and then continuously, all the while making a special effort to mentally visualize your surroundings.

_____ 2. Stand with your feet shoulder width apart with eyes open, looking straight ahead at a target on the wall. Progressively narrow your base of support from:

feet apart to

feet together to

a semi–heel-to-toe position to

heel almost directly in front of the toes

Note: change your foot position one inch at a time.

Continued

Box 23-4

EXERCISES TO IMPROVE POSTURAL STABILITY—cont'd

Do the exercise first

_____with arms outstretched and then

_____with arms close to your body and then

_____with arms folded across your chest.

Hold each position for 15 seconds and then move on to the next most difficult exercise.

_____ Repeat exercise #2 with eyes closed, at first intermittently and then continuously, all the while making a special effort to mentally visualize your surroundings.

Repeat # _____ above but while standing on a foam pillow

(Note: unusual for patients with BVL).

_____ 3. Walk close to a wall with your hand braced available for balancing.

Walk with a narrower base of support.

Finally, walk heel-to-toe.

Do this with eyes _____ (open/closed).

Practice for _____ minutes.

_____ 4. Walk close to a wall and turn your head to the right and to the left as you walk. Try to focus on different objects as you walk.

Gradually turn your head more often and faster.

Practice for _____ minutes.

_____ 5. Walk and turn your head to the right and to the left as you walk while you count backwards out loud from 100.

Try to focus on different objects as you walk.

Gradually turn your head faster.

Practice for _____ minutes.

Practice turning around while you walk. At first, turn in a large circle but gradually make smaller and smaller turns.

Be sure to turn in both directions.

Box 23-5

ADJUSTMENTS TO EXERCISES BECAUSE OF SEVERE DIZZINESS

- Decrease the number of times they perform the exercises each day.
- Move the head more slowly.
- Perform each exercise for a shorter period of time.
- Rest longer between each exercise.

Outcome and Expectations

Several studies report factors that might affect recovery in patients with BVH. Gillespie and Minor (1999) found that patients with poor outcome had a greater number of comorbidities and lower gain and shorter time constant on rotary chair testing than did patients with better outcomes.[2] Krebs et al (2003) noted that some patients with BVH developed depression that prevented participation in vestibular rehabilitation, which would affect outcome.[4] Herdman et al (2007) found that age and time from onset did not affect

■ Table 23-4 **SUGGESTED PROGRESSION FOR GAZE STABILITY EXERCISES**

Exercise	Duration	Frequency	Position
X1 viewing paradigm against plain stationary background; horizontal or horizontal and vertical head movement	Maybe for less than 1 minute each time	Two or three times daily	Sitting until can perform the head movements easily and then standing
X1 viewing paradigm against plain stationary background; horizontal or horizontal and vertical head movement	Increase to 1 minute each exercise	Increase to five times daily	Standing[a]

■ Table 23-4 SUGGESTED PROGRESSION FOR GAZE STABILITY EXERCISES—cont'd

Exercise	Duration	Frequency	Position
X1 with target held in hand against a plain background; horizontal and then vertical head movements	1 minute each exercise	Up to five times daily	Standing[a]
Add eye-head exercises, horizontally and vertically	No specific duration	Two to three times daily	Sitting at first and then standing
X1 with target held in hand and also with target at distance	1 minute each exercise	Two or three times daily	Standing[a]
Eye-head exercises, horizontally and vertically	No specific duration	Two or three times daily	Standing[a] if possible
X1 with target held in hand and also with target at distance	1 minute each exercise	Four times daily	Standing[a]
Eye-head exercises, horizontally and vertically	No specific duration	Four times daily	Standing[a]
Add imaginary target exercise	No specific duration	Four times daily	Sitting at first and then standing
X1 with target held in hand and also with target at distance	1 minute each exercise	Four times daily	Standing[a]
Eye-head exercises, horizontally and vertically	No specific duration	Four times daily	Standing[a]
Imaginary target exercise	No specific duration	Four times daily	Standing[a]
Some patients may be able to progress to X2 with target held in hand and also with target at distance	1 minute each exercise	Four times daily	Standing[a]
Eye-head exercises, horizontally and vertically	No specific duration	Four times daily	Standing[a]
Imaginary target exercise	No specific duration	Four times daily	Standing[a]
X1 with target held in hand and also with target at distance	1 minute each exercise	Four times daily	Standing[a]
Eye-head exercises, horizontally and vertically	No specific duration	Four times daily	Standing[a]
Imaginary target exercise	No specific duration	Four times daily	Standing[a]
Add finding numbers written randomly on large (6 ft by 5 ft) checkerboard pattern placed on wall	No specific duration	Twice daily	Stand and step to touch number

[a]The exercise can be made more difficult by changing the base of support (e.g., from feet apart to feet together).

recovery of visual acuity during head movement.[5] A more comprehensive study examined the relationship of patient characteristics, subjective complaints, and physical function to outcome.[10] In agreement with other studies, they found that age did not contribute to whether a patient improved or returned to normal—older patients were just as likely as younger patients to improve. Several specific comorbidities had fair to strong correlations with certain outcome measures. For example, a history of headaches was correlated with a higher percentage of time symptoms interfered with activities, with lower balance confidence and a greater intensity of head movement–induced dizziness at discharge. When lower extremity sensory deficits or Ménière's disease were present, patients walked more slowly at discharge. In addition to the presence of specific comorbidities correlating with outcome, the number of comorbidities affected outcome such that the greater the number of comorbidities, the worse the disability at discharge. Another important finding was that the longer the time from onset until the initiation of vestibular rehabilitation, the poorer the balance confidence at discharge. Loss of both superior and inferior nerve function was correlated with worse complaints of oscillopsia at discharge. Higher intensity of disequilibrium initially was correlated with worse balance confidence, poorer disability, and higher percentage of time symptoms interfered with activities at discharge. Similarly, poor DGI score initially correlated with lower balance confidence, poorer disability scores, and poorer the DGI scores at discharge.

How do you use this information? One way is to first identify your patient's primary goal(s). For example, if the patient states that his or her main problem is that the symptoms (dizziness with head movement, oscillopsia, disequilibrium) interfere with both leisure and work activities and that he or she would like to enjoy these activities more, then the clinician can look at the initial measures of disequilibrium and fall risk (DGI) scores as a guide as to what to expect for outcome. Values that initially are closer to normal would suggest that the patient should improve in the primary goal.

Comparison with Patients with Chronic Unilateral Vestibular Hypofunction (UVH)

One of the surprising findings is that patients with BVH and patients with UVH have similar results on initial assessment (Table 23-5).[10,54] The only differences appear to be that patients with BVH walk more slowly and have a greater percentage of time that symptoms interfere with activities. The primary difference is in the level of recovery reached by the two groups (Table 23-6). For most measures of subjective complaints, a smaller percentage of patients with BVH improve significantly compared with patients with UVH.

Guidelines to Treatment and Prognosis

There are several factors to remember when working with patients with bilateral vestibular deficits:

1. The patient's perception of dizziness can be the major deterrent to the patient's return to normal activities.
2. Recovery following bilateral deficits is slower than for unilateral lesions and can continue to occur over a 2-year period,
3. Recovery is easily upset by other medical problems such as having a cold or receiving chemotherapy.

■ Table 23-5 **COMPARISON OF RESULTS FROM INITIAL ASSESSMENTS IN ALL PATIENTS WITH UVH AND BVH**

Outcome measures	oVAS	dysVAS	DRS	% TSI	ABC	GS	DGI	DVA
UVH: Baseline score	3.1 ± 2.7	3.97 ± 2.5	2.9 ± 0.95	59 ± 29.8	51.5 ± 16.9	0.80 ± 0.15	14.2 ± 4.0	.327 ± 0.134
BVH: Baseline score	3.0 ± 2.2	4.2 ± 2.2	2.9 ± 1.1	72.7 ± 27.0	49.4 ± 20.5	0.69 ± 0.22	13.5 ± 3.6	.323 ± 0.107

(From Herdman et al, 2013.[10])

■ Table 23-6 COMPARISON OF OUTCOMES FOR PATIENTS WITH BVH AND UVH

Outcome Measures	oVAS	dysVAS	DRS	% TSI	ABC	GS	DGI	DVA
UVH: discharge	0.79 ± 1.65	1.58 ± 1.91	1.39 ± 1.4	24 ± 30.1	78.5 ± 16.0	1.01 ± 0.17	19.8 ± 3.0	0.210 ± 0.138
UVH: % improved significantly	78%	81%	75%	80%	78%	85%	88%	78%
BVH: discharge	1.1 ± 1.5	2.1 ± 2.2	2.1 ± 1.4	31.4 ± 24.9	68.3 ± 18.7	0.84 ± 0.18	18.1 ± 3.0	0.195 ± 0.102
BVH: % improved significantly	32.1%	17.1%	43.5%	81.3%	81.3%	61.3%	70.2%	78.3%

(From Herdman et al, 2013.[10])

4. To maintain recovered function, patients may always need to be doing some exercises, at least intermittently.
5. Postural stability will never be completely normal. The patient may have a negative Romberg and may be able to maintain the Sharpened Romberg position with eyes open, but not with eyes closed.
6. Initially, the patient may need to use a cane or a walker while ambulating. Some patients, especially older patients, need to use a cane at least some of the time. Most patients, however, are eventually able to walk without any assistive devices. Ambulation during the acute stage will be wide-based and ataxic with shortened stride length and side-stepping to the right and left. The patient will turn en bloc, and turning the head will cause increased instability. Ambulation will improve, but again, it will not be normal.
7. Patients will be at increased risk for falls when walking in low-vision situations, over uneven surfaces, or when they are fatigued.

Future Treatment

Various devices and technologies, such as auditory cues, tactile cues applied to the torso, and stimulation of the tongue (see Chapter 28: The Role of Emerging Technologies in Vestibular Rehabilitation for a detailed review) have been employed in an attempt to replace the lost vestibular function. Although these devices have shown potential benefits, the devices are solely focused on improving postural control, not on improving gaze stability during head movements. With the success of cochlear implants, several labs have been working on developing a vestibular prosthesis. The basic design is that sensors will detect and measure the directions of rotation and then electrically stimulate the appropriate ampullary nerves. The majority of the work to date has been performed in bilaterally vestibular–deficient chinchillas and monkeys. Della Santina and colleagues have demonstrated partial restoration of the angular VOR to horizontal, as well as RALP (right-anterior, left-posterior) and LARP (left-anterior, right-posterior) rotations with unilateral implantation of the vestibular prosthesis in chinchillas treated with gentamicin.[55] In addition to the angular VOR improvements, they noted improved postural stability in the animals as well. Similar improvements in angular VOR responses were observed in rhesus monkeys with bilateral vestibular deficits after unilateral implantation of the vestibular prosthesis.[56] Even though the responses were not normal, the responses were strong and had similar dynamics to normal animals, and the implanted animals had not undergone any adaptation treatments with the vestibular prosthesis. These studies bode well for future implantation of a vestibular prosthesis in humans with bilateral vestibular hypofunction, which may, in combination with rehabilitation, lead to improved function and decreased disability.

CASE STUDY 23-1

A 74-year-old woman with a history of imbalance is referred for treatment. Four months before this clinic visit, she had a chronic bladder infection that had not responded to other antibiotics and was placed on IV gentamicin at home for 10 days. This effectively treated her bladder infection and she returned to all normal activities. Two months ago, she had another bladder infection and was again treated with IV gentamicin. She began to complain of imbalance 2 days after the last dose of gentamicin. There is no history of renal failure, nor was she on other antibiotics or a loop diuretic at that time. She had no complaints of hearing loss, tinnitus, or vertigo. Her balance was severely affected, and she is now using a walker. She says she is unable to walk independently. Standing, walking, and moving her head exacerbate her balance problem.

Comment

Her history suggests an ototoxic reaction to gentamicin. Fewer than 15% of all individuals treated with gentamicin develop a BVH, and she had no known risk factors that would increase the likelihood of an ototoxic reaction such as renal failure or being on a loop diuretic, but was over 65 years old.

Pertinent History

Past Medical History: Osteoarthritis, hypertension, hypothyroidism. She has been treated for depression and anxiety. Current medications include Lisinopril, Synthroid, Zoloft, Valium.

Neurological Examination

Normal except for: visual acuity of 20/20-3 OU with head stationary that increases to 20/80-4 during 2-Hz head oscillations (clinical DVA test); patient makes corrective saccades with slow and rapid head thrusts bilaterally.

Comment

The large decrease in visual acuity during head movement (greater than 4 lines) is consistent with a severe vestibular deficit. The presence of corrective saccades with slow head rotations and with rapid head thrusts also suggests a profound deficit. She does not appear to have compensated at this time based on the positive Romberg test and the poor VOR during slow head rotations. Her gait pattern may reflect fear as well as the vestibular deficit. Her history of depression and the number of comorbidities may be a factor in the final level of recovery.[4]

Caloric Test

No response to either cool or warm irrigation of either ear (Fig. 23.3). Ice water irrigation of both the right and left ears resulted in nystagmus with peak slow-phase eye velocities of 4 deg/sec. The direction of the nystagmus reversed when the patient was moved from supine to prone.

Comment

Ice water is a stronger stimulus than either cool or warm water. The nystagmus generated by ice water irrigation may represent either a response of the peripheral vestibular system or an alerting response to the extreme cold. If the nystagmus were because of excitation of the hair cells in the inner ear, the direction of the nystagmus would reverse when the patient is moved from supine to prone because the direction of endolymph flow would reverse. If the nystagmus were because of an alerting response, it would not reverse when the patient was moved from supine to prone. The test results for this patient suggest there is some residual function in each ear.

Vestibular Rehabilitation

The patient was seen 1 week later to institute a vestibular rehabilitation program. She was still using a walker. She reported that her imbalance occurred with movement, especially while walking, and was worse in the dark. She denied any falls but reported that she had numerous near falls daily. She also reported staggering and side-stepping and tends to drift to both the right and the left when she tried to walk without her walker.

Social History

She lived with her husband. Her home had no stairs. She did not smoke or use alcohol.

Functional Level

She was independent in self-care activities and enjoyed gardening, playing cards, and church. She was no longer driving. She has been inactive since the onset of this problem.

CASE STUDY 23-1

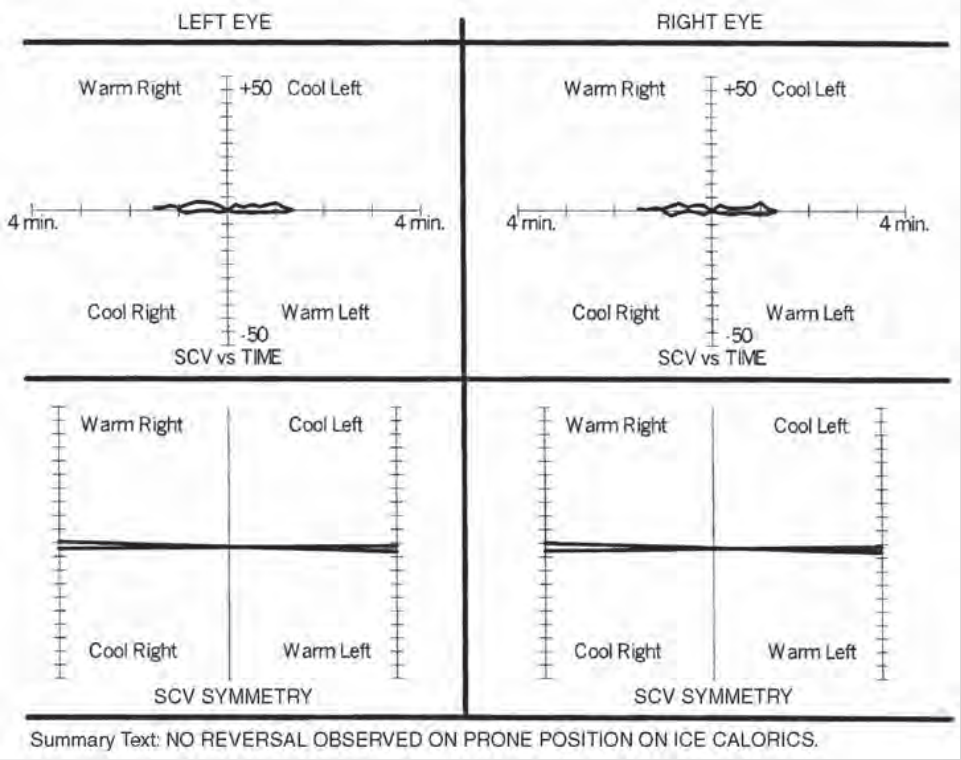

Figure 23.3 Results from bithermal caloric irrigation of the external auditory canals in a patient with profound bilateral vestibular hypofunction. SCV = Slow component velocity.

Subjective Complaints

She stated her symptoms disrupted her performance of both her usual work and outside activities (rated a 3 on the Disability Scale). Her dizziness interferes with her activities 80% of the time. She scored a 56 on Jacobson's Dizziness Handicap Inventory, indicating that she would not go for a walk by herself, could not walk in the dark, and was limited in her ability to travel or participate in social activities. She also reported feeling frustrated, embarrassed in front of others, and handicapped. Her balance confidence score (ABC) was a 67%. She rated her oscillopsia as a 5.7, her disequilibrium as a 9.3, and her head movement–induced dizziness as a 7.8 on an analogue scale of 0 to 10 (10 worse).

Quantitative Dynamic Visual Acuity

Her visual acuity during 120 deg/sec horizontal head movements was LogMAR = 0.450 to the right and left (approximately 20/50-3; normal dynamic visual acuity for age LogMAR = 0.258 based on mean + 2 standard deviations for healthy subjects).[31]

Gait

She has a positive Romberg. Gait is extremely slow and cautious without the walker. She stops frequently and cannot turn around without stopping. When asked to walk without the walker, her cadence appeared to be slow, and she had a variable base of support. Her step length was decreased but symmetrical. Arm swing and head and trunk rotation were markedly decreased, especially when she had to turn around. She could not walk a straight path and tended to reach toward the hallway walls as she walked. Her gait pattern was affected by her scoliosis (arm swing asymmetry). When asked to try to turn her head horizontally while walking, her cadence slowed significantly, and she side-stepped occasionally. She could not step over a shoebox without stopping and even then almost lost her balance. Her Dynamic Gait Index (DGI) score is a 14/24. Gait speed is 3.12 ft/sec (normal for age and gender is 2.79 ft/sec.[57]

Comment

She does not appear to have compensated at this time based on the positive Romberg test and the poor VOR

Continued

CASE STUDY 23-1

during slow head rotations. Her gait pattern may reflect fear of falling as well as the vestibular deficit. The patient's Disability Scale score and her Dizziness Handicap Inventory score both indicate a moderate perception of disability/handicap. Shepard et al [58] showed that patients who score a 5 on the Disability Scale are less likely to show improvement with vestibular rehabilitation than those who rate themselves as a 3 or lower, as she did (see Chapter 21). Although she has been using a walker, she was able to walk without the walker. Head movements (turning her head or turning around) both increased her instability. Her dynamic visual acuity was consistent with other patients with bilateral vestibular loss.

Goals

The short-term goals for this patient were (1) to perform the vestibular exercises without a significant increase in her symptoms, (2) to walk daily for 20 minutes, and (3) to no longer use her walker or touch walls or furniture in her home. The long-term goals were (1) to return to all normal activities except possibly driving, (2) reduce her subjective complaints by 30%, (3) improve her balance confidence to 65%, (4) to improve her gait speed to 2.9 ft/sec, (5) to reduce her fall risk as indicated by a DGI score of 18/24, and (6) to improve her visual acuity during head movements by one line (by LogMAR 0.100) on the quantitative DVA test and to 20/50 on the clinical DVA test.

Plan

The patient was placed on a home exercise program, which included the X1 viewing paradigm to be performed with both horizontal and vertical head movements for 1 minute each four times a day. The target was to be placed against a plain wall. Initially, she was to perform this exercise seated and then was to perform the exercise standing with her feet apart. She was also instructed to practice walking in a hallway at least twice daily for 5 minutes each time without touching the walls. Finally, she was told to walk for 20 minutes daily outside with her husband. She was given a calendar to fill in to help ensure compliance. The total duration of her exercises at this point was 36 minutes. The program was limited until the patient's response could be determined. The patient was seen at 1-week intervals.

On the first follow-up, the patient was doing well with the exercises and was no longer using her walker at all. Her exercise program was changed to include the eye-head exercises both vertically and horizontally, and the X1 paradigm using a near target held in the hand was added to her program. She was to practice walking and turning her head horizontally for 5 minutes, twice daily, resting as needed because the head movement would likely make her feel more dizzy. She was also instructed in a static balance exercise in which she would stand while gradually decreasing her base of support. This was to be performed with eyes open and then eyes closed. She was to continue walking for 20 minutes daily. Total exercise time was increased to 45 minutes.

On the next follow-up visit, the patient reported that she initially had difficulty with the exercises involving head movements, all of which increased her dizziness. However, she reported that after 2 days of performing the exercises, she was able to perform them without a significant increase in dizziness. Review of the exercises showed that she was performing them all correctly and was maintaining fixation on the X1 viewing paradigms, although her head movements were slow. Her exercise program was modified to include performing the X1 paradigm using a target on a checkerboard, and the "remembered target" exercise was added. For these exercises, she was instructed to attempt to move her head more rapidly while maintaining focus on the target. She was also instructed to add walking and moving her head vertically. The X1 viewing paradigm using a target placed against the wall was discontinued. Eventually, in her program, she was instructed in the X2 viewing paradigm.

One month after the initiation of her exercise program, the patient was walking with an increased cadence and a more narrow base of support. She still had difficulty walking a straight line, but she was able to turn her head while walking and turn around without stopping. She no longer used a walker at any time. Her rating of her subjective complaints of disequilibrium was a 0.6 (down from 9.3) and of oscillopsia was a 1.5 (down from 5.7). Her quantitative DVA score was LogMAR 0.330 (initial DVA was LogMAR 0.450), a one-line improvement. The plan was for the patient to continue with the rehabilitation process to further improve her gait and to enable her to return to more of her normal activities. A 6-month follow-up visit was scheduled.

CASE STUDY 23-2

A 34-year-old woman with a history of diabetes and renal failure has been on peritoneal dialysis for 1 1/2 years. She had been treated with gentamicin 9 and 6 months ago for peritonitis. She had no complaints of disequilibrium after either of those drug courses. Two months later she again developed peritonitis and again received IV gentamicin. After a few days, she complained of vertigo and tinnitus, developed disequilibrium, and could not walk unassisted. She also complained that she was not able to see clearly when her head was moving. She was admitted to the hospital for a workup of her vertigo and disequilibrium.

Clinical Examination

Significant findings on clinical examination included a spontaneous nystagmus with fast component to the left, poor VOR to slow head movements bilaterally, and large corrective saccades with rapid head movements bilaterally, worse with head movements to the right than to the left. The test for head-shaking nystagmus was not performed because the patient complained of severe nausea following even gentle head movements and vomited. She also had a positive Romberg test. The Sharpened Romberg and Fukuda's Stepping Tests were not performed. The patient could not ambulate without the assistance of two people.

Comment

This patient's signs and symptoms (vertigo, disequilibrium, oscillopsia, spontaneous nystagmus, and poor VOR) certainly were suggestive of a vestibular disorder. Furthermore, her history included multiple treatments with gentamicin, an ototoxic medication. With bilateral dysfunction from treatment with an ototoxic drug, the symptoms of oscillopsia and disequilibrium develop over time and may not appear until after the drug treatment is finished. Once the symptoms appear, they may continue to become worse for several weeks. With some patients, there is a partial reversal of symptoms with time. Depending on the aminoglycoside used, vestibular symptoms may be accompanied by hearing loss. Gentamicin has primarily vestibulotoxic effects. It is unusual for systemic gentamicin to result in vertigo and nystagmus, because that indicates asymmetrical vestibular function. Although gentamicin usually results in BVL, symptoms of asymmetric effects on the vestibular and auditory systems have been reported.[6] Her poor VOR to slow head movements and the presence of corrective saccades during rapid head thrusts bilaterally suggested a bilateral vestibular deficit, which was confirmed by the rotational test results. This patient's gait disturbance appeared to be unusually severe, and further testing showed a moderate loss of proprioceptive and kinesthetic perception in her feet, which would contribute significantly to her problem. This sensory loss was probably caused by her diabetes. This finding was particularly important in developing her exercise program and in predicting her final level of recovery.

Caloric tests showed a poor response bilaterally to warm or cool water, although ice water in the left ear did result in a weak but appropriate response. There was a directional preponderance to the right. Rotational chair test showed a severe bilateral vestibular deficit. There was little optokinetic after-nystagmus and the VOR Tc was 2.4 seconds to 60 deg/sec step rotations. At 240 deg/sec step rotation, some vestibular response was evident—the gain of the response was 0.15, and the Tc was 2.4 seconds (Fig. 23.4).

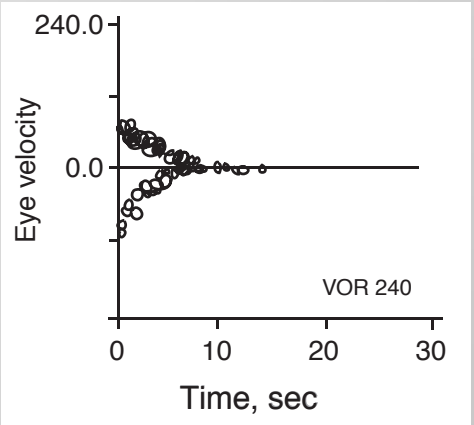

Figure 23.4 Plot of the decay in slow-phase eye velocity with time during VOR after nystagmus. Patient is first rotated in a chair in complete darkness for 2 minutes, and then the chair is stopped. The slow-phase eye movements that occur are caused by the discharge of the velocity storage system and represent the function of the vestibular system. These results show the poor peak slow-phase eye velocity (35 deg/sec) and the short time constant (less than 2 sec) in a patient with bilateral vestibular loss. A step rotation at 240 deg/sec was used. Eye movements were recorded using electro-oculography.

Continued

CASE STUDY 23-2

Treatment

At this point, the patient was started on a vestibular rehabilitation program. She performed the X1 viewing paradigm exercise (see Box 23-3) first with horizontal head movements for 1 minute and then vertical head movements for 1 minute. Because head movement exacerbated her nausea, she rested for 10 minutes or more between each of these exercises. Initially, she performed these exercises while sitting, up to five times a day. She also practiced standing unsupported, first with her feet apart and her eyes open and then gradually moving her feet together and briefly closing her eyes by blinking and then gradually keeping her eyes closed for longer periods of time. She was instructed on how to use a walker, and an emphasis was placed on increasing her endurance. Initially, she needed contact guarding while using the walker and would occasionally lose her balance, especially when trying to turn or if she moved her head too quickly. After 4 days, she was able to walk independently with the walker and was discharged from the hospital. At that time, she no longer had nausea with gentle head movements.

Comments

Although this patient had a bilateral vestibular deficit, the caloric and rotary chair tests showed that she had remaining vestibular function (response to ice water caloric on the left and the responses to 60 deg/sec and 240 deg/sec step rotations). Her initial exercise program, consisted of vestibular adaptation exercises and ambulation training. Her balance exercises were designed to gradually increase the difficulty of maintaining balance by slowly decreasing her base of support, changing her arm positions (arms out, arms at side, arms across the chest) and then altering her use of visual cues. Although she had decreased sensation in her feet, subtracting visual cues was used as a treatment approach to facilitate her ability to use the remaining somatosensory and vestibular cues.

Follow-up

The patient continued to be followed as an outpatient. Exercises designed to facilitate the substitution of alternative strategies to maintain gaze stability as well to improve her static and dynamic balance were added to her program. The patient no longer needed to use a walker, but she had a wide-based gait and used momentum to walk a straight path down a hallway. She had to stop walking before turning around or she would lose her balance. She had a negative Romberg but could not perform a Sharpened Romberg. Although her vision improved and she could read if she was sitting quietly, she could not see clearly while in a car and had not resumed driving. Approximately 2 months later, the patient had a retinal hemorrhage in her left eye. She already had retinal damage in the right eye from her diabetes which essentially meant that she had only partial visual, vestibular, and somatosensory cues for balance and, as a result, she could no longer keep her balance even in well-lighted conditions. For 1 week, she either used a wheelchair or, at home, a walker. Fortunately, her vision recovered, and she was again able to walk independently. On her last visit she reported that she had returned to most activities except driving. Her base of support while walking was more narrow, and her stability while turning had improved. Her Romberg was clinically normal, but she could not perform a Sharpened Romberg with eyes open. She was seeking part-time employment and was waiting for a kidney transplant.

CASE STUDY 23-3

Your patient is a 73-year-old man who was seen by a neurologist for balance problems that had become worse over the past 5 years. Past medical history was positive for high cholesterol, a mild bilateral sensorineural hearing loss (Fig. 23.5), and mild osteoarthritis primarily affecting his knees and back. History was negative for ototoxic medications, episodes of vertigo, and head trauma. Current medications include lisinopril, celecoxid, and dulocet. His neurological examination was normal for his age except for positive head impulse tests bilaterally, abnormal gait with a widened base of support, and an inability to stand with feet together with eyes closed. Vestibular function tests: no response to ice water irrigation bilaterally, VOR gain is less than 0.05 with rotation to the right and 0.09 with rotation to the left bilaterally, and Tc is less than 1 second bilaterally to 60 deg/sec step rotation, and there is low gain and high phase at all frequencies on sinusoidal rotational chair testing, absent cervical VEMPs

CASE STUDY 23-3

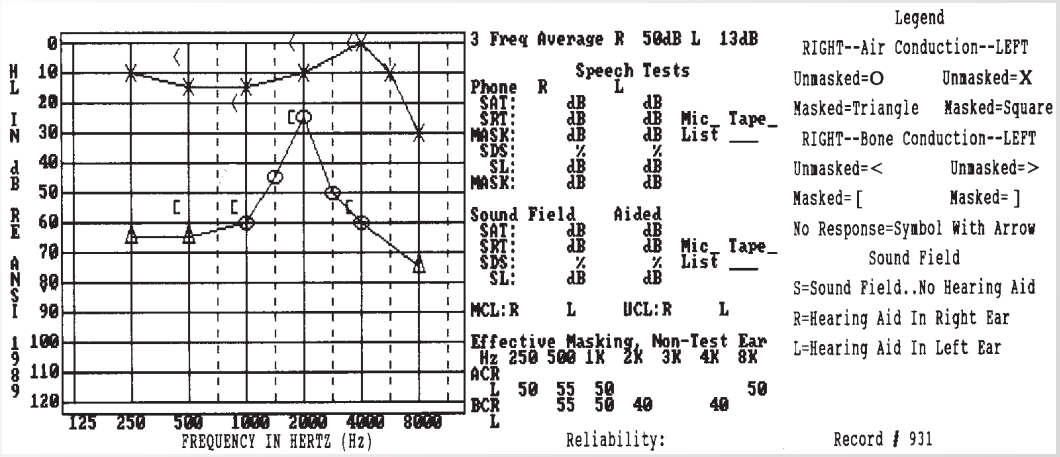

Figure 23.5 Hearing test results from patient with gentamicin ototoxicity. Circles and triangles indicate right ear, and Xs and squares indicate left ear, respectively. Note asymmetry in hearing loss for this patient.

bilaterally. His diagnosis w as progressive bilateral vestibular loss.

Comment

The patient has involvement in the distribution of both the superior and inferior branches of the v estibular nerve with little evidence of any residual function.

Rehabilitation

On his first visit for therapy, he reported that he lives with his wife and that her brother , who is disabled, lives with them. They live in a one-story home with four steps to enter the house with no railing.The laundry room is in the basement. He is retired b ut has a part-time job that requires driving. He is independent in his activities of daily living and still drives day and night but avoids night driving if possible. The only activity he has stopped is ballroom dancing. The patient reports that he staggers when he walks and is worried that people will think he is drunk. Therefore, his primary goal is to walk without staggering.

Subjective Complaints

PANAS is normal for depression and anxiety. He rates his visual blurring while seated as a 0.4/10 and while walking as a 2.2/10; his disequilibrium while seated is 0.4/10 and while walking is 4.6/10. His balance confidence is 72%. Symptoms interfere with activities 10% of the time. His disability score is a 3/5.

Balance and Gait

Romberg with eyes open can be maintained for 30 seconds and with eyes closed for 3 seconds; single le g stance time with e yes open is 3 seconds, and he cannot maintain his balance with e yes closed; gait speed is 3.79 ft/sec (normal for age and gender is 3.08 ft/sec) and his DGI score is 14/24. He slows his gait and staggers when moving his head horizontally or vertically; he has to take several steps to regain his balance when he turns 180 degrees and stops walking; he is unable to step over a shoebox without loss of balance, and he goes up and down stairs putting two feet to a stair.

Visual acuity during head movement is LogMAR 0.458 for head mo vement to the right and 0.420 for head movement to the left (normal for age is LogMAR 0.258 based on mean + 2 standard deviation of healthy subjects).[31]

Treatment

The goals for this patient include decreasing his per - ception of oscillopsia and disequilibrium by at least 10% in four weeks; decrease his f all risk based on his single leg stance time and his DGI score; that he can move his head while walking without losing his balance or staggering; that he can step o ver low objects safely; and to improve his visual acuity during head movement. Exercises were aimed at decreasing head mo vement–induced symptoms, improving gaze stability, and improving his balance especially while walking. He is able

Continued

CASE STUDY 23-3

to perform the X1 viewing exercise while standing, so his exercises start at that level, five times a day. He also starts standing balance exercises with eyes open and closed, practicing walking with a more narrow base of support twice daily, and walking outside for 20 minutes daily. In addition, several suggestions were made to the patient concerning modifications in his home to ensure safety, including emergency lighting that would come on automatically if there were a power failure, railings for all stairways, and safety bars in the bathroom.

Progress

In the first week, his oscillopsia and disequilibrium intensity scores (VASs) were worse. This may have simply been because of the increase in his head movements with the exercises and did not persist past the first week. Some modifications to his exercise progression were necessary. For example, he had pain in his back when standing and performing the X1 viewing exercises with his feet together, so he performed them with his feet apart. We were able to add the eye-head and remembered target exercises and to challenge his balance when walking with head movements and eventually head movements while counting backwards. He was able to return to some of the ballroom dancing he enjoyed with his wife, noting that the jitterbug was difficult but the waltz was fine. He fell at one point while dressing standing up and was counseled to sit down to take off his shorts. He showed improvement in his subjective complaints, gait speed, and fall risk scores. At 6 weeks, his brother-in-law became suddenly worse, and he was helping with his care. During that time, all his test results became worse. Once he did not need to help his brother-in-law any more, he felt better and his scores again approached normal. He was fitted with an offset straight cane with a light imbedded in it for his use at night (Pathlight®) and reported that he found it useful when he went to the movies.

Summary

Studies have shown that the worse disability is in part dependent on the number of comorbidities the patient has as well as greater initial intensity of disequilibrium, lower balance confidence, and poor fall risk scores. Patients with bilateral vestibular problems can be expected to return to many activities but will continue to have problems in certain areas. They will be able to ambulate without the use of a cane or walker, at least when they are in well-lighted environments.[59] Patients should be able to return to work, but it may be necessary to find a different occupation. Patients often report difficulty driving, particularly in the rain or snow or at night or on high-speed roads, but actual driving habits and accidents do not differ from subjects with normal vestibular function.[60,61] Treatment approaches include increasing the function of the remaining vestibular system, inducing the substitution of alternative mechanisms to maintain gaze stability and postural stability during head movements, and modifications of the home and working environment for safety.

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CHAPTER 24

Physical Therapy Management of Children with Vestibular Dysfunction

Rose Marie Rine, PT, PhD ■ *Jennifer Braswell Christy, PT, PhD*

The vestibular system develops relatively early in gestation.[1-4] The vestibular system is composed of vestibulo-ocular and vestibulospinal subsystems, each with distinct tasks and each requires different methods of assessment. Labyrinthine reflexes, which are mediated by the vestibulospinal system, provide postural tone necessary for the emergence of early motor milestones (e.g., rolling, sitting, standing).[5-7] The role of the vestibulo-ocular system in visual stabilization, acuity, and the development of visual spatial and perception abilities has been well documented.[8-11] It is therefore logical to deduce that individuals with deficits of the vestibular system since birth or very early in life will present with difficulties in either motor or visuospatial abilities or both.[12,13]

The functional integrity of the vestibular system is rarely tested in young children, and thus impairments can be undetected and untreated. This may be in part a result of the child's inability to describe symptoms or even know that what they are experiencing is not "normal."[14,15] One of the major difficulties in the identification of vestibular dysfunction in children has been the unavailability or the omission of the use of traditional and more recently developed tests of vestibular function with this population. This chapter will provide a review of the types and incidence of vestibular deficits in young children, related impairments, the development of postural control and oculomotor abilities, and appropriate testing and intervention for this population.

Incidence of Vestibular Deficits in Children and Related Functional Impairments

Common Diagnoses of Vestibular Dysfunction and the Etiology of Vertigo in Children

In the last decade, there is increased evidence in the literature of vestibular dysfunction or vertigo of various etiologies and consequent functional impairments in children (Table 24-1).[15-24] Lesions of the vestibular system may be peripheral (either unilateral or bilateral) or central in nature. Each has differing signs and symptoms that can change from the acute to chronic stage.

Vertigo

Vertigo is typical of acute unilateral peripheral vestibular hypofunction but is not common in bilateral peripheral deficits. Furthermore, symptoms can be acute, progressive, or chronic. The most comprehensive study completed to

■ Table 24-1 REPORTED ETIOLOGY OF VERTIGO IN CHILDREN

	AUTHORS				
	Choung et al[16]	Erbek et al [17]	Wiener-Vacher [15]	Riina et al[25]	Gruber et al[26]
Causes of Vertigo					
Otitis media	—	—	4.0%	10%	—
Migraine	30.9%	34.0%	25%	14.0%	32%
BPVC†	25.5%	12.0%	20%	19.0%	7.0%
BPPV††	<1.0%	12.0%	2.0%	<1.0%	—
Mondini malformation	—	—	8.0%	—	—
Ménière's disease	<1.0%	2.0%	1.0%	2.0%	2.0%
Perilymphatic fistula	—	2.0%	—	—	—
Delayed endolymphatic hydrops	<1.0%	—	—	—	—
Vestibular neuritis/ labyrinthitis	<1.0%	4.0%	10.0%	12.0%	22.0%
Trauma	<1.0%	—	10.0%	5.0%	2.0%
Metabolic	—	6.0%	—	—	—
Epilepsy	<1.0%	6.0%	1.0%	2.0%	—
Psychogenic	—	10.0%	4.0%	5.0%	22.0%
Tumor	<1.0%	—	<1.0%	—	—
Demyelination	—	—	—	—	5.0%
Amblyopia/vision deficit	—	2.0%	10.0%	—	<1.0%
unclassified	18.2%	10.0%	—	5.0%	5.0%

† Benign Positional Vertigo of Childhood
†† Benign Paroxysmal Positional Vertigo

date regarding the varying etiologies of vertigo and their incidence was done by Wiener-Vacher,[15] who investigated the causal mechanisms of vertigo in over 2,000 children seen in otolaryngology. Similar to other reports, [15-17,27] Wiener-Vacher[15] reported that the most common causes of vertigo and/or imbalance were migraine-associated dizziness (25%) and benign paroxysmal vertigo of childhood (BPVC; 20%). It should be noted that in all these studies, only children referred to otolaryngology were included. Many children and/or parent(s) may not identify vertigo or imbalance, which suggests an underestimation of the problem.[20]

BPVC, BPPV, and Migraine

BPVC has been identified as a common cause of vertigo in young children. It is therefore important to note that BPVC is not the same as benign paroxysmal positional vertigo (BPPV). BPVC typically is seen in children less

than 7 years of age, with vertigo lasting less than 10 minutes and no associated headache. Vestibular and otological exams are typically normal. A child may not complain of dizziness, but autonomic signs (pallor, nausea) may be noted. Children may or may not present with typical migraine later in life.[28] Although BPPV has been reported in children,[21,29] its incidence is much lower in children than in adults.[30] Vestibular migraine is a common cause of vertigo in children.[15,27,30,31] Vertigo may last hours or days and might be independent of headache. A double-blind, dose-comparison trial showed that preventative therapy with topiramate reduced the frequency of basilar migraine attacks in children by 50% and the duration of attacks by as much as 89 minutes.[32]

Otitis Media

Otitis media with effusion (OME) is typically associated with conductive hearing loss and speech delay in children. However, evidence exists that OME may also cause balance impairments and dizziness. The mechanism for these impairments is unclear.[33,34] Balance impairments in children with OME are often reversed following insertion of tympanostomy tubes.[33,35-38] Abnormal electronystagmography (ENG) to include spontaneous and positional nystagmus in children with acute OME also resolved following insertion of tympanostomy tubes.[35,38] However, Casselbrandt et al[39] reported that children with a history of OME had persistent decreased gain for rotary stimuli at 0.1 Hz compared with controls without a history of OME. This demonstrates that OME, even when treated with tympanostomy tubes, might cause long-lasting changes to the vestibular system.

Sensorineural Hearing Loss

Often, children with severe or profound sensorineural hearing loss (SNHL) have concurrent bilateral vestibular hypofunction.[15,22] Tribukait et al[22] reported that 70% of children with SNHL since birth had some form of peripheral vestibular dysfunction, typically hypofunction, (as confirmed by rotary, caloric, and vestibular evoked myogenic potential [VEMP] tests. Unfortunately, these investigators did not report associated impairments in function. Others have reported motor and balance deficits in young children with SNHL and evidence of vestibular hypofunction as measured by rotary, caloric, and/or VEMP tests).[13,40-43] Because children with sensorineural hearing impairment (SNHI) typically have bilateral vestibular hypofunction, this group does not present with nystagmus or vertigo. In spite of these reports, children with SNHI are not routinely tested for vestibular function. The importance of testing is particularly relevant for the children with SNHI who are candidates for cochlear implantation. Jacot et al[44] followed over 200 children who received cochlear implants. Fifty percent had normal vestibular function before surgery. Cochlear implantation altered vestibular function in 71% of the children who previously had vestibular function (10% had areflexia). Cochlear implantation altered vestibular function in 51% of the entire cohort of children with and without vestibular function before surgery. This necessitates that vestibular function testing should be performed both before and after cochlear implantation so that functional impairments can be addressed.

Developmental Disabilities and Prematurity

Recent reports suggest that learning disabilities and other diagnoses may have vestibular involvement.[45,46] Franco and Panboca[45] examined vestibular function in children underperforming in school and found that 67.4% of them had unilateral or bilateral irritative peripheral vestibular deficits. Compared with children without learning disorders, these children had values above the normal range with cold thermal stimulation and abnormal accuracy with saccadic testing. In another study,[46] children with learning disabilities and irritative peripheral vestibular findings also had behavioral abnormalities to include difficulties skipping, playing hopscotch, and riding a bicycle. Although the exact neural mechanism behind vestibular hyperexcitability has not been studied, the behaviors exhibited by these children are similar to those with developmental coordination disorder (DCD). Children with DCD have poor balance, poor coordination, and difficulty organizing movements. Imaging studies have shown that the corticospinal pathways, posterior thalamic pathways, and the parietal lobe are affected in children with DCD.[47,48] However, peripheral vestibular function has not been tested in this population. Eviatar and Eviatar[49] used per-rotary and cold caloric stimulation to examine vestibular function in full-term and premature infants. They reported that in premature, low birth weight infants, there was a maturational delay in vestibular system function evidenced by longer latency and smaller frequency, amplitude, and speed of slow component eye movements compared with the normative sample. Complete maturation was attained in all infants by 12 months of age.

Other Potential Causes of Vertigo or Vestibular Related Impairments

One of the risks of aminoglycoside antibiotics is peripheral vestibular hypofunction.[50-52] In one study,[51] peripheral hypofunction, as measured by ENG with caloric irrigation, was reported in 30.4% of young adults (mean age 23 years) with cystic fibrosis who received tobramycin

(0.87% unilateral and 21.7% bilateral). Another study of infants who received gentamicin at birth[52] reported no significant differences in vestibular responses as measured by rotary chair between patients and controls at 12 months of age. The conclusion was that the vestibulotoxic effects of gentamicin in controlled doses at birth had less of a vestibulotoxic effect than in older children or adults.

Benign paroxysmal torticollis of infancy (BPTI) is characterized by recurrent episodes of head tilt with associated vomiting, pallor, agitation, and ataxia that subsides within a few hours or days. After 5 years of age, BPTI typically evolves into a migraine syndrome such as benign paroxysmal vertigo of childhood (BPVC).[53,54] Formal vestibular function testing has not been done in this population.

Although children can have vestibular dysfunction, the results of some reports should be questioned, because they lack diagnostic vestibular testing or lack positive test results. Furthermore, identification of vestibular dysfunction may be confounded by the plasticity of the nervous system in response to injury.

Plasticity of the Nervous System Confounds Identification of Vestibular Dysfunction

Investigations of plasticity and recovery of function support the idea that age at the time of injury and age at the time of testing affect results obtained on tests of function.[55] Recovery of function is well documented and evidenced on vestibular tests of adults with unilateral lesions of the peripheral vestibular apparatus, in spite of the fact that the peripheral apparatus does not regenerate or recover. Based on evidence that neural changes occur in individuals with nervous system damage at birth or during childhood, as compared with those who sustain damage later in life, it is logical to assume that neural substrate changes will also occur in young children with vestibular deficits. Furthermore, these changes may mask the deficits typically noted with traditional testing methods. It is therefore important to clearly document impairments.

Vestibular-Related Impairments in Children

Impairments With Peripheral Vestibular Dysfunction

Tsuzuku and Kaga[56] and others[12,14,45,57] have reported learning disabilities and delayed development of walking and balance abilities in children with peripheral vestibular hypofunction as measured with caloric and/or rotary chair tests. Rine et al[58] reported that children with SNHI and vestibular hypofunction as measured by rotary chair presented with delayed maturation of vestibular and vision ratios on posturography testing (Fig. 24.1). These children also had a progressive delay in gross motor development as measured by the Peabody Developmental Motor Scales Test. Inoue et al[43] published a retrospective chart review of 89 children with profound SNHI who were to undergo cochlear implant surgery. The children were tested using rotary chair, caloric, and cervical VEMP tests. Twenty percent showed abnormal rotary chair tests, 41% had an abnormal caloric test, and 42% had an abnormal VEMP test. Children with abnormal vestibular function tests developed head control and independent walking significantly later than typically developing children. Shall[13] reported that children with hearing loss since birth and deficits on VEMP testing scored lower on tests of motor development than those with hearing loss and normal saccular function based on VEMP testing. These findings concur with the report by Weiner-Vacher,[4] who reported that the acquisition of walking ability was related to measures of otolith, not canal, vestibular function.

Horak et al[59] found that adults with bilateral peripheral vestibular hypofunction since birth did not demonstrate the lack of leg muscle responses to perturbations evident in adults who had adult onset bilateral vestibular loss. However, activation of trunk muscles was delayed and smaller in amplitude in the early onset group as compared with either those without a deficit, or those with

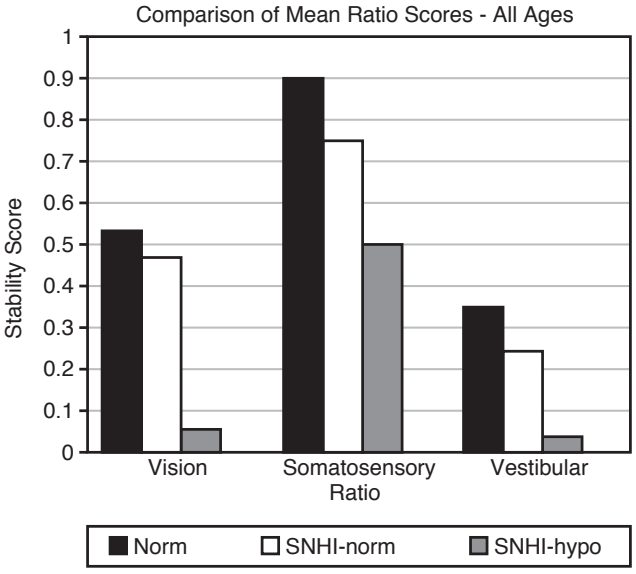

Figure 24.1 Children with SNHI and concurrent vestibular hypofunction (SNHI-hypo) achieved significantly lower somatosensory vestibular and vision effectiveness ratios than those with SNHI without vestibular deficit (SNHI-n) and those without deficit (Norm). This is indicative of deficient sensory organization for postural control.[41]

adult onset deficit. These results suggest that somatosensory inputs from the cervical and upper trunk areas compensated for the vestibular loss in the early onset but not the adult onset group. In contrast, Rine et al [41] noted delayed leg muscle responses on dynamic balance testing, and deficient re-weighting of sensory cues during posturography testing in young children with bilateral vestibular hypofunction since birth (see Fig. 24.1).

In addition to the motor and balance impairments, Braswell and Rine[11] reported that children with confirmed vestibular hypofunction (rotary testing) failed tests of dynamic visual acuity and had impaired reading acuity. This impairment of gaze stability may contribute to the problems in school performance that have been reported. Despite the documented functional relationship of the visual and vestibular systems, and the effect vestibular deficiency may have on gaze stability and oculomotor control,[10,11] children with visuomotor deficits on educational or developmental testing are rarely tested for concurrent vestibular function or gaze stabilization deficits. However, interpretation of test results may be compounded by changes caused by adaptation or plasticity.

The variation of clinical symptoms seen with a peripheral vestibular deficit sustained early in life, versus in adulthood, may also be evident in children with central vestibular deficits.

Impairments with Central Vestibular Deficits

Although vestibular and postural control deficits have been documented in adults with central nervous system involvement, similar investigations of children are scarce (see Chapter 5 on central vestibular disorders).[60-63] It is logical to assume that children with traumatic brain injury or other acquired central nervous system conditions will present with vestibular deficits similar to those of adults. However, because of maturation issues involved in the development of postural control, and particularly the effectiveness of vestibular function in postural control, the consequences to motor recovery may differ, and varied mechanisms of compensation and adaptation may mask vestibular deficits.

Liao et al[63] reported poor static postural control under various sensory environments in children with spastic cerebral palsy. The children had normal vision and somatosensation. Although it is expected that the muscle tone impairment involved is to a great deal responsible, the fact that postural sway was significantly different between children with and without impairment only on the swayed surface (SS), eyes closed—swayed surface (ECSS), and sway ed vision—swayed surface conditions (SVSS), and similar in all other conditions, suggests sensory organization impairment of a central nature. Nashner, Shumway-Cook, and Marin [64] and others[61,65] reported similar results, as well as a reversal of muscle activation patterns in children with cerebral palsy. However, in none of these studies was vestibular function formally tested.

Benign Paroxysmal Vertigo of Childhood (BPVC) is a migraine-related vertigo syndrome occurring most often in children between 2 and 6 years of age. It causes vertigo lasting seconds to minutes with associated nystagmus, postural imbalance, and nausea and vomiting. Vestibular impairments are not evident between attacks.[66] However, a study by Zhang et al[30] showed that of 56 children with BPVC studied, 14.3% had abnormal bithermal caloric tests and 32.1% had abnormal cervical VEMP tests. High-stimulus auditory brainstem responses and Transcranial Doppler sonography tests were abnormal in 48.2% of the cases. These results suggest that BPVC is a vascular event and related to migraine syndrome. Authors also suggest that the inferior vestibular pathway is more damaged than the superior vestibular pathway.

The identification of a vestibular deficit is important to the development of appropriate intervention and for prognostic purposes. Further research is needed in this area, particularly because early identification and intervention during critical periods may affect recovery.

In summary, children, like adults, can and do have central and peripheral vestibular deficits. Reportedly, a delay in acquisition of motor skills and learning disabilities is evident in children with peripheral and central vestibular dysfunction. To understand the implication of vestibular deficits early in life, and to enable the selection and interpretation of testing and the development of appropriate intervention for children, a review of the development of postural and oculomotor control as they relate to vestibular function is warranted.

Development of Postural and Oculomotor Control as They Relate to Vestibular System Function

Functional maturation of the vestibular and visual systems for postural control continues through 15 years of age[67,68] with the following noted: (1) although visual system effectiveness in postural control is less mature than that of the somatosensory system before age 7.5 years, it is the dominant source of information for postural control in standing[69-71]; (2) vestibulo-ocular mechanisms are intact and mature by 1 year of age [72]; (3) vestibular system effectiveness for gaze stabilization is mature by 3 years of

age[14]; (4) the vestibulospinal mechanism, or the effectiveness of the vestibular system in postural control, continues to develop beyond 15 years of age[73]; and (5) the sensory integrative capacity required for postural control evolves between 7 and 15 years of age.[69,74,75]

Numerous reports on the development of postural control support the idea that although postural response synergies and the ability to use each of the sensory components for postural control are evident in young children learning to sit and walk, the muscle responses, weighting of different sensory information, and integrative abilities of postural control are not like those of adults until adolescence.[2,69-71,76,77] Forssberg and Nashner[78] and others[2,76,77] reported that although the automatic responses to perturbation seen in adults were present and functional in young children, response latencies (e.g., short-, medium-, and long-latency) were significantly longer and matured at different rates. Haas et al[76] claimed that the change in short-latency responses to adultlike levels by 5 years of age reflected improved nerve conduction velocity. The long-latency response, which is reportedly an indicator of vestibulospinal function,[68,79] was not reduced to adultlike levels until 15 years of age. The authors attributed this to acceleration of central polysynaptic transmission and myelinization. In addition, Shumway-Cook and Woollacott[74] reported that the attenuation of response seen in 7 year olds and adults was not observed in younger children, with increased variability of responses in 4- to 6-year-old children. Additional studies demonstrated that the development of sensory integrative capacity and experience within a posture affect maturation of these muscle responses.[69,75]

Results of investigations by Forssberg and Nashner[78] revealed that, although children 3 through 10 years of age could maintain balance on all posturography test conditions, children less than 7.5 years of age swayed significantly more than older children and adults on conditions dependent on vestibular function (eyes closed and sway surface, and sway referencing of both the visual surround and support surface). Furthermore, Rine et al[71] reported increased variability of responses on posturography testing in children 4 to 6 years of age. These investigators and others[70] also noted that somatosensory effectiveness was adultlike by 4 years of age (Fig. 24.2).

Several conclusions may be gleaned from reports on the development of postural control: (1) experience within a posture is critical for the development of postural control abilities[75]; (2) by 3 years of age, somatosensory effectiveness in postural control emerges, but in children less than 7.5 years, visual and vestibular system

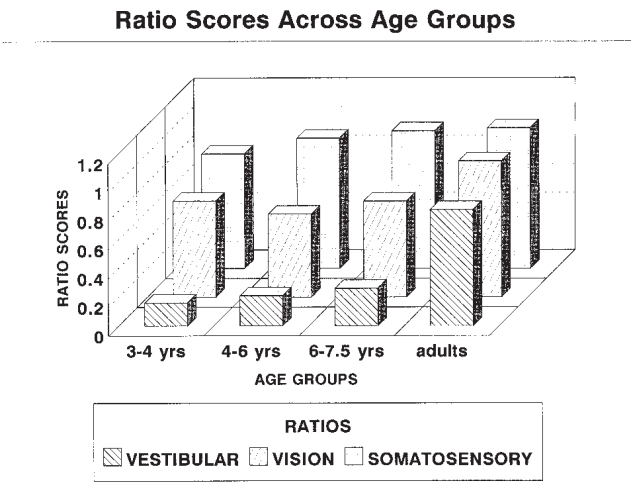

Figure 24.2 Median of ratio scores attained by individuals of varying age without deficits.[71]

effectiveness in postural control is immature (see Fig. 24.2)[69,70,78,80,81]; (3) maturation of postural responses occurs in a stage-like pattern[75,76]; and (4) the time period between the ages of 4 and 6 years is a transition period in which mature patterns are emerging.[70,71] Investigators have suggested that these changes are a result of maturation of the effectiveness of individual sensory systems and the sensory integrative mechanisms involved in postural control.[70,71,80,81]

The findings that development of postural control is affected by the integrity of all systems involved, as well as practice and experience in a posture, suggest that deficits in any one system, in central processing capabilities, or in the ability to experience various upright postures will impede development. Furthermore, findings that vestibular system effectiveness in oculomotor ability is adultlike on tests of dynamic visual acuity by 3 years of age,[14] but that its effectiveness in postural control is not adultlike until adolescence,[67,68,73] indicates that the two vestibular systems mature at different rates and function separately. Therefore, a comprehensive assessment of vestibular function in children should include tests of both vestibulo-ocular (VO) and vestibulospinal (Vsp) function. Normative data are available for tests of VO and Vsp function. This is important for (1) the identification of deficits, and (2) the development of appropriate early interventions for children with motor, oculomotor, or postural control deficits, secondary to vestibular dysfunction.

It is important to note that the causes of vertigo and/or imbalance may be a result of central or peripheral lesions or pathology that may or may not be vestibular related. To determine if the cause of imbalance or vertiginous symptoms

is a vestibular pathology requires a team effort and comprehensive testing.

Evaluation of Children with Vestibular Dysfunction

A comprehensive examination of vestibular function includes functional and diagnostic tests of vestibulo-ocular (VO) and vestibulospinal (Vsp) function.[82,83] Because diagnostic procedures are costly and may be uncomfortable, particularly for young children, screening for appropriate referrals for in-depth testing is warranted. Based on the literature, the following children should be tested with regard to motor and balance development and oculomotor control.[19] Children with:

- SNHI,
- identified learning disability with evidence of functional impairments related to Vsp or VO function,
- recurrent and chronic inner ear infections who also present with difficulties of coordination, reading, or developmental delay, and
- complaints of dizziness or visual stabilization difficulties.

Therapists can complete developmental functional testing of motor and postural control abilities, perceptual and oculomotor abilities, and screening of VO and Vsp function.[14,84,85] The purpose of these tests is to:

1. establish that a functional balance and/or oculomotor deficit(s) exists,
2. isolate the contributions of the various components of the postural control and gaze stability systems, and thus determine which component(s) is (are) problematic, and
3. provide a basis for referral for further diagnostic testing and the development of remedial programs.

Children who attain scores below age-appropriate levels and those in whom screening identifies VO or Vsp dysfunction should be referred for more comprehensive testing (e.g., oculomotor, rotary chair, caloric, VEMP).

Testing Balance and Gaze Stability

Normal postural control for balance requires intact vestibulospinal abilities and integrated labyrinthine reflexes. Clinical functional measures of balance that have normative data and are standardized for children include:

- Functional Reach Test,[85,86]
- Locomotion subscale of the Peabody Developmental Motor Scales, 2nd edition (PDMS-2),[87]
- Balance subscale of the Bruininks-Oseretsky Test of Motor Proficiency, 2nd edition (BOT-2),[88]
- Pediatric Balance Scale,[89,90]
- Dynamic Gait Index,[91]
- Pediatric Clinical Test of Sensory Interaction for Balance.[92,93]

Care should be taken in the selection of tests, because not all are appropriate for all ages (Table 24-2). The BOT-2 and PDMS-2 are valuable in documenting general gross and fine motor developmental status and providing more specific measures of balance and visual motor development. Tests of sensory integration dysfunction (i.e., The Test of Sensory Functions in Infants[94] and the DeGangi-Berk Test of Sensory Integration[95]) are appropriate for children with only mild motor delays. Specifically, these tests examine for the persistence of primitive reflexes, such as the tonic labyrinthine and asymmetric tonic neck reflexes (see Figs. 24.3 and 24.4). Persistence of these responses purportedly suggests involvement of the vestibular system. However, the literature remains unclear on the relationship between peripheral and central vestibular function and sensory integration dysfunction as measured by these tests. Therefore, the use of the tests of sensory integration dysfunction and intervention for motor planning, dyspraxia, and sensory integrative dysfunction will not be further addressed in this chapter. However, these issues are appropriately and completely reviewed elsewhere.[5]

For children with or without balance impairments, gaze stability should be examined. The functional measure of gaze stability, and more specifically, the contribution of vestibular information to gaze stability, is the dynamic visual acuity (DVA) test.[14,96,97] For children, visual acuity charts are available with symbols (Fig. 24.5) to complete Dynamic Visual Acuity Testing, which has excellent sensitivity and specificity for the identification of vestibular hypofunction in children.[14] In addition, a computerized DVA test was recently developed. Normative data was gathered for children, and the test was determined to be reliable and valid.[96]

Children who present with deficits in balance or gaze stability on any of the tests noted above, or more specifically, on the balance, reflexive, or visual motor subtests, or dynamic visual acuity test, should be further examined for sensory (i.e., vision, somatosensory, and vestibular) and postural control system effectiveness, to include

■ Table 24-2 ASSESSMENT OF PROBLEMS ASSOCIATED WITH VESTIBULAR DYSFUNCTION IN CHILDREN

Assessment Tool	Test Type	VFx	Age Group	Items/Behaviors Tested
Peabody Developmental Motor Scales, 2nd edition[87]	Motor development Balance ability/ development Visual Motor Reflexes, Stationary, Locomotion, Object Manipulation, Grasping, Visual-Motor Integration Subtests	Vsp VO	Birth through 71 months	Developmental reflexes Single leg stance eyes opened Jumping, hopping, skipping Tandem stand and walk Visual track, perception, tracing Catching, throwing, and kicking balls to targets
Bruininks-Oseretsky Test of Motor Proficiency, 2nd edition[88]	Balance and gross motor ability Manual dexterity, upper-limb coordination, bilateral coordination, balance, running speed and agility, strength subtests	Vsp VO	4 through 21 years of age	Balance on beam in tandem and single leg stance Single leg eyes opened and closed Dribbling a ball, target finger to nose, bilateral coordination tasks
DeGangi-Berk Test of Sensory Integration[95]	Sensory integration and motor planning Postural control, bilateral motor and reflex integration subtests	Vsp VO	3 through 5 years of age	Pivot prone and supine flexion Postural tone Dysdiadochokinesi Tapping, jumping, and bilateral tasks ATNR and STNR testing (Figs. 24.3 and 24.4)
Test of Sensory Function in Infants[94]	Sensory processing and reactivity in infants	VO Vsp	4 through 18 months of age	Responses to tactile, vestibular and visual, ocular motor control
The Sensory Integration and Praxis Tests[98]	Sensory integration and motor planning abilities Visual perception	Vsp VO	4 through 8 years of age	Form and space perception Praxis Tactile discrimination Vestibular integration Prone extension test
Southern California Post Rotary Nystagmus Test[99]	Visuo-vestibular system		5 through 11 years of age	Nystagmus duration after rotation Entire test done in room light
Emory Clinical Vestibular Chair Test (ECVCT)[100,101]	Vestibular ocular system test	VO	adults	Nystagmus duration after rotation Entire test done with fixation removed

■ Table 24-2 **ASSESSMENT OF PROBLEMS ASSOCIATED WITH VESTIBULAR DYSFUNCTION IN CHILDREN—cont'd**

Assessment Tool	Test Type	VFx	Age Group	Items/Behaviors Tested
Functional Reach[85,86]	Balance ability in standing	Vsp	5 through 15 years of age	Forward reaching, standing
Pediatric Balance Scale[89,90]	Balance	Vsp	2 through 15 years of age	14 items (e.g., standing with eyes closed, tandem standing, turning 360 degrees, placing alternate foot on stool, forward reach)
Pediatric Clinical Test of Sensory Interaction for Balance[102]	Balance under varying sensory conditions	Vsp	4 through 9 years of age	Measures of sway in double leg stance with eyes opened and closed while standing on foam or floor, or with conflict dome
Dynamic Posturography Testing[103,104]	Functional test of balance Ability to use vestibular, visual and somatosensory systems	Vsp	Normative data 3 years through adult on SOT DPT component: 1.5 years through adult	SOT—computerized measures of sway DPT—measures EMG responses to perturbation, standing
Dynamic Visual Acuity Test (Clinical);[14] Computerized (NIH toolbox)[96,97]	Gaze stabilization	VO	3 years through adult	Visual acuity with head stable and moving at 2 Hz.
Canadian Occupational Performance Measure (COPM)[105]	Goal setting	n/a	All ages	Semi-structured interview format. Parents and children identify important problems and rate performance/satisfaction.

Abbreviations: VFx = Vestibular Function; Vsp = Vestibulospinal system; VO = Vestibulo-ocular system; ATNR = Asymmetrical Tonic Neck Reflex; STNR = Symmetrical Tonic Neck Reflex; SOT = Sensory Organization Test; DPT = Dynamic Perturbation Test (i.e., toes up tip test)

screening of neurological, oculomotor, and musculoskeletal systems.

Screening Motor and Sensory Systems

The musculoskeletal system must be examined to determine if restrictions in range of motion, pain, reduced strength, or limited endurance are present, because any of these may affect postural alignment or the availability of movement strategies to maintain equilibrium. Furthermore, these measures may assist in differential diagnosis of normal central nervous system response to an abnormal musculoskeletal system or an abnormal central nervous system response to a normal musculoskeletal system. Subtests of the PDMS-2, BOT-2, and the sensory integration tests can provide screening of neurological status (e.g., Romberg testing with eyes open and closed, finger to nose with eyes open and closed, developmental tonic reflex integration). Sensory screening should include an examination of the visual, somatosensory, and vestibular systems.[106]

SENSORY AND MOTOR SCREENING

- Lower extremity strength
  - Manual muscle testing
  - BOT-2 strength subtest
- Coordination
  - Heel to shin and rapid alternating toe taps
  - Finger to nose with eyes opened and closed
  - BOT-2 coordination subtests
- Deep tendon reflexes
- Equilibrium reactions
  - Tilting
  - Postural fixation
  - Protective extension
- Somatosensory screening in lower extremities
  - Light touch
  - Proprioception (i.e., position sense)
  - Cortical sensation (e.g., graphesthesia)
- Visual screening
  - Visual field capabilities
  - Oculomotor tests
- Saccades, smooth pursuit
- Clinical Vestibulo-ocular Reflex Testing (see Table 24-2)

Clinically, vestibular screening can be done by noting the presence of abnormal responses such as gaze-evoked or positional nystagmus and corrective saccades on the head thrust test.[83,107] Other screening tools for vestibular canal function include the DVA and the Emory Clinical Vestibular Chair Test (ECVCT, Box 24-1). Unlike the Southern California Post Rotary Nystagmus Test (SC-PRNT), the ECVCT is measured with fixation removed and has been proposed as a clinical measure of pure vestibular hypofunction.[100,101] Screening of vestibular otolith function is typically done by testing the perception of vertical.[108] The Bucket Test[108] was recently developed for adults as a reliable, valid, and inexpensive version of the test of subjective visual vertical (SVV). If a balance or a visuomotor deficit is evident, concomitant with evidence of VO or Vsp deficit, further diagnostic testing of vestibular function is warranted.

Diagnostic Postural Control Testing

To test postural control, dynamic posturography testing is used.[41,73,79,103,109] Dynamic posturography testing involves both a sensory organization test (SOT) and dynamic perturbation test (DPT), which complement and expand on the information provided by traditional clinical testing, and yield an objective, functional measure of postural control.[71,73,103,109] Although sensory and motor tests enable the clinician to isolate the integrity of the different components of postural control, they do not measure the functional use and effectiveness of the sensory modalities, the integrative capabilities necessary for postural control, which the SOT provides (see Chapter 11 for details on the SOT). In addition, sensory system ratios calculated from results obtained from the SOT may be used to monitor dominance or maturation of sensory system effectiveness in postural control.[71] Normative values of SOT measures have been reported for children 3 through 15 years of age (Table 24-3).[70,71,103,109,110] A clinical, less-sophisticated form of the sensory organization test, one that requires inexpensive materials and is portable, has been developed.[92,102,111] The Pediatric Clinical Test of Sensory Interaction for Balance (P-CTSIB) has fair to good reliability when combined sensory conditions scores were used.[102] Using the P-CTSIB, the examiner documents duration of balance (up to 30 seconds) and body sway under the six SOT conditions using a dome and medium-density foam. Recently, the National Institutes of Health (NIH) developed the NIH toolbox, which includes a reliable and valid balance test called the Balance Accelerometry Measure (BAM).[112] The BAM uses an accelerometer and

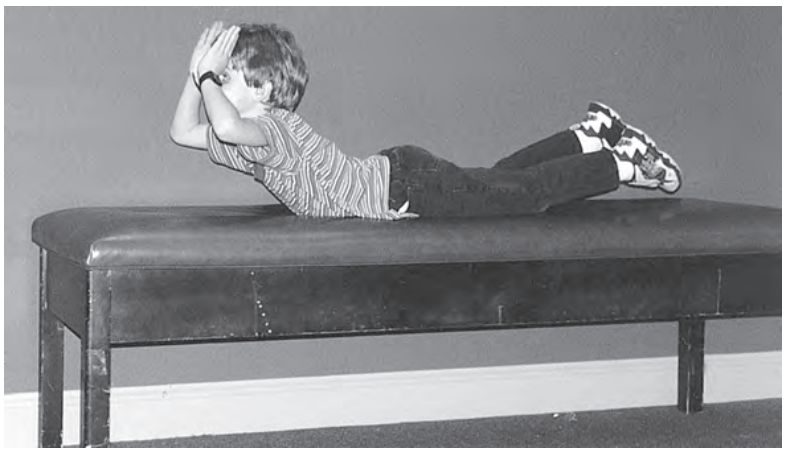

Figure 24.3 The prone extension test examines the child's ability to assume and maintain a pivot prone position. Inability to do so is indicative of persistence of the tonic labyrinthine reflex.

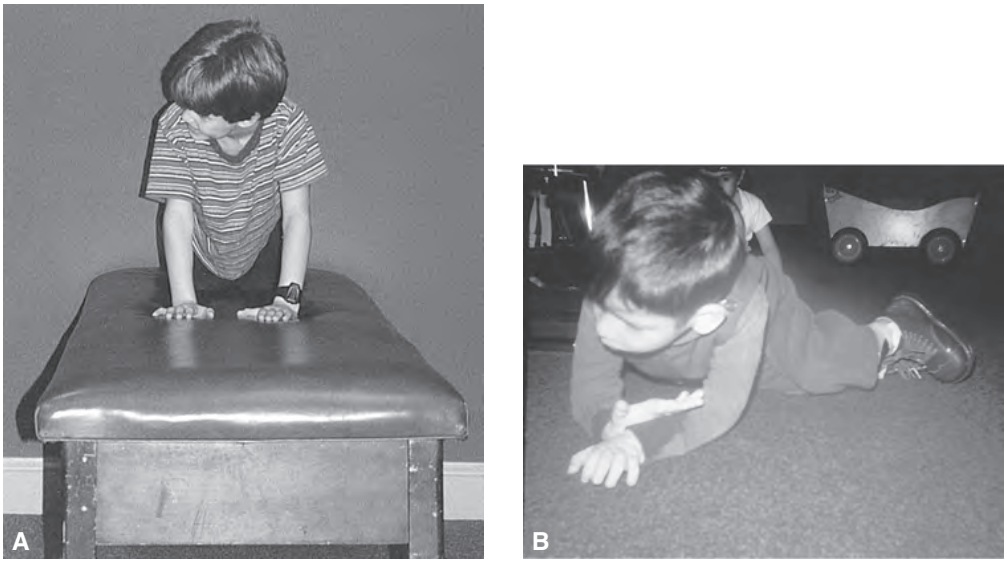

Figure 24.4 One method of testing whether the asymmetrical tonic neck reflex is interfering with function is to request that the child maintain quadruped with elbows extended as he/she looks to the left or right: **(A)** the child without deficit is able to do so without difficulty; **(B)** the arms collapse as the child attempts to look to his right.

medium-density foam to objectively measure sway. Normative data has been collected for children.[97]

The DPT component of posturography may be used to determine if neurological dysfunction is evident, and at what level, and provides an indirect measure of Vsp function.[59,79,113] Furthermore, maturation changes in the neuromotor component of postural control can be measured and monitored. Muller and colleagues [2] and Dichigans and associates[79] reported that responses are evident with minimal maturation changes evident in the very early stages of learning to stand and walk. Harcourt[114] reported that although posturography testing may not be as

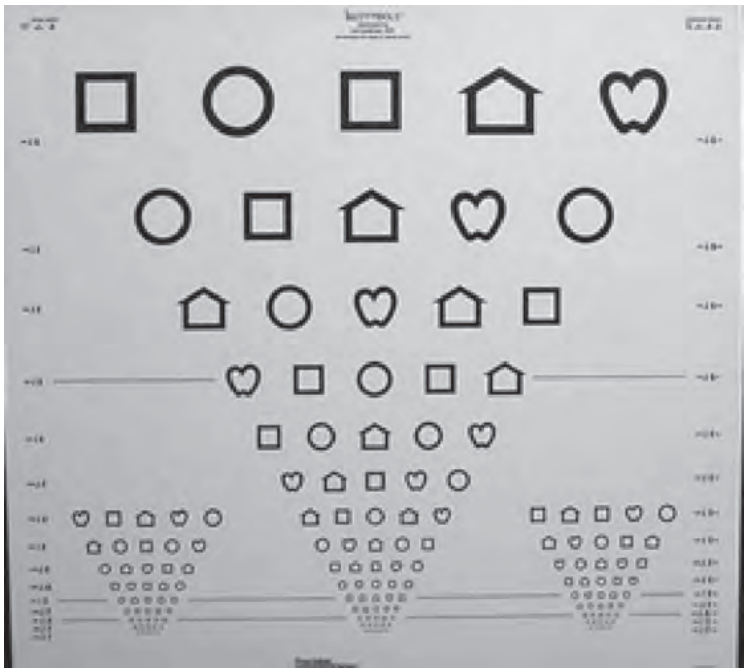

Figure 24.5 For children who do not know letters, vision charts with symbols can be used. (Good-lite Co. 1155 Jansen Farm Drive Elgin, IL 60123; www.good-lite.com/).

Box 24-1

POINT AND COUNTERPOINT: Should The Southern California Post-Rotary Nystagmus Test (SCPRNT) Be Used To Identify Peripheral Vestibular Hypofunction?

The SCPRNT was designed by Jean Ayres[99] as a test of the global vestibular system to include the vestibular receptors and central vestibular structures. It was intended to be used in the evaluation of children with learning disabilities and other sensory processing disorders. *It was not intended to identify peripheral vestibular hypofunction*. The test is done in room light with eyes open. The child sits cross-legged on a spinning board as the examiner spins the child to the left 10 rotations at 0.5 Hz. Following spinning, the examiner times the duration of the nystagmus as the child looks at a white sheet. After resting for 1 minute, the test is repeated to the right. The examiner also notes behaviors (e.g., pleasure or stress) and maintenance of head and body posture during the test.[115,116] The gold standard tests of peripheral vestibular function continue to be caloric irrigation and rotary chair testing with ENG or VNG. These tests are often unavailable to clinicians and the caloric test might be uncomfortable for children. No studies have been done to determine the sensitivity or specificity of the SCPRNT to detect vestibular hypofunction as measured by the gold standard tests. However, some investigators have used the SCPRNT to screen for pure vestibular hypofunction or to note hyperactive vestibular responses.[12,117] The problem with this is that the gain of responses to rotation with eyes open in room light includes a contribution from the optokinetic system as well as the vestibular system. After rotation in room light, nystagmus can be suppressed through the visual system. Therefore, pure vestibular responses to rotation must be tested in darkness.[115]

Clinicians can screen for peripheral vestibular hypofunction using the Emory Clinical Vestibular Chair Test (ECVCT) developed by Hall and colleagues.[100,101,118] This test, as modified for children, requires an office chair, the head tipped forward 30 degrees, metronome to maintain proper speed of spinning, and infrared goggles. The child is rotated for 30 seconds at 0.5 Hz with eyes closed. When the chair is stopped, the goggles are placed over the eyes, the eyes open, and nystagmus is timed from the time of the chair stopping until there is no observable nystagmus. The patient rests for twice the duration of nystagmus, and the test is repeated to the other side. The cutoff that provided optimal sensitivity and specificity for children with hypofunction was post-rotary nystagmus in the dark lasting less than 29.2 seconds total (left and right).[118]

■ Table 24-3 **NORMATIVE DATA FOR THE SENSORY ORGANIZATION TEST[70,103,109]**

Age (years)	*SOT 1	*SOT 2	*SOT 3	*SOT 4	*SOT 5	*SOT 6	*COMP
4 (n = 51)[109]	79(7)	74(11)	74(9)	52(13)	38(15)	40(15)	56(8)
5 (n = 76)[109]	84(7)	77(8)	76(13)	55(15)	40(16)	42(16)	60(9)
6 (n = 74)[109]	85(6)	80(8)	78(8)	62(14)	43(15)	46(16)	64(8)
7 (n = 69)[109]	86(7)	81(9)	82(7)	62(14)	47(15)	48(16)	66(9)
8 (n = 38)[109]	87(7)	84(6)	80(7)	64(17)	44(13)	40(19)	66(8)
9 (n = 18)[109]	90(7)	86(5)	85(7)	61(19)	57(14)	54(18)	69(11)

■ Table 24-3 NORMATIVE DATA FOR THE SENSORY ORGANIZATION TEST—cont'd

Age (years)	*SOT 1	*SOT 2	*SOT 3	*SOT 4	*SOT 5	*SOT 6	*COMP
9–10 (n = 58)[103]	90.3(4.6)	88.7(6.7)	87.5(9.8)	65.8(19.3)	46.3(22.4)	42.3(25.1)	67.6(16.0)
11–12 (n = 72)[103]	93.4(2.5)	91.9(2.8)	90.7(5.4)	75.7(10.5)	51.8(20.5)	48.8(20.8)	73.7(11.0)
14–15 (n = 19)[70]	91.8(2.8)	90.4(2.2)	88.2(3.0)	78.0(6.4)	49.4(12.6)	49.4(11.9)	70.0(5.0)
20–59 (n = 26)[70]	93.2(1.7)	91.0(3.6)	87.7(4.7)	83.2(11.1)	63.5(10.6)	59.0(14.7)	75.7(7.2)

Equilibrium score; standard deviation in parentheses.

sensitive as caloric testing in the identification of individuals with peripheral vestibulopathy, it does provide additional information and is most valuable for identifying patients with central abnormalities.

Diagnostic Testing of Vestibular Function

Diagnostic tests of canal vestibular function include caloric and rotary chair testing. Normative values are available for children as young as 1 year.[49,110,119] Otolith vestibular testing for children, to include vestibular evoked myogenic potentials (VEMP), off-vertical axis rotation, and SVV, has also been reported.[4,22,110,120,121]

Once appropriate testing identifies functional impairments related to vestibular dysfunction, appropriate intervention can be developed and administered.

Treatment of Vestibular Dysfunction

Peripheral Disorders

As with adults with peripheral dysfunction, once a diagnosis is complete, medical management and rehabilitation should begin. This may include medication, surgery, or gaze stability and balance training activities (see Chapters 17, 21, 22, and 23).

Evidence of Treatment Efficacy

Peripheral Vestibular Hypofunction

Rine and colleagues[58] completed a randomized double-blind controlled study to determine the efficacy of exercise intervention for the improvement of motor development and postural control in children with bilateral vestibular hypofunction. Children participated in an exercise intervention focused on substitution three times weekly for 30 minutes, under the direction of a physical therapist (Table 24-4). The previously noted progressive motor development delay was halted and visual and somatosensory effectiveness ratios that were previously deficient improved to within normative ranges. Braswell and Rine[122] reported improved scores on dynamic visual acuity tests and improved subjective reports of gaze stability following exercise intervention in children with vestibular hypofunction. Medeiros and colleagues also showed improvement of symptoms and balance on SOT conditions 5 and 6 in children with peripheral vestibular hypofunction who received vestibular rehabilitation.[123]

Benign Paroxysmal Positional Vertigo (BPPV)

Evidence on treatment efficacy in children with BPPV is limited to case studies. D'Agostino, Melagrana, and Taborelli[21] reported a case of horizontal canal BPPV in a 10-year-old child, with "spontaneous" recovery following short-term hospitalization and repeated mobilization (e.g., roll side to side) in supine. Saka et al[124] published two cases with BPPV. An 11-year-old boy presented with apogeotropic nystagmus following the roll test, implying cupulolithiasis in the horizontal canal. A 3-year-old girl presented with geotropic nystagmus following the roll test, implying canalithiasis of the horizontal canal. In both cases, treatment was not done and positional nystagmus disappeared 1 week later at the follow-up visit. Shetye et al[125] reported about a 13-year-old girl who developed BPPV following fairground rides. She had cochlear implant surgery 2 years prior. The case was successfully treated using the Epley maneuver and Brandt Daroff exercises. In a retrospective chart review, Song et al[126] reported that five patients including three children with enlarged vestibular aqueduct syndrome also presented with recurring BPPV and multiple canal involvement. They were treated successfully with the Epley maneuver.

■ Table 24-4 DESCRIPTION OF EXERCISE PROGRESSION[58]

Sessions are 30 minutes; 3X/week; 12 weeks. For each session, choose 1 exercise from 3 categories and spend 10 minutes working on the exercise. Document success and progression.

Categories	Examples of Specific Exercises	Progression (80% Success of Trials)
Eye-hand coordination	Tap small pictures on a balloon as it moves; bat a balloon with a badminton racket; throw bean bags to a target; pick up Velcro pictures with a tennis ball (bouncing it on the floor); catching balls	Balloon: begin with balloon still, then move it with increasing speed and direction; Badminton: begin with a balloon and progress to a birdie; Bean Bag Target & Velcro ball: begin close to the target and progress to standing further away; Catch: begin with larger balls, progress to small rubber ball.
General coordination	Side-stepping; braiding; dancing; running, skipping, galloping, and hopping races; obstacle courses that include crawling, climbing, and jumping	Begin with side-stepping, easy dance steps, galloping and progress to more advanced steps (e.g., braiding, skipping); Time the obstacle course and race and try to beat the previous time; measure the distance of long jump and try to increase it.
Visual-motor training	Identify pictures, letters, and numbers while: (1) swinging in a net swing; (2) jumping on a trampoline; (3) spinning on a sit-and-spin; (4) walking quickly.	Decrease the optotype size; increase the speed of head movement
Balance training	Walking on a narrow path; walking or standing on a balance beam; single legged stance; walking on various surfaces (e.g., thick mat); sitting on a T-stool (i.e., a stool with a single leg); assuming a posture on a scooter board and holding a position while being pulled around the room.	Decrease the size of the path to a tandem walk; balance in various lighting conditions (e.g., dim light, Vaseline goggles, eyes closed, varying depth of compliant surface); T-stool: begin with feet apart while catching large balls and progress to feet together catching smaller balls.

Considerations for Treatment of Pediatric Patients

Balance and substitution training activities must be modified to the child's level of cognitive maturation and interest level, with particular consideration to the caregiver. Unlike the adult who will be responsible for his/her own exercise regime, the child is dependent on caregivers and therapists to carry out the program and ensure compliance. To maximize functional recovery similar to that of adults, the program should be completed several times daily throughout the day in short but intense bouts. The use of toys, books, games, and other items to facilitate visual tracking, or the use of swings to provide the movement during visual stabilization activities, is important for cooperation. Instead of letters, which are not motivating or fun, line pictures, moving balls or toys with symbols or letters, or interactive computer or video games may be used.[122] If the child can attempt to grasp or point to specific letters or symbols while he/she is moving the head, VOR exercise becomes a game, is fun, and may maximize effort and cooperation. This cooperation and effort is critical to the effectiveness of the exercise regime. Typically, when acute symptoms have subsided, and appropriate medical treatment is rendered (e.g., surgery to repair fistula), children are eager to resume play and other age-appropriate activities. The exercise program for children born with bilateral hypofunction might need to be of longer duration than that for children who once had vestibular function and lost it.[122] It is critical to monitor progress and provide short-term training to be carried out daily at home with caregivers (e.g., visual tracking exercises, visual

stabilization regime, balance, and movement activities to resume age-appropriate levels of activity).

Central Vestibular and Postural Control Deficits

Few studies have been conducted to examine vestibular function in children with central nervous system disorders. However, reports in the literature do note the following: children with autism spectrum disorder and dyslexia present with sensory organization and balance deficits,[127,128] postural control deficits are evident in children with cerebral palsy, and vestibular stimulation does improve motor and visual abilities in children with central nervous system deficit, autism, and low birth weight premature infants.[9,57,129-131] Specific clinical signs and symptoms of central deficits have been delineated elsewhere in this volume. Reportedly, postural control deficits are the most consistent finding. Therefore, children with deficits of balance, postural control ability, and visual motor function should be evaluated for VO and Vsp function and differential diagnosis of the multiple factors involved. Treatment can then be developed to either facilitate the use and integration of systems intact but not used, or to facilitate compensatory mechanisms. For example, a child with hypertonicity and developmental delay, as well as evidence of VO and Vsp dysfunction, should participate in programs that include facilitation and improvement of visual stabilization ability, movement tolerance and balancing during visual stabilization, and balance training under varying sensory environments to encourage the use of intact systems and facilitate integration of information.

CASE STUDY 24-1

This 7.5-year-old female was admitted to the hospital with complaints of dizziness, nystagmus, spinning sensation, and inability to sit or stand. She was diagnosed with vestibular neuritis, hospitalized, and treated for 3 days. Upon discharge, she was referred to physical therapy for evaluation and treatment of symptoms. Evaluation revealed the following:

- Equilibrium reactions: intact; single leg stance with eyes open (EO) 2 seconds, eyes closed (EC) 1 second either leg (norm for this age is 10 seconds with EO or EC); unable to walk on 3.5-inch balance beam in forward or sideways directions (should be able to walk tandem on beam without loss of balance); able to stand in tandem 3 seconds, but not walk without side-stepping (should be able to tandem walk 10 feet).
- Neurological screening (to rule out confounding central factors): Deep tendon reflexes intact; finger-to-nose negative; rapid alternating upper and lower extremity movements intact.
- Vision and VO testing: Upon testing, optokinetic nystagmus intact; gaze-evoked nystagmus was noted (abnormal), with a tendency to maintain head tilted to the right; there was evidence of favoring the left eye by semi-closure of the right during dynamic visual acuity test, which was normal except for this "squint."
- Posturography testing: Loss of balance noted on conditions 5 and 6; vision ratio within normal limits; vestibular ratio well below normative values for age (more than 2 standard deviations below mean).

Parent was trained in visual stabilization training using flash cards the child enjoyed and single words the child was familiar with: (1) child to read card as it is moved to right and left at 1 Hz (smooth pursuit); with card stabilized at midline, child to read card as head is turned to the left and right at 1 Hz (VOR and smooth pursuit); once the child is able to do this without difficulties, the frequency can be increased to 2 Hz (practice 2 minutes each, twice daily); (2) with four "word" cards taped on wall centered in front of child, child sitting 6 feet away, practice reading cards as head is rotated right and left (binocularly and monocularly to strengthen use of each eye); (3) encourage play as before and practice on balancing (walking between tape lines 4 inches apart on floor, gradually reducing to 3 inches; progress to walking heel to toe on line or foot prints (practice 2 minutes daily); and (4) hopping games (e.g., hop scotch, jump rope swung slowly back and forth by Mom and child to jump over it). Because of difficulties in transportation, weekly monitoring of progress and adjustment of exercises was done via phone contact. In 2 months, all symptoms were relieved. The child was able to complete single leg stance for 10 seconds with eyes open or closed, tandem stand, and tandem walk 5 feet. Performance was within normative values on all posturography conditions. No difficulties were noted with reading or visual fixation either with card or head movement.

CASE STUDY 24-2

A 7-year-old male was referred for assessment because of recent complaints of dizziness, vertigo, and vision difficulties. Child was the product of premature birth, with low birth weight (1.5 pounds). Sensorineural hearing impairment was diagnosed at 18 months of age. However, with use of hearing aids, audiological testing was within normal limits and the child did comprehend spoken language and spoke clearly. He also had a history of frequent ear infections with drainage tubes inserted twice (bilaterally). This child had been recently placed in a special education program for learning disability because of poor reading test results (falling below grade level). He was referred for testing to establish vestibular functional status and to determine if further testing or treatment was warranted. Parent reported typical activity levels until recently to include playing T-ball and basketball with peers (intramural sports). Review of records indicated treatment with gentamicin and amoxicillin during the past few years. *(Although amoxicillin does not affect the vestibular apparatus, gentamicin is known to be ototoxic in children, similar to what is seen in adults.*[50-52]*)*

- Vision and VO testing: Subjective reports of dizziness with moving visual stimuli (e.g., looking up at sky and watching clouds, watching television, changing visual direction in classroom); child and parent reported that "eyes do funny things"; when asked to demonstrate this, child moved his eyes left to right in an attempt to replicate nystagmus; difficulties with smooth pursuit were evident, with saccadic intrusions when attempting smooth pursuit, and he attempted head turns to maintain fixation on the moving target; dynamic visual acuity was normal; although nystagmus was not elicited on head shake test, child reported increased dizziness; a modified version of the SC-PRNT was done (i.e., eyes closed for the rotations and eyes opened in room light following rotations). A hyperactive response to this test was noted. Nystagmus persisted for 35 seconds following cessation of chair rotation, and the child had severe vertigo. This was seen with rotation in either direction. This result suggests an irritative lesion. An irritative lesion (e.g., fistula) is one that causes hyperactive vestibular responses.
- Motor and balance ability: The child was able to single leg stance eyes opened for 6 seconds, with eyes closed for 3 seconds; he was able to walk 3.5-inch balance beam, but not balance standing across the beam in double or single leg stance; on Balance subtest of BOTMP achieved age-equivalent score of 4 years 11 months; on visual-motor coordination subtest, age-equivalent score of 7 years 8 months. Significant developmental delay in balance abilities evident.
- Neurological Screening (to rule out confounding factors and determine appropriate treatment approach): Deep tendon reflexes within normal limits, as were rapid alternating movements, integration of labyrinthine reflexes, and finger-to-nose test.
- Posturography (SOT) results: Patient relied on stepping strategy to maintain upright on conditions 5 and 6; rigid posturing of arms and legs noted on condition 4; vision ratio of 0.29 (more than 2 standard deviations below norm for age) and vestibular ratio of 0.45 (2 standard deviations below norm for age).

Patient was referred for further medical diagnostic testing for possible fistula, which was substantiated. The fistula was of unknown etiology. Surgical correction was performed. Physical therapy was resumed 1 month following surgery. Reassessment had similar results as presurgical assessment. Physical therapy was provided focused on visual stabilization, adaptation, and balance retraining. Within 2 months, all vertigo and dizziness was eliminated. Reading ability improved such that the child was removed from the learning disability program and placed in a gifted program. Within 6 months, the child resumed all activities, to include playing basketball with peers.

CASE STUDY 24-3

A 9-year-old female with a diagnosis of spastic hemiplegia since birth presented with gait and balance deviations and delayed gross motor functioning. IQ testing was within normal limits, and thus she was placed in a fully integrated classroom with physical and occupational therapy provided 1:1 twice weekly each.

- Neurodevelopmental status: Hypertonicity was evident in the right upper and lower extremity; gait deviations included toe-heel contact on the right, knee flexion during mid-stance on right, lack of arm swing, and wide base of support; child was unable to run or balance in single leg stance on

CASE STUDY 24-3

right with EO or EC; equilibrium reactions were intact but delayed: reliance on tilting reactions in quadruped and kneeling, and reliance on protective extension (stepping) in upright; balance in all positions was challenged by head rotations; gross motor functional level was 30 months. Tonic neck reflexes were easily elicited and affected alignment: ATNR and STNR were evidenced in quadruped with increased arm flexion in the direction of head turning, and inability to lean forward with less than 120 degrees flexion at the hips if head was extended to look forward; tonic labyrinthine evident with inability to assume pivot prone extension. A 1-inch leg length discrepancy was noted with the right leg being shorter.

- Vision and vestibular testing: Intolerance to movement in net swing—fearful and complaints of dizziness; hyperactive nystagmus response on PRNT (90-second duration in room light in either direction); subjectively, patient reported that she felt "upside down" for the duration of the nystagmus following PRNT; child was able to balance in double leg stance with eyes open or closed with minimal sway but could not balance with dome on head and staggered if asked to stand on foam with eyes open or closed or when also using dome (indicative of failure on posturography conditions 4 through 6)[111]; visual tracking intact in all directions with difference of two lines on dynamic visual acuity testing (negative test).
- Because of the lack of dizziness or complaints of vertigo without stimulation, negative DVA test, and the hypersensitivity to movement (prolonged nystagmus), a central vestibular deficit was identified and no further diagnostic testing was performed.

Physical therapy treatment continued as before with the addition of facilitation of integration of tonic reflexes by use of vestibular stimulation on scooter board and vestibular stimulation in net swing with and without visual fixation, gradually increasing velocity and directions of movement to tolerance. Balance training activities were added to include balancing in kneeling and upright on compliant surfaces (high-density foam), the use of Romper Stompers in upright, and swinging baseball bat at ball on "Tee" (rotation with visual fixation on ball). A 3/4-inch full sole lift was placed on the right shoe.

Initially, the child was unable to locate items on a visually complex poster when swinging in a net swing less than 1 Hz. Gradually, this improved until she could locate very small (0.5- to 1-inch diameter) items on the poster while swinging in net swing in all directions in the prone position. Within 6 months, this was also done in sitting. Balance improved tremendously with patient being able to walk on dense foam with eyes open and closed within 9 months. Gait improved with heel-toe progression within 6 months. Gross motor performance increased 24 months in 9 months' time. At the end of 15 months, she could roller skate and participated in all physical activities with peers. Gait deviations were essentially undetectable to the untrained eye. She was discharged from physical therapy services at school.

CASE STUDY 24-4

The child is an 8-year-old male with congenital profound bilateral sensorineural hearing loss of unknown origin. He wears hearing aids and usesAmerican Sign Language for communication. He attends an elementary school for children who are deaf. The child's mother complains that he has always been "clumsy" and has difficulty keeping up in school. He experienced delays in gross motor development of head control at 5 months, independent sitting at 9 months, and walking at 21 months. Typically, developing children develop head control by 3 months, sit independently by 6 months, and take their first steps by 15 months.[85] He reads below age level. The child states that he would like to be able to keep up with his peers while playing soccer.

- The therapist administered the *Canadian Occupational Performance Measure (COPM)*.[105] This test is a semi-structured interview in which the child and/or parent identify the most important "problems" to them. Following the interview, they identified three important problems:
  - falls often during soccer and kickball;
  - has difficulty reading the board at school;
  - inability to walk on curbs or balance beams.

Continued

CASE STUDY 24-4

- The child's mother scored each problem on her perception of "Performance" ("How well does he perform the task on a scale of 0 = unable and 10 = extremely well?") and Satisfaction ("How satisfied are you with his current level of performance on the task on a scale of 0 = not satisfied to 10 = extremely satisfied?"). For Performance and Satisfaction, scores are added and divided by the number of problems (n = 3) yielding a score between 0 (worst) and 10 (best). The Performance score was 4/10 and the Satisfaction score was 3/10.
- *Neurological screen*: Coordination (rapid alternating movements, heel to shin and finger to nose) and strength and range of motion were all normal. No problems were noted with sensation. Muscle tone was overall decreased. Oculomotor exam was normal (smooth pursuit and saccades). No resting or gaze-evoked nystagmus was noted in room light or with fixation blocked.
- *VOR screen*: *Head thrust* was positive bilaterally. Hypoactive response (only a few beats of nystagmus) bilaterally was noted on *ECVCT* (i.e., spin at 0.5 Hz for 30 seconds with eyes closed observe post-rotary nystagmus with fixation removed). *Clinical DVA* was 5.5 lines of difference between Static Visual Acuity (i.e., SVA: visual acuity with the head still) and Dynamic Visual Acuity (i.e., DVA: visual acuity with the head moving at 2 Hz). Normal for his age is less than 3 lines of difference. On the Bucket Test, the child was able to discern when the line was within 1 to 2 degrees off vertical indicating normal perception of vertical, which is normal.
- *Motor Development testing*: On the *Bruininks-Oseretsky Test of Motor Proficiency, 2nd Edition (BOT-2)*, the child scored <10th percentile for bilateral coordination and balance subscales.
- *Balance testing*: On the *Pediatric Clinical Test of Sensory Interaction for Balance* (CTSIB), the child was unable to stand on foam with eyes closed and could only stand for 15 seconds with eyes open. On the *Pediatric Balance Test*, the child had difficulty with items involving standing tandem or on one leg, stepping with alternating feet onto a stool, and forward reach (total score: 50/56). Typically, developing children 7 years of age and older scored 55.2 (SD = 1.74).[90] Single-legged stance time was 8 seconds with eyes opened and 2 seconds with eyes closed. Typically developing children 8 years of age should be able to stand 10 seconds. He reached 5 inches on the *functional reach test*. Typically developing children 8 years of age should be able to reach 9.5 (SD = 4.5) inches.[85] On the *SOT*, he was unable to complete conditions 5 and 6 without stepping or falling. On the *toes up tip test*, he stepped and demonstrated abnormally increased tibialis anterior latency on EMG. Based on these results, the child was referred to an otolaryngologist where rotary chair and VEMP confirmed a diagnosis of bilateral vestibular hypofunction.

Intervention: PT intervention consisted of a 16-week program. The child was treated in the clinic for 30-minute sessions, once per week and given a daily home program. Exercises and progression were similar to what is described in Table 24-4 except that the parent was instructed to carry out specific exercises at home. The home program was done for 10 minutes, twice per day. It focused on reading with head movement for 1 to 2 minutes in pitch and yaw as well as balance exercises (e.g., standing on thick foam, single-legged stance, tandem stance). The child and parent were instructed to keep a log of exercises completed each day. At each session, exercises were progressed as needed.

Outcome: See Table 24-5 for posttest outcomes. The child made substantial gains with motor development and balance. Although he continued to score below published normative data, the child did not step or fall on conditions 5 and 6 of the SOT or on the toes up tip test. He also improved his functional reach by 3". His gaze stability (DVA) improved only slightly. However, the child reported that he was reading better in the classroom. The child and his mother also reported that he was falling less while playing sports or when he was in challenging balance situations.

■ Table 24-5 **CASE STUDY 24-4 PRE AND POST INTERVENTION OUTCOMES**

Name of Test	Result Pre-intervention	Result Post-intervention	Published Normative Data
COPM Performance and Satisfaction	Performance: 4/10 Satisfaction: 3/10	Performance: 6/10 Satisfaction: 8/10	n/a
Clinical DVA test	5.5 lines of difference	4.5 lines of difference	<3 lines of difference between SVA and DVA[14,118]
BOT-2	<10th percentile for balance and bilateral coordination subscales.	50th percentile for balance and bilateral coordination subscales	Standardized test[88]
P-CTSIB (modified)	Foam EO (15 sec); Foam EC (unable) all other conditions 30 sec	Foam EO (30 sec); Foam EC (25 sec)	30 seconds for all conditions[102]
PBS	50/56	54/60 (improved placing feet on stool and single-legged stance)	Children age 8: 55.2 (SD = 1.74)[90]
Single-Legged Stance	EO (8 sec); EC (2 sec)	EO (10 sec); EC (6 sec)	10 seconds EO and EC[87]
Functional Reach	5"	8"	9.5 (SD = 4.5) inches[85]
SOT	Stepped or fell on conditions 5 and 6	No stepping or falling but continued to have scores lower than age matched typically developing peers	See Table 24-3
Toes Up Tip with EMG	Increased latency for tibialis anterior response; stepped	Continued to have abnormally increased latency for tibialis anterior; no stepping	No stepping; adult-like EMG responses.

SUMMARY

Children presenting with vertigo, delays in motor development and postural control, learning disabilities, and/or impaired gaze stability may have vestibular dysfunction. Children suspected of ha ving vestibular dysfunction should be screened for oculomotor , motor, and sensory impairments. The peripheral vestibular system should be screened using the Head Thrust Test, Emory Clinical Vestibular Chair Test, and Bucket Test. Gaze stability and balance should be tested using the Clinical Dynamic Visual Acuity Test, Modified Clinical Test of Sensory Interaction on Balance, and balance subscales of the BOT-2 or PDMS-II. Children with abnormal tests should be referred for diagnostic testing. Gaze stability, balance, and motor impairments can be treated using adaptation and substitution exercises, modified for children.

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CHAPTER 25

Physical Therapy Management of the Older Person with Vestibular Dysfunction

Susan L. Whitney, DPT, PhD, NCS, ATC, FAPTA
Gregory F. Marchetti, PT, PhD

The complaint of dizziness is one of the most common reasons that older adults visit the doctor's office.[1] The incidence of dizziness increases with age and accounts for 1.3% of all visits to internists in people 45 to 64 years old, 2.9% in people older than 65, and 3.8% in people older than 75. Although dizziness can be caused by many different medical conditions, it is estimated that as many as 45% of cases are a result of vestibular disorders.[2] Aktas and colleagues[3] have reported that in 31% of their subjects with hip fracture, vestibular disease was a comorbidity. In 80% of their patients with falls of unknown cause, Pothula and associates[4] found symptoms of vestibular dysfunction. In a recent report of persons who reported dizziness who were over the age of 40 (n = 5,086), there was a 12-fold increase in their odds of falling within the last year.[5] The vestibular insult or injury may be the same as in a younger individual, but the functional consequences may be very different because of the person's comorbid health status. Older adults with vestibular disorders, therefore, often present with very different problems from those of their younger counterparts. This chapter provides information about the normal changes in the vestibular, visual, and somatosensory systems with aging as well as the pathological changes that can occur in each system. Practical suggestions are made as to how older adults may be treated differently because of their age.

Normal Changes of Aging

Vestibular Function

To appreciate the effect of aging on the mechanisms and potential for recovery in vestibular disease, one must understand the normal anatomy and physiology of the vestibular system (see Chapters 1–4). Several concepts are particularly important and will be emphasized here.

Semicircular Canal Function

The input from the receptors in the semicircular canals produces compensatory oculomotor responses (vestibulo-ocular reflex [VOR]) and compensatory postural responses (vestibulospinal reflex [VSR]). In the ideal situation, the eye movement produced by the head movement is equal (but opposite) to the head movement, and the VOR is said to have a gain (eye velocity ÷ head velocity) equal to 1. With aging, there is a decrease in the numbers of both hair cells and vestibular neurons.[6-8] Functionally, VOR gain decreases, resulting in reduced visual compensation in response to head movement. These changes, however, appear to be frequency- and velocity-dependent.[9,10] Baloh and coworkers[11] found that older subjects had lower visual-VOR gain as velocity increased than younger subjects. If the visual-VOR gain decreases with age, especially at

higher velocities,[12] it would lead to greater retinal slip and therefore poorer visual acuity during head movement. The changes in vestibular function have been compared with a progressive bilateral vestibular deficit[10,11,13] and may contribute to the complaints of disequilibrium (an "off-balance" sensation) and to the gait ataxia, without any true vertigo, that many older people have. Paige[12] suggests that this gradual, mild bilateral vestibular loss results in a mismatch in the visual and vestibular mechanisms. Interestingly, Nadol and Schuknecht[13] suggested that degenerative changes might occur in only one labyrinth, producing sudden, severe vertigo or leading to chronic vertigo and disequilibrium.

Another change related to aging is a decreased ability to adapt the gain of the vestibular system.[10] This would affect the ability of the system to adjust to loss of function or other stresses of the system.

Preexisting vestibular disorders can cause balance problems that manifest slightly differently in older individuals. Subclinical vestibular dysfunction may have developed in an older adult when he or she was younger. Then, with the decrease in the number of hair cells and vestibular neurons with rising age, along with the changes in the vestibular and other systems' ability to compensate, the patient experiences symptoms of vestibular dysfunction. Thus, the problems are caused by decompensation rather than to a new vestibular disorder.

Bilateral vestibular disorders are associated with ototoxicity, autoimmune, otologic, and neoplastic disorders plus ototoxicity.[5] The previous conditions that are associated with bilateral vestibular disorders are more common in older than in younger adults. The most common cause of bilateral vestibular loss is antibiotic therapy after a major infection (see Chapters 4 and 23). The presence of visual and somatosensory changes with aging complicates the functional recovery for patient with a bilateral vestibular loss. Older patients who present with instability of gait and functional mobility are more likely than younger patients to require the use of a walker or cane and will always have difficulty walking in the dark.[14,15] These functional deficits may require environmental and behavioral adaptations to prevent falls and injuries in the older person. One practical suggestion is that the older person increase the lighting in the house, especially nightlights and power-outage lights, and that he or she have numerous flashlights available at night so as never to have to walk in total darkness.[14] Installation of grab bars in the bathroom, especially in the tub or shower, has been shown to be helpful in decreasing falls in older persons. Older persons with bilateral vestibular loss often experience limitations in activities and participation, because many persons with bilateral vestibular loss either choose not to drive or by law are limited in their driving abilities.[16-18] Others have suggested that driving is not compromised with persons with bilateral vestibular hypofunction.[19] On the road, driving did not appear to be impaired in three people with long-term bilateral loss (range of bilateral loss was 7 to greater than 20 years).[19]

Utricular and Saccular Function

The presence of calcium carbonate crystals (otoconia) on the maculae of the utricle and saccule make these structures sensitive to the pull of gravity and to linear acceleration. These crystals are held in place by a glue-like substance, and in all people there is a normal degeneration and regeneration of the crystals.[20] In addition, degenerative changes in the otoconia of the utricle and saccule increase with aging,[20,21] with less otoconia present in older adults.[22] This process may contribute to the relatively high incidence of benign paroxysmal positional vertigo (BPPV, see Chapter 20) in the elderly. The incidence of idiopathic BPPV has been reported as peaking in the sixth and seventh decades of life.[23] Baloh and coworkers[11] reported that, of 116 patients aged 70 or older that they saw for complaints of "dizziness," 25% had BPPV. Whitney and colleagues[24] found that 23% of patients referred to a vestibular clinic had the diagnosis of BPPV and a mean age of 61 years.

Oghalai and associates[25] suggest that older persons with undiagnosed BPPV have more reports of falls, are more likely to have had past depression, and have impairments of activities of daily living (ADLs). In their sample of older persons seen in a general medical clinic, 9% had a positive Dix-Hallpike test result, although none of the older adults were being seen for a balance or vestibular disorder. Gamiz and Lopez-Escamez[26] found that 82% of the adults in their survey older than 60 years who were treated for BPPV had a negative Dix-Hallpike test result at 30-day checkup, suggesting that older persons are successfully treated with canalith repositioning. They also reported that older adults with BPPV showed significant changes in the Medical Outcomes Short-Form Medical Survey (SF-36) and short-form Dizziness Handicap Inventory (DHI) values 30 days after undergoing the canalith repositioning maneuver, suggesting that they had improved. Symptoms of BPPV are often missed in the elderly, because the Dix-Hallpike test is often not performed even when the older adult complains of positional dizziness/vertigo.[27,28]

Several theories have been suggested in relation to why older persons appear to have a higher incidence of BPPV than younger persons. BPPV has now been associated with higher rates of mild head trauma and diabetes,[29] osteoporosis and osteopenia,[30] fissures in the otoconia,[21] and giant cell arteritis.[31] All of these conditions are more

common in older adults. Cause-effect relationships with BPPV are very difficult to determine in older adults, because aging affects many systems, and many older people have a tendency to develop BPPV. Gizzi and colleagues[32] have even suggested that the presence of BPPV is five times more common in persons for whom a blood relative also had reported BPPV in the past, suggesting that BPPV may be associated with a genetic link.

A newly developed BPPV subscale of the DHI may be helpful in identifying BPPV in older persons who present to a balance clinic.[24] Determining whether the older patient becomes dizzy or experiences vertigo when moving from supine to sitting position or while rolling over are two of the key questions included in the new subscale; a "yes" answer significantly increases the likelihood that the person has BPPV.

Even if a patient does not present with a diagnosis of BPPV or with complaints consistent with BPPV, it is recommended that the older adult always be assessed for BPPV during the clinical examination, because of its prevalence.[25] Because the Dix-Hallpike test result may be false-negative in some patients, Viirre and coworkers[33] have suggested that clinicians perform the Dix-Hallpike test, then the roll test, and then repeat the Dix-Hallpike test if results of the first two tests are negative. Whitney and associates[24] also suggest repeating the Dix-Hallpike maneuver as well as performing the test at different speeds with older adults to dislodge the otoconia. Also, the examiner is more likely to find evidence of BPPV earlier in the day.[24]

When performing assessment or treatment for BPPV in older persons, however, it is important to consider the possibility that the patient may have cervical spine, lumbar spine, and cardiopulmonary disorders as well. Cervical spine range of motion should be assessed before position testing. Great care should be exercised in extending and rotating the older adult's head because of possible structural changes caused by arthritis or other cervical disorders, or the position used should be modified by tilting the treatment table so the labyrinth is below the horizontal but the neck is not extended. Alternatively, the side-lying test may be used. Also, the patient should be questioned about vertebral basilar compromise, because great care should be used when deciding how to position the older adult to both test and treat BPPV in someone with suspect vertebra-basilar insufficiency. If the patient presents with a long history of cervical disease (e.g., rheumatoid arthritis or Paget's disease), a tilt table is useful during the testing and repositioning maneuvers. For a patient with normal-pressure hydrocephalus, the neurosurgeon should be consulted about possible problems with placing the patient in the head-down position. In addition, Bhattacharyya et al suggest that concern should be exercised with persons with Down syndrome, cervical radiculopathies, ankylosing spondylitis, those with low back pain, and older adults with morbid obesity.[34]

It is important to move more slowly with the repositioning maneuvers in adults older than 80 years and with the adult who has multiple medical problems.[35] The ultimate success rate of repositioning in older adults is similar to that in younger persons but may require more than one visit. We think that the reason for the difference is that it is difficult to obtain sufficient neck extension and/or rotation for an effective treatment in older patients. Care should also be taken with individuals who have metastatic cancer to avoid pathological fractures or who have had a cerebrovascular accident to avoid vascular insufficiency. We often perform the repositioning maneuver with two clinicians present to help the patient with position changes if he or she is severely impaired.

Another special group is older adults with mental and/or physical disabilities. Having a family member present is helpful to assure the older adult that he or she will not be harmed by the maneuvers. It is very frightening for older persons with mental challenges to undergo the canalith repositioning maneuver. As a precaution, we also routinely monitor blood pressure in people older than 75 years who have abnormally high or low blood pressure.

Vestibular Function Tests

It is difficult to determine whether dizziness is truly because of a vestibular deficit without sophisticated testing (see Chapters 10 and 11). The results of these tests in older persons must be interpreted carefully and should be based on age-related normal values.[36] For example, in people in their 90s, caloric testing may document bilateral vestibular loss, but they may not actually have a bilateral vestibular deficit.

Visual Deficits

Visual acuity, the ability to accommodate, and smooth pursuits normally decline with age.[37,38] These normal changes associated with aging can make rehabilitation after a vestibular insult more difficult.

An inability to adjust to the dark has been shown in the literature to be one of the reasons why older adults may fall.[39] Combining the dark adaptation problem with vestibular dysfunction can make it dangerous for older adults with vestibular disorders to move from areas with ample light to darkened areas or vice versa. This change in light has been shown to cause temporary blindness in older adults for more than a minute.[39]

In addition to dark adaptation, visual acuity and contrast sensitivity have been related to falls in older adults[40-44] and may contribute to imbalance after a vestibular disorder. Older adults may have other eye disorders, including cataracts, glaucoma, and macular degeneration that impair vision.[45] Cataracts typically cloud the lens and may cause blurred vision. In patients who have macular degeneration, near and distant vision are affected without adversely affecting peripheral vision.[45] Individuals with glaucoma have difficulty with peripheral vision.[45] Depth perception disorders, such as cataracts in one eye, and double vision make maintaining upright stance more difficult. Multifocal lenses have also been implicated in falls in older persons.[41,44] Lord and associates[44] reported that wearers of progressive, bifocal, or trifocal lenses were more likely to have described a fall, especially descending stairs, as well as more trips and falls outside their homes.

A home inspection would be very helpful for the person with a visual disorder to ensure that the home is free of hazards. The Home Safety Checklist developed by the U.S. National Safety Council is an excellent tool for identifying fall hazards in the home (Box 25-1).[46] Any of the visual disorders described can potentially raise the risk of falls and complicate the patient's rehabilitation course. A home visit by the physical or occupational therapist (low vision specialists) to reduce hazards may be a very helpful intervention for people who have visual impairments.[45]

Somatosensory Changes

Older adults appear to have a greater chance of reduced somatosensory function. Age-related electrophysiological and functional declines in the peripheral nervous system have been described in groups of healthy subjects

Box 25-1

HOME SAFETY CHECKLIST FOR DETECTION OF FALL HAZARDS

Housekeeping

1. Do you clean up spills as soon as they occur?	yes	no
2. Do you keep floors and stairways clean and free of clutter?	yes	no
3. Do you put away books, magazines, sewing supplies, and other objects as soon as you're through with them and never leave them on floors or stairways?	yes	no
4. Do you store frequently used items on shelves that are within easy reach?	yes	no

Floors

5. Do you keep everyone from walking on freshly washed floors before they're dry?	yes	no
6. If you wax floors, do you apply two thin coats and buff each thoroughly or else use self-polishing, nonskid wax?	yes	no
7. Do all small rugs have nonskid backings?	yes	no
8. Have you eliminated small rugs at the tops and bottoms of stairways?	yes	no
9. Are all carpet edges tacked down?	yes	no
10. Are rugs and carpets free of curled edges, worn spots, and rips?	yes	no
11. Have you chosen rugs and carpets with short, dense pile?	yes	no
12. Are rugs and carpets installed over good-quality, medium-thick pads?	yes	no

Bathroom

13. Do you use rubber mat or nonslip decals in the tub or shower?	yes	no
14. Do you have a grab bar securely anchored over the tub or on the shower wall?	yes	no
15. Do you have a nonskid rug on the bathroom floor?	yes	no
16. Do you keep soap in an easy-to-reach receptacle?	yes	no

Continued

Box 25-1

HOME SAFETY CHECKLIST FOR DETECTION OF FALL HAZARDS—cont'd

Traffic Lanes

17. Can you walk across every room in your home, and from one room to another, without detouring around furniture? yes no
18. Is the traffic lane from your bedroom to the bathroom free of obstacles? yes no
19. Are telephone and appliance cords kept away from areas where people walk? yes no

Lighting

20. Do you have light switches near every doorway? yes no
21. Do you have enough good lighting to eliminate shadowy areas? yes no
22. Do you have a lamp or light switch within easy reach from your bed? yes no
23. Do you have nightlights in your bathroom and in the hallway leading from your bedroom to the bathroom? yes no
24. Are all stairways well lighted? yes no
25. Do you have light switches at both the tops and bottoms of stairways? yes no

Stairways

26. Do securely fastened handrails extend the full length of the stairs on each side of stairways? yes no
27. Do rails stand out from the walls so you can get a good grip? yes no
28. Are rails distinctly shaped so you're alerted when you reach the end of a stairway? yes no
29. Are all stairways in good condition, with no broken, sagging, or sloping steps? yes no
30. Are all stairway carpeting and metal edges securely fastened and in good condition? yes no
31. Have you replaced any single-level steps with gradually rising ramps or made sure such steps are well lighted? yes no

Ladders and Stepstools

32. Do you have a sturdy stepstool that you use to reach high cupboard and closet shelves? yes no
33. Are ladders and stepstools in good condition? yes no
34. Do you always use a stepstool or ladder that's tall enough for the job? yes no
35. Do you always set up your ladder or stepstool on a firm, level base that's free of clutter? yes no
36. Before you climb a ladder or stepstool, do you always make sure it's fully open and that the stepladder spreaders are locked? yes no
37. When you use a ladder or stepstool, do you face the steps and keep your body between the side rails? yes no
38. Do you avoid standing on top of a stepstool or climbing beyond the second step from the top on a stepladder? yes no

Outdoor Areas

39. Are walks and driveways in your yard and other areas free of breaks? yes no
40. Are lawns and gardens free of holes? yes no
41. Do you put away garden tools and hoses when they're not in use? yes no
42. Are outdoor areas kept free of rocks, loose boards, and other tripping hazards? yes no
43. Do you keep outdoor walkways, steps, and porches free of wet leaves and snow? yes no
44. Do you sprinkle icy outdoor areas with deicers as soon as possible after a snowfall or freeze? yes no

Box 25-1

HOME SAFETY CHECKLIST FOR DETECTION OF FALL HAZARDS—cont'd

45. Do you have mats at doorways for people to wipe their feet on?	yes	no
46. Do you know the safest way of walking when you can't avoid walking on a slippery surface?	yes	no
47. Do your shoes have soles and heels that provide good traction?	yes	no
48. Do you wear house slippers that fit well and don't fall off?	yes	no
49. Do you avoid walking in stocking feet?	yes	no
50. Do you wear low-heeled oxfords, loafers, or good-quality sneakers when you work in your house or yard?	yes	no
51. Do you replace boots or galoshes when their soles or heels are worn too smooth to keep you from slipping on wet or icy surfaces?	yes	no
Personal Precautions		
52. Are you always alert for unexpected hazards, such as out-of-place furniture?	yes	no
53. If young grandchildren visit, are you alert for children playing on the floor and toys left in your path?	yes	no
54. If you have pets, are you alert for sudden movements across your path and pets getting underfoot?	yes	no
55. When you carry bulky packages, do you make sure they don't obstruct your vision?	yes	no
56. Do you divide large loads into smaller loads whenever possible?	yes	no
57. When you reach or bend, do you hold onto a firm support and avoid throwing your head back or turning it too far?	yes	no
58. Do you always use a ladder or stepstool to reach high places and never stand on a chair?	yes	no
59. Do you always move deliberately and avoid rushing to answer the phone or doorbell?	yes	no
60. Do you take time to get your balance when you change position from lying down to sitting and from sitting to standing?	yes	no
61. Do you hold onto grab bars when you change position in the tub or shower?	yes	no
62. Do you keep yourself in good condition with moderate exercise, good diet, adequate rest, and regular medical checkups?	yes	no
63. If you wear glasses, is your prescription up to date?	yes	no
64. Do you know how to reduce injury in a fall?	yes	no
65. If you live alone, do you have daily contact with a friend or neighbor?	yes	no

After identifying a fall hazard, the hazard should be eliminated or reduced. One point is allowed for each No answer. Score 1 to 7, excellent; 8 to 14, good; 15 and higher, hazardous.

This checklist was developed by the U.S. National Safety Council in cooperation with AARP, Itasca, IL, 1982.
(Used with permission.)

and patients with neuropathological disorders. These age-related somatosensory changes can result in diminished vibratory and passive motion sense and an increase in lower extremity reaction times.[47,48] Pathological changes such as axonal degeneration because of neuropathy result in abnormalities in distal somatosensation.[49,50] These somatosensory changes have functional implications. Ducic and colleagues[51] and Reid and associates[52] have reported that there are changes in static standing balance (sw ay profiles), because one has less sensation distally from peripheral neuropathy.

Older adults have more difficulty sensing vibration in the distal extremities and less sensitivity for detecting smaller monofilaments. Diabetes is one of the most common conditions in older adults that causes changes in distal somatosensation and vision.[53]

Musculoskeletal Deficits

Another potential difficulty in rehabilitating the older adult with a vestibular disorder involves the musculoskeletal system. An assessment of grip strength is one of the most effective ways to obtain an overall idea of strength in older adults.[54] A systematic review indicated low handgrip strength to be associated with several adverse events, including mortality, onset of disability, and adverse outcomes after illness.[55] Older adults may have weakness or even muscle paralysis of various etiologies. Older adults who have preexisting conditions, such as polio and cerebral palsy, *and* in whom late-onset vestibular disease develops are more difficult to treat.

Weakness is very common in the lower extremities, especially the ankles, in older adults. Careful attention to ankle strength is very important in the patient's rehabilitation. The inverted pendulum model of postural control shows that the ankles play a critical role in the maintenance of postural control.[56] Foot and ankle muscles appear to be very weak in many older patients, and strength training may be indicated. Toe flexor strength and toe deformities have been related to increased risk of falling in a recent prospective study.[57] Waddington and coworkers[58] found that training with a wobble board enhanced older adults' ability to discriminate movement at the ankle.

Postural Hypotension

Patients who have vestibular disorders often complain of dizziness and/or imbalance. This dizziness needs to be differentiated from lightheadedness because of postural hypotension associated with changes in position. Postural hypotension and vestibular-induced dizziness can be easily confused if one does not make a careful examination. Typically, patients with postural hypotension become lightheaded or dizzy when standing up, and the symptoms last for seconds. There is also a 20-mm Hg drop in systolic pressure from supine to standing if blood pressure is measured immediately after rising. The patient is asked to lie in supine for up to 10 minutes, then is asked to stand with the blood pressure cuff secured to an extremity. A drop of 20 mm Hg or more indicates postural hypotension.[59] Many drugs commonly taken by older adults can produce postural hypotension, including diuretics and antihypertensive medication.[60] Postural hypotension alone can put a person at risk for falling because of the significant dizziness that the patient experiences when changing positions quickly.[60] Syncope, which can occur from postural hypotension, never occurs exclusively as a result of a vestibular disorder. Another form of postural intolerance is called postural orthostatic tachycardia syndrome (POTS) which occurs when moving from supine to standing resulting in tachycardia.

Cerebellar Atrophy

Older adults may have disturbances in coordination and tend to move slowly. Patients with cerebellar disease appear to improve with balance therapy (see Chapters 29 and 30).[61,62] It is not uncommon for cerebellar atrophy to be a sign of abnormal aging in the older adult. Patients with cerebellar atrophy often do not complain of dizziness, vertigo, or hearing loss. Their chief complaint is often that their balance has been getting worse over a period of years. Working on the rhythm of gait is very helpful for such patients, because their step lengths can be variable, making them unstable while ambulating.

White Matter Disease

Studies have now suggested that older adults who have significant white matter disease are at greater risk of falling.[63-65] This is a relatively new area of research that raises the possibility that white matter disease may be a factor in falls in older persons. These higher-level gait disorders have been associated with disease of the frontal lobe plus their connections with the basal ganglia, brainstem, and cerebellum.[66] Authors have suggested that white matter disease may account for up to 20% to 30% of gait difficulties in older persons.[66,67] Higher-level gait disorders have been described via the following characteristics: a wide-based gait, shuffling steps (small), slow gait speed, difficulty with walking and performing a secondary task, walking with stiff lower extremities, difficulty initiating the gait cycle, and even what has been described as a magnetic gait. Slowed gait speed and semi-tandem stance time have been reported in older adults with prefrontal and cerebellar atrophy.[68] Reduction in white matter correlates with changes in cognitive measures that is often noted on T2-weighted images.[69] As imaging improves, greater evidence will amass related to gait and cognitive functioning in older persons with white matter disease.

Fear of Falling

Another common problem experienced by older individuals who have vestibular disorders is fear of falling (see Chapters 29 and 30).[70] Balance performance and confidence are related in community-living older people.[71] Older adults with vestibular loss often experience fear of falling[72] and, as a result, may reduce their activity level.[73-75] This fear of falling is extremely disabling to older adults and may actually prevent optimal functioning. Tinetti and

associates[73] suggest that therapists work with patients to reduce their fear of falling, thus also enhancing their function. Older adults with vestibular disorders who expressed fear of falling also report a history of depression.[76]

When asking a patient about a fall, the therapist must make sure to be using the same definition of a "fall" as the patient. In a recent systematic review, no standardized method was noted for the definition, measurement, or documentation of injurious falls.[77] The therapist should determine whether the patient has many "near falls"—coming close to, but not actually, hitting the ground. Falls with no known cause are of concern to the therapist and must be investigated further to determine their cause. Noting whether the patient was injured during a fall and required medical attention is also very important to a better understanding of the seriousness of the fall reports.

Attention

The role of attention in postural control in the elderly is important.[78-83] Shumway-Cook and associates,[78] studying postural control in older adults while standing, have found that older adults allocate their resources differently from younger adults. These investigators found that the older adults' balance was more affected than the young adults' when they were concurrently performing a balance task and a simple cognitive task. Those older people who had a history of falling had significant changes in balance, as measured by center of pressure. Ludin-Olssen and colleagues[79] determined, in the nursing home setting, that older adults who talked as they walked with assistive devices were more likely to fall than those who did not talk as they walked.[79,83]

The concept of "attentional resources" is interesting, and one that can be incorporated into practice. We now instruct older patients who have great difficulty walking in the clinic to try not to walk and talk at the same time. They are instructed to "stop walking while talking," to paraphrase the title of the Ludin-Olssen article.[79] Recent evidence suggests that persons with bilateral vestibular loss, while walking and talking, have poorer gait performance compared with age matched controls.[84]

Depression

Older adults who have vestibular disorders may be experiencing clinical depression. A simple screening examination can be performed with the Geriatric Depression Screening Scale.[85] This scale consists of 30 yes-or-no questions asking the patient to answer how he or she has felt over the past week. The test is simple to use and can help the therapist decide whether to make a mental health referral.

Both cognition and depression affect the ability of the patient to follow through with an exercise program. The older adult who is depressed or who has little support at home may have to be seen more frequently in the clinic and will need closer monitoring by the therapist. The patient who displays an indifferent or negative attitude toward therapeutic intervention should not be merely dismissed as "lacking motivation." Coexisting depression may be the cause of the indifference. Anxiety has been reported to be more common than depression in persons with vestibular disorders.[86] Jacob[87] and Clark and associates[88] have suggested that anxiety is strongly related to vestibular dysfunction. The Dizziness Handicap Inventory appears to relate to the Beck Depression Inventory and may be helpful for screening persons with vestibular disorders who might be experiencing depression.[89]

Risk of Falling in Older Adults with Vestibular Disorders

The actual risk of falling in older adults who present to a vestibular clinic is unknown. In a sample of 247 persons who presented to a vestibular clinic with a mean age of 62 years, 36.8% reported having one or more falls in the last 6 months.[90] Various studies have reported a reduction in fall rates after physical therapy intervention.[14,91-93]

There are standardized tools that one can use to determine risk of falling in older adults. Specific tools that can be used to assess balance are the Berg Balance Scale (BBS),[94-101] the Dynamic Gait Index,[100,102] and the functional reach test.[103-108] The BBS has been used extensively to assess balance in older adults who have Parkinson's disease or stroke or who are frail (Box 25-2). It has been validated for use in persons with vestibular dysfunction.[101] This 14-item examination assesses the patient's balance in increasingly difficult positions and has a maximum score of 56. As the scores decrease from 56 to 36, the risk of falling increases.[100] Shumway-Cook and associates[100] determined that scores of 36 and lower on the BBS relate to 100% risk of falls in community-living older adults.

Others have reported that older adults who had scores greater than 45 were considered less likely to fall than those who scored below the cutoff score.[109] The BBS and the mini-BESTest were recently compared and neither tool demonstrated ceiling or floor effects in a group of participants with a mean age of 66.[110]

The Dynamic Gait Index consists of eight gait tasks that have the older adult move his or her head while walking, walk over and around objects, stop quickly, change speeds, and go up and down stairs.[102] It has been shown to be very helpful in identifying fall risk and is reliable.[91,111,112]

Box 25-2

BERG BALANCE SCALE

1. Sitting to Standing

Instruction: Please stand up. Try not to use your hands for support.

Grading: PLEASE MARK THE LOWEST CATEGORY THAT APPLIES.

(4)	(3)	(2)	(1)	(0)
able to stand with no hands and stabilize independently	able to stand independently using hands	able to stand using hands after several tries	needs minimal assist to stand or to stabilize	needs moderate or maximal assist to stabilize

2. Standing Unsupported

Instruction: Stand for 2 minutes without holding.

Grading: Please mark the lowest category that applies.

(4)	(3)	(2)	(1)	(0)
able to stand safely 2 min.	able to stand 2 min with supervision	able to stand unsupported	needs several tries to stand 30 sec	unable to stand 30 sec unassisted

If Subject Able to Stand 2 Min Safely, Score Full Marks for Sitting Unsupported. Proceed to Position Change Standing to Sitting.

3. Sitting Unsupported Feet on Floor

Instruction: Sit with arms folded for 2 minutes.

Grading: please mark the lowest category that applies.

(4)	(3)	(2)	(1)	(0)
able to sit safely and securely 2 min	able to sit 2 min under supervision	able to sit 30 seconds	able to sit 10 sec	unable to sit without support 10 sec

4. Standing to Sitting

Instruction: Please sit down.

Grading: Please mark to lowest category that applies.

(4)	(3)	(2)	(1)	(0)
sits safely with minimal use of hands	controls descent by using hands	uses back of legs against chair to control descent	sits independently using uncontrolled descent	needs assistance to sit

5. Transfers

Instruction: Please move from chair to bed and back again. One way toward a seat with armrests and one way toward a seat without armrests.

Grading: Please mark the lowest category that applies:

(4)	(3)	(2)	(1)	(0)
able to transfer safely with minor use of hands	able to transfer safely; definite need of hands	able to transfer with verbal cuing and/or definite need of hands	needs one person to assist	needs two people to assist or supervise to be safe

Box 25-2

BERG BALANCE SCALE—cont'd

6. Standing Unsupported with Eyes Closed

Instruction: Close your eyes and stand still for 10 seconds.

Grading: Please mark the lowest category that applies.

(4)	(3)	(2)	(1)	(0)
able to stand 10 sec safely	able to stand 10 sec with supervision	able to stand 3 sec	unable to keep eyes closed 3 sec but stays steady	needs help to keep from falling

7. Standing Unsupported with Feet Together

Instruction: Place your feet together and stand without holding.

Grading: Please mark the lowest category that applies.

(4)	(3)	(2)	(1)	(0)
able to place feet together independently and stand 1 min safely	able to place feet together independently & stand for 1 min with supervision	able to place feet together independently but unable to hold for 30 sec	needs help to attain position but able to stand 15 sec feet together	needs help to attain position and unable to hold for 15 sec

The Following Items Are to Be Performed While Standing Unsupported.

8. Reaching Forward with Outstretched Arm

Instructions: Lift arm to 90 degrees. Stretch out your fingers and reach forward as far as you can. (Examiner places a ruler at end of fingertips when arm is at 90 degrees. Fingers should not touch the ruler while reaching forward. The recorded measure is the distance forward that the fingers reach while the subject is in the most forward lean position.)

Grading: Please mark the lowest category that applies.

(4)	(3)	(2)	(1)	(0)
can reach forward confidently >10 inches	can reach forward >5 inches safely	can reach forward >2 inches safely	reaches forward but needs supervision	needs help to keep from falling

9. Pick Up Object from the Floor

Instructions: Pick up the shoe/slipper that is placed in front of your feet.

Grading: Please mark the lowest category that applies.

(4)	(3)	(2)	(1)	(0)
able to pick up slipper safely and easily	able to pick up slipper but needs supervision	unable to pick up but reaches 1–2 inches from slipper & keeps balance independently	unable to pick up and needs supervision while trying	unable to try/ needs assist to keep from falling

Continued

Box 25-2

BERG BALANCE SCALE—cont'd

10. Turning to Look Behind over Left and Right Shoulders

Instruction: Turn to look behind you over or toward left shoulder. Repeat to the right.

Grading: Please mark the lowest category that applies.

(4)	(3)	(2)	(1)	(0)
looks behind from both sides and weight-shifts well	looks behind one side only; other side shows less weight shift	turns sideways only but maintains balance	needs supervision when turning	needs assist to keep from falling

11. Turn 360 Degrees

Instruction: Turn completely around in a full circle. Pause, then turn a full circle in the other direction.

Grading: Please mark the lowest category that applies.

(4)	(3)	(2)	(1)	(0)
able to turn 360 deg safely in <4 sec each side	able to turn 360 deg safely one side only in <4 sec	able to turn 360 deg safely but slowly	needs close supervision or verbal cuing	needs assistance while turning

Dynamic Weight Shifting While Standing Unsupported.

12. Count Number of Times Step Touch Measured Stool

Instruction: Place each foot alternately on the stool. Continue until each foot has touched the stool four times.

Grading: Please mark the lowest category that applies.

(4)	(3)	(2)	(1)	(0)
able to stand independently and safely and complete 8 steps in 20 sec	able to stand independently and complete 8 steps >20 sec	able to complete 4 steps without aid/with supervision	able to complete >2 steps; needs minimal assist	needs assistance to keep from falling/unable to try

13. Stand Unsupported One Foot in Front of the Other Foot

Instruction: Place one foot as close as possible in front of the other foot.

Grading: Please mark the lowest category that applies.

(4)	(3)	(2)	(1)	(0)
able to place feet tandem and holds 30 sec	able to place one foot ahead and holds 30 sec	takes small step independently; holds 30 sec	needs help to step in place; holds 15 sec	loses balance while stepping or standing

Box 25-2

BERG BALANCE SCALE—cont'd

14. Stand on One Leg

Instruction: Please stand on one leg as long as you can without holding onto anything (knee does not have to be bent).

Grading: Please mark the lowest category that applies.

(4)	(3)	(2)	(1)	(0)
able to lift leg independently; able to hold for >10 sec	able to lift leg independently; or needs assist holds 5 to 10 sec	tries to lift leg; unable to hold 3 sec; remains standing independently	needs help to step in place; holds 15 sec	unable to; tries or needs assist to prevent falling

Total Score _____

Maximum Score _____

Berg K, Wood-Dauphinee S, Williams J, Gayton D. Measuring balance in the elderly: preliminary development of an instrument. *Physiotherapy Canada*. 1989;41(6):304-311.

Another tool that is helpful for assessing risk of falls in older individuals is the functional reach test.[103] The test was developed in older male veterans. It yields a difference score in the patient's willingness to reach forward without taking a step. To be able to perform the test, the patient must be able to stand for 30 seconds without support in flat or no shoes and must have at least 90 degrees of shoulder flexion. Patients are typically instructed to raise the arm, make a fist, and then reach forward along a yardstick as far as they can without touching the wall or the yardstick. They are permitted to use any strategy they choose to complete the trial. Duncan and associates[104] have determined that scores of 6 inches or less on the functional reach test show a significant increase in the risk of falling in older adults. Individuals who reach between 6 and 10 inches are at moderate risk for falling.[104] This test has some drawbacks. Functional reach is related to height; taller patients have longer functional reach scores than those who are shorter in stature. Functional reach scores have been shown to change over the course of rehabilitation.[108] Functional reach is sometimes administered to the patient who has dizziness,[105] although Wernick-Robinson and colleagues[113] have suggested the test may have little value in persons with vestibular dysfunction. The functional reach test appears to be helpful in older adults who complain of balance problems, but it may have less discriminative value in persons with vestibular disorders.

Questionnaires for Balance Assessment

The Activities-Specific Balance Confidence (ABC) Scale is a 16-item questionnaire that can be completed by the patient or administered by the caregiver (Box 25-3).[114,115] The patient's perceived confidence in performing 16 activities that are performed in and outside the house are rated by the patient from a range of 0 (no confidence) to 100 (100% confident). Scores that are closer to 100 are a better score on this scale. The ABC scale and the DHI have been shown to have a moderately negative correlation ($r = 2.64$), indicating that the ABC scale is a valid tool for use in persons with vestibular dysfunction.[116] The ABC scale has shown to be sensitive to change over the course of rehabilitation.[92,117,118]

The ABC scale has been compared with the Modified Falls Efficacy Scale.[75] Both scales are sensitive tools that can be used in community-based older adults, and both discriminate individuals who are high-functioning from those who are low-functioning. Lajoie and Gallagher[119] have reported that older adults who score 66% or less on the ABC are at high risk for falling. Myers and coworkers[114] previously reported that persons who scored less than 80% on the ABC scale were somewhat impaired and that those who scored less than 50% were often home-bound individuals. Older adults should be able to perform most of the 16 ABC activities with great confidence.

Box 25-3

THE ACTIVITIES-SPECIFIC BALANCE CONFIDENCE (ABC) SCALE

For each of the following activities, please indicate your level of self-confidence by choosing a corresponding number from the following rating scale:

0% 10 20 30 40 50 60 70 80 90 100%

no confidence completely confident

"How confident are you that you will not lose your balance or become unsteady when you. . .

1. . . . walk around the house? _____%
2. . . . walk up or down stairs? _____%
3. . . . bend over and pick up a slipper from the front of a closet floor? _____%
4. . . . reach for a small can off a shelf at eye level? _____%
5. . . . stand on your tiptoes and reach for something above your head? _____%
6. . . . stand on a chair and reach for something? _____%
7. . . . sweep the floor? _____%
8. . . . walk outside the house to a car parked in the driveway? _____%
9. . . . get into or out of a car? _____%
10. . . . walk across a parking lot to the mall? _____%
11. . . . walk up or down a ramp? _____%
12. . . . walk in a crowded mall where people rapidly walk past you? _____%
13. . . . are bumped into by people as you walk through the mall? _____%
14. . . . step onto or off of an escalator while you are holding onto a railing? _____%
15. . . . step onto or off an escalator while holding onto parcels such that you cannot hold onto the railing? _____%
16. . . . walk outside on icy sidewalks? _____%

Instructions to Participants

For each of the following, please indicate your level of confidence in doing the activity without losing your balance or becoming unsteady by choosing one of the percentage points on the scale from 0% to 100%. If you do not currently do the activity in question, try and imagine how confident you would be if you had to do the activity. If you normally use a walking aid to do the activity or hold onto someone, rate your confidence as if you were using these supports. If you have any questions about answering any of these items, please ask the administrator.

Instructions for Scoring

The ABC is a 16-point scale, and ratings should consist of whole numbers (0 to 100) for each item. Total the ratings (possible range = 0 to 1600) and divide by 16 to obtain each subject's ABC score.

If a subject qualifies his/her response to items #2, #9, #11, #14, or #15 (different ratings for "up" versus "down" or "onto" versus "off"), solicit separate ratings and use the lowest confidence of the two (as this will limit the entire activity—for instance, likelihood of using the stairs).

From: Powell and Myers, 1995.[86] Reprinted with permission of the Gerontological Society of America, 1030 15th Street, NW, Suite 250, Washington, DC 20005, via Copyright Clearance Center, Inc.

The Vestibular Activities of Daily Li ving (VADL) Scale is another questionnaire that is helpful in determining the functional capabilities of people living with vestibular disorders.[120,121] The VADL scale is further discussed in the chapter by Cohen on disability (see Chapter 7).

The Modified Fast Evaluation of Mobility, Balance, and Fear Baseline (MFEMB AF) questionnaire is e xtremely helpful in determining risk f actors for falling (Box 25-4).[122,123] The therapist fills out "yes" or "no" answers to the risk f actor questions either as he or she

Box 25-4

MODIFIED FAST EVALUATION OF MOBILITY, BALANCE, AND FEAR BASELINE QUESTIONNAIRE

Name_____ Age____/____/____ Gender____ Height____ Weight_____
Blood Pressure _____
Lives at Home, Alone_____ Lives with Somebody_____ Lives in an Institution_____

Risk Factors

	YES	NO
1. Needs aid for two (or more) basic activities of daily living (washing, cooking, dressing, walking, continence, feeding)	____	____
2. Needs aid for two (or more) instrumental activities of daily living (money management, shopping, telephone, medications)	____	____
3. Has had a fracture or articular problems at hips, knees, ankles, feet	____	____
4. Has visible articular sequela in the mentioned joints	____	____
5. Uses a walking device (e.g., cane, walker)	____	____
6. Limits physical activity to basic activities of daily living at home	____	____
7. Self-defines as anxious	____	____
8. Complains of vertigo	____	____
9. Complains of imbalance	____	____
10. Makes complaints suggesting an existing postural hypotension	____	____
11. Fell one or two times in the current year	____	____
12. Fell more than twice in the current year	____	____
13. Required nursing after the fall	____	____
14. Had a fracture after the fall	____	____
15. Is afraid of falling in general	____	____
16. Is afraid of falling indoors (e.g., bathtub, kitchen)	____	____
17. Is afraid of falling outdoors (e.g., bus, stairs, street)	____	____
18. Avoids going outside for fear of falling	____	____
19. Presents three or more somatic pathologies that require regular medical supervision	____	____
20. The pathologies require home-based medical-social supervision	____	____
21. Shows a specific pathology likely to induce falls: • neurological (e.g., cancer, peripheral neuropathy, multiple sclerosis, lupus) • cardiovascular (e.g., postural hypotension) • musculoskeletal (e.g., total joint replacements, arthritis) • sensory (e.g., visual impairment) • other (amputation, Parkinson's disease, Alzheimer's disease)	____	____

Continued

Box 25-4

MODIFIED FAST EVALUATION OF MOBILITY, BALANCE, AND FEAR BASELINE QUESTIONNAIRE—cont'd

22. Takes medications that are potentially dangerous in regard to falls: ____ ____
    - hypotensives
    - neuroleptics
    - hypnotics/anxiolytics
    - antiarrhythmics
    - antiparkinsonians
    - analgesics/anti-inflammatory drugs
    - various vasoregulators

Risk Factors (= total of "yes" answers): _____

Task Completion

Scores are determined for fear, pain, mobility difficulties, and lack of strength for each of the following:

TASK	SCORE (3, 2, 1)
1. Sitting on a chair, with folded arms, raises both legs horizontally	____
2. Sitting on a chair with armrests, stands up without aid, without using banister	____
3. Sitting on a chair, stands up without aid, walks five steps, turns around, goes back and sits down	____
4. One-footed standing (left foot): stands on left foot without aid during 5 seconds minimum	____
5. Repeat with one-footed standing (right foot)	____
6. Romberg Test: stands with heels together, eyes closed, remains steady for 10 seconds	____
7. Squatting down: without aid, squats down until buttocks reach knee level, then stands up	____
8. Picking up a pencil from the ground without aid or support	____
9. Standing jumping without losing balance, over a distance equal to one's own foot	____
10. Stepping over an obstacle (foam or cardboard, 10-cm wide × 15-cm high) without touching it; the foot to arrive past the obstacle at a distance equal to its own size (left)	____
11. Repeat with overstepping to the right	____
12. Shoving forward to trunk; subject to remain steady following a nudge between shoulder blades (examiner's arms stretched out, nudge realized by a sudden bending of hand on trunk)	____
13. Repeat with shoving backward (nudge on the sternum)	____
14. Climbing stairs without losing balance, without aid or using banister (five steps minimum)	____
15. Repeat with descending stairs (five steps minimum)	____
16. Transfer from standing-kneeling (both knees on the ground); stable, no assistance for rising	____
17. Managing the "eyes-closed forward fall"; the subject lets himself/herself fall, eyes closed, onto the examiner standing 50 cm from him/her	____
18. Repeat with eyes-closed backward fall	____

FEMBAF TOTAL TASK COMPLETION SCORE: ____

FEMBAF TOTAL SUBJECTIVE COMPLAINT SCORES:

fear _____ pain _____ mobility difficulties _____ lack of strength _____

3 = successfully completed without imbalance

2 = task initiated but unsteady or partially completed

1 = unable to perform or initiate task

From DiFabio and Seavy, 1997.[94] With permission of the American Physical Therapy Association.

interviews the patient or after having obtained information from the patient's medical chart. This questionnaire has a comprehensive list of fall risk questions, answers to which help guide intervention.

The Vestibular Activities and Participation (VAP) scale[124] was developed using only activity and participation items from the International Classification of Functioning (ICF). The VAP consists of 55 items but is currently undergoing analysis to determine if the scale can be shortened. The ICF core set for vertigo, dizziness, and balance disorders was recently developed at a consensus in 2012, and the core sets should have value in future design of studies so that the world is using the same terminology for function.[125] The largest number of items chosen were in the activities and participation area (40) followed by environmental items (29), body functions (25), and body structures (6).[125]

Dizziness Assessment

The Dizziness Handicap Inventory (DHI) is extremely helpful in determining what type of intervention will most benefit the patient with vestibular dysfunction.[72] This tool helps determine the self-perceived handicap of the individual completing the form. The patient answers 25 questions related to dizziness and/or handicap. There are three subdivisions of the test, and sub-scores can be calculated. The test has been divided into emotional, physical, and functional subsets. Information from this tool can significantly direct the treatment of the patient. If the patient checks only the physical symptoms, one needs to look at assessing positions and specific movements that predispose or increase the patient's dizziness and/or falls.

Older adults who have lost mobility as a result of dizziness often check a significant number of the emotional questions on the DHI. One then must determine the actual activity level of the person. Scores on the DHI have also been related to falls in persons with vestibular disorders.[118] Scores of 60 or higher were associated with increased reports of falls in persons with vestibular disorders. Use of the SF-36 form can be helpful in assessing the activity level of the patient; the DHI, however, is more responsive to changes after a 6- to 8-week course of vestibular rehabilitation.[126,127]

Typical Balance Tests

Gait speed is the most powerful measure of balance available and should be recorded on all older adults seen in a balance clinic.[128] A recent prospective study of 34,000 people over the age of 65 suggests that gait speed, age, and gender can predict how long a person will live, with faster gait speeds related to better health.[128] Others have suggested that faster gait speeds improve survival rates at 8-year follow-up.[129] Generally, gait speed is recorded from 3 to 5 meters with a stop watch.

If the patient is complaining of balance problems, the single-leg stance (SLS), Romberg, and tandem Romberg tests are also performed. Bohannon and associates[130] have found that SLS times significantly decrease as people get older.[130] Single-leg stance, Romberg, and tandem Romberg tests help the therapist determine what kind of functional movements might be difficult for the patient.[130-134] Generally, the patient who is unable to stand in SLS usually has great difficulty going up and down stairs without holding onto the railing. Such a patient may also have strength deficits if unable to stay in SLS. The strength deficits can be determined through further testing. The patient who has difficulty with the Romberg and tandem Romberg test is often challenged while walking through tight spaces. Standing with the feet close together may be very destabilizing for such a patient. Some older adults may live in homes with narrow hallways or small rooms with little clearance, and the therapist must consider the patient's environment in designing the treatment program.

The Short Physical Performance Battery (SPPB) was developed to assess risk of falling in older adults.[135] The SPPB has 3 components: (1) the Romberg, semi-tandem Romberg, tandem Romberg, (2) repeated sit to stand, and (3) gait speed. Scores range from 0 to 12, which has been norm-based on over 10,000 older adults (higher scores indicate better function).[136] The SPPB is easy to complete and provides important information about overall health, balance, and strength.

Home Assessment

Preparing the patient to function independently in his or her own home is very important. Occasionally, a home visit is necessary for older persons with vestibular dysfunction, if the therapist is concerned about their safety. Determining how many stairs the patient must ascend or descend is critical. Some extrinsic environmental hazards that have been identified are items such as poor lighting, uneven or slippery surfaces, loose rugs, steep stairs, objects in the pathway, long bathrobes, inappropriate furniture, and lack of handrails, especially in the bathroom.[137]

Modification of the home environment so that the patient does not have to reach excessively either up or down to perform ADLs is helpful. The investigators at the Center for Studies in Aging in Toronto developed a device to assist persons in the bathroom or kitchen who are very unsteady. The device, made of a material that makes it easy to grip, is secured easily without damaging walls. Persons

who have difficulty reaching without holding on may significantly benefit from this type of appliance in the home. Primary care physicians, especially in managed care situations, should be made aware that such services are available to their patients.

Many of the older adults seen in our clinic with complaints of dizziness and balance dysfunction do not have a specific diagnosis. A total vestibular workup and a neurology assessment of such patients are helpful and should be performed so that the physical therapist knows what interventions may be most effective.

Driving Function in Older Adults

According to the National Highway Traffic Safety Administration (NHTSA), the proportion of drivers in the United States who were age 65+ increased 13% between 2000 and 2009. Older drivers accounted for 12% of fatal traffic crashes and 8% of all injuries nationwide.[138] Fatal crash rates per mile traveled increase starting at age 75 and increase significantly after age 80.[139]

Many of the age-related issues that can contribute to increased likelihood of imbalance and falling, such as vision, cognition, physical function, and medical factors, may also contribute to the decline in driving skills.[140] Because driving helps older adults remain mobile and independent, issues related to driving safety often create difficult decisions for older adult patients, their families, and clinicians. These issues are further complicated by a lack of precise, easily administered and validated clinical screening tools for driver safety, insufficient resources and expenses for on-road evaluation, and poorly understood guidelines for clinicians in reporting unsafe drivers that differ between states.

Against this background of clinical uncertainty, the American Medical Association and the NHTSA have jointly published guidelines that can assist physicians and other clinicians to identify impairments that may limit driving safety.[141] These guidelines include the Assessment of Driving Related Skills (ADReS). The ADReS is an easily administered battery that allows screening of vision (acuity and peripheral fields), cognition (clock drawing and trail-making) and motor/somatosensory function (walking speed, strength, range of motion, and proprioception). Scoring standards for the ADReS are provided as well as recommendations for further evaluation on substandard patient performance. It is important to note that results of ADReS screening have not been specifically validated against crash outcomes. Thus, the results cannot be used to specifically infer risk of a future crash but does identify limitations that could potentially increase crash risk. The complete guideline documentation with description of state-by-state unsafe driver reporting standards can be obtained at http://*www.ama-assn.org/go/olderdrivers*. Cohen et al[142] have reported that some persons with vestibular disorders have difficulty with driving, yet a recent report of 3 persons with bilateral vestibular loss over the age of 60 suggests that people with bilateral loss may be capable of driving without additional risk. When uncertain about the capability of the person's ability to drive, a driver examination is warranted.

Duration of Treatment

Older adults frequently need to be treated for a longer time than a younger patient. The longer duration of treatment is related to the number of risk factors present in such patients and to their fear. They may be seen on a more traditional schedule of one to three times per week because of their multiple medical problems and the risk of falling when unsupervised at home. Many older patients have difficulty being transported to physical therapy. This fact can complicate rehabilitation; the older adults' cancellation rate is often higher. If their transportation system breaks down or if the weather is bad, many older adults will be forced to cancel their appointments; referral to a local agency for the aging may be indicated to help the patient obtain dependable transportation. Evidence now suggests that treatment outcomes in older adults are similar to those in younger persons with the same vestibular disorder.[117]

What to Do Once the Risk Factor Has Been Identified in an Older Adult

After the older person with vestibular dysfunction has been assessed, it is important to determine what problems identified in the evaluation must be addressed, referral being one alternative. Patients who have visual problems should be referred to an appropriate physician for further eye testing. People with undiagnosed vestibular disorders should be referred to a neurologist or otolaryngologist. If a neuropathy is suspected, a referral to a neurologist, a physical therapist specializing in electromyography, or a physiatrist is recommended.

The physical or occupational therapist can also provide an environmental assessment with specific recommendations, determine whether the patient needs an assistive device, and teach the patient about safety and clothes to wear to reduce the risk of falling. In addition, shoe type and wear can be determined, and recommendations can be made to the patient.The older adult who may

have chosen to wear inappropriate shoes can sometimes be counseled to change.

During the environmental assessment, lighting in the patient's home may be identified as a major risk factor. Many older adults use low-wattage light bulbs or keep the lights off most of the day to save money. Use of nightlights is strongly suggested, especially in the bathroom. Motion detector lights, which are automatically activated when one passes through the plane of the sensor, may also have some value. Additionally, the layout of some patients' homes may cause significant changes in contrast of light levels, which has been identified as a contributor to falls.[42,43] It is also advisable to have battery-operated lights that will come on when there is a power outage placed around the house. Proper lighting must be addressed with the patient and family.

Motor weakness is most often assessed by performance of a manual muscle test or through the use of a dynamometer. Strength deficits can be addressed, because older patients have the potential to improve their strength, although it may take 6 to 10 weeks for improvement to be evident.[143-145] Range of motion is a major factor that can be improved through rehabilitation. Patients can be significantly at risk for falls if they lack adequate distal range of motion in their feet.[146] Having normal plantar flexion and dorsiflexion is extremely helpful in preventing falls and achieving normal gait, because the feet are the only part of the body touching the ground when one is walking. Assessing flexibility of the toes and foot musculature may be of added benefit; having strong dynamic stabilizers distally may make the patient more stable.[57] If the patient has an extremely immobile foot, performing normal balance reactions will be difficult.

CASE STUDY 25-1

Mrs. H is a 91-year-old woman referred to physical therapy with a diagnosis of bilateral BPPV. She has been seen by a neurotologist who wants to schedule Mrs. H for a joint visit with himself and the physical therapist for repositioning.

The patient is a well-oriented and extremely pleasant older woman. Her chief complaint is that she became very dizzy 3 weeks ago when she looked up at her clock at home and also when she sat up or went from sitting to lying down. Her daughter is very concerned and worried about her mother, stating several times during the examination that she believes Mrs. H should move in with her. Mrs. H lives alone in a small one-bedroom apartment. She normally takes the van that leaves daily from her apartment complex to the grocery store, and she loves to shop! Mrs. H cleans her own apartment but has someone come in once a month to do the heavy cleaning. She arrives at the outpatient clinic carrying a straight cane while seated in a wheelchair. Patient reports that she does not use a cane in her apartment and that she has used a cane elsewhere for the past 4 years. She holds onto furniture as she ambulates around the apartment, and rarely uses a wheelchair except for long distances when a wheelchair is available. She reports that when she shops, she uses the shopping cart like a wheeled walker.

Mrs. H is taking no medications except vitamins but does have a 39-year history of Paget's disease. Her laboratory findings were as follows:

Caloric testing: severely reduced responses bilaterally with absence of iced water responses. Oculomotor screening is normal. Rotational chair response, abnormal with moderately decreased gain and a mild directional preponderance. Positional testing is normal. She is not ataxic and does not have oscillopsia.

Patient's timed "Up & Go" score is 30 seconds. She moves slowly while carrying her straight cane. Patient has a very kyphotic posture. She has decreased neck and shoulder range of motion, and her overall strength is F+ to G–.

The patient has already been diagnosed with bilateral BPPV. She is more symptomatic in the left Dix-Hallpike position than in the right, so the left ear is treated first. Four people are present for the repositioning, including the physician, because of the patient's age and Paget's disease. Paget's disease produces excess bone, which can result in narrowing of the vertebral foramen. It is decided to use a high-low table with two movable parts. Her trunk and head are lowered as a unit and at the same time her feet are elevated to put her in the Trendelenburg position. The patient is initially brought down to the left and then is log-rolled from her left side to her right side. Movements are coordinated among the persons helping to perform the maneuver. Her head is slowly brought up as her feet are returned to the horizontal position. This positioning avoids excessive neck extension and excessive torque to her back during the modified canalith repositioning maneuver. Infrared goggles are in place throughout the procedure. The patient has classic torsional and upbeat nystagmus that fatigues within 20 seconds. The second time she

Continued

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is repositioned during the same session, she has no symptoms. She is told to stay upright and to not move her head up or down for 1 hour.

Patient is scheduled to return in 1 week for repositioning of the other ear; she could not make it back in any sooner because of her daughter's schedule. When the patient returns, there is no evidence of BPPV in either ear. She can look up to her clock at home and lie down without symptoms. She has returned to her normal shopping excursions and says that she feels great.

Her daughter is very concerned about how active her mother is and wants her to stop many of her activities. The daughter is strongly encouraged to allow her mother to stay active and to enjoy her trips out of her apartment.

During her last visit, Mrs. H is instructed in lower extremity strengthening exercises so that she can maintain her strength distally. Patient is discharged after being seen for two physical therapy visits. She no longer has any dizziness, and she is satisfied with her walking abilities.

CASE STUDY 25-2

Mrs. M was a 68-year-old woman seen in physical therapy with a presenting diagnosis of multisensory deficit. Mrs. M was well oriented and very cooperative. She stated that she had been having difficulty walking and that she had fallen twice within the last few weeks. Patient was seen by an otolaryngologist because of her falling. Quick head movements and bending made her unstable.

Her past medical history included a silent heart attack, hypertension, cirrhosis of the liver without a history of alcoholism, mastoiditis, obesity, claustrophobia, uterine cancer, tinnitus, and stomach ulcers. In addition, the patient took medication for her knee arthritis. Past surgical history included a hysterectomy and two operations for cancer.

Mrs. M was taking the following medications: potassium chloride, famotidine, aspirin, a multivitamin (Centrum Silver), and furosemide.

Vestibular testing results showed that she had a normal oculomotor battery, normal static positional testing, severely reduced vestibular responses bilaterally with present iced water caloric responses, and reduced gain on rotational chair testing. Patient had two previous infections necessitating IV antibiotics. She had osteomyelitis of a toe 10 years ago and again 2 years ago, which were treated with IV antibiotics. Furosemide can be ototoxic, so the patient was counseled to consult with her physician about whether another medication could control her lower extremity swelling without the same side effects.

The patient stated that she occasionally got dizzy with changing positions. Her DHI score was 12/100. She stated that the onset of her gait instability was gradual and that it was getting worse.

Patient lived alone in a condominium on one floor with an elevator in the building. She formerly worked as a superintendent of schools in her area. Mrs. M was widowed at an early age and raised two children alone.

Walking with head turns and quick head movements increased her symptoms. Her ABC score was 51%. She did not use an assistive device.

She had fallen twice in the last few weeks. She tripped over a box the first time, and the second time she got tangled in a chair cover and lost her balance. Patient stated that she also has had many near falls. She stated that she almost fell the morning of the evaluation while sitting down on the commode. Mrs. M reported difficulty getting up from the floor.

Patient's strength and range of motion were generally within normal limits for her age. She had diminished vibration sense but had intact proprioception distally at her ankles. She became short of breath with exertion during functional activities and gait.

Mrs. M's timed "Up & Go" score was 12.5 seconds. Her repeated 5 times sit-to-stand test score was 16.2 seconds. Her Sensory Organization Test composite score on the EquiTest (Neurocom International, Inc.) was 77. Her Berg Balance Score was 55/56, and her Dynamic Gait Index score was 19/24. The patient was able to stand in SLS for 15 seconds on the right and 10 seconds on the left.

Overall, it appeared that the patient's balance was fairly good during testing except for during dynamic gait activities. She also reported falling two times in

CASE STUDY 25-2

the last 4 weeks, which put her at high risk for another fall.

Goals for Mrs. M included improving the DGI from 19/24 to 22/24 and the EquiTest composite score from 77 to 85, decreasing the DHI score from 12 to 5, and raising the ABC score from 51% to 70%.

The plan was to see the patient for 3 or 4 visits over the next 2 to 3 months to improve her dynamic balance, increase her stamina, and decrease her fear of falling. She agreed to the stated goals. The plan was to discuss a pool exercise program with her to attempt to have her increase her strength and mobility in a non–weight-bearing exercise program that she might enjoy, to avoid any worsening of knee pain associated with the more intense activity level.

Mrs. M had been prescribed the following exercises: walking with head turns to the right and left, stepping up to a stool but not onto it and down, bending down toward the floor from the sitting position, a walking program, and walking with 180-degree turns. She was instructed to do the exercises two times a day.

The patient was seen four times in physical therapy. During her second visit, she reported that her physician had changed her diuretic. She had not fallen since her last visit to physical therapy. During her second physical therapy visit, it took her 20 seconds to rise from the chair 5 times, which was worse than on the first visit. Her ABC score had increased by 11% to 65%, and her DHI score decreased from 12 to 2. Her DGI score had increased from 19 to 21/24. Her composite sensory organization test value on the EquiTest was 82, an increase of 5. Patient was prescribed standing plantar flexion next to the kitchen sink, the trace-the-alphabet exercise with her foot, walking and turning 180 degrees, walking with head turns, and walking and making 360-degree turns. She was told to try to do the exercises two times a day.

During her third physical therapy visit, Mrs. M stated that she had been having difficulty finding time to do the exercises. She could do the alphabet exercise without holding onto the kitchen sink. Walking and looking up was a problem for her, but walking backwards was easier. Mrs. M complained of swelling in her feet. She was instructed to keep them elevated but was also shown how to perform ankle pumps and ankle isometrics. SLS times had improved to 21 seconds on the left and 20 seconds on the right. Her 5 times sit-to-stand test time was 17.3 seconds, her composite sensory organization test score was 77/100, and her ABC score was 64%.

Mrs. M was given written ankle exercises, an SLS exercise, a seated exercise in which she rolled a rolling pin under her foot, standing weight-shift exercises, and walking with head movements.

During the patient's fourth and final visit, she stated that she had difficulty with the standing weight shifts and that moving back onto her heels while balancing was difficult. The alphabet exercise and walking backward were not a problem for her. SLS continued to be a challenge for her balance. Patient reported that she had not been walking as the therapist had requested. Her DGI score had increased to 23/24. Her DHI score had remained at 2. Her sensory organization test score remained at 76, and her ABC score was 61%. Her 5 times sit-to-stand test time had improved to 12 seconds, and her timed "Up & Go" test time was now 11 seconds. SLS times had improved to 28 seconds on the left and 30 seconds on the right.

Her home exercises for the fourth physical therapy visit consisted of standing in SLS and moving her head slightly to the right and left, walking on her toes in plantar flexion, and standing in SLS on a pillow.

Mrs. M was discharged at the end of her fourth clinic visit. She had made great strides with her walking and was no longer falling. The four visits were spaced out at 3-week intervals over a 3-month period. She had met one of her goals and had partially met three of the other four goals that were initially developed in her plan of care. She was satisfied with her progress and was instructed to rejoin the cardiac exercise group that she had belonged to after she had her silent heart attack; she hated to exercise alone. One of her neighbors from her condominium was also attending the cardiac group program, so she was encouraged to join her neighbor to improve her compliance. She preferred to read and perform less physically demanding activities.

The physical therapist encountered Mrs. M's daughter-in-law 6 weeks after discharge, who reported that Mrs. M still had not rejoined the exercise group but that she had not been falling. Her daughter-in-law was encouraged to "remind" Mrs. M to restart the cardiac exercise program because of her shortness of breath.

Summary

Older adults with vestibular disorders have some unique differences from younger adults. The normal physiological changes associated with aging in the vestibular apparatus, the eye, and somatosensation can complicate the rehabilitation of the older adult with a vestibular disorder. Older adults can improve with vestibular rehabilitation but may need special care. Comorbid medical problems that may be seen in older adults with v estibular disorders require the physical therapist to think carefully before initiating an intervention program. Patient safety and encouraging compliance with the interv ention are essential. Careful identification of the patient's functional limitations enables a therapeutic program to be de vised to restore the older adult's function safely.

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CHAPTER 26

Physical Therapy Management of the Patient with Vestibular Dysfunction from Head Trauma

Laura Morris, PT ■ *Kim Gottshall, PT, PhD*

Traumatic Brain Injury (TBI) has been defined as damage to the brain, resulting from an external force such as an impact, penetration from a projectile object, rapid acceleration/deceleration forces, or blast waves. Brain function can be disrupted temporarily or permanently, and structural damage may or may not be identified using current imaging techniques. Given the trauma to the head and the location of the vestibular end organs, TBI can result in vestibular impairment. The impairment may be caused by peripheral damage to the end organ, central damage at the cerebellar or cortex level, or a combination of both. This chapter outlines the unique considerations for the management of those with vestibular impairment and TBI.

Traumatic brain injury results in damage that ranges from mild to severe injury. Classification of the severity of TBI can be described as mild, moderate, or severe based on Glasgow Coma Scale (GCS) ratings, the presence of loss of consciousness (LOC), or post-traumatic amnesia (PTA). See Table 26-1 for the classification structure that is most commonly described. Neither the location of the lesion, nor the nature of resulting impairment factors into the severity classification.[1]

The term *concussion*, used interchangeably with mild traumatic brain injury (mTBI), describes a complex pathophysiological process that affects the brain. This process is induced by traumatic biomechanical forces, but does not require direct contact to the head. For example, blast exposure can cause concussion, as can a fall that results in rapid acceleration/deceleration of the head but no direct contact between the head and the ground. Concussion causes a rapid onset of short-lived impairments of neurological function that usually resolve spontaneously. Loss of consciousness may or may not be present with a concussion.[2,3]

The diagnostic criteria for post-concussive syndrome (PCS) have been defined by several groups, including the World Health Organization (WHO) for the ICD-10 coding system, and the American Psychiatric Association for the DSM-IV. These criteria differ in their inclusiveness (Box 26-1) and have since been found to be flawed.[4-6] The Centers for Disease Control (CDC) defines PCS as having a prolonged recovery, and generally the period of 3 months is used to separate acute concussion from PCS.[3] There is little consensus or published literature that states a clear delineation between mTBI or concussion and PCS.

■ Table 26-1 SEVERITY CLASSIFICATION FOR TRAUMATIC BRAIN INJURY

	Mild	Moderate	Severe
Glasgow Coma Scale	13–15	9–12	3–8
Loss of consciousness	0–30 minutes	<30 minutes to <24 hours	>24 hours
Post-traumatic amnesia	<1 day	>1 day but <7 days	>7 days

Classification of the severity of TBI can be described as mild, moderate, or severe based on Glasgow Coma Scale ratings, presence of loss of consciousness, or post-traumatic amnesia.

Box 26-1

CURRENT DEFINITIONS OF POST-CONCUSSIVE SYNDROME (PCS) FROM THE WORLD HEALTH ORGANIZATION (ICD-10) AND AMERICAN PSYCHIATRIC ASSOCIATION (DSM-IV)

ICD-10 Definition: Post-concussional syndrome

Listed below are the diagnostic criteria for PCS from ICD-10. Note: the nosological status of this syndrome is uncertain, and criterion A of the introduction to this rubric is not always ascertainable. However, for those undertaking research into this condition, the following criteria are recommended:

A. The general criteria of F07 must be met. The general criteria for F07, Personality and Behavioral Disorders Due to Brain Disease, Damage and Dysfunction, are as follows:
   G1. Objective evidence (from physical and neurological examination and laboratory tests) and/or history, of cerebral disease, damage, or dysfunction.
   G2. Absence of clouding of consciousness and of significant memory deficit.
   G3. Absence of sufficient or suggestive evidence for an alternative causation of the personality or behavior disorder that would justify its placement in section F6 (Other Mental Disorders Due to Brain Damage and Dysfunction and to Physical Disease).
B. History of head trauma with loss of consciousness, preceding the onset of symptoms by a period of up to four weeks (objective EEG, brain imaging, or oculonystagmographic evidence for brain damage may be lacking).
C. At least three of the following:
   1. Complaints of unpleasant sensations and pains, such as headache, dizziness (usually lacking the features of true vertigo), general malaise and excessive fatigue, or noise intolerance.
   2. Emotional changes, such as irritability, emotional lability, both easily provoked or exacerbated by emotional excitement or stress, or some degree of depression and/or anxiety.
   3. Subjective complaints of difficulty in concentration and in performing mental tasks, and of memory complaints, without clear objective evidence (e.g., psychological tests) of marked impairment.
   4. Insomnia.
   5. Reduced tolerance to alcohol.
   6. Preoccupation with the above symptoms and fear of permanent brain damage, to the extent of hypochondriacal over-valued ideas and adoption of a sick role.

DSM-IV Definition: Post-concussional disorder

A. A history of head trauma that has caused a significant cerebral concussion.
   Note: the manifestations of concussion include loss of consciousness, post-traumatic amnesia, and less commonly, post-traumatic onset of seizures. Specific approaches for defining this criterion need to be refined by further research.

Continued

Box 26-1

CURRENT DEFINITIONS OF POST-CONCUSSIVE SYNDROME (PCS) FROM THE WORLD HEALTH ORGANIZATION (ICD-10) AND AMERICAN PSYCHIATRIC ASSOCIATION (DSM-IV)—cont'd

B. Evidence from neuropsychological testing or quantified cognitive assessment of difficulty in attention (concentrating, shifting focus of attention, performing simultaneous cognitive tasks) or memory.
C. Three (or more) of the following occur shortly after the trauma and last at least 3 months.
   1. becoming fatigued easily,
   2. disordered sleep,
   3. headache,
   4. vertigo or dizziness,
   5. irritability or aggression on little or no provocation,
   6. anxiety, depression, or affective liability,
   7. changes in personality (e.g., social or sexual inappropriateness).
   8. apathy or lack of spontaneity.
D. The symptoms in criteria B or C have their onset following head trauma or else represent a substantial worsening of preexisting symptoms.
E. The disturbance causes significant impairment in social or occupational functioning and represents a significant decline from a previous level of functioning. In school age children, the impairment may be manifested by a significant worsening in school or academic performance dating from the trauma.
F. The symptoms do not meet criteria for Dementia due to Head Trauma and are not better accounted for by another mental disorder (e.g., Amnestic Disorder due to Head Trauma, Personality Change Due to Head Trauma.

Second Impact Syndrome (SIS) is an unusual and often fatal phenomenon in which an individual sustains a second head injury before the resolution of the first head injury. The first concussion may have been minutes, days, or weeks prior. The second blow may be minor, with a direct blow to the head, or an indirect injury that results in accelerative forces to the brain. The patient will suddenly collapse and undergo rapid neurological decline and respiratory failure in the first few seconds to minutes after the injury. SIS results in death or severe neurological impairment. The pathophysiology of SIS is thought to be caused by either a subdural hemorrhage and/or dysautoregulation of the cerebral vasculature that causes widespread hyperemic brain swelling and increased intracranial pressure. Adolescents and young adults are primarily affected.[7-10]

Pathophysiology of Mild TBI

Injury to the brain following a mild TBI has been studied extensively over the past decade. Our understanding of the pathophysiology of mTBI is based on the work of Giza and colleagues, which was described in 2001 (Fig. 26.1). [11,12] Unlike moderate to severe injuries, which cause focal vascular or axonal lesions, mild injury results in a cascade of neurometabolic events that damage the brain's processing ability. Upon onset of a biomechanical injury , excitatory neurotransmitters such as glutamate cause neuronal depolarization that leads to an efflux of potassium from the neuron and an influx of calcium into the neuron. The sodium-potassium pump in the cell membrane must then work overtime to restore proper ionic balance within the cell. The pump requires increased amounts of adenosine triphosphate (ATP). The increased demand for ATP causes a large metabolic requirement for glucose. This sudden metabolic need is accompanied by a general decrease in cerebral blood flow. The resulting decrease in the supply of energy in the form of glucose, and the increased demand by the NA/K pump leads to a cellular metabolic crisis.

Once the initial period of energy crisis has passed, the concussed brain enters a period of depressed metabolism. Persistently high levels of calcium in the cell are toxic, leading to intracellular edema and diffuse axonal death. In addition, higher calcium levels have been shown to disrupt posttraumatic neural connectivity.[11,12]

Mechanism of Injury: Blast Injury Versus Blunt Head Trauma

Blast wave exposure occurs from explosions of all types, including industrial accidents and military operations. Blast-induced injuries have been recognized as

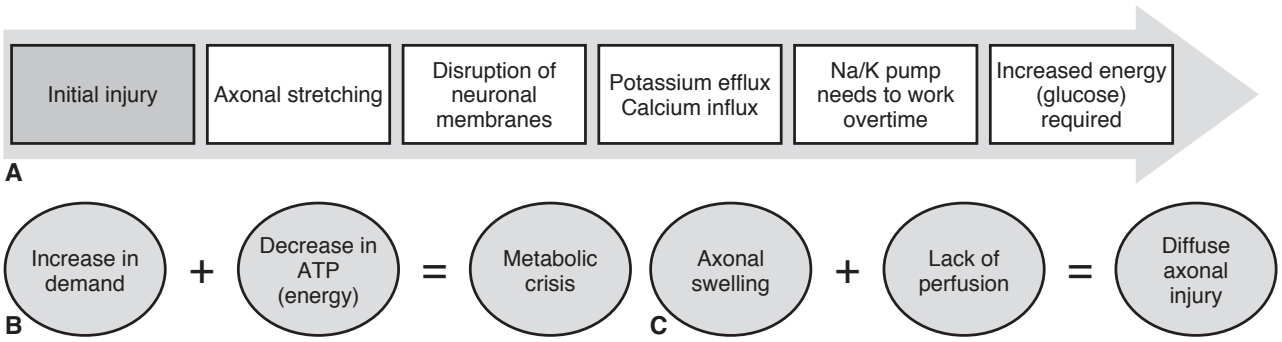

Figure 26.1 Pathophysiology of mTBI. **(A)** Acute effects of a mild TBI caused by disruption of neuronal membranes. **(B)** A metabolic crisis occurs because of a rapid increase in energy needs, coupled with a decrease in available adenosine triphosphate (ATP). **(C)** Once the initial period of energy crisis has passed, metabolism is depressed. Persistently high levels of calcium in the cell are toxic, leading to intracellular edema and diffuse axonal death.

a major cause of head trauma in recent combat operations, accounting for more than 80% of all battlefield injuries.[13] Blast exposure is defined as an event in which the individual feels or is exposed to a pressure wave before they hear a noise. The pressure wave may be regarded as a primary causative factor of much of the brain injury seen after the exposure. Individuals who experience this type of injury experience different symptoms and injury patterns than those who have blunt head trauma.[13-17]

The patterns of injury in survivors of blast injury can be complex because of multisystem involvement and the severity of injury. A large percentage of these individuals suffer limb loss, orthopedic trauma, burns, gastrointestinal injury, visual impairment and post-traumatic stress disorder.[16] The pressure wave from the blast can cause significant middle and inner ear damage, with resulting vestibular pathology. Shupak studied Israeli soldiers following blast exposure, and found that hearing loss, tinnitus, and tympanic membrane perforation occurred in 80% of subjects, and 60% had persistent dizziness.[17] Hoffer et al found that 98% of those individuals experiencing blast injury reported dizziness acutely, and 80% of the individuals reported dizziness in the subacute and chronic phases.[13] Hoffer and colleagues also reported that those individuals with blast injuries had increased rates of headache and greater symptoms of disequilibrium than those with blunt head trauma. There was a reported difference in the nature of the symptoms, as the individuals who suffered blast injuries reported that their symptoms tended to be constant, as compared with the individuals with blunt head trauma, who had more intermittent symptoms. Individuals with blast injuries also showed higher rates of hearing loss, post-traumatic spatial disorientation, and cognitive dysfunction as compared with those with blunt head trauma.[13]

Symptomatology in Head Trauma

Unlike the focal neurological changes that occur with moderate to severe brain injury, the diffuse nature of the metabolic impairment and axonal death leads to the broad symptom picture that occurs with mild brain injury. Figure 26.2 details the symptoms that are typically described, and highlights the broad categories of impairment.[18] Dizziness is a very common symptom after traumatic brain injury, present in 23% to 81% of cases in the first few days after injury.[19-22] In the sports population, 55% of athletes report dizziness

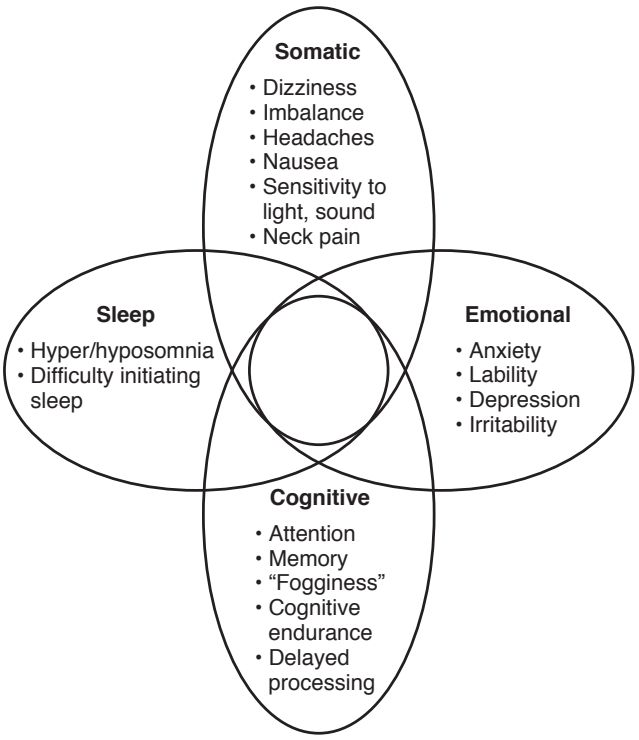

Figure 26.2 Symptoms from m (mild) TBI: Categories of impairment and typical symptoms associated with mTBI.

after concussion.[23] When military personnel report symptoms following blast-related mTBI, dizziness is the most common post-injury symptom.[13]

The etiology of dizziness following head trauma is diverse. The vestibular pathology found in head trauma can be from temporal bone fractures, labyrinthine concussion, benign paroxysmal positional vertigo (BPPV), vascular lesions, central lesions, and perilymphatic fistula.[14,24,25] Gottshall studied service members to describe type of injury and potential for recovery. She found that impairments in gaze stability during head movement and disequilibrium were common, as was exertional dizziness.[14] Scherer et al studied blast exposure in service members with the intention of determining the etiology of dizziness.[26] Using scleral search coil methods to study eye movements during rapid head rotations, they found impairment in the angular VOR (aVOR), and hypothesized that this could be caused by differential destruction of Type I hair cells, or a disruption of the irregular afferent VOR pathways. This would account for normal vestibular function for low-velocity movements that are detected by Type II cells, and lower than normal aVOR gain with high-velocity movements. Significantly, elevated symptom severity during exertion was also found, consistent with work by Gottshall and Hoffer. Scherer and colleagues found deficits in pitch plane aVOR, which may contribute to the elevated dizziness during running, because running invokes greater head movements in the pitch plane as compared with walking, and thus places more of a demand on the pitch plane aVOR.[26]

There are other causes of dizziness with TBI that are not related directly to peripheral vestibular loss. Post-traumatic BPPV, anxiety related dizziness, migraine-associated dizziness, autonomic dysregulation, and cervicogenic dizziness have all been implicated as sequelae of mTBI. The orthopedic injuries associated with head injury often involve the cervical spine, so a cervical etiology to dizziness should be assessed.[27-29] The evaluation and management of cervicogenic dizziness is discussed in Chapter 31.

Post-traumatic BPPV is a common occurrence following head trauma. The estimated incidence of post-traumatic BPPV in the TBI population with symptoms of dizziness varies widely, from less than 5% to 28%. [13-30] The differences in the reported incidence rates may be a result of the nature and extent of the TBI (concussion vs. blast exposure), as well as length of time between the head trauma and the time that patients are first assessed. Spontaneous resolution can occur with BPPV, which may account for the lower incidence of BPPV reported in some studies. The incidence of bilateral BPPV is higher in post-traumatic as compared with idiopathic BPPV.[31]

Anxiety-related dizziness has been found to be present in both military and non-military populations following mTBI. In one group of non-military subjects with mTBI, 41% reported anxiety 5 years after injury.[32] In military studies, as many of 44% of soldiers in the Iraqi conflict that had a loss of consciousness during their head injury met the criteria for post-traumatic stress disorder and endorsed symptoms of anxiety.[33] The pathophysiology of anxiety-related dizziness has been well described, and is believed to be caused by common central pathways in the brainstem that perceive dizziness and vertigo and also control anxiety.[34,35] Migraine–anxiety related dizziness has also been suggested as a separate entity, which may be seen as a sequela of mTBI.[36,37] Careful interviewing of the patient and collaboration with other team members may help identify anxiety in patients referred for vestibular rehabilitation, and the presence of anxiety will have implications in the rehabilitation process.

Post-traumatic headache, both migrainous and non-migrainous, is a common sequela of mTBI.[38] In addition to photophobia, phonophobia, nausea, and visual changes, dizziness and vertigo are common symptoms in migraine, occurring in 25% to 30% of the individuals with migraine.[37] These symptoms can occur without or without the headache. In the military population, Hoffer et al found that 41% of soldiers with mTBI met criteria for migraine-related dizziness.[13] The pathophysiology of migrainous vertigo is not well understood, but may be a result of neuroanatomical connections between the trigeminal nuclei and vestibular nuclei.[37] The presence of migrainous headache in athletes with mTBI has been identified as being associated with significantly decreased neurocognitive function, as compared with those with non-migrainous headache or no headache.[38] The management of migraine by the medical team is important to the overall positive outcome of the rehabilitation process. Well-managed migraine will allow patients to participate more fully in the rehabilitation process.

Dizziness may also occur in mTBI caused by autonomic dysregulation. Patients with autonomic dysregulation typically report dizziness with position changes such as sit to stand, bending over, and climbing stairs and often note that any form of exertion may provoke dizziness. The symptoms experienced from autonomic changes may mimic BPPV or movement intolerance, so care must be taken to assess symptoms closely. There is occasionally clear evidence of orthostatic hypotension, but more often the changes are transient and may not be detectable with blood pressure monitoring. Later in the recovery process, patients may have better tolerance for position changes but may experience dizziness during exercise or any activity that increases heart rate.[39]

Postural instability and impaired balance reactions may be caused by sensory organization dysfunction and/or vestibulospinal reflex impairment. Sensory organization

impairment may be a result of abnormal central processing of the input from peripheral end organ, or impairment of the end organ itself in the case of peripheral vestibular lesions. Guskiewicz et al studied concussed athletes and found impairment in sensory organization using computerized and clinical assessment tools.[40,41]

In addition to somatic symptoms, cognitive symptoms are a hallmark sign of mild brain injury. Typical impairments include limited attention, a sensation of mental "fogginess," impaired immediate and delayed recall, decreased cognitive endurance, and delayed cognitive processing. Multitasking and the ability to alternate tasks are also impaired. Because of decreases in metabolism and increased energy demands, new learning is often difficult.[42]

Sleep disturbance complicates many other functions, and sleep is often impaired in individuals with mTBI. The disturbances may be in initiating sleep, maintaining sleep, or sleeping too much (hypersomnia).[43] Lack of adequate sleep exacerbates many of the same symptoms that are caused by the initial brain injury, including irritability and anxiety, depression, decreased concentration and attention, delayed reaction time, decreased energy levels, and fatigue.[44]

Emotional symptoms are also widely described with concussion; these symptoms include irritability, anxiety, depression, and emotional lability. Healthy adolescents tend to have difficulty regulating their emotions, which is often worsened with concussion.[45] The strong comorbidity between anxiety and dizziness should trigger therapists to pay close attention to the presence and persistence of anxiety in their patients.[6,46]

As therapists managing individuals with brain injury, we must take into account these other areas of symptoms that may affect our patients' abilities to recover from their vestibular impairment. Assessing the patient in a holistic manner should improve treatment effectiveness and ultimately result in better outcomes and less frustration during the rehabilitation process. Managing all aspects of the patient's impairments are not within our scope of practice, but knowledge of a patient's symptom picture and overall challenges will allow us to refer patients, as appropriate, to other practitioners and allow for more realistic expectations of the patient. Working with a multidisciplinary team will also allow for better management of this broad scope of recovery.

Examination

Diagnostic Imaging

The process of identification of moderate to severe brain injury is relatively straightforward, using imaging and clinical neurological testing. The presence of vestibular dysfunction may be less clear, and may be difficult to determine in the presence of severe oculomotor deficits or central patterns of nystagmus. In mild brain injury, the diagnostic process is less clear, and a standard of assessment has been developing over the past two decades.

Numerous imaging techniques hold promise as diagnostic tools and as measures of recovery. Diagnostic imaging, using standard CT or MRI scanning, is not sensitive enough to reveal mTBI/concussion in most cases. Because the injury is metabolic in nature and the dysfunction is diffuse rather than focal, no abnormalities can be identified. There are several imaging techniques that can be used to reveal changes associated with mTBI. The development of single-photon emission CT (SPECT) imaging has allowed researchers to identify changes in blood flow and perfusion of cortical tissue. Abu-Judeh et al, were able to identify abnormal perfusion as part of the pathophysiological changes following mild to moderate TBI.[47] Positron emission tomography (PET scans) measures metabolic changes in the brain. This makes PET scans particularly helpful in identifying the changes that occur in mTBI. In one study by Chen et al, subjects with mTBI appeared normal at rest.[48] When PET scans were measured after performing a cognitively difficult task, there was a significant difference in images between controls and the subjects with mTBI. The use of fMRI, which shows local blood flow and metabolic changes in real time, was used as well and found the same results.[49] In a study performed on high school athletes, there was a positive correlation between the extent of abnormal brain hyperactivation found on the fMRI studies and the length of the clinical recovery, as measured by cognitive testing and reported symptoms.[50] In the future, it may be possible to use fMRI to give more objective data to support return to play, or removal from play, for athletes.

Proton magnetic resonance (MR) spectroscopy measures brain metabolites, and this has provided an objective basis to confirm the presence of a metabolic imbalance in brain tissue. Vagnozzi et al imaged concussed athletes via proton MR spectroscopy and found the ratio between N-acetylaspartate (NAA) to creatinine (Cr) was reduced for 30 days after the first mild brain injury, and 45 days after two injuries.[51,52] Diffusion Tensor Imaging (DTI) is an imaging technique that measures white matter changes in the brain. In a recent prospective study by Mayer et al, those with mTBI were found to have abnormal DTI imaging that was sensitive to change over time, and this may serve as a potential objective biomarker of recovery in the future.[53]

Although most patients do not receive these advanced imaging techniques, the tests may be used more commonly as technology improves and more is learned

about the diagnostic utility of these imaging techniques in the future.

Acute Testing

Accurate and objective clinical assessment of mTBI should be multifactorial to be sensitive to the broad range of dysfunction that is commonly found in this population. Much of the work in developing assessment tools has been focused on athletic injuries, with the goals of (1) establishing an on-field test to determine if an athlete has sustained an mTBI, and (2) establishing a test to determine whether the athlete should return to play following an mTBI. An international group of practitioners developed a consensus statement that outlined the management of concussion in sport. As part of this consensus statement, the Sports Concussion Assessment Tool, or SCAT3, was recommended for sideline assessment. It consists of seven subtests: (1) the Glasgow coma scale (GCS), (2) an orientation to game/play called the Maddocks score, (3) a structured symptom scale, (4) cognitive assessment, (5) cervical examination, (6) balance assessment using either the modified BESS (Balance Error Scoring System) test or a tandem gait test, and (7) a coordination test. Imbedded in the SCAT3 is the Sideline Assessment of Concussion, or SAC, which is a cognitive assessment test. It consists of tests of basic orientation, immediate memory, concentration, and delayed recall. A pediatric version of the test has also been developed, called the Child SCAT3. The seven areas of assessment are similar, with modifications to appropriately assess children from 5 to 12 years old.[18,54-58]

One limitation in the currently used assessment is the lack of dynamic balance tasks as opposed to static testing, and the lack of dual task assessment. Catena et al have proposed that dual-task balance testing may be a better screening tool for identifying dysfunction in the concussed athlete.[59-61] This research focused on dual tasks such as answering questions while walking and during obstacle crossing tasks. In these studies, walking while answering questions was found to be sensitive to concussive impairment if assessed within 6 days of injury.

Subjective Symptom Scales for Assessment of mTBI/Concussion

In addition to the subjective scale mentioned previously, there are several subjective symptom scales that are used for both acute and subacute assessment of TBI. These scales measure the global symptoms of post-concussive symptoms and are not focused on vestibular symptoms. They all contain some reference to dizziness and balance problems, but are not as specific as the Dizziness Handicap Inventory[62] or the Activities-specific Balance Confidence Scale.[63] The Rivermead Post Concussion Symptom Questionnaire,[64] the Post Concussion Symptom Scale (PCSS),[65] the Graded Symptom checklist,[56] and the Concussion Symptom Inventory[65] are all used to measure symptom severity. These assessment tools vary from 12 to 22 items and use either 5 or 7 point Likert scales. Caution should be used when making management decisions based solely on a patient's symptoms, because symptoms may be under- or over-reported, depending on the patient's situation.[66,67]

Neurocognitive Testing

Assessment of neurocognitive function is an integral part of the management of the post-concussive patient, and has been endorsed by leaders in the field as part of the assessment battery.[2,68] There are many forms of assessment, both computerized testing and paper and pencil testing. Regardless of the actual test, the testing domains are the same and are listed in Box 26-2. The testing can provide information on changes in cognitive function if performed serially. Some practice models endorse regular testing to determine improvement in function over time. There is, however, some controversy over whether pain, emotional distress, and depression may influence the outcomes of neurocognitive assessment.[69-72] There is a trend toward performing baseline testing, which allows for pre- and post-injury comparison. This data may be helpful in determining level of impairment and management of the patient.[73] There has been some suggestion that the baseline data and subsequent neurocognitive post-injury data are not always used effectively when making return-to-play decisions, and that

Box 26-2

TYPICAL COGNITIVE FUNCTIONS THAT ARE ASSESSED IN STANDARDIZED NEUROCOGNITIVE TESTING

- Attention Span
- Working Memory
- Sustained Attention
- Selective Attention
- Nonverbal Problem Solving
- Reaction Time
- Visual Memory and Verbal Memory
- Response Variability

symptom report, rather than objective data, guides decision making. As the field of concussion management grows and education improves, the use of the objective data should increase.[74] As with all assessment tools used for concussion diagnosis and management, neurocognitive testing should not be used as a single measure, but as part of a comprehensive patient assessment.[65]

Clinical Examination

Physical therapy assessment of patients with suspected vestibular impairment caused by mild to severe TBI follows a similar practice pattern to those without TBI, although care must be taken to attend to possible comorbidities. For subjective assessment of symptoms, there are a variety of tools that can be used to assess the impact of the vestibular impairment on the individual's ability to function. Two commonly used tools are the Dizziness Handicap Inventory, which can be used to measure the impact of the dizziness on specific tasks,[62] and the Activities-specific Balance Confidence Scale, which assesses balance confidence over a number of conditions.[63] The Vestibular Rehabilitation Balance Questionnaire (VRBQ) can be used to monitor subjective dizziness and also to measure changes in quality of life.[75] The Vestibular Activities of Daily Living scale measures functional difficulty with various tasks and is one of the few scales that also addresses the issue of participation.[76] Visual-motion induced symptoms can be quantified using the Situational Vertigo Questionnaire[77] or the Visual Vertigo Analog Questionnaire.[78]

Because a wide variety of people have traumatic brain injuries, from the young athlete to the elderly person with TBI as a result of a fall, there is no universally accepted objective measurement tool that will meet the needs of this patient population. Selection of the appropriate assessment tools should take into account the needs and goals of the individual patient. For the athlete, the tests must be challenging enough to make return to play decisions; for the nonathlete, the tests should determine fall risk and the ability to return to previous activity levels. The ability to use different sensory signals for postural control can be measured using computerized dynamic posturography (CDP) testing or the modified Clinical Test of Sensory Interaction in Balance (mCTSIB).[79,80] The Functional Gait Assessment (FGA) measures gait tasks that incorporate head movement, narrow base of support, and changing speeds.[81,82] The test is similar to the Dynamic Gait Index (DGI), but includes more challenging tasks that avoid the ceiling effects found with the DGI when testing higher-functioning patients. The FGA also has an identified fall risk cutoff of 22/30, which is helpful for assessing lower level TBI patients.[82] The mini-BESTest combines these various components in a more global balance assessment tool.[83]

Two balance tests have been designed specifically for this patient population. The Balance Error Scoring Scale (BESS) is recommended for sideline assessment for individuals with suspected concussion, and mTBI and can be used later in the recovery process as well.[84] The HiMAT test was specifically developed for assessing people with traumatic brain injury. It is a measure of high-level mobility, such as dynamic walking tasks, the ability to negotiate stairs, running, skipping, hopping, and bounding.[85,86] This measure can be uniquely helpful in determining return to sport or return to active work duty for physically challenging jobs.

Oculomotor Findings in TBI

A variety of oculomotor abnormalities can be observed in individuals as a result of TBI. Some of these findings may be related to a loss of vestibular function; other findings may be related to deficits in the central oculomotor pathways or damage to the extraocular muscles. Some of the patient's symptoms may be attributable to these oculomotor deficits. The dynamic visual acuity test (DVA), which assesses an individual's ability to see clearly during head movements, is a functional measure of presumed VOR function. Dynamic visual acuity testing can be measured by a clinical test or with a computerized system. Although either method allows for insight into gaze stability, the computerized DVA is more sensitive than the clinical DVA test. Computerized testing may include a battery of tests that are thought to measure perception time (PT), target acquisition (TA), target following (TF), gaze stabilization (GST), and dynamic visual acuity (DVA). Dynamic visual acuity is measured as the difference between static and dynamic visual acuity, and is typically reported as a logMAR score (log minimal angle of resolution). Computerized testing has been used to identify gaze stability impairments in those with mTBI and to track changes with recovery.[14] Computerized DVA testing has been found sensitive and reliable for identifying vestibular impairment.[87] This type of testing may be helpful in identifying subtle impairments in elite athletes or young, higher-level patients.

In addition to the assessment of gaze stability, a thorough oculomotor assessment is critical. There is evidence for saccadic and smooth pursuit deficits with TBI, caused by central nervous system impairment. These impairments are not sensitive as a diagnostic tool, but may have implications for rehabilitation and recovery. A person with saccadic or smooth pursuit deficits may have difficulty reading, tracking people or objects in his/her environment,

or visually scanning.[88,89] There is very limited evidence (only small-size case studies without control groups) to suggest that an oculomotor training program (fixation, saccades, smooth pursuit, and simulated reading) could lead to improvement in reading function in subjects with TBI and CVA.[89]

Vergence and accommodative abnormalities have also been identified as oculomotor impairments in TBI.[90] *Convergence insufficiency*, or the inability to generate the appropriate amount of ocular convergence to maintain clear and single binocular vision with a near target, has been identified in TBI, but the specific etiology of the impairment is unknown. The neurons that control vergence are located near the oculomotor nucleus in the mesencephalic reticular formation, and may be implicated because of injury at the brainstem level during TBI.[91] Some individuals with vergence disorders following TBI endorse symptoms of dizziness that are non-vestibular in nature and have visual motion–provoked dizziness.[90] It is not clear in these cases if the individual's symptoms are a result of the convergence insufficiency (dizziness caused by blurred vision or diplopia) or because of other underlying deficits as a result of the TBI. Screening for convergence insufficiency is important for accurate assessment and efficient management of the rehabilitation process. Headache, fatigue, and loss of concentration are also symptoms reported with non-vestibular oculomotor deficits.[92] There has been some evidence to suggest that vision therapy designed to improve the near point for convergence can be effective.[89,92,93] Vision therapy often consists of clinical visits with a neuro-optometrist and home exercise programs. If convergence insufficiency is identified during the oculomotor exam, and the patient reports an increase in dizziness with convergence tasks, it may be beneficial to incorporate some convergence tasks into the intervention plan. If symptoms or oculomotor abnormalities persist, a discussion with the referring physician and possible recommendation to neuro-ophthalmology or neuro-optometry would be appropriate.

Convergence Spasm is defined as an inappropriate or excessive convergence, accompanied by pupillary miosis, lasting seconds to minutes, and occasionally longer. This impairment was first described as a psychosomatic phenomenon, often accompanied by anxiety or panic. An organic etiology to convergence spasm was implicated with TBI, likely caused by brainstem involvement.[94-96] The spasm is brought on most often by vertical eye movements. Patients who experience convergence spasm describe symptoms of vague discomfort, disequilibrium, or dizziness, especially with reading or looking down. Until recently, the standard treatments involved atropine, which dilates the pupil and breaks the spasmodic pattern, corrective lenses, or occlusion. There has been suggestion that convergence spasm is similar to other dystonias and that botulinum toxin treatment may be effective.[97] There is no evidence that any oculomotor exercise will help with recovery from convergence spasm.

As in patients with symptoms of dizziness not related to head trauma, findings of oculomotor abnormalities during the clinical exam should be discussed with the referring physician or medical team because of the implications for central nervous system involvement.

Intervention Considerations

Problem-oriented Approach

Those patients with physical therapy needs following traumatic brain injury progress at different rates. Following a problem-oriented approach and assessing patients carefully may maximize recovery in the most efficient manner. When developing a treatment plan, the therapist may need to prioritize the impairments, because of the diversity and number of impairments associated with TBI (Fig. 26.3). In addition, the therapist will need to maintain flexibility and be able to modify the interventions to maintain an optimal level of challenge to the patient.

Recovery typically happens in a progressive fashion, and the rehabilitation process follows in a similar progressive fashion (Fig. 26.4). The general process starts with medical management and physical therapy intervention as appropriate, incorporating musculoskeletal intervention and vestibular rehabilitation techniques. The next step in the process is aerobic conditioning and the addition of strength/power training as appropriate. Once the patient is able to tolerate aerobic training and strength/power exercises without exacerbation of his or her symptoms, functional training for return to sport or work can be initiated. This may take the form of high-velocity ball tasks, hopping, jumping, running, climbing ladders, or repetitive tasks that require rapid movement. Customization and creativity are necessary to individualize the intervention to the patient's specific functional needs. This is often the last step in the physical therapist's involvement with the patient.

The recovery process is not necessarily over at this point for athletes, members of the military, or other individuals, depending on their job requirements. Athletes will likely need to work on sport-specific training. Return to practice with the team may be appropriate if symptom-free. There should be no return to play until symptoms are fully resolved and neurological and cognitive testing is normal. For service members, combat-specific training will be needed, incorporating virtual reality with a moving support surface to simulate combat situations. For those who work at heights, like electrical workers, iron workers,

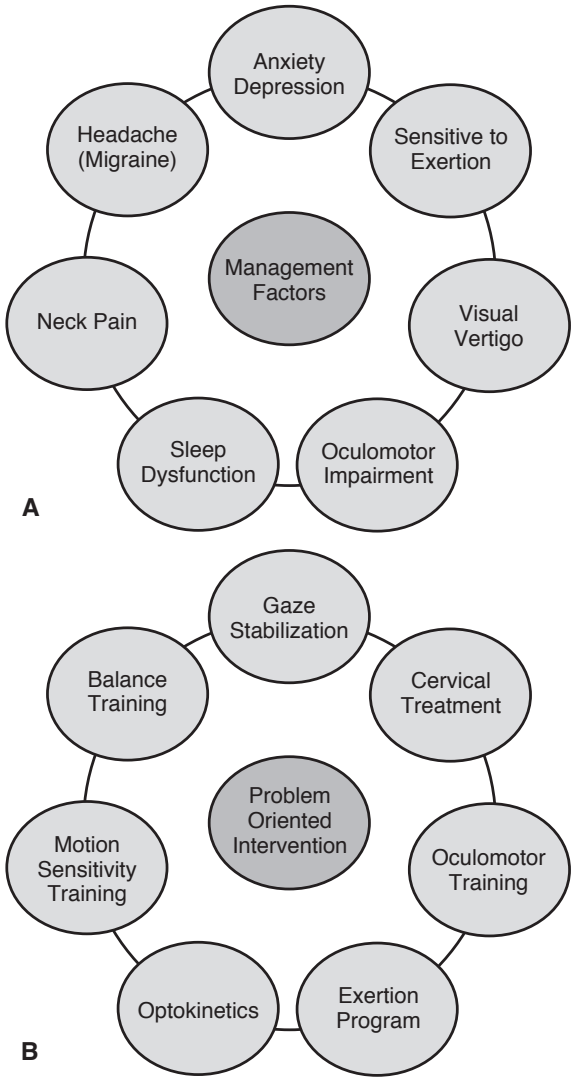

Figure 26.3 **(A)** Management factors and **(B)** problem-oriented intervention techniques for mTBI.

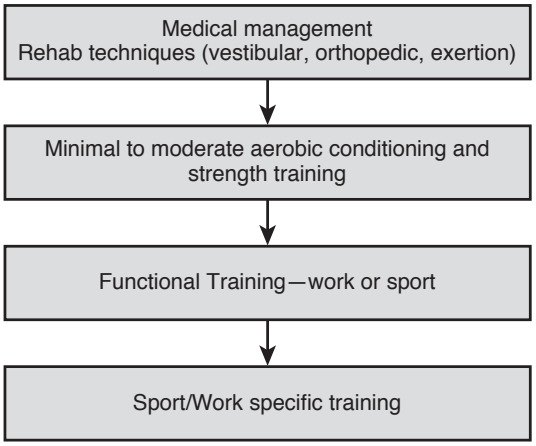

Figure 26.4 Step-wise progression of intervention in mTBI.

or firefighters, continued training with specific equipment may be necessary before return to work.

Gaze Stability Deficits

Impairment of gaze stability during head movement may be caused by peripheral vestibular loss, focal central lesion, or mixed central and peripheral vestibular deficits. For those patients who experience gaze stability deficits following TBI, as assessed by the DVA test, exercises that focus on improving gaze stabilization and eye-head coordination are recommended within the patient's tolerance, taking into account that the head movements will likely exacerbate the patient's symptoms. There should be a timely resolution of the exercise-provoked symptoms to baseline levels after completion of the exercise session. This time-to-resolution is not clear-cut or well defined in clinical practice or scientific literature. Most expert clinicians use 15 to 30 minutes as a general guideline, while taking into account the patient's needs.

Visual Motion Intolerance

Decreased tolerance of complex visual environments and visual motion is a common problem in individuals with vestibular disorders. For individuals with mTBI, the added complications of slowed processing and impaired ability to shift attention or multitask, which occur post-injury, makes these environments even more challenging. A careful, gradual reintroduction of visual motion via optokinetic training will help the transition back to community environments that may be overwhelming. Training can be initiated with static complex backgrounds, then transitioned to observing visual motion on a computer screen, and finally to full-field stimulation with lighted disco balls. Once tolerance to visual motion has improved, then patients can start to be exposed to more challenging community environments. Frequent daily exposure is recommended, with the duration of the exposure set to be within the patient's tolerance.[98]

Motion Sensitivity

Exercises based on the principles of habituation, typically larger amplitude, fast movements, may be used to treat symptoms of motion sensitivity. As with the exercise approach for treatment of visual motion sensitivity, a careful, gradual reintroduction of head motion training will help to decrease the sensitivity to movement. Exercises may begin in a seated position and involve simple head movements. The velocity of the head movement and the number of head movements should be dosed to induce increased

symptoms with the same time-to-resolution as discussed above. As head motion tolerance increases, the exercises can be increased in terms of velocity, number, and complexity. The exercises may be performed in standing and may involve trunk and whole-body movements. One of the challenges in treating motion sensitivity in individuals with TBI is that the exertion level is often increased as one moves with greater speed and larger body movements. Because exertion can provoke symptoms as well, these two problems can interfere with one another. Cervical dysfunction and migraine can complicate the treatment of motion sensitivity, and occasionally, habituation exercises must be postponed.

Postural Instability

Postural demands vary across individuals and will vary with age, lifestyle demands, and activity levels. An individual involved in athletic endeavors will have greater postural stability needs that an individual who has a sedentary lifestyle. For individuals with TBI, recovery of postural stability can be a complicated issue given the diffuse nature of their injuries. Whereas individuals with peripheral vestibular disorders generally have difficulty with the appropriate use of the different sensory cues for the maintenance of balance, individuals with TBI and damage to the central nervous system may also experience disruption of the automatic postural responses and anticipatory postural adjustments. Treatment of postural stability deficits in these cases will need to move beyond the typical exercises used to promote the utilization of vestibular cues for balance, and will need to incorporate exercises to work on balance reactions to expected and unexpected perturbations. Static and dynamic balance exercises using the dual-task paradigm are often beneficial. In developing interventions to treat the postural instability, the therapist should be mindful of the needs of the patient in terms of the postural demands of work and normal activity levels.

Exertion Intolerance

Exertion intolerance is a unique problem in TBI. It is unclear whether the dizziness reported is a result of movement in the pitch plane or caused by the exertion itself. Scherer and colleagues found that service members with mild to moderate TBI secondary to blast exposure and complaints of dizziness had greater exertion-induced symptoms and decreased angular VOR gains to pitch plane head impulses, as compared with those without symptoms of dizziness following blast exposure.[26] Leddy and colleagues[39,99] theorize that there is ongoing central and systemic physiological regulatory dysfunction that causes symptoms with exertion. Leddy et al studied both athletes and nonathletes who were more than 1 month post-concussion. The symptoms that patients identified upon exertion assessment were headache, dizziness/balance, sadness/depression, irritability, fatigue, and concentration/memory impairments. The investigators found that these symptoms could be reduced or alleviated by an individualized, controlled aerobic exercise program that trains patients with a sub-symptom threshold paradigm. The subjects underwent an exertion assessment on a treadmill, in which the exertion level (as measured by heart rate) that provoked symptoms (symptom threshold) was identified. Exercise was started at 80% of symptom threshold for 5 to 6 days per week, with the exertion level increased gradually as long as symptoms stayed under control. Duration was also increased within the patient's tolerance. Using this intervention plan, Leddy et al found that 72% of patients returned to full daily functioning.

In following a training program like this, it is important to ensure that patients are being honest in the reporting of symptoms, because many individuals are motivated to return to sport or normal activity levels and may underreport symptoms. If patients have persistent baseline symptoms of any sort before the aerobic exercise training, these symptoms need to remain stable without exacerbation for multiple treatment sessions before progression of the intensity or duration of the training program.

Acute Management Issues

Immediately after injury and for the first few days, those with concussion should be monitored closely and encouraged to rest if symptoms are present. Rest includes both physical and cognitive rest, which includes avoiding social interaction, as well as not using telephones and television. Playing video games should be avoided because of the cognitive engagement and visual motion stimulation. Because of impairments in sensory processing and limited cognitive resources for managing complex environments, busy community or social events may cause space and motion discomfort or visual motion intolerance. These environments may be overwhelming and cause increased symptoms, and as such, these experiences should be avoided in the early stage. As symptoms resolve, graded exposure to complex sensory environments is recommended. Majerske et al found that individuals who returned to play or work and experienced high levels of exertion had increased symptoms and cognitive impairments.[100] Limiting activity in the short term after concussion will likely result in a quicker resolution of symptoms and return to full function.

Education is a critical component of successful recovery at this stage. When written information about concussion was supplied to injured subjects at the onset, the subjects

reported significantly less stress 3 months post-injury.[101] A clear understanding of the risks of repeated concussions and the physiological rationale behind appropriate levels of rest will help to avoid prolonged or incomplete recovery. Involvement of parents, coaches, and trainers in the education process, as appropriate, should improve the consistency of the activity restrictions. Because of the cognitive and processing deficits, both written and verbal communication should be used to improve information retention. In clinical settings, patients may appear to understand and assimilate your verbal recommendations, but delayed recall may still impair their ability to remember the recommendations when they leave the clinic. When exercises are prescribed for a patient, written instructions and an exercise log may be beneficial to enhance compliance with the exercise program.

In the acute stage, activity level (such as returning to school), exercise, or social interaction should only be restricted if the individual reports symptoms. Progression of the individual's activities should occur when the individual is symptom-free with the current level of activity. Although exertion should be restricted if exacerbation of symptoms occurs, all types of activity should be moderated according to symptom levels. If, for example, brisk walking causes an increase in symptoms but static postural control activities do not, then the static postural control activities tasks should be recommended. All subjective symptoms should be considered, including headache, mental "fogginess," dizziness, blurred vision, fatigue, or nausea.

In the acute and subacute stages, function may be limited secondary to pain from musculoskeletal involvement. Occasionally, back and neck pain are not identified until the patient begins to move more in therapy. Upper cervical spine instability can be a consequence of head injury, so a careful assessment of the cervical spine is important.[102] If cervical involvement is noted, a cervicogenic basis for symptoms may be present. See Chapter 31 for more information on the management of cervicogenic contributions to dizziness. If vestibular exercises are indicated, they may need to be postponed, or modified, until cervical limitations allow freer movement. Smaller amplitude cervical movements that remain within the individual's pain-free range may be performed, which will still allow for appropriate stimulation of the vestibular system.

BPPV should be ruled out in all patients following head trauma, because head trauma is a common cause of BPPV.[31] Patients may move slowly and carefully after TBI and may not be aware of their positional symptoms right away, so therefore may not report dizziness with position changes.

For individuals with symptoms of dizziness following head trauma, referral for vestibular function testing may occur at this time, depending on the practice pattern of the program. Some facilities will perform vestibular function tests automatically with any report of dizziness, but others wait until there is clear delineation between vestibular symptoms and non-vestibular dizziness. If there is any indication that a vestibulopathy is present during physical therapy assessment, a referral may be appropriate. If a patient has dizziness that is vestibular in origin, it is appropriate to initiate physical therapy intervention at this time as long as the interventions are within the patient's tolerance. A conservative approach may be wise initially, to ensure that the patient does not experience a marked and prolonged exacerbation of symptoms. If a patient demonstrates motion sensitivity and postural stability impairments, initiating balance tasks is a good place to start the intervention. Eye-head coordination tasks may be recommended if they are well tolerated. Exercises will most likely provoke symptoms, but the resulting dizziness should measure no more than a 3 to 4 (out of 10) using either a numerical rating scale or visual analog scale, and the symptoms should resolve within a few minutes after completion of the task.

Management Considerations with Chronic TBI

There is no clear delineation for what is considered an acute or chronic phase of recovery with TBI. The medical literature generally categorizes the chronic phase as anything longer than 1 to 3 months. Although metabolic processes may still be slowed, the true metabolic crisis that is present post-injury has resolved by approximately 3 months.[11] The resulting symptoms and functional impairments may be caused by axonal death, or altered neural connectivity.

The rehabilitation process continues, again following a problem-oriented approach. The therapist should be aware that individuals with TBI often have problems beyond those seen in the typical patient with a unilateral or bilateral vestibular deficit. Migraine headaches are common, and if present, medical management is important for resolution of the headache, which would otherwise interfere with exercise and activity tolerance. If musculoskeletal injuries exist, especially cervical, one must address them via physical therapy and/or medical management. Emotional distress tends to be evident at this point, because people experiencing a prolonged recovery may be affected by long periods of inactivity or a lack of productivity. If anxiety issues are present, medical or psychological management may be indicated.

Stress management, which is usually situational because of the recent injury, but can be a result of pre-injury factors, can become integral to recovery. Hanna-Plady et al monitored individuals with mild TBI and post-concussive symptoms exposed to high stress and low stress conditions

as compared with non-injured individuals.[103] The investigators found increased symptoms, decreased speed of information processing, and subtle memory deficits during the high stress condition that were greater in the individuals with mild TBI as compared with the non-injured individuals. Although relaxation training and stress-management training are not typical functions of the physical therapist, there are resources available that may help patients manage these issues. A myriad of relaxation training resources are available in podcast and CD formats. Mantra repetition, the use of a short word or phrase as part of meditation, may be incorporated into meditation training and then be available for use when patients find themselves in stressful or symptom-producing situations.

Evidence for Efficacy of Vestibular Rehabilitation with TBI

The literature related to mild TBI has been growing but has primarily been focused on sports-related injuries and trauma in the military. There is still much to be learned in the areas of diagnostic imaging, etiology of dizziness, measurement of recovery, efficacy for medical management, standards of medical care, and rehabilitation. The studies regarding vestibular rehabilitation for individuals with TBI have thus far been retrospective studies without control groups or descriptive studies without control groups.

In two studies that focus on service members in the military with mild TBI, Hoffer et al characterized patients by the etiology of their dizziness and then monitored their recovery following vestibular rehabilitation.[13,104] In the first study, 58 military members post-injury were categorized by the etiology of their dizziness into three categories: BPPV, migraine-related dizziness, and spatial disorientation. Twenty-eight percent of the individuals had BPPV, which was cleared in 1 week via canalith repositioning maneuvers, with resulting resolution of their symptoms. Forty-one percent of the individuals were categorized as having migraine-related dizziness, and 19% of the individuals were diagnosed with spatial disorientation. These two groups underwent physical therapy with a customized intervention program using vestibular rehabilitation techniques. The migraine-related dizziness group had resolution of symptoms within 8 weeks. The spatial disorientation group required up to 39 weeks for resolution of symptoms. In this study, 9% of subjects could not be classified. In the second study, a fourth etiological category was identified: exercise-induced dizziness, which accounted for 7% of the individuals.[104] This group was also treated with customized physical therapy interventions and required approximately 4 weeks for resolution of symptoms. In a prospective study, Gottshall and Hoffer studied soldiers returning from combat with mild TBI from blast injury. The subjects underwent physical therapy utilizing vestibular rehabilitation techniques for gaze stabilization training, balance training, and habituation exercises for motion sensitivity.[14] The investigators documented significant improvement in gaze stability, gait, and balance after 12 weeks.

There is only one study thus far that includes subjects that were not exclusively athletes or the military. Alsalaheen et al reviewed 84 patients in a retrospective study of those referred for vestibular rehabilitation following mTBI.[30] The patients underwent customized physical therapy interventions using vestibular rehabilitation techniques, such as eye-head coordination exercises, habituation exercises, static and dynamic postural control training, and dynamic gait tasks involving walking with head turns and direction changes. The patients improved significantly in the areas of subjective dizziness, gait, and balance within 5 weeks.

There is clearly a need for continued research in the area of vestibular rehabilitation for those with traumatic brain injury. Studies with appropriate control groups are needed to determine treatment efficacy. In addition, studies related to dosage of the exercises, timing of the exercises, and progression of the rehabilitation program are needed.

Coordination of Care and the Team Approach

Given the diverse nature of impairments that can occur with TBI, a multidisciplinary team approach that addresses all aspects of recovery is an important aspect in the care and rehabilitation of the individual with a TBI. In a multidisciplinary team, neurologists and physiatrists may work to control headaches and manage pain and orthopedic comorbidities, and psychiatrists can manage preexisting mental health needs or developing anxiety. Neuropsychologists may monitor and address cognitive deficits, and counseling psychologists can work with patients on anxiety management, relaxation training, and issues related to adjustment. Occupational therapists and speech therapists may work on executive level skills and address cognitive and communication deficits. Physical therapists address the dizziness, balance, exertion, and orthopedic components of rehabilitation. Case managers assist in bringing team members together for regular collaboration or conferences, and also act as liaison for external case management, employers, team trainers, teachers, and school officials.

With the increased incidence of TBI among service personnel as a result of recent armed conflicts, the military has developed an integrated, multidisciplinary team approach that may serve as a model for comprehensive care. Patients presenting to a military vestibular clinic with mTBI and symptoms of dizziness undergo a detailed history and

standardized physical examination by a team composed of a neurotologist and a physical therapist. In addition to the physical examination, the patients undergo complete auditory and vestibular test batteries, including rotational chair testing, VNG test battery including caloric examination, vestibular-evoked myogenic potentials (VEMP), an audiogram, and electrocochleography (ECOG). MRI is obtained as indicated by the patient's physical examination and test results. Military centers have instituted a program called the POWER: *P*rogram *O*f *W*ellness *E*ducation and *R*ecovery. The program uses medical providers from physiatry, neuropsychology, primary care, physical therapy, occupational therapy, speech therapy, mental health, and case management. There are many program goals, but through organized and coordinated services as well as structured programs, the overriding goal for the program is to focus on wellness and education with a positive expectation for recovery and return to duty. As part of the educational program, the Vestibular Balance Awareness and Safety Course includes an overview of vestibular anatomy and function, factors contributing to balance, strategies for decreasing dizziness and unsteadiness, how drugs affect motion sickness, and how alcohol affects vertigo. The physical program includes exercises specific to the individual's impairments and strategies for relaxation, activities to increase endurance and decrease fatigue, exercises and activities to promote core strengthening, overall strength and flexibility, and agility.

Prognosis: Factors That Contribute to Outcome

According to several studies of sports concussion, mTBI resolves in most cases within 2 weeks.[23,105,106] Dizziness at the time of injury was the sole on-field sign or symptom that was associated with a protracted recovery, which was defined as greater than 20 days.[107] Ten to fifteen percent of those diagnosed with concussion will have a prolonged recovery (greater than 7 days).[108,109] Several factors have been associated with longer recovery times and greater impairments:

- Age: Adolescents require more time and rehabilitation services following mild TBI.[23,110,111]
- Amnesia: There is some evidence to suggest that loss of consciousness is not a predictor for recovery.[112] However, amnesia (retrograde or anterograde) on the sideline is a predictor for longer recovery and presence of neurocognitive deficits.[112]
- Exertion: High levels of exertion or activity level post-injury is associated with greater impairment in neurocognitive scores and greater symptoms 1 month post-concussion.[100]
- Headaches: Athletes with post-concussion headaches or migraines have higher rates of amnesia, lower neurocognitive scores, and higher reported symptoms.[38,113]
- Symptoms: Individuals with symptoms of "fogginess" following a concussion have higher scores for other symptoms and lower scores on neurocognitive testing.[114]
- History: Amateur athletes with multiple concussions (3 or more) have greater baseline symptoms, have increased chance of a significant drop in memory performance with subsequent concussions, are more likely to sustain another concussion, and have prolonged recovery times.[115,116]
- Gender: Female athletes report more post-concussive symptoms compared with males and demonstrate more impairment on computerized neurocognitive testing.[117,118]

CASE STUDY 26-1

The patient was a 24-year-old Caucasian male on active duty in the United States Navy. The mechanism of injury was a fall from 40 to 60 feet, resulting in multiple fractures of his skull including a left temporal bone fracture with otic capsule sparing, hemotympanum, and dislocated ossicles. He also sustained fractures of the left mastoid, right maxilla (suboccipital region), and four left ribs, as well as a right pulmonary contusion. The patient denied any loss of consciousness but was stunned right after the fall. Five days after the initial injury, the patient began experiencing weakness and sensory loss in the left side of his face and was diagnosed with axonal neuropathy of all three branches of cranial nerve VII. A gold weight was implanted in the left eyelid to assist with eye closure. The patient had a profound mixed hearing loss between 250 and 8,000 Hz with absent acoustic reflexes in the left ear. This hearing loss did not resolve with intratympanic steroid injections. The patient underwent ossicular chain reconstruction surgery to restore the correct anatomical position and obtain normal mobility of the ossicular chain.

The patient presented to physical for evaluation 19 days after injury. The patient's chief complaints

Continued

CASE STUDY 26-1

were dizziness with positional changes and head motion, unsteadiness with walking, and spatial disorientation that was worse in the dark. He also complained of persistent loss of hearing on the left and tinnitus. He also reported drooling when attempting to drink from a cup. He was taking no medications.

The patient reported that the following activities exacerbated his dizziness or unsteadiness: looking up or down, sit to stand transitions, head movements while walking, walking in the dark, aerobic exercise if not seated, rapid head movements, loud noises, and bumps in the road when riding in the car. The patient's goals for physical therapy were to resolve his dizziness and unsteadiness during activities so he could return to full duty status as a Navy Seal.

The clinical oculomotor exam revealed right gaze-evoked nystagmus, convergence insufficiency, saccade slowing, abnormal smooth pursuit, and a positive head thrust with head rotation to the left. Upon clinical exam, the Weber test tone lateralized to the right. Caloric testing revealed a 58% asymmetry with reduced responses in the left ear. Rotational chair testing revealed decreased gains with rotation to the left. Subjective Visual Vertical (SVV) testing was abnormal, consistent with left utricular dysfunction.

The patient completed the Dizziness Handicap Inventory (DHI) and Activity-specific Balance Confidence Scale (ABC). Computerized dynamic posturography was performed, including the Sensory Organization Test (SOT), the SOT Head Shake Test, Motor Control Test (MCT), and Adaptation Test. The Functional Gait Assessment (FGA) and High-level Mobility Assessment Tool (HiMAT) were performed as clinical measures of dynamic balance. Functional assessment of the VOR was performed using the Dynamic Visual Acuity Test (DVA) and Gaze Stabilization Test (GST).

The patient's test results indicated abnormalities in VOR function, deficits in somatosensory integration, impaired gait, and motion sensitivity.

Plan of Care

Based on the initial evaluation and test data, short-term and long-term goals were established. The short-term goal (3 weeks) was to be able to walk 20 feet with head turns and without path deviation, as measured by the FGA. The long-term goal was for the patient to decrease DVAT LogMAR loss to no more than 0.20 in 6 weeks. The patient's self-goal was to return to full performance in his assigned position.

Intervention

The patient received vestibular physical therapy four times per week, for 6 weeks, each session lasting for 1 hour. The patient was retested at 3-, 6-, and 9-week time points. After 9 weeks, GST deficits remained. Physical therapy continued for 3 more weeks before resolution of the GST deficits. Physical therapy interventions are listed below.

To improve gaze stability during head movements:

- X1 and X2 viewing exercises: The gaze stability exercises were progressed from sitting, to standing, to walking. As the patient progressed, the velocity of the head movement was increased and more visually challenging targets were used. The progression ended with performing exercises several stories above ground level on walkways between buildings.
- Remembered target exercises: The exercises were performed initially in sitting and progressed to standing and then walking.

To improve convergence:

- The patient stood with arms against a wall, while looking at a fixed object in front of him on the wall. The patient's task was to maintain focus on the object while doing a push-up against the wall.
- Active slow convergence: The patient started looking at the tip of his finger held at arm's length. He slowly brought the finger closer to his nose, trying to keep it in focus. This was practiced for 1 minute, first in sitting and then progressed to a standing position.
- Active fast convergence: looking from far to near targets of varying sizes
- Brock string exercises

To improve standing balance:

- Standing balance exercises with arms crossed, eyes closed, and a decreasing base of support. Performed initially with head level and then with head tilted back.
- Standing balance exercises with arms crossed, eyes closed, and a decreasing base of support. Performed with active weight shift forward, backwards, and side to side (without loss of balance), and holding each position for a short period of time before switching to the next position.
- Standing balance exercises with arms crossed, eyes closed, a decreasing base of support and a slight knee-bend.
- These exercises were progressed by changing the support surface from the floor to a softer, compliant surface.

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To improve dynamic balance:

- Walking with head turns horizontal, vertical, and diagonal
- Turning corners and pivot turns while walking
- Skipping, hopping, lunging
- Stair ascent and descent

To decrease motion sensitivity:

- Visual tracking: To decrease visual motion sensitivity, the patient held a visual target at arm's length, and then moved the target side to side, up and down, and in diagonal directions, tracking the object with his eyes and keeping his head still. This exercise was performed initially in sitting, but progressed to standing, and then to walking.
- Rapid, large-amplitude head movements: To decrease the symptoms of dizziness that occurred with head movements and position changes, the patient performed the following movements: sit to supine; sit to bending forward, sit to stand with head turn, standing full body diagonals, and standing pivot turns.

To decrease exertion-induced symptoms and improve aerobic conditioning:

- Aerobic exercise: Patient exercised first on a stationary bike, and was then progressed to an elliptical machine, and finally to swimming, with steadily increasing demands. Progression from one activity was based on the patient's ability to perform the activity without provocation of dizziness.
- Job specific activities: Given the patient's job requirements, the swimming exercises progressed from distance swims to sprints with flip turns in daylight and in dark. In addition, the patient attended surf clinic and used the surf kayak, stand-up paddle board, and surfboard.

The patient w as also follo wed by audiology , where he was fit with hearing aids and was taught tinnitus adaptation strategies.

Outcomes

After 9 weeks of vestibular physical therapy, the patient had met his short-term goal, and had partially met his long-term goal. Table 26-2 summarizes the changes in

■ Table 26-2 SUBJECTIVE AND OBJECTIVE OUTCOME MEASURES FOR CASE STUDY 26-1

*Measures	Initial Visit	9 Weeks
DHI	44/100	12/100 (CSI)
ABC	54%	93% (CSI)
FGA	24/30	30/30 (WNL)
SOT Composite Score	65 (abnormal)	83 (WNL)
DVA – Left	0.4LogMAR loss (impaired)	0.38LogMAR loss (impaired)
DVA – Right	0.4LogMAR loss (impaired)	0.1LogMAR loss (WNL)

*Measures and abbreviations
DHI – Dizziness Handicap Inventory (0–100, 100 = worst)
ABC – Activities-specific Balance Confidence Scale (0%–100%, 100% = best)
FGA – Functional Gait Assessment (0–30, 30 = highest score <22 = fall risk in older adults)
SOT – Sensory Organization Test (composite score <70 = abnormal)
DVA – Dynamic Visual Acuity Test (normal ≤0.2 LogMAR difference)
VRBQ – Vestibular Rehabilitation Benefits Questionnaire (0%–100%, 100% = worst)
VVAS – Visual Vertigo Analog Scale (0–10, 10 = worst)
HiMAT – High-level Mobility Assessment Test (HiMAT) (0–54, 54 = best score)
CSI – clinically significant improvement
WNL – within normal limits

Continued

CASE STUDY 26-1

the outcome measures between the initial visit and the 9-week time point.

Discussion

This patient's success can be seen in the normalization of most outcome scores throughout his treatment. He did not meet his GST long-term goal of achieving 180 degrees per second in all directions, nor did his DVA for head rotation to the left return to normal levels. Standing and dynamic walking and aerobic function improved by 6 weeks. The patient was able to return to full active duty with his unit 7 months after injury.

CASE STUDY 26-2

The patient was a 52-year-old male, who was working as a police officer when he was involved in a motor vehicle accident. His car was hit on the passenger side, and forced off the road onto an embankment. He did not lose consciousness but has no memory of approximately 30 minutes after the accident. He was extricated from the vehicle and transported to the nearest emergency department. The initial injury report identified cervical whiplash, dizziness, headache, nausea, vomiting, and irritability. He was released home to follow up with neurologist within 1 week.

Before his initial physical therapy appointment, he had been evaluated by neurology and neuropsychiatry. His headaches met criteria for migraine, and the neurologist had prescribed amitriptyline and zolmitriptan for headache management. A neuropsychological assessment identified impairments in sustained attention, selective attention, response time, and verbal memory. Based on these findings, it was recommended that all information and instructions given to the patient be written. Neither vestibular function tests nor an audiogram had been performed. A driver's evaluation was recommended when appropriate.

The patient presented to physical therapy for evaluation 35 days after injury. The patient's chief complaints were headache, photophobia, phonophobia, dizziness with any head motion, blurred vision, and imbalance. His spatial disorientation increased in the dark or with eyes closed. He reported visual motion intolerance in any environment other than home. He noted marked exacerbation of symptoms when he attempted to attend his daughter's lacrosse match. He reported difficulty initiating sleep; he had been working nights and was having difficulty with adjusting his sleep cycle before the MVA. The patient also reported irritability and anxiety, as well as increased frustration at being unable to exercise.

The patient was married, lived with his wife and three teenage daughters, and before the MVA had been running 4 to 6 miles per day and was active in the community in addition to working full time. At his initial physical therapy visit, he was not driving and not working. He was unable to exercise and had been sedentary.

The patient reported that the following tasks exacerbated his dizziness or unsteadiness: walking with head motion, standing or walking in dark conditions, aerobic exercise, moving his head rapidly, loud noises, reading/computer work, and visual motion. He could not read his phone. He rated his symptoms of dizziness as follows: at rest: 0/10, with typical activities: 4/10, at worst: 7/10.

The clinical oculomotor exam revealed direction changing, gaze-evoked nystagmus, a 4-line degradation on the clinical DVA test, and an abnormal near point of convergence at 20 cm with symptoms of dizziness during testing. All other oculomotor tests were normal.

Results for subjective tests and balance tests are summarized in Table 26-3. There were no limitations in extremity or trunk ROM, strength, coordination, or sensation. No cervical pain or limitations were identified; the symptoms associated with the initial cervical injury had resolved.

The following problems were identified based on the patient's report and clinical examination:

- Visual motion intolerance
- Visual dependence for balance
- Gaze stability deficits during head movements
- Convergence insufficiency
- Head motion sensitivity
- Exertion-provoked symptoms

Because of the patient's ongoing dizziness and abnormal results of the oculomotor exam, the patient was recommended for vestibular function testing. The test

CASE STUDY 26-2

■ Table 26-3 **SUBJECTIVE AND OBJECTIVE OUTCOME MEASURES FOR CASE STUDY 26-2**

*Measures	Initial Visit	12 Weeks
DHI	56/100	22/100 (CSI)
ABC	65%	95% (CSI)
VRBQ	38%	12% (CSI)
VVAS	7.2/10	2.4/10
Dizziness Severity (0–10, 10 = worst)	0 to 7/10	0 to 2/10
FGA	22/30	30/30 (WNL)
HiMAT	Unable to tolerate	50/54
Clinical DVA	4-line difference (impaired)	2-line difference (WNL)
Romberg eyes closed / compliant surface	5 seconds, abnormal with increased sway	30 seconds, normal stability

*Measures and abbreviations
DHI – Dizziness Handicap Inventory (0–100, 100 = worst)
ABC – Activities-specific Balance Confidence Scale (0%–100%, 100% = best)
VRBQ – Vestibular Rehabilitation Benefits Questionnaire (0%–100%, 100% = worst)
VVAS – Visual Vertigo Analog Scale (0–10, 10 = worst)
FGA – Functional Gait Assessment (0–30, 30 = highest score <22 = fall risk in older adults)
HiMAT – High-level Mobility Assessment Test (HiMAT) (0–54, 54 = best score)
DVA – Dynamic Visual Acuity Test (normal ≤0.2 LogMAR difference)
CSI – clinically significant improvement
WNL – within normal limits

results were as follows: ENG test battery revealed a 38% asymmetry to caloric testing with decreased responses in the right ear, persistent direction changing, gaze-evoked nystagmus; rotary chair testing revealed low VOR gain for rotations to the right and a right directional preponderance.

Plan of Care

The patient verbalized the following goals: resolution of dizziness and unsteadiness, return to exercise, be able to participate in daughter's sports activities, and return to full-duty police officer. The following short- and long-term goals were set:

Short-term Goals (4 weeks)
- FGA improved to 27/30
- Static standing eyes closed in semi-tandem for 30 seconds
- Able to tolerate 60 seconds of gaze stabilization task with dizziness rated as less than 4/10
- Able to exercise on stationary bike for 15 minutes at a level of intensity 80% of the symptom threshold level

Long-term Goals (12 weeks)
- FGA improved to 30/30
- A 2-line or less difference between static and dynamic visual acuity in the clinical DVA test
- HiMAT score greater than 46
- Able to exercise via jogging for 30 minutes without an increase in dizziness or headache
- Able to tolerate 30 minutes of complex visual environments that include noise and visual motion without increase in dizziness and headache
- Return to all work duties

Continued

CASE STUDY 26-2

Intervention

The patient was scheduled to receive vestibular physical therapy twice a week for 3 weeks, once a week for 5 weeks, then once every 2 weeks for 4 weeks. Therapy sessions were 1 hour in duration. The patient was retested at 4 weeks, 8 weeks, and 12 weeks.The patient cancelled two appointments because of migraine; the sessions were rescheduled within the week. He attended 13 sessions total.

A portion of the first four visits was devoted to patient education regarding migraine management, rationale for vestibular rehabilitation, pacing, and reinforcement of previously recommended relaxation techniques. All the information was also provided in the form of written handouts.

A 10-point symptom scale to assess dizziness and headache was used at the be ginning and end of each session. The scale was used periodically during the session to monitor the patient's tolerance of symptom-exacerbating tasks. The specific physical therapy interventions are listed below.

To improve convergence:

- Brock string exercises
- Pencil push-ups
- Note: Near point of convergence resolved to 10 cm without symptoms within 2 weeks. The patient still reported dizziness with computer scrolling but found that reading on the paper or phone had improved. Based on the normal performance on convergence, no further referral was recommended for this condition.

To improve gaze stability during head movements:

- X1 and X2 viewing exercises: The gaze stability exercises were started in standing with feet apart. As the patient progressed, the exercises were performed with a narrow base of support and then while walking. Increasing the velocity of the head movement, the use of busy backgrounds, and performing the exercises in complex visual and auditory environments were added to the exercise program as tolerated.

To improve standing balance:

- Standing balance exercises with eyes closed on firm and compliant surfaces: Decreasing the base of support, weight shifting with eyes closed, the use of a rocker board and bosu ball were used as exercise progressions.

To decrease motion sensitivity:

- Visual motion exercises: The patient began the visual motion exercises at 4 weeks. The initial exercises consisted of watching videos taken while individuals walked through busy urban environments. Initial exposure time was 5 minutes, which was progressed to 15 minutes. When the patient could tolerate this visual motion exposure, he started graded exposure to community environments including the grocery store, observing daughter's lacrosse practice, watching his daughter's game, and then busy restaurants.
- Head motion exercises: The habituation exercises consisted of progressively more complex and challenging movements—gait with head turns and whole body turns, combined eye and head tracking in standing then while walking, ball toss in standing and then while walking. As the patient progressed, repetitive motion tasks using high and low surfaces, agility drills, and rapid random direction changes were added to walking activities.

To decrease exertion-induced symptoms and improve aerobic conditioning:

- Stationary bicycle: 10 minutes duration. The duration was increased when the patient had dizziness rated as no more than a 3/10 that lasted for less than 10 minutes after completion of the stationary bicycle exercise.
- Jogging on a treadmill: This was progressed by increasing the speed and duration of the exercise, using the same guidelines as described for the stationary bicycle exercise. Jogging outside: Once patient was able to jog 15 minutes on treadmill with good tolerance, he began to jog outside.
- High-level agility tasks were added for the last 6 weeks, including hopping, jumping, obstacle climbing (low wall), and carrying up to 50 lb.
- NOTE: Patient was instructed to avoid these exercises if he was experiencing a migraine.

Outcomes

During the course of recovery, patient's headaches resolved gradually with 1 to 2 headaches per month by discharge, which he was able to control with Zolmitriptan. He reported significant irritability, lack of patience

CASE STUDY 26-2

with family, and situational anxiety (financial, change in life role). These symptoms gradually resolved without medical intervention, using relaxation techniques and aerobic exercise. His sleep had also improved with the use of relaxation podcasts, and he had returned to nighttime sleeping.

After 12 weeks of vestibular physical therapy, the patient had met all goals (Table 26-3). He had passed the driver's evaluation and had returned to regular exercise, although he was not back to previous fitness levels. His symptoms of dizziness had resolved. The patient still reported some difficulty navigating in the dark, but had incorporated compensatory techniques. He had returned to work in an administrative position, daytime shift, and had elected to not return to his previous position because of safety risks.

CASE STUDY 26-3

The patient was a 21-year-old female, collegiate basketball player and premedical student, enrolled in her junior year. During a basketball game, she was struck on the head with an elbow and knocked to the floor hitting the back of her head on impact. She did not lose consciousness and reported no amnesia. She was pulled from the game, was given ice and ibuprofen, and the athletic training staff monitored her status. Sideline testing was administered but not available for review at the time of her physical therapy evaluation. She reported that her initial symptoms were headache, dizziness, fogginess, and blurred vision. The patient returned home and sought medical attention through student health services the next day. The patient was referred for CT scan and a consult with neurology. The CT scan was performed 2 days later and was read as normal. The neurology consult recommended vestibular rehabilitation and identified occipital neuralgia. The patient was treated with occipital injections that resolved her headaches.

The initial neuropsychological assessment identified mild impairments in attention span, selective attention, visual memory, and working memory. The patient was also experiencing hypersomnia and fatigue. Neither vestibular function tests nor an audiogram had been performed.

The patient presented to physical therapy for vestibular rehabilitation for evaluation 15 days after injury. Her chief complaints at that time were dizziness with rapid head motion; blurred vision; imbalance; spatial disorientation, which was increased in the dark or with eyes closed; and an inability to exercise on a treadmill for more than 5 minutes without provocation of her dizziness.

The patient lived in student housing, had been practicing or playing basketball 6 days per week, and was a full-time student with 3.8 GPA. Her past medical history included left ankle fracture at age 10, which was fully resolved. Her head injury happened at the end of the season. She had completed final exams the week before the injury, and she had 6 weeks until start of the new semester.

The patient reported that the following tasks exacerbated her dizziness or unsteadiness: walking with head movements, standing or walking in dark conditions, aerobic exercise greater than 5 minutes, rapid head movements, reading/computer work, and visual motion. She rated her symptoms of dizziness as follows: at rest: 0/10, with typical activities: 4/10, at worst: 5/10.

The clinical oculomotor exam revealed a 3-line difference between static and dynamic visual acuity on the clinical DVA test with provocation of her dizziness to a 4/10. The remainder of her clinical oculomotor testing was normal, but VOR cancellation test also reproduced 4/10 dizziness. In both cases the dizziness resolved within 2 minutes.

Results for subjective tests and balance tests are summarized in Table 26-4. There were no limitations in her extremity or trunk ROM, strength, coordination, or sensation. No cervical pain or limitations were identified. The patient reported generalized stiffness, likely from inactivity.

The following problems were identified based on the patient's report and clinical examination:

- Visual motion intolerance
- Visual dependence for balance
- Gaze stability deficits during head movements
- Head motion sensitivity
- Exertion-provoked symptoms

Continued

CASE STUDY 26-3

Plan of Care

The patient verbalized the following goals: alleviation of the dizziness, return to school, and return to sport. The following short- and long-term goals were established:

Short-term Goals (2 weeks)

- FGA improved to 27/30
- Able to tolerate 30 seconds of the X1 gaze stabilization exercise with busy background with provoked symptoms of dizziness less than 4/10
- Able to tolerate 10 minutes of watching an urban walking video with provoked dizziness less than 4/10
- Able to exercise on stationary bike for 15 minutes at a level of intensity 80% of the symptom threshold level

Long-term Goals (4 weeks)

- FGA improved to 30/30
- A 2-line or less difference between static and dynamic visual acuity on the clinical DVA test
- HiMAT score greater than 52
- Able to exercise via jogging for 30 minutes without an increase in dizziness
- Able to tolerate 30 minutes of complex visual environments that include noise and visual motion without an increase in dizziness
- Return to training with team, progression toward return to play

Intervention

The patient was scheduled to receive vestibular physical therapy twice a week for 4 weeks. Therapy sessions were 1 hour in duration. She attended seven sessions total.

Initial visits included patient education regarding the rationale for vestibular rehabilitation and pacing recommendations to avoid excessive exertion. The patient began a general stretching and 20-minute daily walking program. All information and instructions were also provided as written handouts.

A 10-point symptom scale to assess dizziness was used at the beginning and end of each session. The scale was used periodically during the session to monitor her tolerance to symptom-exacerbating tasks. The specific physical therapy interventions are listed below.

To improve gaze stability during head movements:

- X1 and X2 viewing exercises: The gaze stability exercises were started in standing with feet apart. As the patient progressed, the exercises were performed with a narrow base of support and then while walking. Increasing the velocity of the head movement, the use of busy backgrounds and performing the exercises in complex visual and auditory environments were added to the exercise program as tolerated.

To improve standing balance:

- Standing balance exercises with eyes closed on firm and compliant surfaces: Decreasing the base of support and weight shifting with eyes closed were used as exercise progressions.

To decrease motion sensitivity:

- Visual motion exercises: The initial exercises consisted of watching videos taken while individuals walked through busy urban environments. Initial exposure time was 5 minutes, which was progressed to 15 minutes. When the patient could tolerate this visual motion exposure, she started graded exposure to community environments including walking through the grocery store, attending a basketball game, and then using the subway.
- Head motion exercises: The habituation exercises consisted of progressively more complex and challenging movements—gait with head turns and whole body turns, combined eye and head tracking in standing then while walking, ball toss in standing and then while walking. As the patient progressed, repetitive motion tasks using high and low surfaces, agility drills, and rapid random direction changes were added to walking activities. Finally, she began sport-specific training that included tossing a basketball with rapid turns, hopping, and jumping.

To decrease exertion-induced symptoms and improve aerobic conditioning:

- Stationary bicycle: 10 minutes duration. The duration was increased when the patient had dizziness rated as no more than a 3/10 that lasted for less than 10 minutes after completion of the stationary bicycle exercise.
- Jogging on a treadmill: This was added after 2 weeks of using the stationary bicycle, and was progressed by increasing the speed and duration of the exercise, using the same guidelines as described for the stationary bicycle exercise.
- Jogging outside: Once patient was able to jog 15 minutes on treadmill with good tolerance, she began to jog outside.

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Outcomes

After 4 weeks of vestibular physical therapy, the patient had met all goals (Table 26-4). Her symptoms of dizziness had almost completely resolved. The patient still reported occasional difficulty with rapid agility moves. Reading and computer work no longer provoked her symptoms, and she had resumed attending school. She had returned to training with the team, but she was not yet cleared for return to contact/play. Her course of recovery was uncomplicated. Her hypersomnia and cognitive symptoms resolved over the 4 weeks. She was reassessed by neuropsychologist and had returned to baseline scores.

■ Table 26-4 **SUBJECTIVE AND OBJECTIVE OUTCOME MEASURES**

*Measures	Initial Visit	4 Weeks
*DHI	28/100	2/100 (CSI)
*VRBQ	76%	94% (CSI)
*VVAS	4.5/10	2.1/10
Dizziness Severity (0–10)	0 to 5/10 (frequently with typical tasks)	0 to 3/10 (occasionally with rapid movements)
*FGA	25/30	30/30 (WNL)
*HiMAT	42/54	53/54
*Clinical DVA	3-line difference (impaired)	1-line difference (WNL)

*Measures and abbreviations
DHI – Dizziness Handicap Inventory (0–100, 100 = worst)
VRBQ – Vestibular Rehabilitation Benefits Questionnaire (0%–100%, 100% =worst)
VVAS – Visual Vertigo Analog Scale (0–10, 10=worst)
FGA – Functional Gait Assessment (0–30, 30=highest score <22 = fall risk in older adults)
HiMAT – High-level Mobility Assessment Test (HiMAT) (0–54, 54=best score)
DVA – Dynamic Visual Acuity Test (normal ≤0.2 LogMAR difference)
CSI – clinically significant improvement
WNL – within normal limits

Summary

In conclusion, traumatic brain injury involves a complex sequelae of symptoms and comorbidities. The etiology of dizziness may be vestibular or non-vestibular in nature. The field of rehabilitation for mild brain injury is in its infancy, and there is much to be learned about recovery from vestibular dysfunction following mild TBI. Management of the whole patient, using a team approach, appears to be the most effective path toward effective rehabilitation.

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CHAPTER 27

Physical Therapy Diagnosis: Clinical Decision-Making for Vestibular Disorders

Susan J. Herdman, PT, PhD, FAPTA

The advent of direct access for physical and occupational therapists means that clinicians must be able to screen for and identify for a multitude of problems and make decisions about treatment and referral. At a minimum, therapists need to know when they should refer the patient to a more qualified therapist or to a physician. Sometimes the decision to refer to another health-care provider is made easily, such as when the patient's problems are clearly not under the purview of a therapist (e.g., headache or hearing loss). In other situations, the patient's problem may or may not be something appropriate for physical or occupational therapy, but the underlying cause needs to be managed by a physician. "Dizziness" is one of those patient problems. "Dizziness" is one of the most prevalent complaints for which people seek medical help,[1] with an estimated 90 million Americans over the age of 16 years having experienced it.[2] It can have significant consequences for an individual, with 33% of patients reporting that their professional activities are affected by dizziness and 14% changing or abandoning their profession.[2] Although dizziness can be caused by many different medical conditions, it is estimated that as much as 45% is caused by vestibular disorders.[2]

In this chapter, we discuss an updated paradigm for helping the therapist arrive at the proper physical or occupational therapy diagnosis for a vestibular disorder or to the decision to refer the patient to a physician. The diagnostic schematic presented here is a "work in progress" and is meant as a guide only for the therapist. It is offered as a framework for arriving at a physical diagnosis for patients with vestibular problems. Each of the physical therapy diagnoses presented should demonstrate commonalities across all persons with that diagnosis. There are two phases to making the physical therapy (PT) diagnosis. The first is in the history of the patient's complaints; the second consists of some simple clinical tests of the vestibulo-ocular system.

Diagnosis can be defined as "the art of distinguishing one disease from another."[3] In medicine, the identification of a particular disease leads to specific medical and/or surgical treatment. A physical or occupational therapy diagnosis differs from a medical diagnosis in that, rather than attempting to identify a particular disease, a constellation of symptoms and signs toward which physical and occupational therapy will be directed is identified. Once the PT diagnosis is achieved, the vestibular exercise approach can be identified (Table 27-1). Certainly, there will be times when the medical diagnosis and the PT diagnosis are the same—for example, benign paroxysmal positional vertigo (BPPV). However, there will be times when the diagnoses

■ Table 27-1 **DIAGNOSIS-DRIVEN TREATMENT**

Diagnosis	Treatment Options
BPPV	Canalith repositioning, Liberatory, Appiani, Gufoni, Brandt-Daroff
Unilateral vestibular hypofunction	Gaze stabilization and postural stabilization exercises
Motion sensitivity	Habituation exercises
Bilateral vestibular loss	Gaze stabilization and postural stabilization exercises
Central vestibular	Habituation

will differ. For example, vestibular neuronitis would be a medical diagnosis. As therapists, however, we do not treat the inflammatory process of vestibular neuronitis. A more appropriate diagnosis for physical therapy is unilateral vestibular hypofunction.

Clinical Decision-Making Paradigm for Vestibular Disorders

The original matrix has been modified several times since it was first introduced 14 years ago in the second edition of *Vestibular Rehabilitation*.[4] This most recent version reflects data from a study that identified several problems with the earlier versions.[5] That study examined 400 medical records of patients seen between 2003 and 2007 for complaints of dizziness. From those 400 records, 107 were selected that met the inclusion criteria of the symptoms, history, clinical examination, and vestibular function tests necessary to reach a diagnosis. The medical records were categorized according to six diagnoses made by the treating physician (unilateral vestibular hypofunction [UVH], bilateral vestibular hypofunction [BVH], benign paroxysmal positional vertigo [BPPV], migraine, Ménière's and central vestibular disorder). This then became the "gold standard" against which the clinical decision-making paradigm was compared. Each chart, with the physician diagnosis removed, was reviewed using the clinical decision-making paradigm question-by-question. The results identified two problems in the clinical decision-making paradigm. First, not all patients used the term "vertigo" to describe their symptoms, even when they had BPPV or a sudden onset of UVH. Second, patients were vague about the duration of symptoms during the initial episode, leading to both false positive and false negative assignment of diagnoses. These problems led to rather poor sensitivity of the paradigm to lead to the correct diagnoses of BPPV and UVH. As a result, the current clinical decision-making paradigm has been modified by adding new symptom complaints (episodic dizziness), a different time frame for BPPV (1 to 2 hours as well as less than 1 minute) and the clinical examination tools (e.g., head thrust test). Further study is needed to confirm the usefulness of these changes in the decision-making paradigm.

Physical Therapy Diagnosis and the International Classification of Functioning, Disability and Health (ICF) Model of Disablement

The ICF was developed by the World Health Organization specifically to provide a framework for the "description of health-related states"; it consists of three domains that can be used to describe the effect of different disorders or diseases on a person's health (Fig. 27.1).[6]

Each of the three domains is affected by both environmental and personal factors (Table 27-2).

In the decision-making paradigm presented in this chapter, PT Diagnosis is determined by information gained at the level of the body structure and function.

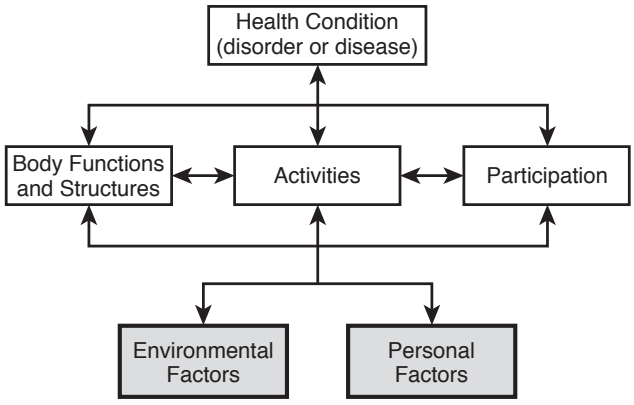

Figure 27.1 ICF Model of Functioning and Disability showing relationships between the health condition and its effect on structure and, indirectly, quality of life, as they are modified by environmental and personal factors.

■ Table 22-2 ENVIRONMENTAL AND PERSONAL FACTORS THAT INFLUENCE THE DOMAINS OF THE ICF MODEL

Environmental Factors		Personal Factors	
e.g.	Natural environment	e.g.	Gender, age
	Support and relationships		Comorbidities
	Attitude of family		Social background
	Attitude of society		Education and profession
	Services, systems, policies		Past experience
	Products and technology		Coping and character style

Modifiers for treatment are identified at the level of the individual (activities) and from environmental and personal factors. Outcome of treatment would then be measured at the level of the individual and participation levels. The paradigm uses four criteria in the decision-making process: Symptom, Tempo, Circumstance, and Signs. Of special importance in the patient's history are the nature of the patient's symptoms and the temporal quality (Tempo) of those symptoms when the problem first occurred. The nature of the symptom refers to whether the patient is complaining of vertigo (an illusion of movement, typically vertigo or spinning), dizziness (vague terminology that can include vertigo, lightheadedness), disequilibrium (the sense of being off-balance), motion sensitivity (sensitive to movement of the visual environment or of self) and pain, specifically neck pain. The temporal quality of the symptoms refers to whether the symptoms are episodic or continuous in nature. If the symptoms are episodic, it is important to establish the duration of the episode. For example, episodes of vertigo lasting seconds or less than 1 minute suggest BPPV. The circumstances in which the symptoms first appeared (positional, pressure induced, spontaneous, or movement induced) also help distinguish different diagnoses. Signs are determined through observation and performance of basic clinical tests. The first of these is positional testing. A positive response to positional testing would consist of vertigo and nystagmus being provoked when the patient's head is in specific positions. The duration and direction of the nystagmus is used to diagnose positional vertigo that is peripheral (e.g., BPPV; see Chapter 20) or central in origin. The second clinical test is the assessment of the gain of the vestibulo-ocular reflex (VOR) using rapid (high-acceleration) head movements (head-thrust test; see Chapters 8 and 10). The presence of the corrective saccade (positive head-thrust test) indicates the low gain of the vestibular system. If the person makes a corrective saccade following a head thrust to the right, the vestibular loss is on the right. If the person makes corrective saccades with head thrusts to the right and to the left, it indicates a bilateral vestibular loss. The sensitivity of the head-thrust test for identifying patients with unilateral vestibular loss is actually fairly low (35%), but its specificity is very high (95%).[5] That is, many patients with unilateral vestibular loss will not have a positive head-thrust test, but if the patient does have a positive test, it is likely that the patient has a vestibular deficit. The third clinical test is the observation of spontaneous nystagmus in room light and in the dark (Frenzel lenses or infrared goggles) and the determination as to whether the nystagmus is direction-fixed (suggestive of unilateral peripheral vestibular hypofunction) or direction-changing, which suggests a central lesion. Other observations include an ocular tilt reaction and lateropulsion, which can occur with a variety of deficits (Chapters 5 and 12). The last clinical test is whether body movement with the head stationary induces symptoms as would occur in cervicogenic dizziness (Chapter 31).

Diagnostic Flowchart

The first question asked is whether the patient has complaints of vertigo (Fig. 27.2). If the patient has a history of vertigo (spinning), the next step is to determine the duration of the vertigo. Spells of vertigo lasting for brief periods of time (less than 1 minute) suggest BPPV, the most common cause of vertigo resulting from a peripheral vestibular problem (see Chapter 20 for the diagnostic flowchart for BPPV). It is important to distinguish whether the nystagmus is typical for BPPV (combined torsional and vertical, or horizontal and torsional), is typical for a central lesion (pure vertical or pure horizontal), or if the history suggests another problem such as a perilymphatic fistula. Note that positional testing may not provoke vertigo and nystagmus. However, this

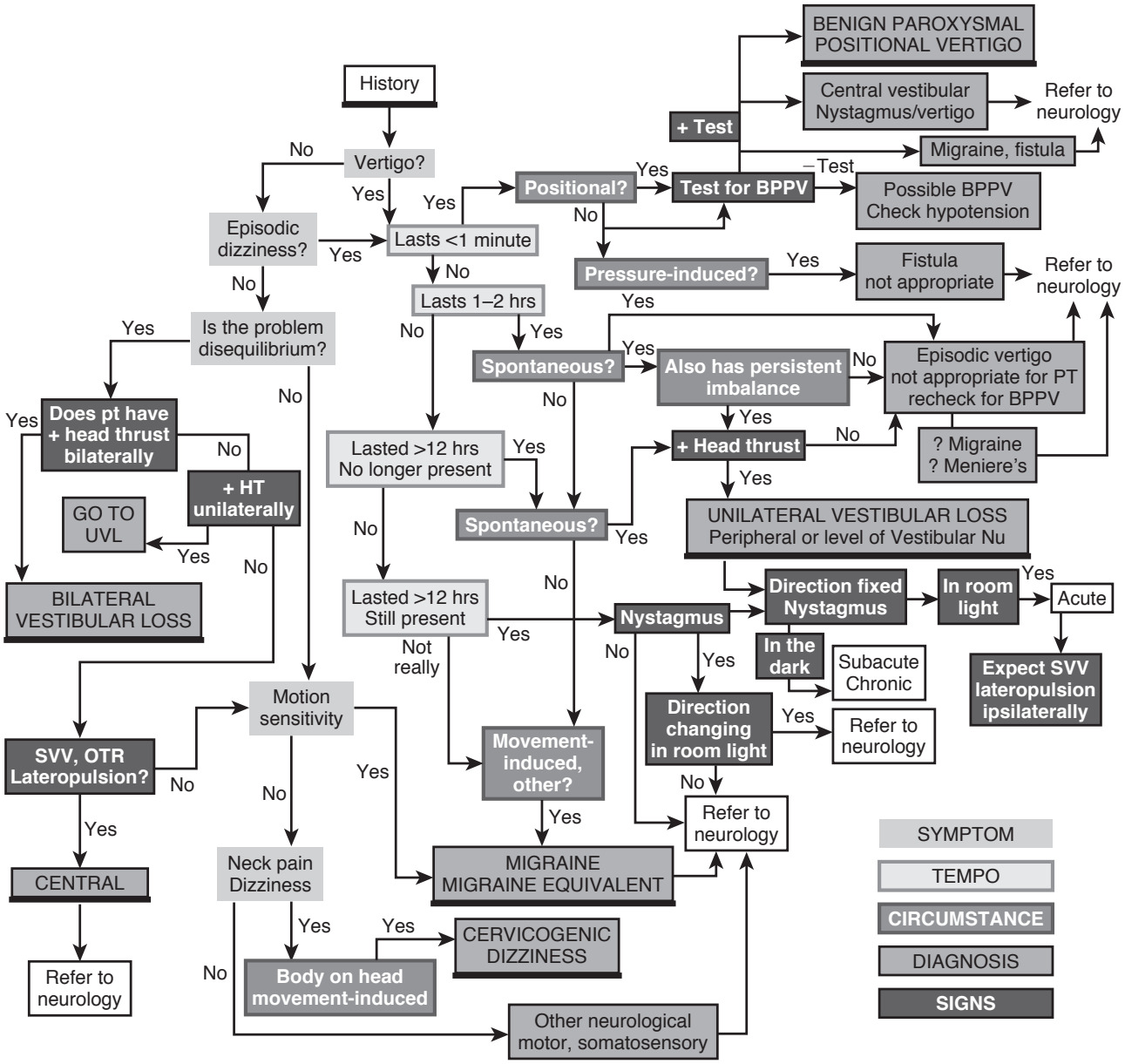

Figure 27.2 This flow diagram begins with the history (center). The patients should be asked if they have experienced vertigo (a sense of spinning). If the answer is "yes," the next series of questions deals with the temporal nature of that vertigo. (Column to the right: <1 minute, 1 to 2 hours, >12 hours, continuous). The temporal quality of the vertigo then leads to diagnoses of BPPV, episodic vertigo, UVL, continuous vertigo, and motion sensitivity. Note that some of these diagnoses are appropriate for PT treatment, and others are not. If, on history, the patient states that he or she has not experienced vertigo but rather has complaints of disequilibrium, the path leads to diagnoses that must be distinguished based on clinical examination. Note how the presence or absence of corrective saccades to rapid head thrusts is used to identify unilateral and bilateral vestibular loss. (UVL = unilateral vestibular loss, SVV = subjective visual vertical, OTR = ocular tilt reaction.)

does not mean that the patient does not have BPPV. The appearance of vertigo and nystagmus during testing is notoriously inconsistent. A diagnosis of BPPV may be reached, at least tentatively, by history alone. Also note that the possibility of a diagnosis of BPPV is raised regardless of the patient's description of the duration of the spell of vertigo. Patients are often poor historians and may believe the vertigo lasted for extended periods of time because they continue to feel poorly even after the actual spinning has stopped. In fact, all patients should be tested for BPPV, including those without complaints of true vertigo. BPPV is very common and has been identified in patients who also have UVH or BVH (see Chapters 22 and 23).

Vertigo that lasts for a few hours is most likely to be from Ménière's disease or may be a migraine-related event (see Chapters 4 and 15). These disorders are not appropriate for treatment using vestibular rehabilitation, and must be managed medically. The exception would be the patient who has persistent complaints of imbalance between these episodes of vertigo who might have developed UVH with repeated episodes of Ménière's disease. The head-thrust test may help to identify this problem, although formal vestibular function tests may be necessary.

Vertigo lasting 12 hours to days typically signifies the sudden onset of UVH (see Chapter 4). During the acute period following onset, spontaneous nystagmus in the light and possibly a skew deviation would be observed. Both should resolve within 1 to 2 weeks. Failure of spontaneous nystagmus, or of a skew deviation, to resolve strongly suggests central involvement of the structures responsible for compensation. Thus, the presence of spontaneous nystagmus in the light should be correlated with time from onset to determine if there is central involvement. Continuous vertigo has several possible explanations. First, the patient may be in the acute phase following onset of unilateral dysfunction. This can be easily verified by determining when the vertigo started. Additionally, if the problem is acute, the patient should still have spontaneous nystagmus that follows Alexander's Law (Chapter 10). Second, if the patient describes vertigo but there is no nystagmus when fixation is blocked (with Frenzel lenses or infrared goggles), it is not likely that the patient has a vestibular deficit that is treatable with exercises. Therefore, the patient should be referred to a neurologist. Unfortunately, looking for nystagmus with the patient looking at a Ganzfeld (unstructured, uniform visual background) may not be sufficient for this examination unless you can truly attest that the patient could not be suppressing nystagmus by fixating on a target. Third, the patient may not be actually experiencing vertigo continuously or may be misusing the word vertigo. For example, the patient may be experiencing movement-provoked symptoms or motion sensitivity. This implies that movement of the individual or movement of the environment produces symptoms that may include lightheadedness, nausea, and even an illusion of movement. Although this can occur following unilateral or bilateral vestibular loss, it can also occur in patients with other, non-vestibular problems such as migraine. Finally, complaints of continuous vertigo of long duration suggest problems for which the patient should be referred to a neurologist.

The patient may deny a history of vertigo but may complain of episodic dizziness. Note that episodic dizziness leads back to Tempo and specifically "last less than 1 minute." The patient may also complain of disequilibrium. There are many underlying etiologies for the complaint of disequilibrium, both vestibular and non-vestibular. The head-thrust test can be used to identify whether the patient has chronic UVH (positive to one side only) or bilateral vestibular loss (positive with head thrusts in both directions). Patients with apparently normal VOR to rapid head thrusts may still have a vestibular deficit, so further testing (vestibular function tests) may be needed to reach this diagnosis.[7]

The complaint of disequilibrium (a sense of imbalance) may also be an aspect of motion sensitivity. If patients with disequilibrium specifically have complaints of lateropulsion (and also have an abnormal ocular tilt reaction and abnormal subjective visual vertical), they probably have a central vestibular problem above the level of the vestibular nuclei (see Chapters 5, 10, and 12). The complaint of disequilibrium may be used by patients to describe motion sensitivity, leading to questions as to whether the patient is experiencing migraine or migraine-equivalent events. Finally, disequilibrium may be related to other neuromuscular problems.

Identification of Modifiers

Once a PT diagnosis has been achieved, based on history and elements of the clinical examination, it is necessary to perform other assessments to identify other problems that will modify the exercise program for each individual patient. These modifiers include comorbidities, subjective complaints, psychological factors, and the patient's premorbid activity level (Box 27-1). Finally, there are specific assessments that must be performed to establish the patient's baseline performance so that changes in status can be assessed following treatment and the patient's return to improved or normal participation at the societal level can be evaluated (Table 27-3).

Box 27-1

MODIFIERS OF TREATMENT

Activity Level
- Basic ADL—bath, dress, meals, clean
- Balance—gait speed, fall risk, endurance
- Fall history—circumstance, frequency, injury
- Driving—day, night
- Work—harder to perform, changed jobs, not working

Personal factors
- Visual—cataracts, macular degeneration, field
- Somatosensory—peripheral neuropathy
- Musculoskeletal—cervical, back, arthritis, strength, range of motion
- Central nervous system—stroke (e.g., brainstem), cerebellar disease
- Gender

Psychological
- Coping strategy
- Anxiety, depression
- Perception of disability
- Somatoform, conversion
- Severity of subjective complaints
- Secondary gain issues

Environmental factors
- Family support
- Availability of transportation
- Location of stairs, bedroom, bathrooms

■ Table 27-3 **FUNCTIONAL ASSESSMENT**

Problem	Tests
Subjective complaints	Visual analog scales Symptom scale Balance Confidence Dizziness Handicap Scale Anxiety and Depression scales
Visual acuity during head movement	Computerized Dynamic Visual Acuity Test Clinical Dynamic Visual Acuity Test
Balance in stance	Romberg Sharpened Romberg Single-leg stance
Balance while ambulating	Qualitative gait description Gait speed Dynamic Gait Index Gait deviations Gait with head turn
Quality of life	Vestibular Rehabilitation Benefits Scale Dizziness Handicap Inventory Disability Scale Vestibular Disorders – ADL Scale MOS 36-item short-form health survey

Summary

The use of this paradigm should enable the therapist to arrive at a diagnosis in which all patients have a common group of symptoms and signs that will indicate the appropriate treatments for that patient. The complete examination, including assessment of those factors that will result in modifications in how those exercises are applied, should enable the therapist to develop a problem list, goals, and treatment plan for the patient.

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CHAPTER 28

The Role of Emerging Technologies in Vestibular Rehabilitation

Courtney D. Hall, PT, PhD ■ *Dara Meldrum, PT, MSc, PhD* ■ *Susan L. Whitney, PT, PhD, NCS, ATC, FAPTA*

It is an exciting time in rehabilitation. In the past decade, there has been a proliferation of novel technologies designed to enhance rehabilitation outcomes. Virtual reality is one of those emerging technologies that has been applied to a variety of medical purposes including medical education and training, primary care, psychiatry, surgery, radiology, and more recently, rehabilitation. There is considerable excitement surrounding virtual reality across a multidisciplinary group of scientists and clinicians who are developing this new technology for physical, psychological, cognitive, and social rehabilitation purposes. Telehealth, the delivery of health-care services at a distance, has emerged as a significant component of the health-care delivery system with the advent of high-speed, high-bandwidth telecommunication networks. The combination of virtual reality with telerehabilitation holds great promise in increasing our ability to work with patients at a distance, offering greater access to care for rural patients. Although telerehabilitation incorporating virtual reality to train balance in patients following a stroke has been recently tested with good outcomes,[1] there have been no studies to date that have validated the use of telerehabilitation for patients with vestibular disorders.

It is currently estimated that 90% of the world's population now has access to a mobile network. This access creates opportunities that did not previously exist for improving patient care. In addition, the use of smart phones, which have sophisticated built-in sensors (e.g., accelerometers, gyroscopes, global positioning systems, and compasses), may be used to provide feedback and deliver and track precise rehabilitation programs. More recently, applications (apps, as they are commonly known) have been developed that can be used on smart phones and tablets to record gait speed, demonstrate the labyrinth and how the vestibulo-ocular reflex (VOR) works, and even to employ a metronome for use during assessment (e.g., dynamic visual acuity) and treatment. Additional apps for eye movements and demonstration of vestibular treatments are in the process of being developed. With the use of YouTube, one can show patients how to perform vestibular exercises and even the canalith repositioning maneuvers. As with anything, one must always ensure the accuracy of what is being taught or demonstrated. In the future, part of our job may be to identify for our patients the appropriate technology to optimize education concerning their specific condition.

The goal of this chapter is to highlight new technologies, focusing on virtual reality but also including augmented sensory biofeedback and relevant apps that are

currently being tested or newly available to clinicians. These apps may be a useful adjunct to vestibular rehabilitation for improving outcomes or for widening our reach to include patients with limited clinic access. This is a burgeoning area of research that is exponentially expanding as technology costs decrease and research capacity increases.

Key Concepts Related to Virtual Reality

One of the challenges facing therapists is identifying activities that are appealing, meaningful, and motivating so that patients will engage in these activities at a level that is of sufficient intensity and duration to produce long-term compensation. Some of our patients complain that vestibular exercises are boring. The use of virtual reality technology for rehabilitation has the potential to enhance rehabilitation outcomes by being engaging and interesting. Virtual reality is an immersive, interactive experience that occurs in real time and is created by a computer interface.[2] Specific assets of virtual reality related to rehabilitation include the opportunity for experiential learning in challenging but safe environments and the ability to objectively measure behavior while maintaining control over stimulus delivery and measurement.[2] Nazareth et al identified three factors that predicted chronic dizziness: a history of fainting (probably a symptom of panic), vertigo, and avoidance of situations that provoked dizziness.[3] By allowing patients to carefully control the level of exposure, thus symptoms, to situations that provoke their dizziness, virtual reality may encourage early training in provocative situations, which may result in improved outcomes.

Virtual reality systems specific to vestibular rehabilitation have been developed to achieve three primary goals: (1) reduction of symptoms (vertigo, dizziness, space and motion discomfort/visual vertigo), (2) adaptation of the VOR and optokinetic responses, and (3) retraining of postural stability. There is preliminary support for the use of virtual reality to meet these goals.[4-6] In addition, studies have reported that participants enjoy interacting with virtual environments,[7] and importantly, achieve a high number of repetitions,[2,8] which may lead to greater motivation to exercise more, especially if the virtual environment can be used in the home.

One of the approaches commonly used in vestibular rehabilitation, habituation, is very well suited to the virtual reality platform. Habituation is used to reduce symptoms by repeated, graded exposure of the patient to situations that elicit symptoms.[9] The therapist guides the patient through situations or positions that provoke the patient's symptoms and prescribes a home-based program that involves repetitions of these situations in a graded fashion to provoke moderate, but not severe, symptoms. Initially, patients may feel worse as they experience those situations that they may have previously avoided. Little is known about the mechanism of habituation. One hypothesis is that active movement presents a sensory mismatch to the brain that promotes compensation and adaptation for vestibular disorders.[10]

Visually complex environments, such as grocery stores, frequently induce symptoms in patients with vestibular deficits. The difficulty for the therapist is in creating gradations of the visual environment from relatively simple to complex. Virtual reality of the environment can be finely controlled, thus "dosed" to match the stage of recovery of the patient. As mastery is achieved, the complexity of the virtual environment is systematically increased. For example, with the virtual grocery store, shelves can be sparsely populated with products initially, and then as the patient progresses to complex visual environments, the virtual shelves can be more densely populated.[6]

There are numerous reports of using virtual reality, specifically using a game-based platform, for dynamic balance training to improve postural stability in individuals with neurological impairments or older adults.[11,12] It is only recently that an application of virtual reality has been developed specifically to incorporate vestibular adaptation exercises that involve head movement while focusing on a visual target.[13]

Although virtual reality systems have great potential, most of them are not commercially available, require extensive space or specially trained staff, and are prohibitively expensive; thus, virtual reality is inaccessible to the majority of clinicians involved in vestibular rehabilitation and their patients. Recently, the gaming industry has developed portable, low-cost interactive game systems, such as Nintendo Wii Fit® (Nintendo, Kyoto, Japan) and Microsoft Xbox 360 Kinect® systems (Microsoft, Redmond, WA), which monitor movement. The use of these off-the-shelf video games as an adjunct to rehabilitation has garnered much interest with physical therapists over the past few years. In addition, there is increasing evidence that older adults experience a high level of enjoyment from video game experiences. A recent study showed that 20% of seniors over the age of 65 reported playing video games and, in fact, they played more often than their younger counterparts.[14]

Virtual Reality–Based Vestibular Rehabilitation

There has been a considerable increase in research exploring the use of virtual reality in vestibular rehabilitation in the past decade[15,16] coinciding with a general acceptance that virtual reality has a place in rehabilitation.[17-19] Thus far, the research has been primarily laboratory-based because

of the expensive equipment and multiple disciplines (clinicians, computer scientists, engineers) required for this technically demanding process. To date, investigations of virtual reality have involved physiological responses, acceptability of virtual reality to patients, and efficacy of treatment, although efficacy research has been limited to small non-controlled, non-randomized studies.

Virtual reality has demonstrated profound effects on the visual and vestibular systems and can induce both wanted and unwanted oculomotor and postural effects in healthy subjects. For example, virtual reality can cause motion sickness[20-22] and transient oculomotor changes.[23] Increased postural sway[15,22,24] and gait deviations have also been observed with virtual reality.[21] Further research is needed to quantify these effects and identify how they may be applied therapeutically in patients with compromised vestibular systems before recommendations of virtual reality for mainstream clinical use.

Two main types of virtual reality systems have been investigated in patients with vestibular pathology: high-end systems consisting of head-mounted display and wide field of vision, and off-the-shelf systems including Nintendo Wii®, Microsoft Kinect®, and Hybrid systems (Table 28-1). Hybrid systems employ different combinations of components, usually from low-cost systems (such as the Nintendo Wii® remote controller or Microsoft Kinect ®) to create vestibular-specific exercises. These systems are differentiated by levels of immersion, technical specifications, front-end flexibility, availability, cost, and the evidence supporting their application to rehabilitation.

High-End Systems

High-end systems are highly immersive and have front-end flexibility (i.e., the therapist can control and adapt delivery of stimuli, such as the speed and direction of

■ Table 28-1 **SUMMARY OF CURRENT VESTIBULAR REHABILITATION–SPECIFIC VIRTUAL REALITY SYSTEMS**

High-End Systems	Off-the-Shelf Systems
Examples Head-Mounted Display systems • Balance Rehabilitation Unit (BRU) Wide Field of View • Computer Assisted Rehabilitation Environment (CAREN) • Balance Near Automatic Virtual Environment (BNAVE) • Virtual Environment and Postural Orientation (VEPO) Laboratory	Examples • Nintendo Wii • Microsoft Kinect • Hybrid systems (using elements of low-cost systems and specially developed software)
Advantages • Precise measurement of motion and postural stability parameters • Highly immersive • Front-end flexibility	Advantages • Low cost • Readily available • High usability • Easy to incorporate into clinical/home environment
Disadvantages • High cost • Not readily available • Not for home use • Technically demanding • Multiple disciplines required for use	Disadvantages • Not highly immersive • Minimal front-end flexibility • Measurement not as precise • Narrow field of view (no optokinetic stimuli/visual flow)
Levels of evidence • Quasi-experimental* (BNAVE, BRU) • Small RCT** (CAREN)	Levels of Evidence • Quasi-experimental (Wii, Kinect, Hybrid) • RCT (Wii)

*Quasi-experimental design—subjects are not randomized but measured pre and post intervention/exposure
**RCT—Randomized controlled trial

optokinetic stimulus). In addition, these systems allow for precise measurement of motion and postural stability. The disadvantages of these systems are the high cost, technical demands of the systems, and the multiple disciplines that are needed to operate the equipment and interpret data. There are two basic types of high-end systems—head-mounted display and wide field of view—that will be discussed below.

Head-Mounted Displays

Head-mounted displays (HMDs) are considered completely immersive. When wearing an HMD, the external world is no longer visible and is replaced by a virtual world projected onto two internal liquid crystal display screens (LCD) in the HMD (Fig. 28.1*A*, *B*, and *C*). Head position is tracked and the image on the screens is updated in real time (i.e., if the patient turns the head left, the virtual scene moves accordingly). HMDs can be heavy and typically result in a narrow field of view (30 to 60 degrees). The lack of peripheral field of view has been associated with "cybersickness" or "simulator sickness."[25] Simulator sickness can include symptoms of nausea, dizziness, faintness, headache, double vision, and/or fatigue and is thought to be a result of sensory conflict. The time lag between head movement and the visual display update is also known to cause simulator sickness.

Viirre[26,27] and Kramer et al[28] were the first to discuss the use of virtual reality with persons with vestibular disorders suggesting that virtual reality could be used to adapt visual scenes to a person's capabilities, thereby facilitating faster adaptation. Viirre and Sitarz used the time lag between image update and head movement therapeutically in patients with vestibular disorders.[5] As the user moves the head, thus the HMD, the virtual scene updates to correspond to the new point of view. The virtual scene motion was altered by slowing the visual scene to just slightly faster than the patient's VOR gain (i.e., the virtual scene moved proportionally slower than the head moved), and then as VOR gain improved, the virtual scene was gradually speeded up. Nine subjects performed the virtual reality exercises twice a day for 30 minutes for five days, and six controls completed the same exercises but without alteration of the scene speed. The exercise program entailed active search tasks, necessitating active head movements. There was a significant increase in VOR gain for the virtual reality group compared with controls immediately after the intervention, which partially remained after 1 week. In contrast, Di Girolamo et al found a transient *decrease* in VOR gain in healthy subjects playing a virtual game with HMD.[29] They postulated that as the computer was updating the visual scene and doing the work of the semicircular canals, the VOR gain was adaptively reduced. Thus, it appears that VOR gain can be adapted, at least transiently, using HMDs.

In a new application of HMD technology, Pothier and colleagues used immersive virtual reality glasses in

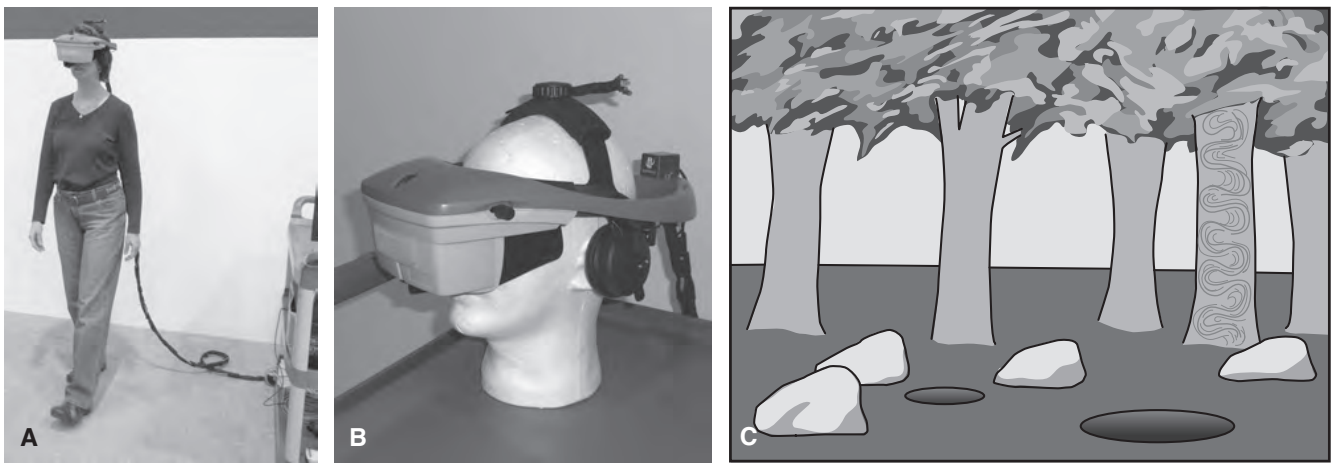

Figure 28.1 **(A)** Head-mounted displays block the person from viewing the external world and replace it with a virtual world that is projected onto internal display screens. Head position is tracked and the image on the screens is updated in real time (i.e., if the patient turns the head left, the virtual scene moves accordingly). **(B)** The head-mounted display is connected via cable to a computer system so that the virtual image is displayed inside the lenses and updated in real time. **(C)** In this application, the patient is navigating through a virtual forest to reach the target tree (with swirls on the trunk) while avoiding obstacles: rocks, holes, and other trees. (Forest Virtual Environment developed by Kathleen Turano, Johns Hopkins University, and Ronald Schuchard, Stanford University/VAPAHCS.)

combination with a digital video camera and customized image-stabilizing software to improve gaze stabilization.[30] With use of this image-stabilizing technology, visual acuity during head motion was immediately improved by more than three lines on the Snellen chart in subjects with bilateral vestibular loss (BVL). This is an interesting finding and may potentially have application to improve patient safety given that patients with BVL are at greater risk for falls and that vestibular rehabilitation involves several months of therapy.[31]

Mora et al[32] developed an HMD vestibular rehabilitation system. Rehabilitation using the system involved a passive phase (optokinetic stimuli were presented) and an active phase (point de mire exercises in a virtual room, i.e., adaptation exercises). They reported 73% improvement in postural stability and 93% improvement in subjective symptoms in an uncontrolled sample of 752 patients with mixed vestibular pathologies. Adverse effects were infrequent (3%) with only headache being reported. This system is not available commercially. An HMD-based computerized system including force plate (Balance Rehabilitation Unit, BRU) has recently been developed and was commercially available for a time, but since has been taken off the market (Fig. 28.2). In this application, virtual environments, such as rooms in a house, can be navigated with or without optokinetic stimuli. Gaze stability exercises can also be performed using this system. This system demonstrates front-end flexibility. One uncontrolled study demonstrated improved postural control in a group of 26 elderly fallers who underwent a 6-week daily program of customized balance rehabilitation using the system.[33] Its use in balance measurement has also been investigated.[34,35] These studies involved patients with Ménière's disease and benign paroxysmal positional vertigo (BPPV) and found the system to be sensitive in identifying postural control abnormalities. Use of these systems is rare in clinical practice, although some assessment systems employing video oculography are incorporating optokinetic stimuli in their systems (e.g., Virtual Environment Glasses at www.micromedical.com).

Figure 28.2 The Balance Rehabilitation Unit (BRU) consists of a computer with evaluation software, force platform, virtual reality goggles, and accelerometer. The head mount display provides virtual reality stimuli re-creating real life situations (e.g., navigating an apartment) or optokinetic stimulus.

Wide Field of View

Wide field of view (FOV) systems are generally high-end systems consisting of large screens arranged to form a room onto which virtual environments are rear-projected. The original wide FOV system (cave automatic virtual environment, CAVE) was created in the Electronic Visualization Laboratory at the University of Illinois, Chicago, and demonstrated publicly in 1992. The Balance Near Automatic Virtual Environment (BNAVE) was developed by the University of Pittsburgh and Georgia Tech and consists of three rear-projected screens on which images are synchronized and controlled by a network server and updated at 30 Hz.[4,36,37] The wide FOV spans the normal visual field (180 deg horizontal and 95 deg vertical) and therefore results in a high level of immersion.[36] One advantage wide FOV systems have over HMDs is in allowing peripheral vision and consequently manipulation of peripheral visual cues, which are important for postural control (Fig. 28.3). A second advantage is that users can see their body parts in relation to the virtual environment.

Wide FOV systems are the system of choice for investigators wishing to avoid the simulator sickness that is common with HMDs. Healthy subjects were asked to rate simulator sickness following performance of eight different gaze tasks in various combinations of the BNAVE virtual environments.[38] Subjects attended six sessions and rated intensity of nausea, oculomotor stress, and disorientation. Overall simulator sickness was minimal and the most commonly reported symptom was oculomotor stress; however, symptoms did tend to increase with more trials.

Figure 28.3 Healthy older subject (74 years old) demonstrates his reaction to motion of the virtual environment while in the Virtual Environment and Postural Orientation Laboratory at Temple University. (Photo used with permission of Max Buten.)

These high-end systems have a number of additional features that are beneficial for studying postural control. The BNAVE can measure and collect electromyography and head motion. A virtual grocery store is one of the environments developed for the BNAVE (Fig. 28.4). The length and width of the virtual aisles in the grocery store and their contents (common grocery items) can be manipulated to adjust complexity of the visual environment. Navigation through the store can be achieved using a joystick (subject is stationary) or pushing on a force-sensitive grocery cart while walking on a treadmill (subject is moving). Pushing the virtual grocery cart with the pressure sensor imbedded in the cart created greater immersion in subjects. Another system, CAREN (Computer Assisted Rehabilitation Environment; Motek technologies), has the capability of tracking three-dimensional movement of body segments and can incorporate a 6 degrees of freedom treadmill into the virtual reality environment, allowing precise measurements of the kinematics of movement during walking or running. Virtual environments can include optokinetic stimuli such as stripes or squares (Fig. 28.5), which can be regulated with respect to stimuli contrast, direction, speed, and spatial frequency.[36]

Figure 28.4 The Balance Near Automatic Virtual Environment (BNAVE) was developed in collaboration by the University of Pittsburgh and Georgia Tech. A virtual grocery store is one of the environments developed for the BNAVE. Navigation through the store can be achieved using a joystick (subject is stationary) or pushing on a force-sensitive grocery cart while walking on a treadmill (subject is moving).

Initial trials of the BNAVE system were focused on the responses of healthy subjects and small numbers of patients with vestibular disorders to establish safety. The research demonstrated that use of the wide FOV system resulted in the expected outcomes, which are closely controlled by the system; that is, postural sway increased when the person standing in the system was exposed to optokinetic stimuli.[39] Gottshall et al recently reported use of wide FOV in subjects with mild traumatic brain injury undergoing vestibular rehabilitation.[40] Their findings were promising, although not superior to conventional rehabilitation. Systems such as these are costly and at present are confined to laboratory research facilities. At this point, it is likely that these systems will not be widely available to therapists because of costs and will remain experimental.

Off-the-Shelf Systems

The computer gaming industry has inadvertently made it possible for the advanced technologies present in some of the high-end virtual reality devices to be made available

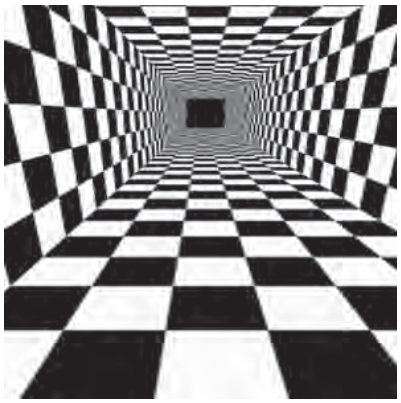

Figure 28.5 Optokinetic displays can be used for rehabilitation with a wide field of view with the use of virtual reality. This scene is similar to the checkerboard pattern commonly used for exercise in persons with vestibular disorders. (Balance Near Automatic Virtual Environment, University of Pittsburgh).

to clinicians at a low cost. These gaming systems, discussed below, have the capacity to track three-dimensional movement and/or center of pressure (COP). Although considered to be non-immersive (they were developed to connect to a television or computer monitor), they can provide auditory and visual feedback in real-time and generate sophisticated virtual environments. Furthermore, these commercially available gaming systems have a wide repertoire of games and exercises. The potential of gaming systems has been realized by the rehabilitation community, and many groups are investigating using these systems off-the-shelf (i.e., as-is) as well as adapting them to meet the specific needs of rehabilitation patients (hybrid systems). One major disadvantage of these systems is that they have a smaller field of view and thus lack the ability to generate visual flow and optokinetic stimuli that are important in postural control and in rehabilitation.[15,41,42] Another major disadvantage for the rehabilitation community is that the gaming systems were designed for healthy populations and lack front-end flexibility; thus, many of the games, as developed, are not appropriate for clinical use.

Nintendo Wii®

Nintendo released the Wii in 2006, and it has become the best-selling gaming console of all time. The Wii consists of a computer interface; a controller (Wiimote), which houses an accelerometer; gyroscope and infrared camera; and a sensor that detects motion in three planes. In 2007, Nintendo released the Wii Fit Plus system, which incorporated a balance board. The balance board is a force plate incorporating four force transducers that compute center of pressure displacement in the mediolateral and anteroposterior directions. Wii Fit Plus has many exercise and balance games that purport to challenge balance and provide visual and auditory feedback of the COP. The accuracy of the Wii balance board (WBB) to detect center of pressure is comparable to a laboratory force plate.[43,44]

A recent study examined perceived usability of the Wii system in patients with balance impairment.[7] In this study, 26 subjects with impaired balance caused by neurological disease underwent a single, 30-minute session on the Wii using selected exercises and balance games (see Case Study 28-1) and afterwards scored the system on usability. Scores indicated high usability, but there was a tendency for older patients to rate the system less usable. Nearly 90% of patients indicated a preference to use the Wii in future therapy, and 70% reported that the Wii was more enjoyable and motivating than conventional balance retraining. Factors such as feedback and motivation were frequently cited as perceived benefits of Wii. Mild adverse events (nausea, headache, and increased dizziness) were reported in a minority of patients. The majority of these patients needed a nearby chair for support when using the Wii. It has also been reported that older adults across a wide range of ages (62 to 90 years old) can safely mount the balance board and quickly learn Wii Fit games.[45]

One key component of balance training is learning to control the center of gravity during a variety of movements. The WBB detects vertical forces applied to the board and then calculates COP, a surrogate measure for center of gravity. The WBB is sensitive enough to detect even small changes in COP that occur during eye movement tasks while standing.[46] By moving and shifting COP, the player controls an avatar in the game environment, which is then displayed in real time on the monitor. For example, the ski slalom game involves the player shifting his or her weight side-to-side to move the avatar laterally or anterior-posterior to control speed down the mountain with the goal of skiing down a virtual ski slope and passing through as many gates as possible.

Through the use of balance games that involve control of COP, the Wii Fit may be a useful adjunct to therapy (Fig. 28.6*A* and *B*). However, Duclos and colleagues found that performing the Wii 50/50 balance challenge using the football game and ski slalom game did not challenge balance (as determined by the distance between COP and the limits of the base of support where a smaller distance is less stable) as much as walking at a fast pace in a small sample of healthy elderly subjects.[47] The static nature of the balance games (feet in place) may not be optimal for retraining dynamic balance. Another important component of balance rehabilitation is multisensory training to improve the ability to integrate sensory input for balance. The incorporation of unstable surfaces and reduced or absent vision (eyes closed conditions) is thought to be a critical aspect in balance training exercises.[48] The limitation of the firm, non-movable surface of the Wii balance board can be overcome by incorporation of a rocker bottom device (Frii Board; Fig. 28.6*C*) that transforms the WBB into a multidirectional unstable surface (Swiit Game Gear) or the Wii Real Balance board (CTA Digital; Fig. 28.6*D*). The games and balance exercises on the Nintendo Wii do not factor in the requirement for performing exercises with eyes closed, although auditory feedback of the COP is built in and can provide feedback regarding postural sway. Some of the exercises/games provide a visual image of COP excursion, which provides knowledge of results feedback. As yet, no studies have investigated these add-on boards or auditory feedback for their impact on rehabilitation.

One randomized controlled trial has demonstrated positive outcomes using a non-modified WBB for balance rehabilitation after acute vestibular neuritis compared with a no-exercise control group.[49] The experimental group performed a variety of exercises including yoga, strengthening,

Figure 28.6 **(A)** Patient with bilateral vestibular loss exercises on the Nintendo Wii Fit Plus® playing the Table Tilt game. The patient stands on the Wii Balance Board (WBB) and, by moving her center of pressure, rolls virtual balls into virtual holes on a virtual tilt table. **(B)** Center of pressure is displayed providing visual feedback to the user. **(C)** A progression of the exercise in which the Frii Board is placed under the WBB to transform it into an unstable (rocker) board. **(D)** The Wii Real Balance Board is another method to transform the WBB into an unstable surface.

and balance games for 10 training sessions, none of which involved specific head movement or gaze stability activities. Sensory organization test and Dizziness Handicap Inventory scores for the experimental group (n = 34) were significantly improved compared with the control group (n = 33). Another randomized controlled trial using the unmodified Wii in vestibular rehabilitation is under way.[50]

The Nintendo Wii is currently being used in clinics for a variety of rehabilitation applications. The motivational and enjoyment aspects of Wii-based programs may result in better adherence to exercise resulting in a higher intensity than conventional rehabilitation. These aspects, plus the low cost of Nintendo Wii and ease of use, make it likely that Wii will have a growing place in vestibular rehabilitation, although there is limited evidence at present to support its effectiveness.[51]

Microsoft Kinect®

Microsoft Kinect® for Sony Xbox was released in 2010 as a computer gaming system. It uses an embedded webcam and infrared system to detect motion, so no handheld controller is required. The accuracy of the Microsoft Kinect

system to detect body motion is comparable to a laboratory motion analysis system.[52] Games that can be played on the system include sports, dance, motorsports, adventures, and workouts. There are only a few reports of its application to rehabilitation in the literature to date. A hybrid system for vestibular rehabilitation has recently been developed in which the Kinect was coupled with the Nintendo Wii Balance Board and a 3D-ready projector with shutter glasses for stereo-vision.[53] Five exercise tasks based on modified Cawthorne–Cooksey exercises were developed. In this pilot study, five patients with vestibular dysfunction used the system for a 5-week, 20-session program. After the intervention, the patients demonstrated decreased postural sway and improved balance function.[53] It is likely that further investigation of the Kinect will emerge over the coming years.

Hybrid Systems

Several research groups have customized the WBB to develop games specific to balance retraining and piloted these games using healthy older adults,[54] patients with neurological disorders,[12] and patients with vestibular disorders.[13,55,56] Che et al coupled the Wiimote signal to a head-mounted infrared device and developed software to project interactive virtual games and environments (including 3D streetscapes, a swinging basket game, and a baseball pitcher) onto three monitors to create a wide FOV.[13] The customized games incorporated head movement and gaze stability similar to conventional gaze stabilization exercises. The virtual reality system has the added benefit of measuring head velocity and incrementally increasing the head velocity required to earn points. Gascuel et al coupled the WBB to a large rear-projected screen on which optokinetic stimuli were projected with increasing speed of flow.[56] Sixteen patients with vestibular loss received eight sessions using the system with resultant decrease in postural sway over time, suggesting improved postural control.

Uptake and Integration of Virtual Reality with Vestibular Rehabilitation

Before recommending virtual reality for widespread use in vestibular rehabilitation, much more research and development is required. As efficacy becomes clearer, therapists will need to be able to assess which system will best benefit patients with vestibular problems. Factors that will contribute to a decision to choose a particular system will include cost, evidence, and usability (Box 28-1).

Box 28-1

FACTORS INVOLVED IN CHOOSING A VIRTUAL REALITY SYSTEM

- Safety
- Efficacy (Superiority, non-inferiority)
- Ethics
- Cost effectiveness
- Usability
  - Therapist
  - Patient
  - Caregiver

Other

- Front-end flexibility
- Space requirements
- Billing issues

Virtual Reality–Based Assessment

Examination of low-cost, interactive game technology as a primary means to assess balance is still evolving. The Wii Fit system consists of the computer interface, monitor, and balance board (WBB), a small force platform on which the player stands during play. Clark and colleagues tested an unmodified WBB with customized software and determined that the WBB has validity and very good test-retest reliability for collecting force data.[43]

Two recent studies examined use of Wii Fit to assess balance and mobility with mixed findings. Hall and colleagues demonstrated that the Wii scores (time and penalties) from the ski slalom game were strongly correlated with functional measures, including gait speed, timed up and go, and dynamic gait index, all of which have been linked to fall risk.[45] In addition, some of the Wii scores were strongly associated with computerized dynamic posturography, specifically sensory organization test composite score and several measures from the limits of stability test. In contrast, Yamada and colleagues did not find the ski slalom game to have utility in identifying fall risk.[57] An important difference in the methodology between the two studies is that in the Yamada study the games were modified to be performed in sitting; in the Hall study both games were performed in standing. Yamada and colleagues modified Wii Fit to increase perceived safety; however, performing weight shifting while sitting may not adequately assess balance required for standing.

Another system has recently been assessed and determined to have good validity and reliability. The Microsoft Kinect system was found to have concurrent validity compared with a gold standard 3D motion analysis system and good reliability for assessing the kinematics of movement during balance testing.[55] Excellent concurrent validity was demonstrated for three postural control tests (forward

reach, lateral reach, and single-leg stance with eyes closed), but there was a tendency for error to be present in larger magnitude movements.

Use of a simple, inexpensive gaming system may provide an alternative method for testing balance in community-dwelling older adults when other resources are not available. The components of the Wii Fit balance board and Kinect sensors are valid and reliable for collecting force and motion data when compared with laboratory force plates and motion analysis systems and thus may have utility in measuring postural control. In addition, off-the-shelf games appear to capture similar information as standard physical performance measures of balance and gait and the gold standard computerized dynamic posturography. Additionally, there is now a commercially available low-cost alternative to computerized dynamic posturography that incorporates the WBB as the force platform component and is paired with customized software to perform assessment of limits of stability and modified clinical test of sensory interaction on balance (CSMI Solutions). Benefits of gaming systems include availability, minimal cost, nonclinical appearance, and potential for assessment of fall risk in community-dwelling older adults.

Technology for Augmented Sensory Biofeedback

A typical sensory substitution system for balance comprises three parts: an inertial measurement unit to measure body motion, a processor to analyze this motion, and a feedback display (e.g., vibrotactile, electrotactile, visual, auditory) to present information regarding body motion to the user. Sensory substitution devices have been shown to decrease trunk sway during real-time operation in a laboratory setting, and preliminary results suggest that persistent improvements in balance performance exist over time periods of minutes to hours following a small number of training sessions. These innovations are primarily designed to help individuals who do not respond well to standard physical therapy interventions, such as individuals with BVL and central vestibular dysfunction. Because the prevalence of falling is so high in persons with BVL, any device that could "signal" to persons that they are putting themselves in a high risk situation might be beneficial.

The hypothesized mechanism for the improvement is sensory reweighting. Feedback of body motion provides the brain with a correlate to the inputs from intact channels of sensory input, so subjects receiving augmented sensory biofeedback learn to increasingly depend on these intact sensory systems (e.g., vision, proprioception). Various types of augmented sensory biofeedback will be discussed including vestibular electrical stimulation, electrotactile feedback, auditory feedback, and vibrotactile feedback.

Vestibular Electrical Stimulation

Recent work suggests that "vestibular electrical stimulation" (VES) was as effective as oculomotor rehabilitation in persons with acute peripheral vestibular disorders.[58] The VES consisted of electrical stimulation that was applied to the paravertebral nuchal muscles, which is purported to stimulate the affected vestibular nuclei through the crossed vestibulospinal pathway. Participants in the electrical stimulation group (n = 14) walked while VES was applied for 40 minutes over a 10-day period. The oculomotor group (n = 14) performed saccadic eye movements to a target, smooth pursuit eye movements, reading a moving paper, and optokinetic stimulation (moving stripes of different colors at a velocity of 30 deg/sec) while standing for the same amount of time as the electrical stimulation group. Barozzi and colleagues found that both groups improved on measures of static posturography and dizziness handicap inventory scores with no differences between the two types of interventions.[58] VES might be an option in the future, but the technology is not readily available for use in the clinic at this time.

Tongue Electrotactile Stimulation

The first sensory substitution system was developed by Bach-y-Rita for persons who were congenitally blind.[59] The tongue electrotactile system translated a visual image from a camera worn by the person into a tactile image projected onto the tongue via an electrode array. The tongue electrotactile device has more recently been described as a type of brain-computer interface technology and was designed to provide direct, noninvasive input to the brain to substitute for vestibular information.[60] The electrotactile device transmits information to the brain about angular head movements via the tongue, thus augmenting sensory information available for balance.

The intraoral device consists of accelerometers to detect angular (pitch and roll) movements of the head and an array of electrodes to stimulate the tongue (Fig. 28.7). Information about the head is conveyed to the tongue via the electrodes corresponding to the head motion.[60] For example, if the head tilts to the right, the electrodes on the right side of the tongue are stimulated. When the head is level, the electrodes in the center of the tongue are stimulated. Vuillerme et al modified the device to provide information from the foot to the tongue to augment propriocepti ve inputs.[61] Use of the tongue electrotactile feedback from the plantar surface of the feet decreased postural sway in standing with the head extended with eyes closed in young

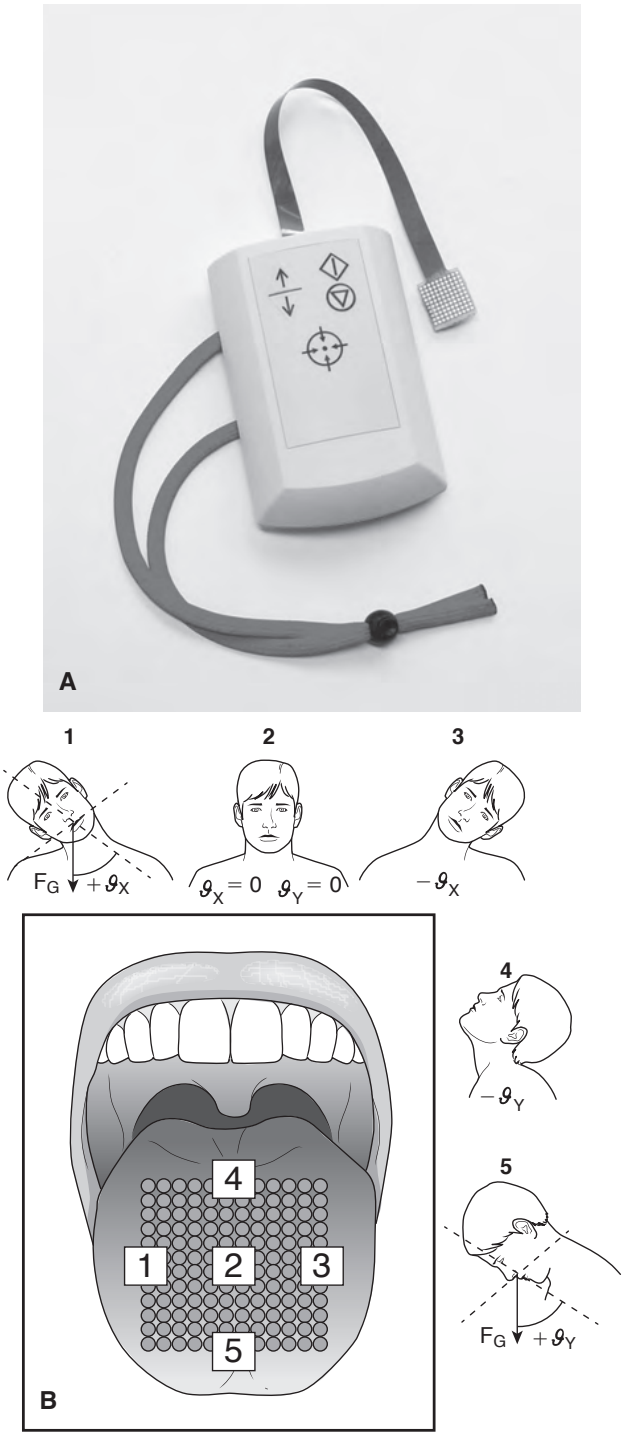

Figure 28.7 **(A)** Tongue electrotactile device and intraoral device; **(B)** stimulus position and movement as a function of head angular motion. (From Danilov et al, 2007. Used with permission.)[62]

healthy people, suggesting that the device might be helpful for persons with neck or vestibular pathologies.[61] Proposed uses for the tongue electrotactile feedback device include the prevention of pressure sores,[61] fall risk reduction in older persons,[61] and balance improvement in persons with vestibular disorders[63] and central imbalance from stroke or traumatic brain injury.[64]

All the studies to date that have used the tongue electrotactile device have had small sample sizes, and none have included a control group. Barros et al trained seven persons with bilateral vestibular hypofunction with the device over 12 sessions.[63] Their findings suggested that balance improved, as measured by the Sensory Organization Test. The only commercial manufacturer (Wicab) of a tongue electrotactile device (BrainPort® balance device) is no longer marketing or manufacturing the device for use in the United States. Furthermore, the company has no plans to pursue FDA approval in the United States for the tongue electrotactile device for the enhancement of balance.[65]

Auditory Biofeedback

Auditory biofeedback to enhance postural control is an inexpensive technology[66-68] that uses accelerometers and has been demonstrated to improve balance in healthy control subjects[66] and in persons with bilateral vestibular hypofunction.[67] Subjects stood with eyes closed on a foam pad on a force platform while wearing headphones. The pitch and volume of the auditory signal increased when subjects moved away from vertical (outside the ±1-deg sway threshold) in any direction. Subjects with bilateral vestibular hypofunction demonstrated less trunk accelerations, a smaller sway area, and greater time spent within the ±1-deg sway threshold after training with the device (Fig. 28.8). Hegeman et al reported that audio biofeedback did not improve sway on a foam pad with eyes closed in people with bilateral vestibular hypofunction but did report that single leg stance time improved with audio feedback.[68] This approach has also been successfully applied to individuals with isolated otolith organ dysfunction.[69,70]

Vibrotactile Biofeedback

Vibrotactile feedback has advantages over lingual, auditory, and visual feedback in that it does not directly compete with tasks that involve speaking, hearing, or seeing. Vibrotactile feedback is similar to the vibration that you would feel from a cell phone in your pocket and is usually provided around the waist area. When vibrotactile feedback was first developed, the vibrotactile devices were quite cumbersome and not user friendly. These research devices used as many as 64 tactors imbedded in a vest worn by subjects.[71,72] Testing of alternative array designs to determine optimal design and signal have resulted in more streamlined products with fewer tactors.

One can think of vibrotactile feedback as an early motion detector system or a "body alarm" that provides the

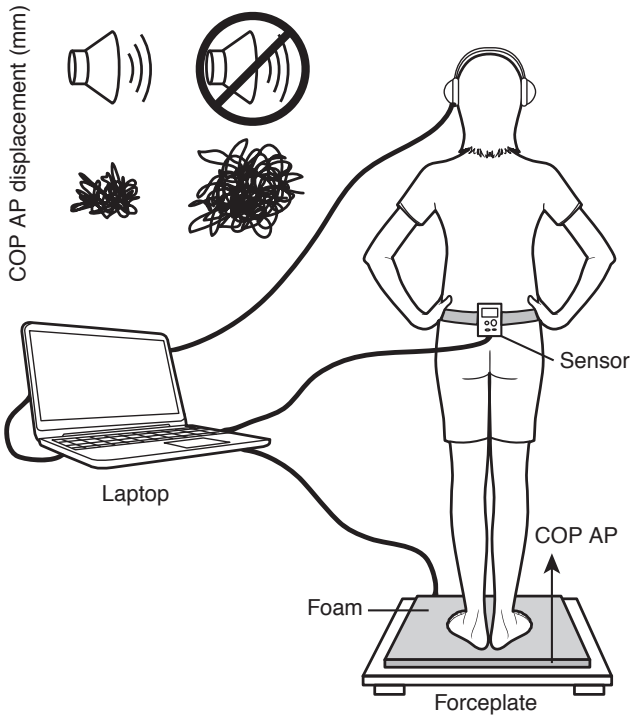

Figure 28.8 A patient with bilateral vestibular hypofunction had less sway while processing the auditory cue than with no auditory cueing while standing on a foam pad. (Modified from Dozza M, Chiari L, Horak FB. Audio-biofeedback improves balance in patients with bilateral vestibular loss. *Arch Phys Med Rehabil*. 2005;86: 1401-1403.)

brain with additional input about body in space and enhanced feedback about a person's limits of stability. The amount of sway that is "tolerated" by the system can be preset for each patient based on his or her abilities and the goal of therapy. To date, most of the studies conducted with the vibrotactile feedback devices have tested subjects in static stance.[71-74] There has been concern that the devices make people stand stiffly; thus, for the devices to have clinical utility they must also be effective during walking or have carryover from standing to walking. There is some evidence to support the usefulness of vibrotactile feedback during gait.[75]

There are at least two commercially available devices developed in Europe for use with patients (VertiGuard® and SwayStar™). Randomized controlled trials suggest that postural control is enhanced while wearing the Vertiguard® device, which is worn around the waist and registers angular deviations and angular velocities of the trunk.[76,77] It is not clear whether future vibrotactile devices will be used at home or only in the clinic to retrain postural control. If they can assist as an early detection system for fall risk, they have great value, especially for the frail elderly, who are at the greatest risk of fall-related injury.

Other Technologies

Platform Perturbations

Typical rehabilitation of patients with vestibular disorders involves balance training with altered visual and/or somatosensory information to improve postural stability in a variety of environments. This training is typically performed on foam cushions and rocker boards; however, there is emerging evidence that the use of computer-driven, motorized force platforms is effective in improving postural stability and in reducing symptoms.[78-80] The rationale is that the subject is not able to rely on somatosensory input, because the platform is moving, and so is forced to rely on vestibular input to maintain balance. This may force reweighting of sensory input so that remaining vestibular function is used to determine position in space and respond to loss of balance.

One paradigm uses a continuously moving platform, translating in the anterior-posterior or medial-lateral direction, on which the subject attempts to maintain balance.[78,79] These studies demonstrated that the platform training was as effective as performance of Cawthorne–Cooksey exercises in terms of postural stability with eyes open or closed on a firm surface and in terms of subjective symptoms of steadiness and dizziness handicap inventory scores. Another perturbation paradigm involves tilt perturbations of the platform.[80] The perturbations involve discrete angular movements of the platform at a range of directions, speeds, and amount of tilt. Winkler and Esses used random tilt perturbations of increasing speed and tilt angle in a small randomized controlled trial.[80] In this study, subjects with chronic vestibular dysfunction were assigned to one of three groups: (1) perturbation only, (2) perturbation + home program of vestibular rehabilitation (including gaze and postural stability exercises), and (3) vestibular rehabilitation only. The perturbation and perturbation + vestibular rehabilitation groups made the greatest improvements in subjective symptoms and perceived function as well as fall risk reduction measured by the dynamic gait index. There has been no direct comparison of the different types of perturbation training, so it is not clear what the optimal training parameters are. Further research is needed to clarify the role of perturbation training in vestibular rehabilitation. Equipment is commercially available that can be used for this type of training: Equitest (NeuroCom®, a division of Natus®) or Proprio® Reactive Balance Systems (Perry Dynamics).

Smart Phones and Tablet Applications

The increasing penetration of smartphones, tablets, and mobile computing in daily life will likely revolutionize health care in the future. These systems are capable of both

storing information and measuring physiological signals in ways that were neither accessible nor portable in the past. They provide opportunities for both patients and professionals to access information readily, and thus have potential to improve the diagnosis and management of vestibular disorders. They can capture, store, and facilitate sharing of multiple types of patient data (e.g., patient databases and reported outcomes, videos, pictures, physiological signals). Most smartphones and tablets have sensors such as accelerometers, GPS, and gyroscopes that could have numerous uses in vestibular rehabilitation. Thus far only a small number of apps specific to vestibular rehabilitation have been developed. There is no governance concerning apps; thus, health professionals should assess each one for merit and applicability before employing them with patients.

At present, apps related to vestibular rehabilitation can be categorized as either measurement or educational. In the measurement category, an iPhone application, "Visual Vertical," is commercially available to measure subjective visual vertical (www.clearhealthmedia.com) based on the bucket test.[81] The iPhone is secured to the inside of the bottom of a bucket. The phone presents a red line against a black background that the subject has to align to vertical and then degrees off-vertical are provided. Two other iPhone apps, "Eye Chart Pro" and "OptOK" provide visual acuity charts that can be used to measure dynamic visual acuity. "OptOK" also provides visual displays of optokinetic stimuli (i.e., moving stripes). There are fewer apps available on Android systems, but developers have devised a balance assessment system using the accelerometer in a smart phone. The phone is secured to a body segment (such as the shank) and the amplitude of movement is recorded over a period of time. These balance assessments include sitting balance, standing on one leg, and standing on an unstable surface. However, no details on the validity or reliability of this app are available.

The University of Sydney, in partnership with Liberty Technology and the Neurology Department at the Prince Alfred Hospital Sydney, have recently developed a free app for medical education, "aVOR" (angular VOR), that can be downloaded from iTunes. It presents a virtual patient showing the semicircular canals in the head in three dimensions. It is interactive in three dimensions allowing translation, rotation, zooming, and pivoting of the virtual head by moving the phone or using the touch screen. The push-pull responses of the canals are demonstrated in response to head movement. The resultant eye responses to moving the head in the presence of various combinations of semicircular canal and/or cerebellar dysfunction are shown, including BPPV and canal repositioning maneuvers. A quiz is available to test knowledge. This application is highly useful for any professional wishing to learn more about the vestibular system.

CASE STUDY 28-1

Nintendo Wii Fit Plus as Adjunct to Vestibular Rehabilitation

This case study reports the vestibular rehabilitation of a 58-year-old male who was referred for physical therapy. He had a diagnosis of unilateral Ménière's disease, which had been diagnosed 33 years ago. Although attacks of vertigo were now intermittent, he complained of constant daily symptoms and rated vertigo as 6/10 and dizziness as 9/10 on visual analog scales. Other subjective symptoms included oscillopsia and constant nausea. He reported that he had disequilibrium particularly when walking and felt like "people thought [he] was drunk." Subjective and objective measures are summarized in Table 28-2. He had abnormal scores in conditions 5 and 6 of the sensory organization test (SOT) on the Equitest, although his composite score was within normal limits (72; normal for age ≥ 70.2). His Activities-Specific Balance Confidence score was 74% and his DGI was 20/24. Gait speed was 1.28 m/sec (normal greater than 0.94 m/sec). His Hospital Anxiety and Depression scale was abnormal on the anxiety subscale (10/21). Dynamic visual acuity was reduced by 2 lines (normal). He was unable to work because of his symptoms and reported that it took him 4 hours to "get going" in the morning because of nausea. His quality of life was severely reduced with these symptoms and his goals for rehabilitation were to be able to wake up in the morning and head straight out the door, and get back to golf.

He underwent a 6-week program of vestibular rehabilitation. On the second treatment, he was provided with a Nintendo Wii on loan for 6 weeks and instructed in its use and safety precautions (Table 28-3). The program on the Wii was detailed in a series of six booklets that he was given to take home at each session. The program was progressively challenging and designed to last 15 minutes per day five days per week. For weeks 4 to 6, he was provided with a Frii Board to create an unstable surface under the WBB. In addition, he

Continued

CASE STUDY 28-1

■ Table 28-2 OUTCOME MEASURES FOR CASE STUDY 28-1

Measure	Before Treatment	After Treatment	Result
Gait Speed (m/s)	1.28	1.43	Significant improvement
Dynamic Gait Index (/24)	20	24	Significant improvement
Vestibular Rehabilitation Benefits Questionnaire			
Symptom Score (/100)	39.5	13.7	Improved
Quality of Life Score (/100)	36.5	0	Improved
Total Score (/100)	38	6.8	Improved
Hospital Anxiety and Depression Score (lower score indicates less anxiety/depression)			
Anxiety (/21)	10	5	Improved
Depression (/21)	6	1	Improved
Total (/42)	16	6	Improved
Activities-Specific Balance Confidence (%)	74	74	No change
SOT Composite Score (/100)	72	85	Improved

■ Table 28-3 EXAMPLE NINTENDO WII EXERCISE™ PROGRAM PROGRESSION PROVIDED TO CASE STUDY 28-1

Wii Exercise	Modification*
Yoga deep breathing	Add in: Body weight shifts, "draw" a circle around displayed limits (yellow circle) Eyes closed Throw a ball in the air Head movements in yaw and pitch planes Incorporate Frii board with eyes open/closed
Standing Knee	Try for single-leg stance at first
Single Leg Extension	No arm movements at first
Sideways Leg Lift	No arm movements at first
Soccer Heading	None

CASE STUDY 28-1

■ Table 28-3 **EXAMPLE NINTENDO WII EXERCISE™ PROGRAM PROGRESSION PROVIDED TO CASE STUDY 28-1—cont'd**

Wii Exercise	Modification*
Table Tilt	Progress to table tilt plus—incorporate Frii Board
Penguin Slide	Progress to incorporate Frii Board
Free Stepping	Advance to step basics, then step plus and incorporate head movements in the yaw and pitch planes.
Snowball Fight	None
Yoga Tree	Raised foot placed lightly on other foot if full pose not possible.
Skateboarding	Progress to incorporate Frii Board
Balance Plus	Progress to incorporate Frii Board
Balance Bubble	Progress to incorporate Frii Board
Snowball Fight	None

*Instructions were given to have a high-backed chair within reach when exercising to provide haptic support.

was given gaze stabilization exercises and a progressive walking program.

Post-treatment he felt much improved, rating both vertigo and dizziness at 2/10. Dynamic visual acuity had not changed. His gait speed had increased to 1.48 m/sec and his DGI was 24/24 (see Table 28-2). At a 6-month follow-up, he reported one further attack of vertigo that had resolved over a 2-week period, and he was out walking dogs and felt able to return to golf. His gait speed at this session was 1.35 m/sec, and his SOT was 80 indicating he had maintained his improvements.

Summary

The use of virtual reality and other complementary technologies in vestibular rehabilitation is an exponentially evolving field. Although at present this field is at a highly experimental stage, findings to date have been encouraging. Future research will likely elucidate the role and efficacy of technology in vestibular rehabilitation. It is likely that in the not too distant future, low cost, portable, vestibular rehabilitation–specific systems will be widely available.

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SIX

Non-vestibular Dizziness

CHAPTER 29

Non-vestibular Dizziness and Imbalance

Ronald J. Tusa, MD, PhD

Non-vestibular dizziness and imbalance can be very frustrating to the clinician, because the symptoms are often vague and the vestibular test results are normal. This chapter discusses the more common causes of these disorders. Disuse disequilibrium with fear of fall, the most common cause of imbalance, readily responds to gait and balance therapy. Other disorders that cause imbalance are leukoaraiosis, normal-pressure hydrocephalus (NPH), progressive supranuclear palsy, Parkinson's disease, large fiber peripheral neuropathy, and cerebellar ataxia. Many of these disorders are associated with disuse disequilibrium (or deconditioning) and, therefore, respond to physical therapy (PT) to a certain extent. Some individuals with these disorders do not have disuse disequilibrium, because they are very active or are too incapacitated to perform PT. In these individuals, one must concentrate on reducing fall risk through the use of assistive devices and education. Finally, we discuss a group of disorders that are best described as dizziness in the head without severe imbalance that are triggered primarily by specific situations.

It is best to have any patient with chronic dizziness first assessed by a physician to determine the diagnosis. After this assessment, PT can be started the same day or as soon as possible. During PT, there is an initial assessment to identify the patient's specific problems. Then the patient is started on a daily home gait and balance program that is reviewed and revised by the physical therapist every week. At each visit, outcomes scores are reassessed. The patient is then reevaluated by the physical therapist, and the outcomes scores are reviewed to determine whether the patient needs more rehabilitation and/or a follow-up appointment in 6 to 12 months. The latter is especially important in patients who have progressive problems (spinocerebellar degeneration, progressive peripheral neuropathy, and progressive supranuclear palsy).

Disuse Disequilibrium (or Deconditioning) and Fear of Fall

Description

Each year, approximately one-third of elderly individuals in the general population experience a fall.[1] In elderly individuals, there is progressive decline in muscle bulk, joint range of motion, and reflex time.[2] Increased exercise can reduce the rate of this decline. Many individuals stop walking and exercising because of recent surgery, fatigue, chronic illness, or a fall to the ground or a near fall. Lack of exercise in the elderly leads to disuse disequilibrium.[3] Fear of fall can occur as a result of disuse disequilibrium and can also exacerbate disuse disequilibrium by reducing the patient's willingness to participate in a home exercise program.[4]

Figure 29.1 shows the age distribution of fear of fall and disuse disequilibrium in patients at our Dizziness and Balance Clinic. The diamonds indicate all patients seen in the clinic (left axis) based on decade of age of the

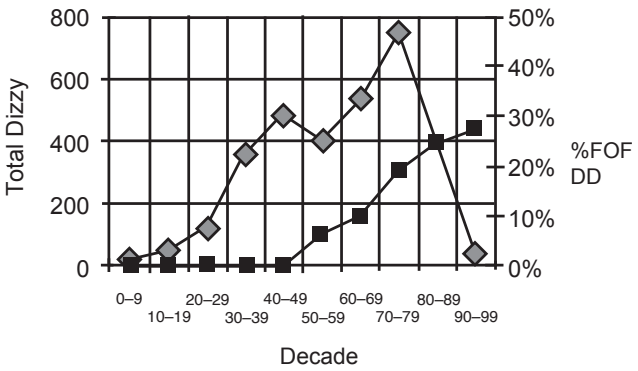

Figure 29.1 Age distribution of disuse disequilibrium and fear of fall in patients of a dizziness and balance clinic. See text for explanation.

patient. The squares indicate the percentage of patients seen within each decade with fear of fall (FOF) and disuse disequilibrium (DD) (right axis).

Useful Outcome Tests

An evidence-based review of fall risk assessment tools from 193 relevant citations was published in 2008.[5] Their conclusions were: (1) a history of recent falls was a predictor of future falls (Level A; 5 Class I and 10 Class II studies); (2) abnormal Get-Up-and-Go, timed Up-and-Go, and Tinetti Mobility Scale were the best screens for falls (Level B; Class II and III studies). See Box 15-4 for description of evidence-based guidelines.

The outcome measurements we use at Emory for patients with imbalance are:

- Fall risk assessments: Tinetti Fall Risk Assessment, Dynamic Gait Index (DGI)
- Timed Gait Tests: 8-ft "Get Up & Go" test, gait speed, Modified Timed Up and Go
- Standing balance tests: computerized dynamic posturography or similar force platform test to quantify standing balance sway
- Subjective tests of gait and balance: Activities-specific Balance Confidence (ABC)
- Scale for confidence in balance, Disequilibrium Visual Analogue Scale (VAS)
- Others: Activities of Daily Living (ADLs)

A full description of these outcome measurements can be found in Chapter 21, Physical Therapy Assessment of Vestibular Hypofunction. A description of posturography SOT can be found in the Chapter 11, Vestibular Function Tests.

Management

There are no large, controlled studies of the management of disuse disequilibrium, but in our experience, imbalance in individuals with disuse disequilibrium and fear of fall readily responds to gait and balance PT. In a recent Cochrane review for fall prevention,[6] there was limited evidence to support any one exercise intervention. This review included 60 randomized controlled trials involving 60,345 participants, half of whom were in care facilities and half in the hospital. Despite this, much smaller studies suggest that a daily home exercise program that increases endurance, balance, and lower extremity strength frequently resolves the problem.[7] Success depends on compliance by the patient and support from the family or friends. This has been our experience as well. Case Study 29-1 describes a patient with disuse disequilibrium and fear of fall.

Leukoaraiosis and Normal-Pressure Hydrocephalus

Description

Leukoaraiosis and NPH are discussed together, because they both manifest with symptoms of cognitive impairment, imbalance, and urinary dysfunction.[8-10] In severe cases, patients with these disorders have gait apraxia, gait initiation defects, and severe retropulsion.

The LADIS (leukoaraiosis And DISability) study group based in Europe has recently published their results from the past decade.[10] Based on those studies, the etiology of leukoaraiosis (ischemic white matter disease) is believed to be significant small vessel disease. Age, hypertension, and lacunar strokes are the major determinants of this entity, and smoking and hypercholesterolemia pose additional risks. Extensive white matter high signal intensity is found in the corona radiata and sometimes in the pons on T2-weighted magnetic resonance imaging (MRI) of the head. They found that in patients with severe white matter changes, the risk of deterioration to a dependent status is doubled after 3 years.

In NPH, there is a triad of symptoms consisting of dementia, imbalance, and urinary incontinence, with communicating hydrocephalus found on computed tomography (CT) or MRI of the head.[9] Many patients with NPH also have parkinsonian features, including masked facies and cogwheel rigidity, but usually no tremor. Cerebrospinal fluid (CSF) pressure measured by lumbar puncture or lumbar drain during the daytime is normal, but at night there are marked CSF pressure elevations that usually occur during periods of sleep apnea.

Useful Outcome Scores

Patients with imbalance and cognitive decline from leukoaraiosis or NPH should undergo gait and balance outcome measures described in the Disuse Disequilibrium section of this chapter. In addition, they should have tests that assess cognitive function along with tests of balance and gait during multitasking. Individuals with dementia have more problems with gait and balance during multitasking than with simple gait tasks. A useful form that the Dizziness and Balance Center at Emory University developed to document outcome scores in this group of patients is shown in Table 29-1.

They are as follows:

- Cognitive tests: Mini-Mental State Examination [MMSE], Trails tests
- Timed gait tasks with various levels of multitasking: average gait speed, Timed Up & Go (TUG) test, Walk While Talk (WWT) test
- Fall risk assessments: DGI
- Standing balance tests: computerized dynamic posturography or similar force platform to quantify standing balance sway
- Test for retropulsion (a tendency to fall backwards when the hips are gently shoved backwards)

Two of the measurements test the ability of the subject to allocate attention toward balance when performing multiple tasks: the modified TUG test and the WWT test. Several studies suggest that an impaired ability to allocate attention to balance during dual-task situations may contribute significantly to falls in older adults. One study of assisted-living residents revealed that the inability to walk while talking was highly predictive of a future fall: 83% of those who stopped walking while talking experienced a subsequent fall.[11] The same prediction is also applicable to non-demented, community-dwelling older adults.[12] This study revealed a 71% positive predictive value for walking while reciting every other letter of the alphabet (WWT-complex) versus 42% for walking only. In the modified Timed Up and Go test, the patient first sits

■ Table 29-1 **OUTCOME MEASURES USED IN PATIENTS WITH IMBALANCE AND DEMENTIA**

	Pre-Drain Date:	2 hr Post-Drain
MMSE (<24 cognitively impaired)		
TUG (<11.8 sec = M + 2 SD nonfallers)		
TUG cognitive (<14.3 sec = M + 2 SD nonfallers)		
TUG manual (<12.9 sec = M + 2 SD)		
diff TUG (TUG manual – TUG) (diff TUG ≥ 4.5 sec at risk for falls)		
Average gait speed (ft/sec) (Normal age/gender = _____)		
WWT (40 ft) (≥18 sec at risk for falls)		
WWT-simple (≥20 s at risk for falls)		
WWT-complex (≥33 s at risk for falls)		
Fall risk: (DGI ≤ 19 = fall risk; BBS ≤ 49 = fall risk; <36 = 95% fall risk)		
Posturography: SOT composite score (normal for age = _____)		
Trails B – Trails A (normal for age = _____)		
Retropulsion on clinical exam		

in a chair and is instructed to stand up and walk as quickly and as safely as possible for 3 meters, turn around, walk back to the chair, and sit down. Then the patient is instructed to perform the test while counting backwards by threes from a randomly selected number between 20 and 100 (TUG – cognitive). Finally, the patient is asked to perform the test while carrying a full cup of water (TUG manual). A difference between TUG manual and TUG (TUG manual – TUG) greater than 4.5 sec indicates fall risk.

In the Walk While Talk test, subjects are timed while they walk at self-selected speed for 20 feet, turn around and return (40 feet total). On the second trial (WWT-simple) the subject walks the same course and recites letters of the alphabet. On the third trial (WWT-complex) the subject walks the same course and recites every other letter of the alphabet (e.g., "a-c-e"). The gait speed for each of those trials can indicate risk for falling as follows:

WWT (40 ft): greater than 18 sec at risk for falls

WWT-simple: greater than 20 sec at risk for falls

WWT-complex: greater than 33 sec at risk for falls

Table 29-1 lists three columns for data insertion. For patients with NPH, we use the table as it exists. "Drain" in the headings refers to performance of a large-volume CSF drainage procedure, as described earlier. For patients with leukoaraiosis, one can substitute the following headings: "Pre-PT," "1 week of PT," and "4 weeks of PT."

Management

Non-controlled studies have suggested improved cognition and balance from 65% to 77% in patients with NPH after insertion of a permanent shunt system to drain the hydrocephalus.[13,14] At our center, the shunt is inserted only if cognitive and gait tests performed after a large-volume CSF drainage procedure have a positive outcome compared with baseline.

Unfortunately, randomized, prospective clinical trials are lacking. In a Cochrane review published in 2002 and updated in 2009,[15] there is no evidence to indicate whether placement of a shunt to remove fluid is effective in the management of normal pressure hydrocephalus.

Current evidence suggests that in patients with leukoaraiosis, vigorous treatment of cardiovascular disease risk factors may prevent the development or progression of the process as well as the associated cognitive and balance decline.[16] Therefore, hypertension, elevations of cholesterol, diabetes mellitus, and smoking should be controlled in these patients. Unfortunately, there are no prospective trials to determine whether improved blood flow to the brain stabilizes or improves function.

All patients with NPH and leukoaraiosis should be referred for PT. These patients are usually at high risk for falls, especially backwards. For patients with leukoaraiosis, some balance improvement may occur with PT, but many patients must be given an aid (cane, walker, or rollator) when ambulating inside or outside the house. A home health evaluation may also be necessary to reduce the risk for falls at home. This may be true for patients with NPH as well, if shunting does not improve balance. Case Study 29-2 describes a patient with NPH, and Case Study 29-3 a patient with leukoaraiosis.

Progressive Supranuclear Palsy, Parkinson's Disease, Large-Fiber Peripheral Neuropathy, and Spinocerebellar Ataxia

Description

The neurological disorders progressive supranuclear palsy, Parkinson's disease, large-fiber peripheral neuropathy, and spinocerebellar ataxia are usually progressive and are associated with imbalance and falls.

Supranuclear palsy (PSP) and Parkinson's disease are a result of degeneration of different portions of the basal ganglia and forebrain. Both manifest as rigidity and masked facies. Retropulsion is mild in Parkinson's disease but severe in PSP. To perform the retropulsion test, the examiner has the patient stand with feet slightly apart and instructs the patient to take no more than one step backwards when the examiner suddenly pulls the patient backwards at the hips using a mild force. The result is positive if the patient must take three or more steps backwards or falls backwards "like a log." A resting and sometimes action tremor is found in Parkinson's disease but is absent in PSP. Both disorders have some degree of upgaze defect, but it is profound in patients with PSP. PSP can be devastating because the average life span of affected patients is 7 years, death being caused by aspiration or complications from falls to the ground.

Large-fiber peripheral neuropathy results in loss of vibration perception and proprioception. Severe loss of proprioception, especially in the ankles, leads to severe imbalance. Individuals with this disorder are extremely dependent on vision to maintain balance. They usually have a positive Romberg test result on clinical examination. Common causes of large-fiber neuropathy are diabetes mellitus, alcohol abuse, inflammatory neuropathy, and hereditary neuropathy.

Spinocerebellar ataxia (SCA) is a genetic disorder that causes degeneration of different portions of the

cerebellum and structures outside the cerebellum, including peripheral nerve and brainstem structures. These result in imbalance, difficulty with gait, and incoordination of the eyes, arms, and legs. There have been up to 60 different types of SCA identified, and at least 29 different gene mutations.[17] Some patients with SCA type 3 have bilateral vestibular hypofunction as well. Other patients with SCA have normal vestibular function but are not able to cancel their vestibulo-ocular reflex (VOR) because of a defect in the cerebellar vermis; this inability can cause greater motion sensitivity during head movements.

Useful Outcome Scores

Patients with PSP and some with Parkinson's disease demonstrate cognitive decline as the disease progresses, so the outcome measures in Table 29-1 become relevant.

Management

Patients with these degenerative disorders often need a longer course of treatment than those who have disuse disequilibrium. Compliance with the home exercise program is often a problem.

Several excellent medications are used to treat Parkinson's disease, including carbidopa-levodopa (Sinemet), which also helps mobility and balance. Eventually, many patients no longer show response to medication and require deep brain stimulation. It is not clear whether deep brain stimulation improves balance. Some patients experience disuse disequilibrium and require PT.

In a recent Cochrane report, 33 controlled trials consisting of 1,518 patients with Parkinson's disease randomized to either PT (general PT, exercise, treadmill training, cueing, dance, or martial arts) or no PT were assessed.[18] There was significant improvement in gait speed and step length for 2- and 6-minute walk tests, Berg balance scale, and Timed Up & Go test. There was no difference in the rate of falls between the two arms.

There are no successful controlled trials for treatment in patients with PSP. There is an ongoing clinical trial in Oregon (NCT01563276) whose purpose is to better understand why patients with PSP fall. With that information, this group hopes to develop better treatment strategies. For now, the standard of care includes a 4-week trial of low-dose Sinemet to determine if this medication improves gait and balance. There is no surgical treatment for this disorder. Gait and balance PT should be prescribed for fall prevention and reduction of disuse. Patients undergoing PT often need a rollator to prevent backward falls.

In a Cochrane review of the literature from 1966 to 2004[19] that was updated in 2009, there are only three randomized controlled trials of the effectiveness of exercise therapy to improve function in patients with neuropathy. The studies found improvement in muscle strength but not in reduction of disability. Currently, the standard of care for patients with proprioception defects caused by peripheral neuropathy or dorsal root/dorsal column disease is to encourage them to use visual cues while standing and walking.

A randomized clinical trial of 42 patients with pure cerebellar degeneration revealed that rehabilitation with physical and occupational therapies can significantly improve functional gains in ataxia, gait, and ADLs. In this study, the patients were randomized into an immediate treatment arm or a delayed-entry control arm. Some level of improvement was shown to be maintained for 24 weeks after treatment stopped.[20] Case Study 29-4 describes a patient with SCA that improved with rehabilitation.

Chronic, Situation-Related Dizziness

There are numerous chronic, situation-related dizziness syndromes described in the literature that are either chronic or situation induced in which all laboratory testing is normal and no consistent clinical abnormality is found on exam (see Table 29-2). These syndromes may overlap with psychogenic dizziness or be a unique syndrome in which we do not understand the organic basis (Box 29-1).

Space Phobia

This term was first used to describe four patients with intense fear of open spaces and absent visuospatial support.[21] Nine more patients were described in 1981, and of these, 11 out of 15 had severe disturbance in walking needing a visual support system close by (e.g., wall). They commonly crawled on the floor to cross a room or walked close to walls. This condition was distinguished from agoraphobia, because the latter problem has additional fear of public places. In my opinion, space phobia overlaps with what we might now call severe fear of fall.[22] This may respond to cognitive behavior counseling.[23]

Motorist Disorientation Syndrome

This term was first used to describe individuals with a misleading sense of movement and disorientation when they drove a car in a particular situation.[24] This leads to the illusion that the driver is falling to the side of the car or the car is turning on its side. These situations include driving on an open motorway, high speed, peak of a hill, or

■ Table 29-2 **CHRONIC, SITUATION-RELATED FORMS OF DIZZINESS**

Nomenclature	Year Term Used and Described in English Language	Description	Reference
Space phobia	1976	"Perception of unsupported space leads to a fear of falling and inappropriate rescue reactions"	21
Motorist disorientation syndrome	1985	When driving in particular situations, the driver has the illusion of falling to the side of the car or the car is turning on its side	24
Phobic postural vertigo	1986, 1994	Subjective imbalance in individuals with obsessive-compulsive personality that have no balance defect	26
Mal de débarquement	1987	Sense of rocking as if on a boat most noticeable when the individual is still. This sensation is masked when individual is moving (e.g., in the car). Usually induced after continuous passive transportation for several days (cruise ship)	28
Visual vertigo	1995	Overreliance on visual cues for perception and postural control	30
Chronic subjective vertigo	2007	Unsteadiness or dizziness present for most days for 3 months or more. Symptoms are severe when walking or standing and exacerbated by motion of objects in the visual surround or self-motion.	32 See also Chapter 19

traveling downhill. Some consider this a form of space phobia or visual vertigo.[25]

Phobic Postural Vertigo

This term was first used in 1986 by German neurologists, Drs. Thomas Brandt and Marianne Dieterich, and later discussed in the English language in 1994. [26] In the neuro-otology clinics, the incidence of this syndrome may be as high as 17%. It consists of a subjective sense of imbalance in individuals that have an obsessive-compulsive personality but no balance problems. These patients do not show psychogenic signs of a gait disorder .[26] Treatment has included reassurance, description of the mechanism,

Box 29-1

POINT AND COUNTERPOINT

There are a number of syndromes of dizziness cited in the literature that are diagnosed strictly by the history, that is, there are no abnormal tests that are used to confirm the diagnosis (see Table 29-2). Some patients may have vestibular hypofunction or central signs on laboratory testing, but others with the same syndrome do not and these findings do not appear to be the cause for the chronic symptoms. Until objective tests are found that can be relied on to diagnose these different syndrome, it remains unclear if these syndromes are really distinct organic disorders in which we have yet to find the pathophysiological cause, or a problem with coping to vestibular cues following some type of traumatic or unusual event or an exaggerated response to vestibular stimulus. For now, most are treated the same way with limited success.

and repeat exposure with regular exercise. In a 2.5-year follow-up, symptoms of dizziness were reduced, but as a whole there were still significant psychological problems requiring psychotherapeutic intervention.[27]

Mal de Débarquement

The term mal de débarquement syndrome (MdDS) was initially used and described in the English literature in 1987.[28] It consists of the sense of rocking as if on a boat, most noticeable when the individual is still. This sensation is masked when the individual is moving (e.g., in the car). It usually is induced after continuous passive transportation for several days (cruise ship). Enhanced activity in certain limbic areas that may be involved in visual motion has been found on functional MRI of individuals with MdDS.[29]

Visual Vertigo

Bronstein used this term to describe individuals with peripheral vestibular, central lesions in posterior fossa, or strabismus that have abnormally high dependence of full field visual motion.[30] As a result, they report dizziness and show signs of imbalance when exposed to certain visual situations. Since then, this nomenclature has been used to include all individualsß including those with migraine or anxiety that appear to have an "over reliance on visual cues for perception and postural control."[31] Case Study 29-5 describes a patient diagnosed with visual vertigo.

Chronic Subjective Dizziness

This term was first used in 2007 by Staab and Ruckenstein based on a series of studies of patients in the USA that resembled phobic postural vertigo.[32] It consists of unsteadiness or dizziness present for most days for 3 months or more. Symptoms are most severe when walking or standing and exacerbated by motion of objects in the visual surround or self-motion. Triggering factors may be acute or recurrent problems that cause dizziness or unsteadiness including (1) peripheral vestibular dysfunction, (2) medical problems, and (3) psychiatric disorders. Exam and testing are either normal or consist of features that do not explain the symptoms. These patients have an increased prevalence of psychiatric disorders, usually anxiety or depression, but they also may not show these features. Case Study 29-6 describes a patient diagnosed with Chronic Subjective Dizziness. See also Chapter 19.

Management

There is no reliable treatment for any of these chronic, situation-related dizziness disorders.

What is generally suggested is reassurance, increased activity, and discussion of the problem with the health provider. One can also add cognitive behavioral therapy, desensitization, and medication including low-dose serotonin selective reuptake inhibitors and long-acting benzodiazepines. The role of physical therapy is discussed in Chapter 19.

CASE STUDY 29-1

Mrs. T, 73 years old, fell from a 3-foot ladder 9 months ago. Although she had no significant injury, she has had chronic dizziness since. She loses her balance occasionally but denies falling. Before she fell from the ladder, she walked 3 miles a day, but now she is afraid to walk. On examination, she has no significant orthopedic or neurological problems. Vestibular findings are normal. She cannot walk tandem and shows fear of fall when standing with eyes closed. She touches the walls while walking in the clinic. The Tinetti Fall Risk Assessment score was 27, identifying this patient as being at moderate risk for fall (Fig. 29.2). This assessment tool is excellent for patients at risk for fall.[33]

Comment

The history and examination findings are consistent with disuse disequilibrium and fear of fall. Mrs. T started on a daily home exercise program coordinated by a physical therapist with a specialty in geriatrics. She saw the therapist in clinic once a week for 4 weeks. During each clinic visit with the therapist, her balance was assessed and her exercises were made more difficult. The exercises included progressive static balance with eyes opened and closed, progressive gait exercises with and without head movements, and eventually, a walking program that increased from 1 to 3 miles a day. At the end of the fourth week, Mrs. T's Tinetti Fall Risk Assessment score improved to normal range (score = 35/37). She returned to her normal activities, and she was discharged from the clinic.

CASE STUDY 29-1

TINETTI FALL RISK ASSESSMENT

Patient MT 64-57-08 **Date 03/26/99**

Sitting Balance	(2) Steady, stable (1) Holds onto chair to keep upright (0) Leans, slides down in chair
Arising from chair	(2) Steady without holding on (1) Uses arms of chair (0) Unable without help or multiple attempts
Immediate standing balance	(2) Steady w/o support (1) Steady, w/ support (0) Unsteady (grabbing moves feet, etc)
Standing	(2) Steady w/ feet together (1) Steady, feet apart (0) Unsteady or holds on
Balance w/eyes closed	(2) Steady, feet together (1) Steady, feet apart (0) Unsteady; holds on
Sternal nudge light pressure 3 times	(2) Steady (1) Moves feet but keeps balance (0) Begins to fall
Turning (360)	(2) Continuous steps no grabbing or staggering (1) Discontinuous, puts foot down completely before raising the other (0) Unsteady or holds on
Neck turning	(2) Horizontal and vertical (at ceiling), steady (1) Decreased ability but no unsteadiness or pain or dizziness (0) Unsteady or is symptomatic
One-legged stance, eo	(1) Able 5 sec w/o holding on (0) Unable
Back extension ask to lean backwards	(2) Good extensions w'o holding on, staggering (1) Tries but decreased ROM or holds on (0) Will not attempt or staggers
Reaching up high	(2) Able to take down object w/o holding on or becoming unsteady (1) Able to reach but needs to hold on (0) Unable or unsteady
Bending down	(2) Able, single attempt, doesn't hold on (1) Able, single attempt but holds on (0) Unable or multiple attempts
Sitting down	(2) Able, one smooth motion (1) Needs to use arms, not smooth (0) Falls into chair, misjudges distance
Gait	
Initiation of gait	(1) Begins immediately, single smooth motion (0) Hesitates, multiple attempts, not smooth
Step height	(1) Completely clears, 1-2 inches, right (1) Completely clears, 1-2 inches, left (0) Does not clear, right (0) Does not clear, left
Step length	(1) Right foot passes left foot by foot length (0) Right foot does not pass left by full foot length (1) Left foot passes right foot by foot length (0) Left foot does not pass right by full foot length
Step symmetry	(1) Same or nearly same on both sides (0) Varies, or advances with same foot on every step
Step continuity	(1) No breaks or stops in stridez (0) Stops between steps, step length varies
Path deviation	(1) Foot follows straight line (0) Foot deviates side to side or in one direction
Trunk stability	(2) Normal (1) Knees or back flexed, arms not abducted to assist (0) Marked sway
Walk stance	(1) Normal base of support (0) Widened base of support
Turning around while walking	(1) Normal, continuous (0) Staggers or stops to turn

Total Balance (25) = **20** Total Gait (12) = **7**

Total = **27** (37) (> 31 OK; 26-31 = moderate risk; < 25 high risk)

4/29/99

Total Score = 35

Figure 29.2 Example of the use of the Tinetti Fall Risk Assessment.

CASE STUDY 29-2

Mr. G is an 85-year-old man with a 2-year history of cognitive decline, imbalance with falls to the ground, and urinary incontinence. He brought MR images of his head with him (Fig. 29.3). No obstruction of CSF flow was found in any of the ventricles, consistent with a diagnosis of communicating hydrocephalus. His outcome scores before and after a large-volume CSF drainage procedure are shown in Table 29-3. Lumbar puncture results were normal, including CSF pressure.

Comment

On the basis of history, lumbar puncture, and head MRI findings, this patient has NPH. His outcome scores before the drainage procedure document dementia, impaired balance and gait, and fall risk. At baseline (pre-drain), his average gait speed, TUG, and WWT scores indicated slight impairment, but his gait speed during multitasking (TUG cognitive and WWT-complex) showed significant impairment.

After the large-volume lumbar puncture drain, Mr. G showed no significant improvement in cognition, gait, or balance. He and his family elected not to proceed with a shunt. Instead, he was given a 4-week course of gait and balance PT. There was modest improvement, but he was still at risk for fall. Therefore, a walker was prescribed. He was instructed to continue with the exercises, and a 6-month follow-up appointment was scheduled.

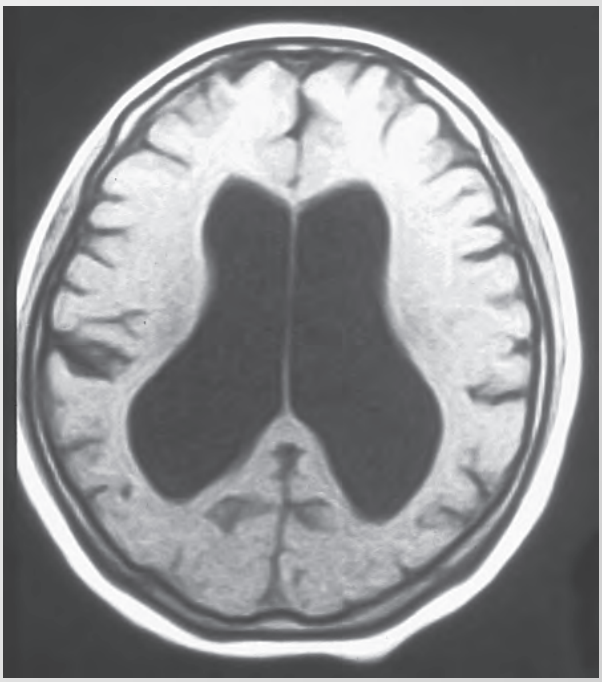

Figure 29.3 Magnetic resonance image of the head in the horizontal plane illustrating hydrocephalus.

■ Table 29-3 **OUTCOME SCORES FOR A PATIENT WITH NORMAL-PRESSURE HYDROCEPHALUS AFTER LARGE-VOLUME CSF DRAINAGE PROCEDURE (CASE STUDY 29-2)**

Pt Name: Mr. G	Pre-Drain	2 Hr Post-Drain
Dx: NPH	Date: 03/31/06	03/31/06
MMSE (<24 = cognitively impaired)	19	21
TUG (<11.8 sec = M + 2 SD nonfallers)	13.21	11.77
TUG cognitive (<14.3 sec = M + 2 SD nonfallers)	25.15	22.75
TUG manual (<12.9 sec = M + 2 SD)	17.83	17.30
diff TUG (TUG manual – TUG) (diff TUG ≥ 4.5 sec = at risk for falls)	4.62	5.53
Average gait speed (ft/sec) (normal) age/gender = 3.08)	2.32 ft/sec	2.20 ft/sec

CASE STUDY 29-2

■ Table 29-3 **OUTCOME SCORES FOR A PATIENT WITH NORMAL-PRESSURE HYDROCEPHALUS AFTER LARGE-VOLUME CSF DRAINAGE PROCEDURE (CASE STUDY 29-2)—cont'd**

Pt Name: Mr. G	Pre-Drain	2 Hr Post-Drain
WWT (40 ft) (≥18 sec = at risk for falls)	18.24	21.64
WWT-simple (≥20 sec = at risk for falls)	26.62	28.53
WWT-complex (≥33 sec = at risk for falls)	41.08	32.53
Fall risk: (DGI ≤ 19 = fall risk)	15	16
Posturography (normal for age = 63.8)	32	36
Trails B – Trails A	472 sec	482 sec

CASE STUDY 29-3

Sixty-year-old Mrs. S complains of chronic imbalance. Her legs feel heavy, "like lead." She has had several falls backwards with injury. She has diabetes mellitus, hypertension, and an elevated cholesterol value. On examination, she has masked facies, mild rigidity, and gait apraxia. When given shoves backwards, she falls "like a log," indicating a poor righting reflex. She does not have tremor or cogwheel rigidity . She has not shown improvement with a trial of Sinemet. An MRI of her head shows leukoaraiosis (Fig. 29.4). She is referred to PT for gait and balance exercises.

Comment

This patient has significant risk factors for small vessel disease of the brain. She has parkinsonian features but does not have Parkinson's disease (no tremor and had no response to Sinemet). She was started on a daily home exercise program and was seen 1 day each week in PT for 4 weeks. There was no significant improvement in fall risk or posturography scores, so she was prescribed a rollator with seat, basket, and hand brakes to improve her independence. She was also instructed in changes she could make in her home to improve her safety.

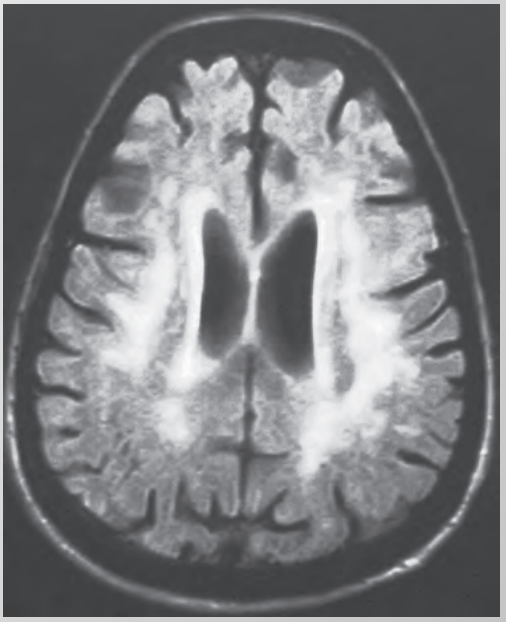

Figure 29.4 Magnetic resonance image of the head in the horizontal plane showing extensive white matter ischemic changes otherwise known as leukoaraiosis.

CASE STUDY 29-4

Ms. P, 50 years old, has had progressive imbalance for 3 years and several spells of spontaneous vertigo lasting for a few seconds while walking. She has fallen three times, and she fractured her left ankle while going down her stairs. She has used a cane for a year. Her father, two sisters, and brother all have the same problem. Examination shows bilateral vestibular weakness based on head thrust, downbeat nystagmus, dysarthric speech, and cerebellar ataxia in the lower extremities. Head MRI shows cerebellar and pontine atrophy (Fig. 29.5). Genetic screening is consistent with SCA type 3, a dominant form of SCA.

Comment

Some patients with SCA type 3, like this patient, have a central cause for bilateral vestibular loss. The physical therapist performed an initial evaluation and identified multiple problems. These included decreased gaze stability during head movements, poor balance in stance when visual cues were diminished and when the support surface was uneven, difficulty maintaining balance when walking on anything other than a firm, flat predictable surface, and a risk for falling. In addition, the patient was unable to successfully perform tasks such as stepping over an object and walking quickly enough to cross a street. The physical therapist instructed Ms. P in exercises for eye gaze stability, balance, strengthening, and gait that were updated weekly. Balance retraining was performed with various sensory conditions (e.g., static balance with eyes open and then closed). The patient practiced balancing on foam with eyes open/closed with supervision and modified single-leg stance activities. The walking program for Ms. P began on level surfaces with cane (walking forward, backwards, and sideways). Eventually, walking while making head turns slowly side-side and walking through an obstacle course stepping over small obstacles, on grass, and slight slopes were added. Strengthening exercises included performing sit to stand without upper extremity assistance and bridges on bed with added hip abduction/adduction to increase hip control. Vestibular adaptation exercises included X1 viewing (see Chapter 20) while sitting, moving her head both horizontally and vertically, and using both near and distant targets. She performed these 3 to 5 times each day but had 1 or 2 days of increased visual blurring; therefore, the range of head movements was decreased and exercises were changed to only 3 times per day. She progressed to standing position, performing exercises for 1 minute, 3 times per day. The patient was seen weekly for 9 visits. She then continued therapy less frequently with follow-up visit 3 weeks later. Table 29-4 shows the improvement in outcome scores with PT for Ms. P.

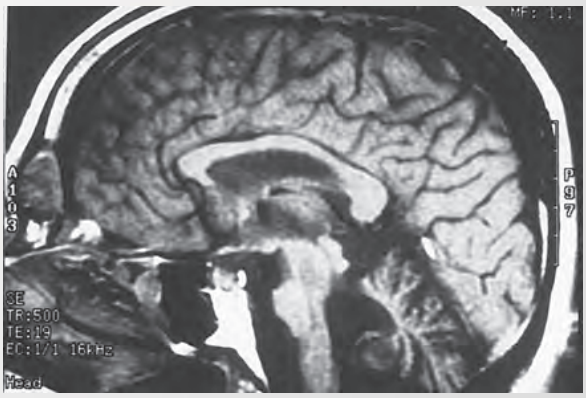

Figure 29.5 Magnetic resonance image of the head in the sagittal plane in a patient with spinocerebellar atrophy type 3.

■ Table 29-4 **OUTCOME SCORES IN A PATIENT WITH CEREBELLAR ATAXIA (CASE STUDY 29-4)**

Outcome Test	Score at Initial Evaluation	Score on Day of Discharge from PT
8-ft Up & Go Test	22.4 sec with cane	11.4 sec without cane
SOT composite score on computerized dynamic posturography	36/72; 6/6 falls	70/72; 1/6 falls
DGI	3/24	17/24
ABC Scale Score	11.2%	71%
Visual Analogue Scale (Disequilibrium)	9.5/10	3.1/10

CASE STUDY 29-5

Visual Vertigo

Forty-one-year-old Mr. T has 2-minute or less spells of "sense of v ertigo/dizziness with blurred vision." He describes this as uneasiness, rocking, and imbalance triggered by a v ariety of visual circumstances. These include when he changes focus points of vision and when indi viduals come up to talk to him, especially after training. "Sometimes feels like when trying to focus on things I may get the spinning/dizzy feeling." There is no nausea, falls to the ground. This has been going on for 5 years since deployment in Iraq as a Ranger . His initial ENG in 2009 suggested mild L UVL but on 5/23/11 it was normal. MRI of head, neurological e xam are all normal. He has had e xtensive physical therapy to treat a presumed v estibular neuritis but has not improved. He was seen by Emory neuro-ophthalmology, who found no problem, and the y referred the patient to me. My neurological and vestibular exam was normal. His rotary chair test in our lab was normal.

Comment

I diagnosed this patient with "visual v ertigo" based strictly on his symptoms of dizziness provoked or aggravated by specific visual contexts (driving, supermarket, or movement of objects). This is an unfortunate term because the patient does not ha ve true vertigo and there is no objective cause, b ut this syndrome has been well described by Dr. Bronstein.[30] I discussed treatment to include medication such as low-dose Paxil in the morning and low-dose Klonopin at night. I discussed beha vioral counseling. Finally, I discussed the study by P avlou et al,[31] a small study of 26 individuals with visual vertigo randomized into no intervention control group and exposure to moving visual stimuli (full-field OKN or rotating disk) for up to 2 min at a time for a total of 4 hr a week for several weeks. The control group spent the same amount of time playing sorts (c ycling, running, swimming). Subjects then were assessed for subjective visual vertigo, postural sway to large rotating disk to assess their visual dependency. Outcomes improved significantly in the experimental group over the control group ($P = 0.04$).

CASE STUDY 29-6

Mrs. M is a 62-year -old female whose chief complaint is "I am always dizzy, ever since heart surgery, 11 months ago." She is extremely bothered by visual motion in her environment, has trouble walking, lightheadedness, double vision, and fear of faint. It is worse when she is walking. She admitted to being a Type A personality. She stopped exercising (treadmill and gym weights) 4 days a week for 1 hour because of the dizziness. She has seen numerous physicians for this including neuro-ophthalmologist, cardiology, primary care, and endocrine, but no causes for her symptoms have been found. She had a normal neurological exam, normal ENG with caloric response of 39 to 41 deg/sec to irrigation of water. Computerized dynamic posturography showed a better than normal composite score (84) compared with age-matched control (68) and no motor defects. There were no aphysiologic features found on this test. Her Tinetti fall risk was normal at 36/37. Her PANAS scale for anxiety and depression was normal. Her MRI of the head was normal.

Comment

Her evaluation satisfies the criteria for Chronic Subjective Dizziness[32] in that her symptoms are daily for at least 3 months, most severe when she is on her feet or walking, and exacerbated by visual motion in her environment. It occurred after sur gery to replace her mitral valve. She lived more than 500 miles away and flew in to see us. I had her see our physical therapist and me, and I ordered ENG and posturography on the same day . The tests were normal as was my exam. PT found that her MSQ was less than 1 and her dizziness VAS score after 1 min of head movements was 7/10 compared with 5/10 just sitting still. She was given habituation exercises to do at home by the PT , and she w as placed on lo w-dose Paxil. The diagnosis was discussed with her, and she was encouraged to read more about it at home. She was encouraged to begin to exercise again and consider cognitive behavioral counseling if this did not help her after 1 month.

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CHAPTER 30

Physical Therapy Management of People with Non-vestibular Dizziness

Lisa Heusel-Gillig, PT, DPT, NCS ■ *Courtney D. Hall, PT, PhD*

Dizziness is among the most prevalent complaints for which people seek medical help, and the incidence increases with advancing age.[1] Dizziness represents a diagnostic and treatment challenge, because it is a subjective sensation, refers to a variety of symptoms (unsteadiness or imbalance, spinning, sense of motion, or lightheadedness), and has many potential contributory factors. As many as 45% of individuals with dizziness are diagnosed with peripheral vestibular disorders;[2] therefore, the majority of individuals have non-vestibular causes for their dizziness and imbalance. Although these patients have similar signs and symptoms to those with inner ear dysfunction, vestibular function testing does not reveal vestibular pathology. It is essential that patients with non-vestibular dizziness and imbalance be referred to specialists in vestibular rehabilitation to achieve optimal outcomes. Many assessments that we recommend, including a thorough oculomotor exam in both room light and with fixation removed, are identical to those used to evaluate vestibular patients. Although they are not commonly performed in inpatient or outpatient settings, they provide important information to the clinician. There is considerable evidence that vestibular exercises are important in the rehabilitation of patients with vestibular pathology.[3] There is also evidence that vestibular rehabilitation is beneficial for patients with non-vestibular dizziness.[4-11]

After excluding patients who have dizziness caused by cardiovascular pathology or medication side effects, patients with non-vestibular dizziness can be divided into two major categories: those with neurological pathology and those without. Individuals with neurological involvement include peripheral neuropathy affecting large fibers with impaired proprioception or kinesthesia in toes or ankles, cerebellar/brainstem disorders (e.g., spinocerebellar ataxia, multiple sclerosis, and stroke), ischemic white matter disease (leukoaraiosis), and mild head injuries and concussions. (See Chapter 26 for assessment and management of individuals with head injuries and concussions; see Chapter 29 for medical management of patients with non-vestibular dizziness.) Individuals with non-vestibular dizziness without neurological involvement include older adults with disuse disequilibrium and/or fear of falls and individuals with motion sensitivity caused by migraines and/or anxiety.

This chapter identifies components of the physical therapy assessment that are the most important as well as outcome measures which are appropriate. Secondly, we discuss therapeutic interventions for patients with non-vestibular dizziness including gait and balance training, vestibular rehabilitation including habituation and gaze stability exercises, and dual-task activities. We suggest recreational activities that are both enjoyable and

challenging for the patient to continue after discharge to maintain gains achieved with formal physical therapy. Each section identifies specific treatments that have been shown to improve balance and mobility, as well as dizziness. Finally, we discuss expected recovery time and factors that may limit functional progress.

Assessment

A comprehensive evaluation is essential to identify the underlying impairments and functional deficits and determine the best treatment approach for patients with non-vestibular dizziness. Our dizziness and balance clinic uses a six-page questionnaire (that is mailed to each patient before the first appointment). This allows the patient adequate time to record relevant history and describe symptoms that help the clinician prioritize the evaluation. A questionnaire should include date of onset, characteristics of symptoms (e.g., acute or chronic, spells or constant), past medical history, medications, and a scale to assess anxiety and depression. Reviewing physicians' notes and previously performed tests also helps the therapist decide which specific assessments to perform.

Taking a good history is the most important part of the assessment. The patient needs to identify exactly what is meant by the word "dizzy." Many may report being "off balance," and others may complain that movements cause their head to feel "lightheaded, woozy, foggy, fuzzy, full, spacey, cloudy" or even express that they are "inside a cotton ball." The therapist needs to ask questions such as "Is there something that makes your symptoms worse?" "Better?" "Is there a time in the day when you have no symptoms?" Additionally, the therapist should have the patient identify the primary goal for physical therapy.

After discussing the questionnaire and history with the patient, reliable and valid outcome measures to assess both symptoms and performance-based balance and mobility should be performed (Table 30-1). Strength, sensation, and coordination also need to be evaluated, but will not be discussed in this chapter.

Therapeutic Interventions

Gait and balance training, vestibular rehabilitation including habituation and gaze stability exercises, and dual-task activities have been shown to improve function, postural stability, and symptoms in patients with non-vestibular dizziness. Studies have shown that including these treatment approaches are beneficial to this population and will be discussed in each section below.

■ Table 30-1 **SUGGESTED OUTCOME MEASURES FOR ASSESSMENT OF PATIENTS WITH NON-VESTIBULAR DIZZINESS (SEE CHAPTER 21 FOR DETAILS REGARDING ICF MODEL)**

OUTCOME MEASURES ACCORDING TO ICF DOMAIN

Tool	Body Structure/ Function (Impairment)	Activity (Limitation)	Participation (Restriction)
Activities-based Balance Confidence (ABC) scale[12]		x	x
Dizziness Handicap Inventory (DHI)[13]		x	x
Dizziness Visual Analog Scale[14]	x		
Motion Sensitivity Quotient[9]		x	
Situational Vertigo Questionnaire (SVQ)[15]			x
Visual Vertigo Analog Scale (VVAS)[16]			x
Preferred Gait Speed[17]	x		
Dynamic Gait Index[18]		x	

■ Table 30-1 **SUGGESTED OUTCOME MEASURES FOR ASSESSMENT OF PATIENTS WITH NON-VESTIBULAR DIZZINESS (SEE CHAPTER 21 FOR DETAILS REGARDING ICF MODEL)—cont'd**

Tool	Body Structure/ Function (Impairment)	Activity (Limitation)	Participation (Restriction)
Modified Timed Up and Go (mTUG)[19]		x	
Walk While Talk Test (WWT)[20]		x	
Tinetti Fall Risk Assessment (Performance Oriented Movement Assessment)[21]		x	
Five Times Sit to Stand (FTSTS)[22]		x	
Computerized Dynamic Posturography (CDP)[23]	x		
Modified Clinical Test for Sensory Integration of Balance (mCTSIB)[24]	x		
Functional Reach[25]	x		

Gait Training

Most patients with non-vestibular dizziness referred to outpatient physical therapy are ambulatory with or without an assistive device, or at a pre-gait level. Difficulty with walking leads to functional decline, loss of independence, and falls in older adults [26]; therefore, gait training is a critical component of rehabilitation. Gait training begins on level surfaces with or without a device and progresses to uneven surfaces. Ambulation should also include a variety of changes in task demands, such as head turns, stepping over and around obstacles, variable speeds, starts and stops, backwards gait, and gait with eyes closed. Practicing specific components of the gait cycle that are impaired can translate into a safer and more efficient functional gait pattern. As part of the home exercise program, patients should walk daily for a specific amount of time to improve endurance, strength, and overall confidence.

Slow or Shuffling Gait

Slow or shuffling gait is common in older adults with fear of fall/disuse disequilibrium and in individuals with neurological disorders such as ischemic white matter disease.[27] White matter disease is common in older adults and often goes undiagnosed.[28] White matter changes are strongly correlated with gait impairments including decreased speed, reduced foot clearance, and minimal heel strike causing a trip hazard.[29] "Step-overs," an exercise involving stepping forward and backwards over a rolled-up towel, increases stride length, foot clearance, and heel strike (Fig. 30.1). Obstacle courses that involve stepping over objects, through ladders, and onto small curbs also address this problem. Stair climbing, turns, and backing up to sit can lead to falls more than forward gait; therefore, these activities should be incorporated into the interventions.[30]

Ataxic Gait

Ataxic gait is common in patients with cerebellar degeneration, infarcts, or tumors. These patients have a wide-based gait with variable step length.[31,32] Individuals with ataxic gait ambulate at a fast pace often because they are unable to balance on one foot and have difficulty slowing down or stopping quickly. The patient should practice stepping over objects as slowly as possible and standing on one foot to improve these impairments. In addition, individuals with ataxic gait cannot smoothly and accurately change directions as needed when walking in tight spaces. Obstacle courses designed to focus on these impairments can be useful.

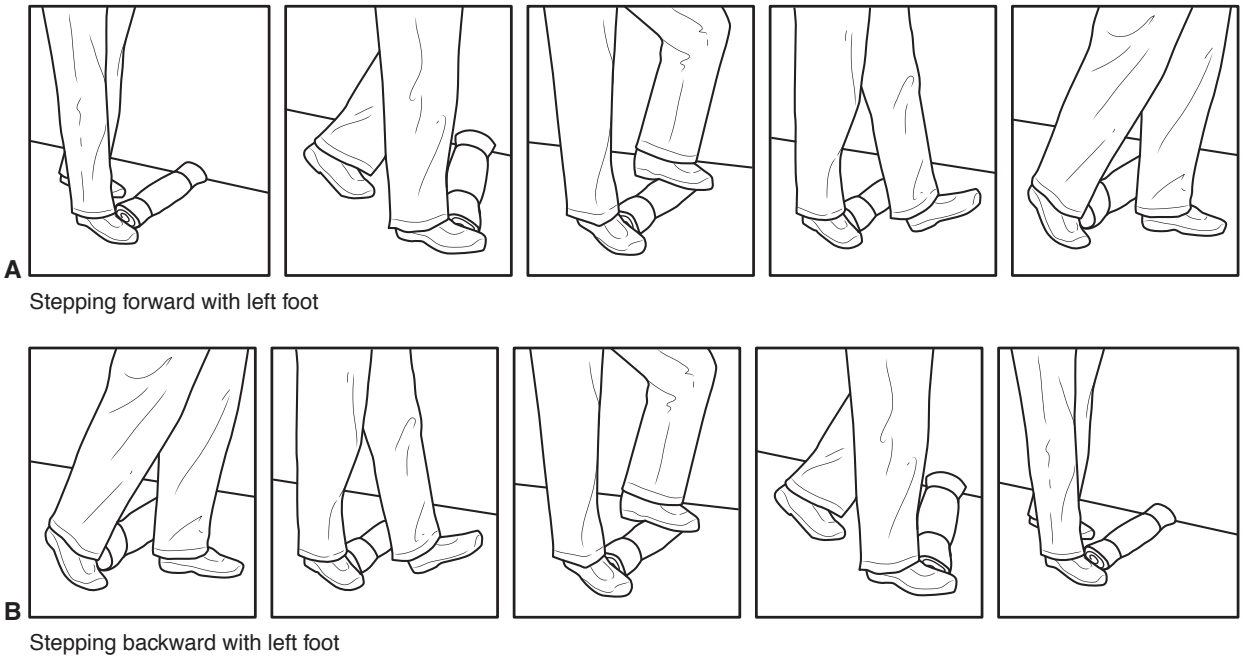

Figure 30.1 Many patients with fear of falls have decreased foot clearance and heel strike. In the "Step-over" exercise, the patient is required to **(A)** step over an object (rolled towel) and shift his or her weight forward as if preparatory to taking another step. The patient then steps backward **(B)** with the same leg and repeats sequence 10-15 times on each leg.

Treatment of Cerebellar/Brainstem Disorders—Supportive Evidence

A retrospective study of patients with cerebellar/brainstem disorders demonstrated that an average of five therapy visits of an individualized program including balance and gait training, sensory integration, and vestibular adaptation exercises resulted in improved balance-related confidence and functional strength, and reduced disability related to symptoms and fall risk as measured by dynamic gait index.[6] The patients included individuals with central vestibulopathy (n = 11), cerebellar deficits (degeneration and stroke; n = 11), non-cerebellar stroke (n = 10), mixed central and peripheral vestibular involvement (n = 9), and post-traumatic central disorders (n = 5). All the patient sub-groups improved significantly; however, those with cerebellar degeneration improved the least.[6] Additionally, a prospective observational study of 152 patients with peripheral and/or central vestibular dysfunction who received customized vestibular rehabilitation showed significant improvement in motion sensitivity and sensory integration as measured by the motion sensitivity quotient (MSQ) and sensory organization test (SOT).[33]

Cautious Gait

"Cautious gait," according to Nutt and colleagues, is a slow, wide-based gait with shortened stride length and "en bloc" turns that occurs in patients with migraines, mild head injuries, and concussions.[34] In our experience, this pattern is also common in patients who have had a recent history of benign paroxysmal positional vertigo (BPPV) who are afraid of provoking symptoms of dizziness and in other patients with head movement–induced dizziness. These individuals should not only practice speeding up their gait and swinging their arms, but also walking with head turns and outdoors on uneven terrains to challenge dynamic balance and improve endurance.

Balance Training

It is well documented that the incidence of falls increases with age. In community-dwelling individuals aged greater than 65 years, the reported incidence of falls varies from 28% to 35%.[35,36] In individuals aged greater than 75 years, this increases to a 42% to 49% incidence of falls.[37,38] Most patients with non-vestibular dizziness are at risk for falls

based on outcomes measures, have fallen at least once in the past year, or have gradually become more cautious and sedentary. Many individuals report holding onto the walls or furniture inside their homes. In the community, the older adults begin using a cane plus the support of their spouse or other family member, or a rollator walker. A combination of slowed motor responses, impaired sensory input, and decreased strength contributes to falls in older adults.[27]

Balance deficits caused by *poor sensory integration* are common in patients with mild cognitive decline, cerebellar ataxia, and peripheral neuropathy.[27,39] To maintain postural stability, the brain must quickly process inputs from the visual, somatosensory, and vestibular systems.[28] When one or more of these sensory systems fail, imbalance and incidence of falls increase.[28] Therefore, balance retraining designed to improve the integration of the three sensory systems should be included as an intervention for non-vestibular dizzy patients. Initial assessments frequently show abnormal findings on the modified clinical test for sensory interaction of balance (mCTSIB) with eyes closed on foam (condition 4) or on conditions 5 and 6 of the SOT using computerized dynamic posturography (CDP).

Balance training for these individuals should focus on increasing the use of the vestibular system by engaging, reducing, or removing visual input (e.g., focusing on a moving object, using dim lighting, or closing eyes) and reducing somatosensory input (e.g., foam surface). If Romberg position with eyes closed (condition 2 on the mCTSIB) is difficult for someone with somatosensory deficits, the patient should be instructed to "feel your weight on the balls of your feet" and practice with feet apart with eyes open and closed and gradually narrow the base of support. This intervention not only forces the use of the remaining somatosensory input, but helps the patient use vestibular and visual inputs more effectively.

Exercises for Fall Prevention—Supportive Evidence

A meta-analysis of fall prevention exercise programs in over 9,000 older adults demonstrated that a program combining a moderate to high level of challenging balance exercises, including reduced base of support, controlled movements of the center of mass, and minimal upper extremity support, as well as an increased dose of exercise time (at least 50 hours) had the greatest effect on fall prevention.[40] Modified single-leg stance (Fig. 30.2), an exercise in which one foot rests lightly on a book or cushion while the patient focuses straight ahead, progressing to head rotations or eyes closed, improves the individual's ability to step up on curbs or over obstacles without support. Exercises that facilitate dynamic postural reactions are also important to prevent falls. The activities should be safe but challenging enough to force the patient to use appropriate automatic balance reactions including ankle, hip, and stepping strategies. If these strategies are not automatic or quick enough to be effective, they need to be practiced initially with voluntary activities and progressing toward involuntary (or reactive) until the patient is able to do them in unpredictable situations.

Patients with *fear of falls* are afraid to shift voluntarily near their limits of stability as determined by

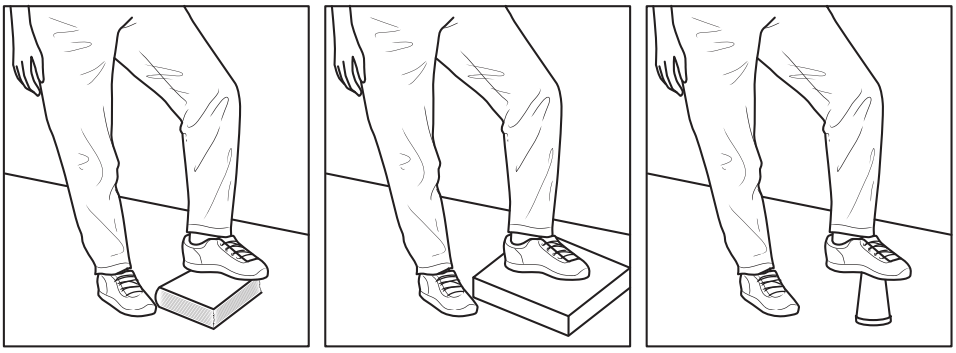

Figure 30.2 Certain activities while walking can require that the patient stand on one foot for brief periods of time without loss of balance. In the "modified single-legged stance" exercise, patients place one foot lightly on a book, or foam or cup, and maintain their balance for a specified period of time. Variations include performing slow head turns with eyes open, or keeping the head stationary and closing the eyes as well as performing cognitive tasks.

the functional reach test or limits of stability on CDP . From our experience, these patients respond well to practicing automatic activities including passing a ball, tapping a balloon, or performing weight shifts using visual feedback (e.g., Equitest or Wii System; Fig. 30.3).

Ataxia, Falls, and Fear of Falling

For individuals with ataxia, falls are a major problem: in one retrospective study, 93% reported falling in the previous year.[41] In addition, patients with ataxia report higher levels of fear of falling than healthy controls (43% compared with 2%).[41] Fortunately, patients with truncal and limb ataxia can benefit from balance retraining.[8] In a randomized controlled trial, subjects with degenerative cerebellar ataxia who underwent an intensive coordination training program that focused on balance improved static and dynamic balance and maintained gains with a prescribed home program at 8-week follow-up testing.[8] Static and dynamic balance exercises included single-leg stance, unpredictable perturbations by the therapist in all directions, and whole-body movements to train trunk and limb coordination. Study results demonstrated that static balance on both firm and compliant surfaces improved.[8] Many individuals with ataxia cannot stand on one foot; therefore, they are prescribed a modified single-leg stance (mod SLS) exercise (see Fig. 30.2) to gradually improve stability.

Figure 30.3 Wii Practice: patients with difficulty shifting weight or who have a fear of falls can benefit from participating in activities that allow them to have fun and focus on visual feedback to help them improve balance.

Migraineurs with motion sensitivity are often visually dependent and sway more than normal when exposed to visually stimulating environments.[42] This group of patients should practice static balance on compliant surfaces with eyes closed forcing them to use their vestibular and somatosensory inputs. Pavlou and colleagues[15] demonstrated that a group of subjects with motion sensitivity who performed customized vestibular rehabilitation (gaze stability and dynamic balance activities) plus simulator training (involving videos of optokinetic stimulus) improved to a greater extent on both postural stability and visual vertigo as determined by SOT and Situational Vertigo Questionnaire (SVQ; Appendix 30-1) than the group that performed customized vestibular training without simulator training. These findings suggest that the use of optokinetic stimulation is beneficial for this group of patients with motion sensitivity.

Habituation Exercises

Many individuals become dizzy and imbalanced with *self-movement* and others experience discomfort, imbalance, and disorientation in *stimulating (visual) environments*, often described as a visual-vestibular mismatch,[43] visual vertigo,[44,45] or space and motion discomfort.[46] Patients with a new onset or exacerbation of motion sensitivity with either self or environmental stimulation respond well to habituation exercises. With repeated exposure to the provoking stimulus, the central nervous system habituates so that tolerance of motion improves.[9] Initially, intensity of symptoms may increase with these exercises but decrease over time with repetition. Persons with oculomotor deficits because of neurological involvement and migraineurs frequently experience motion sensitivity.[6,47,48]

Motion-provoked dizziness should be assessed with valid and reliable measures (see Table 30-1). Suggested scales include Motion Sensitivity Quotient (MSQ),[9] Dizziness Handicap Inventory (DHI),[13] Situational Vertigo Questionnaire (SVQ),[15] and Visual Vertigo Analog Scale (VVAS).[16]

These assessments help determine appropriate exercises for the home exercise program. The MSQ

determines the specific movements that cause the symptoms to increase. This 16-item scale includes movements including rolling, bending over to right and left knees, horizontal and vertical head rotations, and 180-degree turns in standing (see Chapter 21). The SVQ and VVAS help the therapist pinpoint environments and situations that provoke symptoms that can be simulated in therapy and home exercise program (HEP) to reduce symptoms of visual vertigo (see Appendices 30-1 and 30-2). The SVQ and VVAS include items such as walking through a supermarket, watching a movie or action TV, walking on patterned floors, and going on escalators. If the patient is sensitive to self-movements based on MSQ, up to 4 movements are chosen depending on the severity of symptoms (Table 30-2).

Exercises should not provoke more than a mild to moderate intensity for each repetition or set. Each patient is prescribed a specific number of repetitions and sets depending on his symptoms. Duration of the response should last less than 30 seconds per set, and the patient should return to baseline within 15 to 20 minutes of doing all the exercises in the session. Habituation exercises are customized to address the specific provoking stimulus of each patient, and the patient is educated that the movements need to be performed fast enough to provoke the symptoms to ultimately reduce the dizziness. The therapist should start conservatively, so that the patient can carry out his or her daily activities. The exercises need to be done with eyes open and at a speed that provokes mild to moderate symptoms. Rest breaks between each set allow for the symptoms to return to baseline, and ideally, the symptoms after subsequent sets should be less than the first set. If headaches or nausea occur, exercises should be modified (Appendix 30-3).

■ Table 30-2 MOTION SENSITIVITY QUOTIENT (MSQ) (SMITH-WHEELOCK ET AL, 1991)[9]*

Baseline Symptoms	Intensity	Duration	Score
1. Sitting to supine			
2. Supine to left side			
3. Supine to right side			
4. Supine to sitting			
5. Left Hallpike-Dix			
6. Return to sit from left Hallpike-Dix			
7. Right Hallpike-Dix			
8. Return to sit from right Hallpike-Dix			
9. Sitting, head tipped to left knee			
10. Head up from left knee			
11. Sitting, head tipped to right knee			
12. Head up from right knee			
13. Sitting, turn head horizontally 5 times			
14. Sitting, move head vertically 5 times			
15. Standing, turn 180 degrees to the right			
16. Standing, turn 180 degrees to the left			

*MSQ = {(Total score) × (# of positions with symptoms)} + 20.48 MSQ score 0–10 = mild; 11–30 = moderate; 31–100 = severe. Duration: 5–10 sec = 1 point; 11–30 sec = 2 points; greater than 30 sec = 3 points.

If the patient is sensitive to stimulating environments as determined by the DHI, SVQ, or VVAS, the therapist can systematically introduce situations that provoke the symptoms.

Visual Vertigo and Habituation Exercises

Visual vertigo has been shown to respond to habituation exercises using a similar paradigm to habituation of self-movements.[15] Habituation of visual vertigo involves brief doses of visual stimulation for a specific amount of time to provoke mild to moderate symptoms (e.g., 1 minute of an action video), followed by a rest period to allow the individual to return to baseline before introducing another epoch of visual stimulation. Patients can also begin viewing a small screen and progress to a larger screen. Pavlou suggested that optokinetic stimulation is beneficial but needs to be gradual, progressive, and structured.[15] Suggestions for interventions include high- and low-tech options. If the clinic has a CDP, the therapist can use it for training by gradually changing the parameters of the surround movements (Fig. 30.4). Other high-tech systems include virtual reality systems such as the "grocery store"[49] and Balance Rehabilitation Unit (BRU) system.[42] The grocery store model consists of a simulation of walking through an aisle of a supermarket searching for specific items. During assessment and intervention on the BRU system, the patient wears goggles and watches virtual scenes with moving objects, tunnels, and panoramic views while the patient's postural sway is measured on a platform. Low-tech methods that have been used include disco balls, twirling umbrellas, moving stripes or busy tablecloths on the wall, and action videos (Fig. 30.5). Therapists can either provide a list of appropriate videos from YouTube (Appendix 30-4) or design a customized program of videos of appropriate levels of difficulty.

Patients *with migraines* may be sensitive to both self-motion and movements in the environment. These individuals need to begin their HEP at a slow pace so that the movements do not provoke headaches or nausea. Walking short distances outdoors or balance training with slow head movements may be all that can be tolerated initially. Migraineurs need to be educated that habituation exercises should not be performed during migraine spells. Habituation exercises, in addition to medical management for migraines and stress reduction, are essential to optimal outcomes (see Chapter 15).

Many patients with *cerebellar degeneration* experience dizziness as a result of oculomotor impairments including abnormal smooth pursuit, VOR cancellation deficits, and gaze-evoked nystagmus.[31,50] When these individuals turn their heads, the environment may become distorted, causing dizziness, imbalance, and unsteady gait.[31] Habituation exercises including head turns while scanning the environment can decrease motion

Figure 30.4 Patient on Equitest. Patients who have complaints of walking into busy environments may benefit from standing in CDP while the platform and surround are moving randomly at various speeds while searching for numbers on the surround.

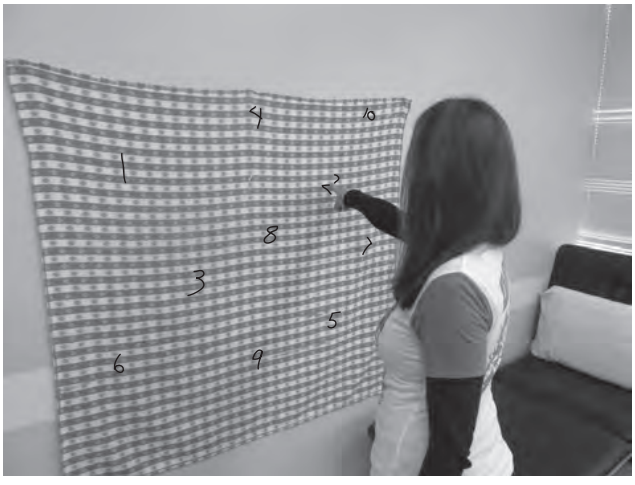

Figure 30.5 Therapists can use low-tech methods to simulate busy environments such as a tablecloth marked with numbers as a treatment in the clinic or part of the home program. The patient stands 6 feet away, walks forward and points to a number, backs up and searches for the next, and continues until c/o symptoms are mild to moderate.

sensitivity.[33,51] These patients can be trained to find the "null" position of the eyes where nystagmus is the least, as well as to turn their head and eyes at the same time to look peripherally while preventing gaze-evoked nystagmus. Following a target with head and eyes together horizontally at a speed at which the target remains in focus (simulating the VOR cancellation testing) can be used as a habituation exercise to decrease dizziness.[11] Cohen demonstrated that patients' gait improved by focusing the intervention on balance exercises and habituation exercises to reduce dizziness.[52]

Based on our experience, habituation exercises can also help older adults with *disuse disequilibrium*, white matter disease, or a history of BPPV who are hesitant to look upward or bend over for fear of provoking dizziness. When this behavior is observed during initial assessment, the therapist needs to include habituation exercises with head and body movements in the home exercise program.

We inform patients with motion sensitivity that they may experience more symptoms with the HEP the first week, because they are doing movements that they have been avoiding. Smith-Wheelock suggested that habituation exercises should be performed only 2 times a day initially to avoid overloading the compensatory system and make symptoms worse.[9] Subjective and objective outcome measures should be repeated in 1 month because motion sensitivity has been shown to significantly improve within 4 to 6 weeks of initiation of habituation exercises.[51] Smith-Wheelock suggested that a decrease in sensitivity and duration of symptoms can occur as quickly as 2 weeks, but may take as long as 6 months.[9] In our experience, patients who may not respond to habituation exercises include those individuals who experience constant vague symptoms of dizziness that do not change with self or environmental movement. Individuals who do not adhere to the prescribed HEP or have psychological causes for their dizziness that have not been addressed may also not improve. For habituation exercises to be the most effective, the therapist needs to perform a thorough evaluation, design an individualized treatment plan based on the findings, and educate the patient of the purpose of the intervention. When there is no improvement or symptoms worsen even with modifications, the patient should discontinue the exercises and return to the referring physician.

Gaze Stabilization Exercises

Two studies have evaluated the use of vestibular gaze stability exercises for patients with neurological (cerebellar) involvement and have shown improved dynamic visual acuity (DVA) and functional mobility.[6,11] However, few studies have looked at the use of gaze stability exercises for patients without neurological pathology.

Gaze Stabilization and Non-vestibular Dizziness—Supportive Evidence

In a randomized controlled pilot study of older adults with non-vestibular dizziness (i.e., normal vestibular function based on clinical exam and caloric testing), subjects in the gaze stability group (n = 20) performed vestibular gaze stability exercises, and subjects in the placebo group (n = 17) performed placebo eye exercises (saccadic eye movement against a plain background).[7] Both groups performed gait and balance exercises based on impairments identified at initial evaluation. Both groups improved on measures of symptoms, balance confidence, gait speed, and sensory integration. The only significant interaction ($p = 0.026$) was for fall risk as measured by dynamic gait index (DGI) with 90% of the gaze stability group demonstrating clinically significant improvement in DGI scores versus 50% of the placebo group. These studies suggest that incorporating vestibular gaze stability exercises in the rehabilitation of individuals with non-vestibular dizziness is beneficial.

Dual-Task Training

Many patients with non-vestibular dizziness, especially older adults, have difficulty walking while performing secondary tasks.[53] Woollacott and Shumway-Cook suggest that the problem with dual-task ability in older adults can be the result of inability to shift attention, a reduction in attentional capacity, impairments in the postural control system, or a combination of these factors.[53] In fact, an impaired ability to allocate attentional resources to balance during dual-task situations is a powerful predictor of falls.[20] Outcome measures such as the modified Timed Up and Go (mTUG) or Walk While Talk (WWT) test can be used to identify difficulty walking and dividing attention (see Table 30-1).

Single-Task to Dual-Task Balance Training—Supportive Evidence

Few studies have compared single-task to dual-task balance training. One such study demonstrated that only participants trained under dual-task conditions showed improvements for gait speed tested under dual-task conditions.[54] In contrast, a retrospective study demonstrated that standard balance rehabilitation with no specific dual-task training improved gait under dual-task

conditions.[55] In this retrospective study, 49 older adults without vestibular or neurological deficits underwent an average of 3.7 physical therapy visits for balance rehabilitation. The patients showed significant improvement in several outcome measures, in addition to a significant improvement in dual-task ability, as measured by the TUG cognitive condition. The data suggest that improvements in underlying balance and gait impairments resulted in increased automaticity allowing more attentional resources to be allocated to the cognitive task.[55]

Further research is needed to clarify the role of dual-task training in balance rehabilitation. However, in our clinical practice we routinely add cognitive dual-tasks to specific balance and gait exercises for non-vestibular patients who are at fall risk. Typically, we incorporate cognitive activities into a walking task; for example, we add a mental task when a patient can walk with horizontal or vertical head turns with only minimal unsteadiness. The tasks can begin very simply, such as counting backwards by 1s, then progress to more difficult tasks, such as counting backwards by 3s, naming flowers or listing boys' or girls' names.

Integration of Exercise into Daily Activities

Because the primary objective of therapeutic interventions is to assist patients to return to the physical activities they enjoy, incorporating these specific interests into the treatment plan is essential. If the initial evaluation determines that a golfer experiences dizziness with head turns or imbalance with weight shifts, practicing a golf club swing is a therapeutic activity that can motivate and prepare the individual to golf without loss of balance. Dancing,[56] water exercise,[57] Wii Fit,[58] Tai Chi,[59-61] Yoga,[62] and Pilates[63] have also been shown to not only improve balance and strength, but also help motivate individuals to exercise.

In our experience, patients who are physically active maintain gains achieved through rehabilitation versus those who are sedentary. Patients are not only instructed to continue their home program and walking routine after discharge from physical therapy, but are also encouraged to return to the activities that they have enjoyed in the past or to learn new activities that challenge balance and will help to maintain gains made in formal therapy. In our clinic, patients are scheduled for a yearly follow-up visit but instructed to return sooner if postural stability or function declines.

Evidence shows that patients often do not continue their prescribed exercises after discharge from rehabilitation; thus, helping the patient to identify recreational activities that can be enjoyed on a long-term basis is important.[64] In follow-up interviews, Cass et al reported that less than 10% of their subjects were performing their prescribed home exercises for reasons including time restraints, boredom, and change of health status.[64]

Time Course for Recovery

There is no magic number of visits for therapy for patients with non-vestibular dizziness. At our clinic, patients with non-vestibular dizziness are scheduled for four weekly visits, with a customized HEP given on the first visit. Reassessment is performed on the fourth visit, and if the goals are achieved and fall risk is low, then the patient is discharged with a modified home program including a walking program or physical activity of his or her choice. If the patient is progressing toward his or her goals, but still at fall risk, more visits are added. If the status unexpectedly plateaus or worsens, the patient needs to return to the referring physician to determine whether psychological or undiagnosed medical causes are contributing to the lack of improvement. If the patient does not return to a level of independence or is at fall risk based on outcome measures, fall prevention and/or training with the appropriate assistive device needs to be performed. In all situations, patients return to physical therapy 6 months later to address any changes in status.

Many factors can affect the time course of recovery to a level at which the patients can be discharged from skilled PT and continue to perform their prescribed HEP and return to their pre-morbid activities.

Factors that may optimize recovery include:

1. *Adherence to prescribed home program including walking, and returning to other physical activities including shopping, chores, and hobbies*. Patients are educated in the purpose of each exercise and are given easy-to-understand handouts in addition to exercise logs, which they return weekly. Mohler showed that use of calendars greatly improved compliance.[65] Hall and colleagues demonstrated that exercise adherence and cognitive status predicted rehabilitation outcomes in older adults with non-vestibular dizziness.[7]
2. *Availability of a dedicated support system*. Spouses or family members who can assist with the home program and motivate the patient are important. In our experience,

individuals who live alone may not be able to do the challenging balance exercises safely, may be fearful of doing the exercises alone, or may not be disciplined enough to do them alone. Also, patients who are themselves caregivers for spouses or parents often do not take the time needed to perform the home program as prescribed.

3. *Absence of neurological pathology*. Older patients with disuse disequilibrium or fear of fall without neurological pathology typically can decrease fall risk and reach goals within 1 month of weekly visits (average of 3.7 visits[7]). Those with cerebellar ataxia[8] and central vestibular[6] require more visits to reach goals (range: 5 to 12 visits).

CASE STUDY 30-1

Patient is a 56-year-old female with a history of migraines who suffered a severe dizzy spell followed by a panic attack 2 years before referral to our clinic. Her physicians were unable to find a cause for the dizzy spell, and she never received a diagnosis. During these 2 years, she avoided moving her head or walking for exercise for fear of provoking another spell. She was diagnosed by the neurologist in our clinic with chronic imbalance from inactivity and motion sensitivity, exacerbated by anxiety. She was placed on a migraine diet (see Chapter 15), a low-dose, anti-anxiety medication, and was referred for gait and balance physical therapy. She participated in four weekly physical therapy sessions and was provided with a daily HEP for gait and habituation exercises. Part of the HEP included walking in her neighborhood with friends for both conditioning and dual-task practice (i.e., looking around and conversing with friends). Habituation exercises were prescribed based on the initial MSQ assessment. She practiced head rotations both vertically and horizontally beginning with 2 sets of 3 repetitions and progressed to 5 repetitions per set. Because she was unable to tolerate the moving, visual surround of CDP and reported avoiding busy environments, she also was exposed to visual stimulus in short doses that exacerbated her visual vertigo. Initially, she practiced finding numbers on a checked tablecloth taped to the wall and progressed to searching for numbers that were taped to the surround of the CDP while both the platform and surround were moving. To address her avoidance of turning her neck during 180-degree turns (she turned "en bloc"), specific turning exercises to separate head and body motion and gait with head turns were also performed. The comparison of her pre-therapy assessment and her post-therapy assessment shows a clear improvement across subjective complaints and physical function (Table 30-3).

Comment

This individual was not given a medical diagnosis for her severe spell of vertigo until she was seen in our clinic. In those 2 years after her first spell, she changed her lifestyle and became sedentary. The combination of being given a diagnosis, understanding the diagnosis of migraine equivalent spells and anxiety, medical management, and rehabilitation helped her achieve optimal recovery and return to her previous lifestyle.

■ Table 30-3 **COMPARISON OF ASSESSMENT BEFORE AND AFTER THERAPY**

Outcome Measures	Initial Evaluation	Discharge Evaluation
ABC (%)	54	90
MSQ	21.1/100 (moderate)	1.4/100 (mild)
DHI	70/100 (severe)	24/100 (mild)
Gait speed (m/sec)	0.72 m/sec (abnl ≤ 1.09 m/sec)	1.25 m/sec
DGI	18/24 (≤19/24 indicates fall risk)	22/24

CASE STUDY 30-2

Patient is a 75-year-old male with a recent diagnosis of idiopathic, late-onset cerebellar ataxia (ILOCA) with a gradual decline of gait stability for 6 years and increased falls. Neurological testing showed gaze-evoked nystagmus, abnormal smooth pursuit, and VOR cancellation, along with peripheral neuropathy. Videonystagmography (VNG) testing showed normal vestibular function. He was seen for five visits for gait and balance PT and given a HEP. Balance training included eyes open and closed on firm and foam surfaces with altered base of support, step-overs, sit to stands to strengthen legs, wall leans to improve hip strategies, and gait training including use of a cane. In subsequent visits, he was also prescribed VOR cancellation exercises, stepping up and down on a 5-inch platform, gait with head turns, and modified SLS using a book.

Comment

Although still at fall risk based on DGI, this patient made a clinically meaningful change in the DGI (≥3 points). Tinetti improved to low risk for falls, and ABC was still low but had a clinically meaningful change (Table 30-4). Because he was still at risk for falls based on DGI and had low balance confidence, continued PT was recommended. He chose to be discharged but agreed to continue his HEP and return if function declined. Because he has a degenerative disease, he was instructed to be active and perform the prescribed HEP to slow the natural progression of the disease.

■ Table 30-4 **PRE- AND POST-INTERVENTION ASSESSMENT**

Outcome Measure	Initial Evaluation	Discharge Evaluation
ABC (%)	32	58
mCTSIB	Firm EO = 30 sec* Firm EC = 22 sec Foam EO = 0 sec Foam EC = 0 sec	Firm EO = 30 sec Firm EC = 30 sec Foam EO = 23 sec Foam EC = 4 sec
TINETTI	22/37 (high fall risk)	35/37 (low fall risk)
Gait speed (m/sec)	0.80 (abnl ≤ 0.94 m/sec)	0.96 m/sec
DGI	10/24 ≤19/24 indicates fall risk	19/24

*30 seconds is normal – average of 3 trials

CASE STUDY 30-3

Patient is a 74-year-old female who became very dizzy with nausea and vomiting 10 months ago. She described her initial dizziness as "out of balance in head," but currently feels "off balance." She reports a fall 2 months ago while walking outside. Her medical history was significant for a stroke in 2000 with late effect vertigo, depression/anxiety, and intermittent diplopia (diagnosed by a neuro-ophthalmologist as episodic, decompensated phoria). Her clinical exam showed normal oculomotor exam, negative head thrust test bilaterally, negative Dix-Hallpike, and normal neurological exam. VNG testing showed normal vestibular function. The neurologist diagnosed her with disuse disequilibrium/fear of falling.

CASE STUDY 30-3

Treatment emphasized (1) vestibular adaptation and substitution exercises with the goal to habituate head movement–induced dizziness and reduce fall risk; (2) balance retraining on altered surfaces with altered visual input; (3) gait training on various surfaces with head turns, change in speed, and stepping over obstacles; and (4) patient education concerning home program, safety issues, and role of physical activity. As the patient progressed, the vestibular exercises were performed faster, balance training incorporated reduced base of support and head movements, and gait training added cognitive dual-tasks (counting backwards by 1s).

Comment

Although this patient had normal vestibular function testing, vestibular gaze stabilization exercises were included as a component of therapy. There is evidence to support this approach in older adults with non-vestibular dizziness.[7] At discharge, the patient's goals were met, and she was at low risk for falls (Table 30-5). She still had head motion–induced dizziness, and for this reason she was encouraged to continue her HEP including gaze stabilization exercises and to follow up in 6 months.

■ Table 30-5 **PRE- AND POST-INTERVENTION ASSESSMENT**

Outcome Measure	Initial Evaluation	Discharge Evaluation
Disability	3/5 (symptoms interfere with both usual and outside activities)	1/5 (symptoms are bothersome)
ABC (%)	56	92
Dizziness VAS	baseline: 1.1/10 after 1 minute head turns: 6.6/10	baseline: 0.4/10 after 1 minute head turns: 3.2/10
SOT (/100)	composite score = 34 (nl ≥ 63.8) • Increased sway conditions 4-6 • 6 falls conditions 5, 6	composite score = 64 • 2 falls conditions 5, 6
Gait speed (m/sec)	0.74 m/sec (abnl ≤ 0.85 m/sec)	0.83 m/sec
DGI	15/24 (≤19/24 indicates fall risk)	21/24

Summary

Patients with non-vestibular dizziness are a challenge to clinicians because of the wide range of impairments and symptoms patients have. These patients can make excellent gains with physical therapy as long as the assessment includes a thorough history, outcome measures to determine fall risk, and questionnaires for balance confidence and dizziness. The plan of care should include a safe, but challenging, daily home exercise program addressing the problems. The clinicians also need to encourage these individuals to stay active by participating in recreational activities after discharge from skilled physical therapy.

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Appendix 30-1 Situational Vertigo Questionnaire

Situational Vertigo Questionnaire

Vertigo is the medical term used for symptoms which patients often describe as feelings of unusual disorientation, dizziness, giddiness, lightheadedness, or unsteadiness.

Please circle a number to indicate the degree to which each of the situations listed below causes feelings of vertigo, or makes your vertigo worse. If you have never been in one of the situations, then for that item ring "N.T ." for "Not Tried."

The categories are:	0	1	2	3	4	N.T.
	Not at all	Very slightly	Somewhat a lot	Quite	Very much	Not tried
Riding as a passenger in a car on straight, flat roads						
	0	1	2	3	4	N.T.
	Not at all	Very slightly	Somewhat a lot	Quite	Very much	Not tried
Riding as a passenger in a car on winding or bumpy roads						
	0	1	2	3	4	N.T.
	Not at all	Very slightly	Somewhat a lot	Quite	Very much	Not tried
Walking down a supermarket aisle						
	0	1	2	3	4	N.T.
	Not at all	Very slightly	Somewhat a lot	Quite	Very much	Not tried
Standing in a lift while it stops						
	0	1	2	3	4	N.T.
	Not at all	Very slightly	Somewhat a lot	Quite	Very much	Not tried
Standing in a lift* while it moves at a steady speed						
	0	1	2	3	4	N.T.
	Not at all	Very slightly	Somewhat a lot	Quite	Very much	Not tried
Riding in a car at a steady speed						
	0	1	2	3	4	N.T.
	Not at all	Very slightly	Somewhat a lot	Quite	Very much	Not tried

(Pavlou, 2004)15
*A lift is an elevator.

Starting or stopping in a car

0	1	2	3	4	N.T.
Not at all	Very slightly	Somewhat a lot	Quite	Very much	Not tried

Standing in the middle of a wide open space (e.g., large field or square)

0	1	2	3	4	N.T.
Not at all	Very slightly	Somewhat a lot	Quite	Very much	Not tried

Sitting on a bus

0	1	2	3	4	N.T.
Not at all	Very slightly	Somewhat a lot	Quite	Very much	Not tried

Standing on a bus

0	1	2	3	4	N.T.
Not at all	Very slightly	Somewhat a lot	Quite	Very much	Not tried

Heights

0	1	2	3	4	N.T.
Not at all	Very slightly	Somewhat a lot	Quite	Very much	Not tried

Watching moving scenes on the TV or at the cinema

0	1	2	3	4	N.T.
Not at all	Very slightly	Somewhat a lot	Quite	Very much	Not tried

Traveling on escalators

0	1	2	3	4	N.T.
Not at all	Very slightly	Somewhat a lot	Quite	Very much	Not tried

Looking at striped or moving surfaces (e.g., curtains, Venetian blinds, flowing water)

0	1	2	3	4	N.T.
Not at all	Very slightly	Somewhat a lot	Quite	Very much	Not tried

Looking at a scrolling computer screen

0	1	2	3	4	N.T.
Not at all	Very slightly	Somewhat a lot	Quite	Very much	Not tried

Appendix 30-2 Visual Vertigo Analog Scale

Indicate the amount of dizziness you experience in the following situations by marking off the scales below. 0 represents no dizziness, and 10 represents the most dizziness.

Walking through a supermarket aisle

0 10

Being a passenger in a car

0 10

Being under fluorescent lights

0 10

Watching traffic at a busy intersection

0 10

Walking through a shopping mall

0 10

Going down an escalator

0 10

Watching a movie at the movie theatre

0 10

Walking over a patterned floor

0 10

Watching action television

0 10

(Adapted from Longridge et al, 2002.)[43]

Appendix 30-3
Sample Home Exercise Program

These sample home programs are for a patient with motion sensitivity.

HABITUATION EXERCISES Do _____daily

1. In a sitting position:
_____Bend over and sit up _____times in a row
_____Wait until symptoms resolve and repeat one more set of _____

2. In a sitting position:
_____Turn head side to side _____times in a row
_____Wait until symptoms resolve and repeat one more set of _____

3. In a sitting position:
_____Move head up and down _____times in a row
_____Wait until symptoms resolve and repeat one more set of _____

4. Stand in a corner:
_____ Turn a FULL ½ turn to the right _____times,
waiting for symptoms to resolve each time
_____Turn a FULL ½ turn to the left _____times,
waiting for symptoms to resolve each time

5. Practice walking in hallway _____times
_____Move head side to side slowly, focusing on the walls each time
_____Move head up and down slowly, focusing on the ceiling and floor each time
_____Move head diagonally slowly, crossing in the center each time

6. Practice tossing a ball hand to hand following it with head and eyes _____

7. Practice handing ball to a person behind you to right and left while walking.
Make sure you rotate your head and body and watch the ball with your eyes

8. Practice rolling in bed
_____roll to the right_____ times, wait for symptoms to resolve
_____roll to the left_____ times, wait for symptoms to resolve

9. Other_____

Appendix 30-4 Suggested Videos to Use in Clinic and Home Program

This sample program is for patients with visual vertigo based on VVAS or SVQ. These videos will make you dizzy and uncomfortable—make sure your symptoms are only mild when you watch these videos. Start with the less-stimulating videos first for only *30 seconds or less*, let the symptoms return to baseline before watching another *30 seconds or less*. Build up from there. Everyone is different, so do not expose yourself for too long or it will not be beneficial. Do only as prescribed by your PT. Sessions should last no more than 10 minutes with symptoms resolving within 20 minutes after stopping. If the symptoms provoked are moderate to severe, do not continue.

EASY

Driving in Manhattan New York: 5:10

http://www.youtube.com/watch?v=Lv0OgDmwC3Q&feature=related

Cruising Lake Erie via Motorcycle 3:06

http://www.youtube.com/watch?v=h2Jz6IdS-iM&feature=related

An evening stroll in the Paris Opera quarter 22:33

http://www.youtube.com/watch?v=i6bDAWqf2RY&feature=related

Ice Skating in High Sierra 6:11

http://www.youtube.com/watch?v=-EG7kLjpzsM

Rue Saint Severin and Rue de la Harpe 4:50

http://www.youtube.com/watch?v=PD_RikPVmyY&feature=related

MEDIUM

An Afternoon Stroll through the Latin Quarter: 14:54

http://www.youtube.com/watch?v=csJupEA1lCI

Just Walking Down The Street In Akihabara Japan: 4:40

http://www.youtube.com/watch?v=5tl-mo2Ob0o

Walk through Harry Potter World: 3:11

http://www.youtube.com/watch?v=-434p3l-2EA

Trail Running (start at 1:00 mark; some pauses built in) 7:16

http://www.youtube.com/watch?v=ENj2yXDMGng

Skiing down mountain in Italy: 12:58

http://www.youtube.com/watch?v=fPZNahPtUD4

Driving: I-95 New Jersey & Henry Hudson Pkwy New York City 5:31

http://www.youtube.com/watch?v=-eQHl9T0W8U&feature=related

Driving: Fort Pitt Tunnel Pittsburgh Pa 1:40

http://www.youtube.com/watch?v=C-Ui0gxYVCg&feature=related

A stroll through the casino at the Monte Carlo hotel in Las Vegas 4:33

http://www.youtube.com/watch?v=AHt_iTuzQNY

Women's Hockey Practice 3:53

http://www.youtube.com/watch?v=ZXXwIiHdrQs

Men's hockey w/ helmet cam 7:40

http://www.youtube.com/watch?v=DqjNyy-ONCU&feature=related

DIFFICULT

Walking through lobby of Venetian hotel Las Vegas 0:54

http://www.youtube.com/watch?v=4imxlc1xEO0&feature=related

Walking through Winn Dixie (grocery store): 6:53

http://www.youtube.com/watch?v=2ojMWHCJKhc

A walk through Times Square: 6:52

http://www.youtube.com/watch?v=5nRVIPyHGR8

Maui: The Road to Hana Timelapse 7:19

http://www.youtube.com/watch?v=5ER2K-ERLlk&feature=fvwrel

Walking through: New Cosmopolitan Casino Hotel Las Vegas 7:24

http://www.youtube.com/watch?v=NsI0vSPKBxs&feature=related

CHAPTER 31

Physical Therapy Management of Cervicogenic Dizziness

Richard A. Clendaniel, PT, PhD
Rob Landel, PT, DPT, OCS, CSCS, FAPTA

Cervicogenic dizziness is a controversial subject at best. The term tends to be used to describe a variety of entities, some of which are theoretically more likely than others, including cervical ataxia, cervical nystagmus, and cervical vertigo. Because *vertigo* is defined as the illusion of movement (rotation, tilt, or linear displacement) and is therefore restrictive, the term *cervicogenic dizziness*, rather than the older term "cervical vertigo," is used in this chapter to refer to symptoms of dizziness (including vertigo, disequilibrium, and lightheadedness) arising from the cervical spine.

Several different processes have been hypothesized to be the cause of cervicogenic dizziness. These pathophysiological mechanisms include irritation of the sympathetic vertebral plexus, vertebrobasilar insufficiency (VBI), and altered proprioceptive afferent signals from the upper cervical spine. This latter potential cause of dizziness is of particular interest because of the large number of patients with either whiplash injuries or neck pain that are seen by physical therapists. This particular cause of dizziness is also perhaps the most controversial.

One of the major problems in identifying patients with cervicogenic dizziness is the lack of a concrete test that is sensitive and specific to this entity. From a therapeutic standpoint, however, the controversy surrounding cervicogenic dizziness may be academic. If an individual presents with cervical symptoms and dizziness, the holistic approach would be to treat the cervical problem as well as the dizziness. The aim of this chapter is to review the anatomical and physiological bases for cervicogenic dizziness, to summarize the scientific findings related to cervicogenic dizziness, to address the clinical methods of assessing cervicogenic dizziness, and to discuss possible management strategies for this condition.

Proposed Etiologies

Posterior Cervical Sympathetic Syndrome

Barré[1] suggested that cervical problems could irritate the sympathetic vertebral plexus, leading to constriction of the internal auditory artery and decreased perfusion of the labyrinth, which would induce vertigo. There is little objective evidence, however, to support this hypothesis. In addition, the intracranial circulation is controlled independently of the cervical sympathetic system. Therefore, it is difficult to see how a cervical injury could lead to restricted blood flow to the inner ear.

Vertebrobasilar Insufficiency

Another possible cause of dizziness arising from the cervical spine is occlusion of the vertebral arteries by osteoarthritic spurs[2] or occipitoatlantal instability.[3] VBI and vertebrobasilar ischemia can arise from a variety of causes, including embolism, large artery atherosclerosis, small artery disease, and arterial dissection, and can occur

at numerous sites along the course of the vertebral and basilar arteries.[4] There are two sites where physical occlusion, or in the extreme, dissection of the vertebral artery can occur. One is in the upper cervical spine, after the vertebral artery exits the transverse foramen and courses around the mobile upper cervical vertebrae. The other potential site for occlusion is in the initial segment of the vertebral artery before it enters the transverse foramen.[5] In theory, the vertebral arteries can be compressed during cervical rotation or extension, as occurs when a person reaches for an object on an overhead shelf, turns the head while backing up a vehicle, or undergoes cervical spine manipulations. In normal individuals, the carotid arteries provide sufficient collateral circulation to prevent symptoms.[6] In individuals with atherosclerotic vascular disease, the cerebrovascular circulation may be compromised to the extent that compression of the vertebral arteries could lead to VBI.

Although there is an anatomical substrate that could link cervicogenic dizziness with VBI, it is not clear that the symptoms of VBI match those of cervicogenic dizziness. The symptoms associated with VBI can be quite diverse. In a study of 65 patients diagnosed with VBI, Williams and Wilson[7] found that vertigo was the initial symptom in 48% of the cases. Vertigo caused by VBI is generally abrupt in onset, is of short duration (several minutes), and may be associated with nausea and vomiting.[8] These symptoms are similar to those of someone with a vestibular deficit, but the vertigo in patients with VBI is generally associated with other symptoms related to ischemia of areas supplied by the posterior circulation—typically including visual hallucinations, loss of vision, ataxia, drop attacks, numbness or weakness affecting both sides of the body, visceral sensations, visual field defects, diplopia, and headaches.[7,9]

Vertigo may be an isolated initial symptom but is typically intermixed with the other symptoms of VBI.[10] Of the more than 400 patients with vertebrobasilar ischemia whose data are entered in the New England Medical Center–Posterior Circulation Registry, fewer than 1% presented with a single sign or symptom, and the initial symptom was not necessarily vertigo or dizziness.[4,11] Individuals with vertebrobasilar ischemia secondary to stenosis or occlusion of the vertebral arteries typically experience transient ischemic attacks (TIAs) with symptoms of dizziness, impaired vision, and loss of balance, which are consistent with ischemia in the vestibulocerebellum and medulla.[12] In cases of dissection of the vertebral artery, the primary symptom is posterior cervical pain that may radiate to the shoulder region. In addition, these individuals typically experience occipital headaches, dizziness, diplopia, and lateral medullary and cerebellar signs including unsteadiness and ataxia.[5,13,14]

Given the typical mixed signs and symptoms of VBI, the presence of episodic, isolated bouts of vertigo in the absence of associated symptoms for more than 3 weeks is believed to be rarely caused by vertebrobasilar disease.[9] In addition, drop attacks, which have been attributed to transient ischemia of the posterior circulation, were never found in isolation in the New England Medical Center–Posterior Circulation Registry cases.[4,11] In summary, although it is unlikely that dizziness as the sole symptom arises from vertebrobasilar ischemia, the possibility of VBI should at least be considered on the basis of the patient's history and symptoms. Clinical tests for VBI, and their usefulness, are described in detail later in this chapter.

Altered Proprioceptive Signals

The mechanism by which cervical pain or dysfunction could lead to symptoms of dizziness has not been identified. One hypothesis is that inflammation or irritation of the cervical roots or facet joints would lead to a mismatch among vestibular, visual, and cervical inputs. This multisensory mismatch would lead to the symptoms attributed to cervicogenic dizziness, and the symptoms would be most apparent during head movements (Fig. 31.1). In theory, once the central nervous system (CNS) has adapted

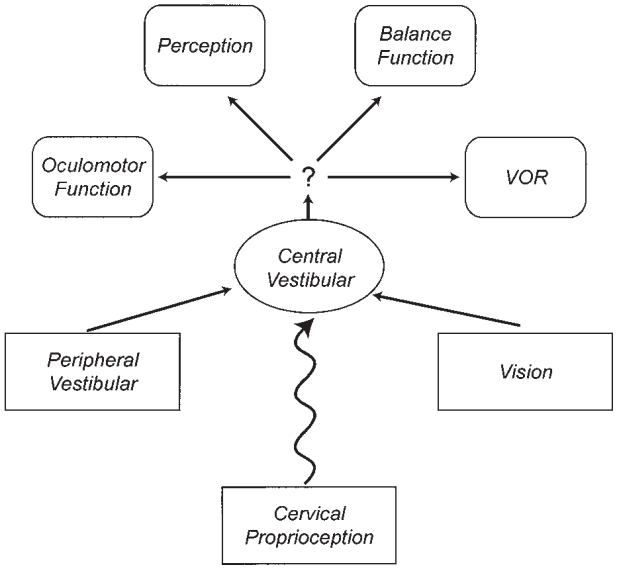

Figure 31.1 Schematic diagram of the multisensory mismatch hypothesis. Peripheral inputs (vestibular, cervical, and visual) converge on central vestibular structures and affect oculomotor function, balance function, vestibulo-ocular reflex (VOR), and perceptions of dizziness. Cervical dysfunction (represented by the curved line) would lead to a mismatch among vestibular, visual, and cervical inputs. This multisensory mismatch would lead to the symptoms attributed to cervical dizziness.

to the altered somatosensory inputs (just as the system is capable of adapting to altered vestibular inputs), the symptoms of cervicogenic dizziness would abate even though the underlying dysfunction remained. Because the symptoms attributed to cervicogenic dizziness can continue for extended periods, this explanation, although enticing, remains controversial. The following sections review the literature that addresses the role of altered cervical proprioceptive signals in the generation of signs and symptoms of dizziness.

Anatomy and Physiology

There is a convergence of cervical proprioceptive and vestibular information throughout the spinal cord, brainstem, cerebral cortex, and cerebellum.[15] A detailed review of the anatomical evidence is beyond the scope of this chapter; however, a brief review of the pertinent physiological evidence, which suggests that cervical inputs may play a role in dizziness, is warranted. Cervical proprioceptive signals important for postural neck reflexes arise from the joint and tendon receptors located in the deep structures of the upper cervical spine.[16] McCouch and colleagues[16] demonstrated that the tonic neck reflexes in cats were mediated through the joint receptors rather than the cervical musculature. The role of the deep paravertebral muscle spindles in this region is not clear and may be species dependent.[17-22]

In addition to its influence on postural neck reflexes, upper cervical spine proprioception is thought to be responsible for the generation of the cervico-ocular reflex (COR), which, when present, complements the vestibulo-ocular reflex (VOR) at lower frequencies of movement.[23] Rubin and associates[24] recorded from neurons in the vestibular nucleus of cats that responded to both vestibular stimulation (whole-body rotation) and movements of the trunk on a fixed head. Further support for the hypothesis that cervical proprioception can influence vestibular function comes from the work of Hikosaka and Maeda.[25] These investigators reported that, in the cat, vestibular excitation of the abducens nerve was inhibited by contralateral and facilitated by ipsilateral electrical stimulation of the cervical dorsal roots or facet joints. In addition, they reported that these effects were seen with stimulations at C2 and C3 but not with stimulations at C5 or lower.

These studies indicate that cervical proprioception, particularly from the upper cervical spine, may play a role in postural control and may have an effect on vestibular function. It is important to remember that the studies were performed in anesthetized or decerebrate animal preparations and that responses in a wake, normally functioning humans may be entirely different. In light of the physiological findings and the anatomical convergence of cervical and vestibular inputs, the following sections explore whether cervical proprioceptive signals can contribute to symptoms of vertigo or disequilibrium and influence vestibular system function or postural stability.

Findings after Cervical Spine Lesions

A number of studies in animals and humans have investigated oculomotor and balance changes following either experimentally induced lesions in the upper cervical spine or cervical injuries. With regard to the cervical injuries, the primary research focus has been on whiplash associated disorders (WADs), which cause a diverse set of symptoms (including headache, dizziness, paresthesia, anesthesia, and cervical, thoracic, lumbar, and upper extremity pain) following a flexion-extension cervical injury.[26] Although most individuals recover from WAD, the symptoms, which can be severe, persist in a significant proportion of these individuals. On the basis of the findings of various studies, cervical injury seems to have little effect on the oculomotor and vestibular system but may lead to disturbances in postural control.

Oculomotor Findings

Cervical proprioceptive ablation has been performed by both sectioning of the dorsal roots and injection of local anesthetics into the neck. Igarashi and coworkers[27] reported on the oculomotor effects seen after local anesthetic injections, or unilateral transection of the C1 and C2 dorsal roots in squirrel monkeys. Lidocaine (Xylocaine) (1 mL of 1% solution) without epinephrine was injected into the deep neck regions unilaterally in the experimental animals, and into either the superficial posterior neck region or abdominal wall in control subjects. The animals were placed in the dark, and their eye movements were recorded. There was no evidence of spontaneous nystagmus in either the experimental or control groups after injection. Optokinetic nystagmus was used as a measure of vestibular function in this study. Optokinetic nystagmus, the reflexive eye movement response to a moving full-field visual stimulus, is a visually mediated response that travels through the accessory optic system, a subcortical neural pathway, to the vestibular nuclei. Here the signals are processed in conjunction with the vestibular afferent input to generate compensatory eye movements. Optokinetic nystagmus was measured pre-injection and postinjection in 10 animals by means of a rotating optokinetic stimulus (1 deg/sec^2 acceleration up to 200 deg/sec). Igarashi and coworkers[27] reported bidirectional declines in both the slow-component and fast-component eye velocities

of the optokinetic nystagmus in the experimental subjects. The control groups also demonstrated declines in the slow-component eye velocity postinjection. The investigators stated that there was a statistically significant difference in slow-component eye velocity changes between the experimental and control groups, and they infer from this finding that cervical proprioception contributes to oculomotor behavior.

Two issues raise questions about their conclusion. First, the investigators reported a statistically significant difference using a *P* value equal to 0.066, which exceeds the generally accepted probability level for statistical significance. Most investigators would not reject the null hypothesis that there is no difference between groups with this *P* value. Second, the declines in velocity of the fast component of the nystagmus in the experimental group and in the velocity of the slow component of the nystagmus seen in the control groups would argue that the local anesthetic has a diffuse, systemic effect on the nervous system rather than a specific effect on the vestibular or optokinetic system through cervical proprioceptive pathways. If the anesthetic were to have a localized effect on vestibular function mediated through the cervical proprioceptive pathways, one would not expect to see changes in the fast component of the nystagmus, which is not mediated through the vestibular system. In addition, one would not expect to see changes in the control groups, in which the anesthetic was administered at a site distant from the deep cervical structures.

The same group of investigators recorded similar measures in five monkeys following left C1 and C2 dorsal root section. They reported no spontaneous nystagmus but did observe asymmetrical optokinetic responses as compared with preoperative responses, which had been symmetrical. Slow-component eye velocity was diminished during clockwise optokinetic stimulation (*P* values ranging from 0.03 to 0.09). The post-rotatory nystagmus (rotation velocity 200 deg/sec) was measured in four monkeys preoperatively and after left C1 and C2 dorsal root sections. During the first 2 postoperative days, the monkeys demonstrated a decrease in the maximum slow-component eye velocity after counterclockwise rotations. The investigators state that this was a statistically significant decrease, and that the effect was likely a result of the loss of proprioceptive input from the cervical spine to vestibular nuclei. As in the previous experiment, however, they used a *P* value equal to 0.06 as the level of significance. This response difference was short-lived. By the fifth day after the dorsal root section, the slow-component eye velocity responses were greater than the control values, and there was no apparent asymmetry in the responses.

In addition to the questionable statistical significance, the responses these investigators noted to the optokinetic stimuli and the rotational stimuli were not consistent. Clockwise optokinetic stimulation induces nystagmus with slow-component eye velocity to the right and the fast-component eye velocity to the left (left-beating nystagmus). In humans, this stimulation also induces a sense of rotation in a counterclockwise direction. The post-rotatory nystagmus following counterclockwise rotation is right-beating (slow-component eye velocity to the left and fast-component eye velocity to the right), and it would be accompanied by a sense of clockwise rotation in humans. If the dorsal root sections were to have an asymmetrical effect on vestibular function, one would expect that the response to optokinetic stimulation would be diminished during clockwise rotations and that post-rotatory nystagmus would be diminished after a clockwise rotation of the chair. Both of these conditions would elicit left-beating nystagmus as well as inducing a sense of leftward rotation in humans. Although Ishigara and coworkers[27] suggest that their findings support the role of cervical proprioception in normal oculomotor and vestibulo-ocular function, their interpretations should be taken cautiously in light of the data.

Care must be taken in extrapolating from the animal studies cited here to humans, because there appear to be species-specific differences. Injection of local anesthetics in the posterior, upper cervical musculature in animals and humans yields different oculomotor responses. De Jong and associates [28] reported that injection of a local anesthetic in the cat, monkey, and rabbit induced nystagmus. The induced nystagmus in the cat and rabbit lasted from several minutes to an hour or more. The nystagmus in the monkey lasted for only several minutes and was suppressed by vision. In rabbits with bilateral labyrinthectomies, injection of the local anesthetic did not induce nystagmus, indicating that the nystagmus is generated through the vestibular system.[29] In monkeys, the local anesthetic had no effect on optokinetic nystagmus, optokinetic after-nystagmus, or caloric nystagmus. These investigators also injected local anesthetic into two human subjects and observed no nystagmus in either subject. Therefore, the nature of the cervical-induced nystagmus appears to be species specific.

On the basis of the anatomical convergence of vestibular, cervical proprioceptive, and visual signals from the accessory optic tract on type II neurons in the vestibular nuclei,[30,31] Karlberg and Magnusson[32] proposed that asymmetrical cervical proprioception may affect optokinetic after-nystagmus (OKAN), the nystagmus present in the dark following the cessation of the optokinetic stimulus.

These researchers measured OKAN in normal, healthy individuals with the head in neutral, passively rotated 70 degrees to either side, and actively rotated 60 to 70 degrees to either side. They found that initial velocity, duration, and cumulative eye position of OKAN were significantly decreased when the optokinetic stimulus was rotated in the direction of passive head rotation in comparison to the OKAN with the head in neutral. When the optokinetic stimulus was in the direction opposite to the head rotation, there was no decrease in the OKAN response. A similar decrease in OKAN duration and cumulative eye position occurred during active head rotation. The researchers suggest that this cervical influence on OKAN may play a role in the provocation of symptoms of dizziness in individuals with cervical pain and dysfunction.

Although Karlberg and Magnusson[32] conducted their study in normal humans, the results may support the findings of Igarashi and coworkers[27] after unilateral C1 and C2 dorsal root section in squirrel monkeys. Recall that the latter investigators reported a decrease in the slow-component eye velocity of post-rotatory nystagmus when the slow component was directed toward the side of the dorsal root section. Karlberg and Magnusson[32] reported a decrease in slow-component velocity when the optokinetic nystagmus was in the same direction as the head rotation. If one assumes that head rotation to the left induces an asymmetrical input from the cervical proprioceptors (greater on the right than the left because of stretching of the muscle spindles), the results of the two studies are qualitatively similar. In both cases, the slow-phase eye velocities were decreased toward the side of decreased cervical proprioceptive input. Whether this asymmetrical OKAN test can be used to identify individuals with cervical vertigo remains to be seen.

Several other studies have assessed oculomotor and vestibular function in humans after flexion–extension injuries to the cervical spine. Oosterveld and colleagues[33] conducted electronystagmographic (ENG) studies in 262 patients who had symptoms of cervical or cervical and upper extremity pain after experiencing acceleration injuries to the cervical spine. Eighty-five percent of these patients had symptoms of lightheadedness or a floating sensation. The investigators report that none of the patients had rotational vertigo. Spontaneous nystagmus was reported in 165 patients (63%); 110 of these patients (67%) also had positional nystagmus. On the basis of the pattern of spontaneous and positional nystagmus in these 165 patients, the nystagmus was thought to be of central origin in 159 cases (96%). Central oculomotor findings, including direction-changing nystagmus, saccadic smooth pursuit, and impaired visual suppression of the VOR, were found frequently. The incidence of individual central signs ranged from 26% to 43%. The investigators also reported that cervical nystagmus was present in 168 patients, which may simply be the normal manifestation of the COR. Because there is no indication of the incidence of cervical nystagmus in normal individuals, one cannot ascertain whether this finding is abnormal. On the basis of results of their oculomotor studies, the investigators contended that cervical whiplash injuries induce diffuse, rather than well-localized, lesions within the CNS.

Toglia[34] reported ENG and rotational chair results in 309 patients who had primary symptoms of dizziness after flexion–extension acceleration injury to the cervical spine. Of these patients, 57% had abnormal caloric test results (40% had significant canal paresis, and 22.5% a significant directional preponderance), and 51% had abnormal rotational test results. These data support the idea that peripheral vestibular system deficits are frequently present in individuals who experience whiplash injuries. One might make the argument that the caloric weaknesses and rotational asymmetries were due not to peripheral vestibular dysfunction but to impaired cervical input to the vestibular nuclei. This possibility is unlikely, however, given the relatively small input from the cervical spine to the vestibular nuclei compared with the input from the vestibular labyrinth. In addition, because there was no cervical movement during the caloric or rotational tests, one would have no physiological reason to expect that the asymmetrical responses arose from the cervical spine.

The smooth pursuit neck torsion test (SPNT) has been proposed to identify cases of cervicogenic dizziness. This laboratory test evaluates the smooth pursuit eye movement system and is conducted with the neck in neutral and rotated 45 degrees to the left and to the right. Smooth pursuit eye movements are recorded in each position. The gain (the ratio of eye velocity to target velocity) of the response is determined for each neck position, and the difference between the smooth pursuit gain in neutral and the average gain in the rotated positions is calculated (the SPNT difference). Several studies have demonstrated a significant increase in the SPNT difference values between subjects with WAD and normal subjects, as well as between subjects with WAD and those with Ménière's disease or central vertigo.[35-37] In addition, these studies have shown a significantly greater increase in the SPNT difference in subjects with WAD and concurrent complaints of dizziness as compared with subjects with WAD but no dizziness, suggesting that the dizziness is a critical component.

It is possible that the severity of cervical pain is the critical component in the SPNT difference values. Both Tjell and Rosenhall[35] and Treleaven and colleagues[36]

noted that patients with WAD and dizziness had greater complaints of pain than patients with WAD but no dizziness. Treleaven and colleagues[36] evaluated the role of pain levels in their analyses of the SPNT. They found mixed results: if they grouped data from all patients with WAD, there was a weak but statistically significant correlation (Spearman rank correlation [ρ] = 0.27) between pain levels and SPNT differences, such that with increasing pain levels, there was an increase in the SPNT difference. On the other hand, there was a significant negative relationship between SPNT differences and reported levels of pain/disability in patients with WAD but no dizziness, such that those with greater pain tended to have a smaller SPNT difference. Although the SPNT difference values were greatest in patients with WAD, individuals with other causes of cervical pain, such as cervical spondylosis and fibromyalgia, had significant SPNT difference values when compared with normal individuals.[37] Regardless of the exact etiology of the changes in SPNT, either nociceptive or proprioceptive factors, the results do suggest a relationship between cervical pain, cervical proprioception, and CNS oculomotor control.

Attempts have been made to determine the sensitivity and specificity of the SPNT. Tjell and Rosenhall[32] determined the sensitivity and specificity of the SPNT using 2 standard deviations from the mean of the SPNT difference value found in healthy controls as the threshold to determine a normal/abnormal test result. For the control group, they combined the healthy controls, subjects with Ménière's disease, and those with central vertigo, because there were no differences in the results among these groups. Comparison of individuals with WAD and dizziness with the "normal" group gave the SPNT a sensitivity of 90% and a specificity of 91%. Comparing the individuals with WAD but no dizziness with the "normal" group gave the SPNT a sensitivity of 56% and a specificity of 91%. Based on these results, the authors concluded that the SPNT test is useful for diagnosing cervicogenic dizziness in individuals with WAD. However, they did not address the issue of discriminating between individuals with WAD and cervicogenic dizziness and folks with WAD and dizziness arising from another cause, such as a concurrent vestibular problem. Using the reported sensitivity and specificity values, one can determine the positive and negative predictive values of the SPNT test for comparing WAD with dizziness and WAD without dizziness. The positive predictive value (PPV) is 62%, and the negative predictive value (NPV) is 81%. With these values, the absence of a significant SPNT difference value does a fairly good job of eliminating cervicogenic dizziness as the cause of symptoms. However, a significant SPNT difference is unlikely to differentiate between individuals with WAD and cervicogenic dizziness and individuals with WAD and another cause of dizziness.

In a second study, Tjell and colleagues[37] compared individuals with WAD to individuals with cervical spondylosis, cervical vertigo, or fibromyalgia. They found significant differences in the SPNT difference values between the individuals with WAD and each of the other conditions, suggesting that the SPNT test may be more appropriate as a test for WAD than cervicogenic dizziness.

Another point to keep in mind is that the SPNT, as described, is a laboratory test where the analysis of the eye movements is performed off-line, and the identification of saccades within the smooth pursuit movements is made by visual inspection of the eye movement data. It is not clear that this test can be replicated with the typical eye movement recording systems used in vestibular function testing clinical laboratories. Kongsted and colleagues[38,39] were unable to identify a significant SPNT difference in individuals with WAD using computerized eye movement analyses similar to those used in clinical vestibular labs. There were other methodological differences between the studies performed by the two groups. In the studies performed by Kongsted and colleagues, a chin rest was used to stabilize the head, which may have had an influence on head movements that occurred during the test and potentially altered the cervical afferent input. There were also differences in the parameters of the smooth pursuit stimulus. The SPNT test shows promise, but it has not been adequately validated for use in identifying individuals with cervicogenic dizziness. In addition, although some have indicated that SPNT can be performed as part of the clinical bedside exam, to date there are no published data to support this claim.[40]

In summary, although there are known cervical inputs to the vestibular nuclei and a convergence of cervical, visual, and vestibular inputs in the CNS, it does not appear that ablative cervical lesions have a profound effect on the oculomotor or vestibular systems in humans. Cervical disorders have been shown to induce changes in smooth pursuit, but the studies to date have not been able to differentiate between individuals with cervical pain only and those with cervical pain plus dizziness.

Postural Control Findings

In contrast to the inconclusive results of various oculomotor tests in individuals with suspected cervicogenic dizziness, the tests of balance and cervical kinesthetic sense may be more beneficial in the diagnosis of this disorder. De Jong and associates[28] described ataxia in all species following injections of a local anesthetic in the cervical region. Cats demonstrated ataxia and hypotonia ipsilateral to the injection and had a tendency to fall to

that side. Rabbits also displayed a complex behavior after unilateral injection; they first fell and rolled to the side of the injection, then developed lateropulsion, and then demonstrated ipsilateral hypotonia. Similarly, when unilateral cervical root sections were performed in rabbits that had previously undergone unilateral labyrinthectomy, the rabbits fell to the side of the labyrinthectomy and rolled along the long axis of their bodies. The direction of the rolling depended on the side of the labyrinthectomy and was independent of the side of the cervical root section. Although the nystagmus induced by the local anesthetic in monkeys was small compared with that in the cat, the ataxia was greater. The monkeys displayed a head and trunk tilt of approximately 10 degrees toward the side of the lesion and marked ataxia of the ipsilateral limbs. Injection of a local anesthetic in human subjects resulted in lightheadedness and a sense of lateropulsion. There was a deviation of stance toward the side of the injection and ipsilateral past-pointing. When human subjects were in supine, the injection produced a sense that the bed was rolling over toward the side of the injection. Although the injection of local anesthetic was no doubt an unusually potent stimulus, it yielded both postural and perceptual findings that one might expect to see in cases of altered cervical proprioceptive inputs.

Several studies have examined balance control in individuals with cervical symptoms both with and without associated complaints of dizziness. These studies have used various manipulations and measurement modes to determine the role that cervical pain may play in standing balance and to establish a test for cervicogenic dizziness. For the most part, these studies have documented that cervical pain leads to a disruption of standing balance, which has been postulated to be the result of a disruption of the proprioceptive inputs.

Alund and colleagues[41] measured standing balance in patients with chronic cervical pain and associated symptoms of vertigo or imbalance, which were compared with those obtained in age-matched, normal healthy controls and to patients with chronic cervical pain but no associated vertigo or imbalance. Postural sway was measured with computerized dynamic posturography. The subjects were tested with eyes open in both stable and sway-referenced platform conditions (sensory organization tests 1 and 4). The subjects performed the tests with the cervical spine in neutral, flexion, extension, lateral flexion to the right and left, and rotation to the right and left. The subjects with cervical pain also performed the tests with the cervical spine in the most painful position. When the subjects performed the tests on a stable platform, there was no difference among groups or test positions. However, with the test performed on a sway-referenced platform, the researchers found differences between groups and with different cervical positions. All three subject groups generally demonstrated increased sway when the cervical spine was out of anatomically neutral position. The patients with cervical pain and dizziness had significantly greater sway than the healthy controls when the cervical spine was in neutral. The subjects with cervical pain and dizziness had greater sway than subjects with cervical pain but no dizziness when the cervical spine was held in the most painful positions. On the basis of these findings, the researchers concluded that dynamic posturography may be an appropriate method of determining the presence of cervicogenic dizziness. They did not determine the specificity and sensitivity of the test, which must be examined before the test can be clinically useful.

Karlberg and colleagues[42,43] have described changes in postural control in individuals with suspected cervicogenic dizziness and in individuals with cervical and radiating upper extremity pain. They did not mention whether the individuals with upper quarter pain had complaints of dizziness or imbalance. Postural control in these studies was assessed using a force platform and measuring the motion (sway velocity and sway variance) of the center of pressure. These investigators induced body sway with vibrators attached to the gastrocnemius muscles or the posterior cervical musculature. Compared with age- and gender-matched controls, the individuals with upper quarter pain had greater sway velocity and increased variance of sway with eyes closed when either site was vibrated.[42]

In a subsequent study, Karlberg and colleagues[43] evaluated postural dynamics (measures of swiftness, stiffness, and damping determined from an inverted pendulum model of stance control) for three groups. Using the vibration-induced body sway described previously, they measured the postural control parameters in individuals with suspected cervicogenic dizziness, individuals with a recent bout of vestibular neuritis, and in normal healthy individuals. The group with a suspected cervical cause of dizziness demonstrated lower values of stiffness than either the normal or vestibular neuritis groups. In addition, the cervicogenic dizziness group had higher values for damping than the normal group. Using Fisher linear discriminant analysis and the swiftness, stiffness, and damping values, the investigators were able to distinguish subjects with cervicogenic dizziness from both the normal subjects and those with vestibular neuritis. They were also able to differentiate the subjects with vestibular neuritis from the normal subjects.

The results of these two studies give further support to the hypothesis that cervical disorders in general lead to disturbances in postural control, possibly through distortion

of the proprioceptive information. Karlberg and colleagues[43] report that a naïve tester using this method could correctly classify 78% of individuals with cervicogenic dizziness, so evaluation of postural control may be a test for cervicogenic dizziness. However, as the investigators note, the specificity of the test has not yet been determined. Two other points should be raised regarding this test. First, the sensitivity of this test was calculated using data from individuals previously identified with cervicogenic dizziness, rather than in a mixed population with the examiner blinded to the diagnosis. Second, although this test may hold promise as a diagnostic tool, it incorporates computations and measures that are not currently part of commercially available balance assessment tools.

Finally, Treleaven and colleagues[44] assessed standing balance using the Clinical Test for Sensory Interaction in Balance in normal individuals and individuals with WAD, half of whom had associated complaints of imbalance and dizziness. The amount of postural sway was recorded by means of a force platform and analyzed with wavelet analysis. As a group, the individuals with WAD had greater sway than the age-matched normal individuals. In addition, the individuals with WAD and complaints of dizziness had greater sway than the individuals with WAD but no dizziness. As noted previously by these investigators, individuals with WAD and dizziness had greater complaints of pain than individuals with WAD but no dizziness. The investigators noted statistically significant, moderate correlations (Spearmans rho correlation coefficients 0.32 to 0.41, $P < 0.01$) between pain levels and sway energy in the various test conditions for the individuals with WAD (combined dizzy and nondizzy groups). No attempt was made to determine sensitivity or specificity for this test. The results of this study further support the proposition that altered cervical proprioceptive input or cervical pain can lead to disturbed balance responses. Care should be taken in interpretation of these results, however, because vestibular lesions were not ruled out in the subjects of the study except by patient history.

There is other evidence to suggest that WAD can contribute to imbalance and postural sway, but it should be noted that in this study there was no mention of whether the patients with WAD had symptoms of dizziness. Stapley et al[45] found that, following whiplash injuries, some patients (7 of 13) were more susceptible to fatigue in the neck extensor musculature. These seven subjects showed an increase in postural sway following isometric contractions designed to fatigue the cervical extensor musculature. The subjects who were not susceptible to fatigue showed no changes in their postural sway following the same isometric contractions. Interventions aimed at reducing the cervical pain, improving range of motion, and extensor muscle endurance resulted not only in reduced fatigability, but improved postural sway following the fatiguing isometric contractions. These findings suggest that cervical extensor muscle fatigue may have a role in the increased postural sway and that the cervical muscle fatigue testing and treatment may be important components in the treatment of individuals with WAD. At this point, it is not clear if cervical extensor fatigue has a role in cervicogenic dizziness.

Cervical Kinesthesia Findings

Another line of investigation provides additional support to the hypothesis that cervical problems, WAD in particular, can lead to altered cervical kinesthetic sense. Several studies have demonstrated a decrease in cervical kinesthesia in individuals with complaints of cervical pain. Revel and coworkers[46] reported that subjects with chronic cervical pain, compared with normal subjects, had a diminished ability to relocate the head on the trunk (in the absence of visual cues) after an active head rotation. Using a threshold value of 4.5 degrees, these researchers determined that the test had a sensitivity of 86% and a specificity of 93%. In a subsequent study, Revel and coworkers[47] found that patients with chronic cervical pain and impaired kinesthetic sense, who were enrolled in a kinesthetic retraining program in combination with medical analgesic therapy, experienced improvements in kinesthetic sense and cervical rotation range and a decrease in pain in comparison with patients who received only medical analgesic therapy. The training program used in this study is described later in this chapter.

Subsequent studies have assessed cervical kinesthesia in individuals with WAD and dizziness. Similar to the findings reported by the Revel group, Heikkilä and Wenngren[48] found that individuals with WAD had greater cervical repositioning errors than normal individuals. They also noted that individuals with WAD and complaints of dizziness had greater cervical repositioning errors than individuals with WAD and no complaints of dizziness. It is not clear whether this greater error is caused by the associated dizziness (and presumed influence on the vestibular system) or the increased cervical pain that is typically found in individuals with WAD and dizziness. Heikkilä and Wenngren[48] did not report the pain levels in the two subgroups with WAD. They did observe, however, that individuals with WAD and radicular symptoms also had greater cervical repositioning errors than individuals with WAD and no radicular symptoms, suggesting that the extent of injury, and the resulting pain levels, may be the critical factor in determining the extent of the cervical repositioning errors.

Treleaven and colleagues[49] also noted greater cervical repositioning errors for rotation and extension in individuals with WAD than in normal individuals. When these investigators compared results in individuals with WAD and dizziness and individuals with WAD but no dizziness, they noted that the individuals with the associated dizziness had significantly greater cervical repositioning errors (4.5 degrees vs. 2.9 degrees) for rotation in one direction. The difference in cervical repositioning errors between these two groups approached statistical significance (3.9 degrees vs. 2.8 degrees) for rotation in the opposite direction. There was no difference between the two WAD groups in the repositioning error for cervical extension movements. These investigators reported that the subjects with WAD plus dizziness had higher scores on the neck pain index, but made no attempt to correlate the level of pain with the extent of the cervical repositioning error.

The results of these studies suggest that cervical kinesthesia is disturbed after injury to the cervical spine. Similar kinesthetic impairments have been reported with pain or injury to other joints, such as the ankle,[50] knee,[51,52] and shoulder.[53] The disruption of cervical kinesthetic sense has been used as a possible explanation for the mechanism behind cervicogenic dizziness. Although this is a plausible mechanism for cervicogenic dizziness, the fact that not all individuals with WAD and impaired cervical kinesthesia have complaints of dizziness detracts from the argument. Certainly the extent of the injury and the level of cervical kinesthetic impairment may be a factor in determining which individuals experience dizziness.

Possible Mechanisms

The lesion-induced ataxia in humans, the changes seen in stance control in individuals with cervical pain, and the cervical repositioning errors associated with cervical disorders lend support to the hypothesis that cervical proprioception has a role in balance control and, possibly, cervicogenic dizziness. The mechanism of the postural disturbance is unresolved. There are several possible explanations for the observed balance/proprioceptive deficits. First, individuals with cervical spine disease or cervical disc disease may have compression of the spinal cord and the spinal tracts relaying the proprioceptive information from the lower extremities, which could cause the observed postural disturbances. Many of the individuals with cervical pain or dizziness, however, have no findings of spinal cord compression on radiological or clinical examination, making this finding an unlikely cause of the postural deficits.[54]

Second, inaccurate proprioceptive input from sensitized receptors in either the joint capsules or the cervical musculature could create a sensory mismatch between the vestibular and proprioceptive inputs, which could lead to symptoms of dizziness and altered postural control.[55] Third, rather than creating a sensory mismatch, the altered cervical kinesthetic sense could lead to inaccurate representation of head position relative to the trunk. Because the vestibular system is physically located in the head, the output of the vestibular system must be modulated in relation to head position on the trunk to properly control balance. If the CNS receives inaccurate information about head position relative to the trunk, the modulation of the vestibular responses may be inappropriate, resulting in the observed imbalance. In both of these instances, the symptoms would be greatest during head movements, when vestibular and cervical proprioceptive inputs would be changing.

Finally, it may not be disruption of the cervical kinesthetic receptors, but the actual pain in the cervical region, that leads to the observed balance and proprioception deficits. Studies by Rossi and colleagues[56,57] have shown that chemically induced tonic muscle pain can alter the position sense of the affected limb in the absence of any actual damage to muscle or joint receptors. The studies have also demonstrated changes in somatosensory-evoked potentials at a cortical level, rather than at the spinal cord, following chemical induction of muscle pain.[57] On the basis of the nature of the stimuli used and the measured effects, these researchers concluded that alteration in the somatosensory-evoked potentials reflected changes in the proprioceptive pathways. A third study demonstrated inhibition of both cortical and spinal level motor excitability as a result of chemically induced muscle pain.[58]

From the results of these studies, one could hypothesize that cervical pain will alter the proprioceptive sense in the neck, leading to inappropriate control of the head on the trunk. Recall from the previously discussed clinical studies that the postural control deficits and cervical repositioning errors increased with rising levels of cervical pain. Regardless of the actual mechanisms involved, it appears that cervical disorders can lead to alterations in balance and cervical position sense that may be related to symptoms of dizziness. It remains to be seen, however, whether any of these tests can be used to identify individuals with cervicogenic dizziness.

Examination

With the lack of a definitive test for cervicogenic dizziness, the diagnosis is based on the individual's signs and symptoms and on the absence of otologic or neurological causes for the clinical findings. A patient with suspected cervicogenic dizziness typically presents with

disequilibrium or lightheadedness, cervical pain, ataxia or unsteadiness, and limited cervical motion. Head movements typically aggravate the symptoms. Other disease processes, such as cerebellar and spinal ataxia, bilateral vestibular loss, benign paroxysmal positioning vertigo (BPPV), and chronic unilateral vestibular loss, can also manifest similar signs and symptoms. These otologic and neurological causes of dizziness may also cause restricted cervical motion and neck pain as a result of muscular guarding of the neck to limit head movements. Consequently, the presence of disequilibrium or lightheadedness, cervical pain, ataxia or unsteadiness, and limited cervical motion is not conclusively indicative of a cervical cause of the dizziness, and the clinical decision-making process should first rule in or rule out otologic and neurological causes of the symptoms. If the symptoms are because of an otologic or neurological cause, this fact should become apparent with a comprehensive clinical examination, vestibular function tests, or radiological evaluation.

If there are no apparent neurological or otologic causes for the symptoms, one should conduct a more detailed evaluation of the upper quarter. Malmstrom et al[59] examined patients with suspected cervicogenic dizziness for musculoskeletal impairments. Subjects were excluded if they had a history of central nervous system disease, head-neck trauma, major injuries of the lower limbs, cerebrovascular diseases, ear diseases, psychiatric disorders, pregnancy, or hyperthyroidism. Vestibular causes of dizziness were excluded via clinical examination, electronystagmography, and pure tone audiometry. Common findings included good cervical mobility but reduced cervical-thoracic mobility. There were postural alignment deviations, and evidence of postural imbalance. Dorsal neck muscle tenderness and tightness was evident in the majority of the subjects, and there was local tenderness at several zygapophyseal joints. It follows that an appropriate evaluation of patients with suspected cervicogenic dizziness should include assessment of postural alignment and control, active and passive cervical and cervico-thoracic range of motion, tests for instability in the upper cervical spine, neurological examination of the upper extremities (strength, sensation, and reflexes), palpation of the cervical spine musculature and facet joints, and segmental mobility testing of the cervical spine. A detailed description of these examination procedures is beyond the scope of this chapter; three specific test maneuvers, however, are addressed here.

The first specific test is the test for VBI. Some authorities recommend testing for VBI during the cervical screening examination before treatment of the cervical spine is initiated. We should emphasize one point regarding VBI testing in the clinic. Studies to date have shown that the clinical tests for vertebral artery compression do not have adequate sensitivity to rule out the disorder (see Richter and Reinking[60] and Childs and colleagues[61] for in-depth reviews of this issue). Therefore, a negative test result does not rule out the possibility of vertebral artery compromise.

Despite its lack of utility as a screening test, local statutes and practice patterns may dictate the performance of VBI testing. Testing for VBI often consists of the vertebral artery compression test (cervical extension and rotation), typically performed in the supine position. It should be noted that this test position is similar to the final position in the Dix-Hallpike test used to assess for BPPV.[62] There are differences in the two tests, at least in patients with normal cervical range of motion. The test for VBI involves full extension and rotation, whereas the Dix-Hallpike test involves 45 degrees of rotation and 10 to 20 degrees of cervical extension. In patients with limited cervical range, however, the Dix-Hallpike test may involve movements at the limits of extension and rotation.

If a patient with complaints of cervical pain and dizziness experiences symptoms of dizziness with either the Dix-Hallpike test or the vertebral artery compression test, differentiating between BPPV and VBI can be problematic. As discussed previously, it is unlikely that VBI would produce isolated symptoms of dizziness or the characteristic patterns of nystagmus one sees in patients with BPPV. However, given the nature and severity of the deficits that can result from VBI, one would like to be able to differentiate between these two entities. This differentiation can be made if one remembers the following two facts: (1) BPPV is brought about by changes in head position relative to gravity, regardless of the position of the cervical spine, and (2) VBI caused by cervical motion is brought on by the position of the cervical spine, regardless of head position relative to gravity.

Consequently, one can test for BPPV by positioning an individual's head (relative to gravity) in a position identical to that for the Dix-Hallpike test without extending the neck by having the person lie supine on a table that is in the modified Trendelenburg position (foot of the table elevated relative to the head). Another method is to perform the side-lying test for BPPV.[63] If cervical rotation must be avoided as well, the individual can lie down in a partial side-lying position. Because the neck is maintained in a neutral position with these test modifications, symptoms brought on with the tests would not be attributed to VBI. In an analogous fashion, one can position the individual's cervical spine in a position identical to the Dix-Hallpike position without changing the

orientation of the head relative to gravity. The patient sits, forward flexes at the hips, and at the same time extends and rotates the neck. This sequence of movements places the cervical spine in a position identical to that obtained in the Dix-Hallpike and vertebral artery compression tests, but the patient's head position remains unchanged relative to gravity. Maintaining the vertical orientation of the patient's head prevents the occurrence of the signs and symptoms of BPPV (which are provoked by changes in head position relative to gravity). Therefore, any symptoms associated with this position change may be attributed to vertebral artery compression and VBI.

Another method of testing for VBI, which is recommended by the Australian Physiotherapy Association, involves having the patient actively assuming the following positions in a sequential manner: extension, rotation, and quadrant position (rotation and extension).[64] Each position is maintained for 10 seconds while the examiner monitors the patient's status. If results of these positions are negative (no dizziness, nausea, tinnitus, headache, blurred vision, slurred speech, slowed responses, or facial or tongue paresthesia), the examiner repeats the positions by using passive movements with overpressure. This testing method, when done in sitting, avoids the severe position changes of the head relative to gravity and may help differentiate between BPPV and VBI. In particular, if this test is performed first and no signs or symptoms are noted, subsequent Dix-Hallpike test findings can be attributed to BPPV.

The second test is a test to differentiate between cervical- and vestibular-provoked symptoms. When patients complain of dizziness associated with head movements on a fixed trunk, one cannot differentiate, from that statement, whether the symptoms are attributable to a vestibular disorder, cervicogenic dizziness, or a combination of the two. To differentiate between a cervical cause of the dizziness and a vestibular cause of the dizziness, one must isolate the two systems. One method is to perform the Head-Neck Differentiation test, a clinical variation of the neck torsion nystagmus test,[65-67] in which one looks for provocation of symptoms, not for nystagmus, as individuals without a cervical disorder can have nystagmus with this test. To test the cervical spine in isolation, one must move the patient's body under a stable head. For example, the patient rotates the trunk in one direction while the examiner stabilizes the head in space (Fig. 31.2*A* and *B*). Symptom provocation (dizziness) with this test would indicate a cervical component to the disorder. To test the vestibular system in isolation, the head and body must move together (en bloc). To test for horizontal rotation, for example, the head and trunk move as a unit, as in a standing pivot or with the patient sitting in a chair or on a stool that will rotate (Fig. 31.2*C*). The examiner must stabilize the head in relationship to the trunk with this test. Provocation of symptoms by this test suggests a vestibular component to the disorder. With this test sequence, it is important to perform the complete sequence to determine whether the symptoms originate from the cervical spine, the vestibular system, or both. A similar approach can be used to differentiate between vestibular and cervical causes of dizziness induced by vertical head movements (cervical flexion-extension).

Another method of assessing whether the symptoms originate from the cervical spine is a trial of manual cervical traction. The theory behind this examination technique is that unloading the cervical spine may alter the cervical somatosensory inputs to the CNS. Although unloading of the cervical spine can be performed in various ways, application of traction while the patient is sitting causes minimal disturbance of the vestibular system. A reduction in symptoms during the traction would suggest a cervical component to the disorder. The test can be modified by placing the patient in a symptom-provoking position before applying the manual traction to test for alleviation of the symptoms. This examination technique has not yet been validated in the literature, but we have found it clinically useful.

The third test is a test of cervical kinesthetic sense. This test has been performed in laboratory settings,[46] but can be performed clinically. The patient starts sitting in a chair with a backrest with the neck in a neutral, resting position. The patient closes his or her eyes and makes a full range cervical rotation to the right or left. After the full range rotation, the patient attempts to relocate his or her head to the initial starting position, still with the eyes closed. The accuracy of this movement is assessed (the actual process will be described in the next paragraph), and the patient will open the eyes and return to the initial starting position. The process will be repeated with cervical rotation to the other side, as well as for cervical extension and flexion.

There are three clinical methods that can be used to assess the accuracy of the relocation task. One method is to use a laser pointer that is attached to a headband or the brim of a baseball cap. The laser pointer is positioned such that the "spot" will appear at eye level, in front of the patient. Another method is to use foveal glasses, which are glasses that block peripheral visual input and only allow vision through a small (1 mm square) aperture. These glasses are constructed by first covering one lens completely with opaque tape. Then, with the patient wearing the glasses and looking at a small object located directly

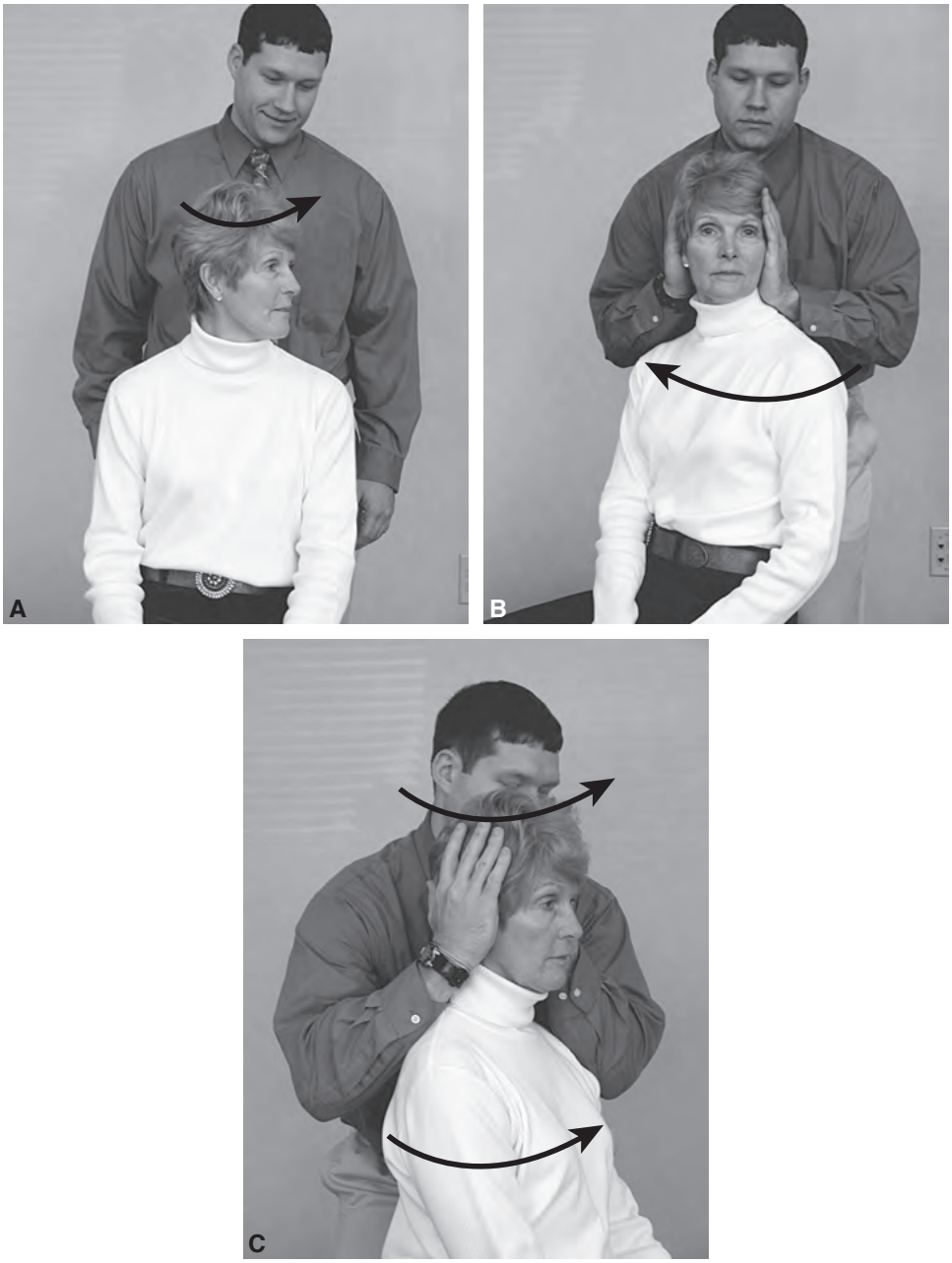

Figure 31.2 **(A)** The patient complains of symptoms of dizziness with head rotation to the left. **(B)** To assess the cervical contribution to her symptoms, her head is stabilized in space, and she turns her body to the right. **(C)** To assess the vestibular contribution to her symptoms, she is rotated en bloc to the left. In this example, the patient is sitting on a stool that rotates. The same examination procedure could be performed with the patient standing.

in front of him or her, the peripheral portions of the other lens are obscured with opaque tape, leaving a small aperture such that the patient can still see the object. To perform the test with the laser pointer, an appropriately sized target is placed in front of the patient, such that the laser pointer is pointing at the center of the target. The patient will perform the cervical relocation task described above. To evaluate the accuracy of the repositioning task, the examiner measures the location of the laser spot before the patient opens the eyes. To perform the test with the foveal glasses, an appropriately sized target is placed in front of the patient, such that the patient can see the center of the

target. The patient will perform the cervical relocation task described above. To assess the accuracy of the repositioning task, the examiner holds the patient's head stationary, and asks the patient to open the eyes and tell the examiner what he or she can see through the glasses. The third method is to attach the Cervical Range of Motion device (CROM; Performance Attainment Associates) to the patient's head and measure the position of flexion/extension/rotation the patient is in when he or she has finished the repositioning maneuver.[68]

The size of the target that is used will vary with the distance between the patient and the target. Using 4.5 degrees as the cutoff for normal cervical joint repositioning error and trigonometry, one can determine the appropriate distance and target size. The relationship between target size and distance can be expressed by the following formula:

$$\text{Tangent (4.5 degrees)} = (0.5W)/D$$

where W = width of the target, and D is the distance from the axis of rotation (top of the head) to the target. Note that we use one-half the width of the target to allow for error to either side of the center of the target. Because the tangent of 4.5 degrees is 0.0787 and the width of the target can be measured, the equation can be rearranged to determine the necessary distance of the patient from the target:

$$D = (0.5W)/0.0787$$

For example, using a standard "sticky-note," which is 3 inches square, the distance that the patient should be from the target is

$$D = (0.5 \times 3)/0.0787$$
$$D = 19 \text{ inches}$$

If one used a sticky-note for the target and the patient could perform the cervical relocation test such that the laser pointer was within the sticky-note, or if the patient could see the sticky-note through the foveal glasses, then the test would be within normal limits. Note that with the foveal glasses, the field of view increases as the distance between the patient and target increases. Consequently, one should use small targets (such as sticky-notes) located closer to the patient. This is not a concern when using a laser pointer.

Management

Treatment in cases of supposed cervicogenic dizziness is directed to the clinical findings and the patient's symptoms. Treatment of the cervical spine should address restricted mobility (because of joint restrictions or muscle tightness), hypermobility, increased muscle tone, trigger points, poor cervical posture, and impaired cervical kinesthesia. Detailed descriptions of the treatment approaches for upper quarter dysfunction are beyond the scope of this chapter; the therapeutic treatments may include cervical spine mobilization, range-of-motion exercises, cervical strengthening exercises, cervical proprioception exercises, soft tissue mobilization, and therapeutic agents.

Because of the apparent role of the upper cervical spine in the generation of cervicogenic dizziness, treatment may need to be focused on this area. Numerous techniques are available to increase joint mobility in the upper cervical spine. The treatment techniques described here are examples of those that have been well tolerated by patients, are relatively easy to perform, and appear to be effective in improving cervical mobility and reducing pain. In addition, the patients may note a reduction in dizziness. The first technique is occipito-atlas distraction, which can be performed in various manners. In one technique, the patient lies supine, and the patient's head is supported by the therapist. The therapist positions his or her hands such that the superior nuchal line of the patient's skull is resting on the therapist's fingertips (Fig. 31.3*A*). The therapist can simply allow the patient's head to rest on the fingertips or can apply gentle traction to the upper cervical spine by either flexing the metacarpal phalangeal joints or leaning away from the patient. Another method of performing the traction is for the therapist to cup the patient's occipital region in the palm and fingers (Fig. 31.3*B*). The therapist applies traction by leaning away from the patient.

The second technique is designed to increase rotation between C1 and C2 (Fig. 31.4). The patient lies supine. The therapist prepositions the patient's lower cervical spine into flexion. This position can be combined with lateral flexion (*not shown*). While maintaining the cervical flexion, the therapist then passively rotates the patient's head in the direction of restricted mobility to the end of passive, pain-free motion. If the prepositioning of the lower cervical spine is maintained, this cervical rotation occurs only in the upper cervical spine. At end range, the patient attempts to gently rotate in the opposite direction. This motion is blocked by the therapist's hand. After 10 to 15 seconds, the patient relaxes, and the therapist gently rotates the patient's neck in the direction of the restricted motion. This treatment technique is analogous to the hold-relax stretching techniques often used to stretch limb musculature. The force generated by the patient during the active, resisted rotation in this technique is minimal. Telling the patient to look in the direction of

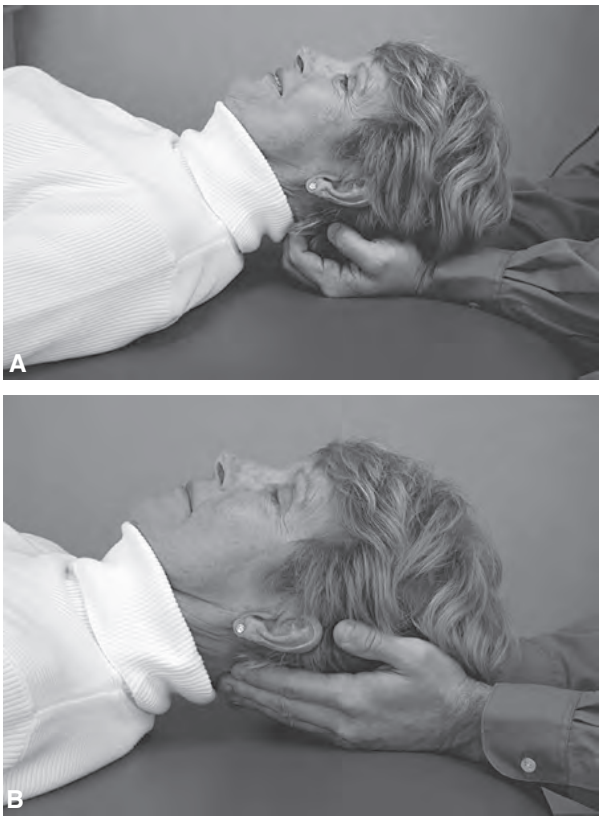

Figure 31.3 Methods of cervical traction: **(A)** The patient rests the head on the therapist's fingertips, and the therapist can lean away from the patient, providing traction. **(B)** A variation of this technique has the therapist supporting the patient's occipital region in the palms and fingers. By leaning away from the patient, the therapist can apply traction to the upper cervical spine.

Figure 31.4 Mobilization of C1–C2 to increase cervical rotation to the left.

the rotation is often sufficient to generate the appropriate level of force.

As with most mobilization techniques, it is important to follow up with active exercises to maintain the increased range. Any findings of imbalance or motion sensitivity can be treated with head movement exercises, positioning or habituation exercises, and balance exercises described elsewhere in this book. In addition, retraining of cervical kinesthesia should be initiated. Exercises such as those described by Revel and coworkers[47] have been shown to be effective. The basic components of this exercise approach are as follows:

Slow passive supine head movement with eyes fixed on a target.

Follow a moving target using alternatively slow pursuit and saccades, with free eye and head movement.

Sitting and standing exercises with restricted peripheral vision:

- Active head movements following a slow-moving target.
- Active head movements to maintain gaze on a fixed target while the trunk is passively moved.
- Fixating on a target and memorizing the head position; then closing the eyes, performing maximal rotation, returning to the starting position, and opening the eyes (angle reproduction).

The exercises performed while peripheral vision is restricted use glasses in which all but foveal vision is blocked (Fig. 31.5*A*). Because any eye movement while the patient is wearing these glasses results in blocked vision, the only way for the patient to visually follow a moving target is to move the head. Head position is driven, therefore, by visual input and must be carefully controlled by the patient to accomplish the task. To maintain gaze on a fixed target while the trunk is rotated, head motion must be disassociated from trunk motion (Fig. 31.5*B*).

Finally, the patient can practice angle repositioning while using the foveal glasses (Fig. 31.6). The patient must reproduce the starting position without visual cues. If an error in head positioning has occurred, the patient will not be looking at the target upon opening the eyes. Errors in performance will be apparent to the observer, because the foveal glasses force the patient to shift the head position to reacquire the target. The patient will receive kinesthetic feedback about the amplitude and direction of the error that can be incorporated into subsequent trials, leading to improved performance. As with the progression of motion sensitivity and habituation exercises, the variables that can be manipulated with these exercises include speed, amplitude, and direction of movement, as well as repetitions and sets.

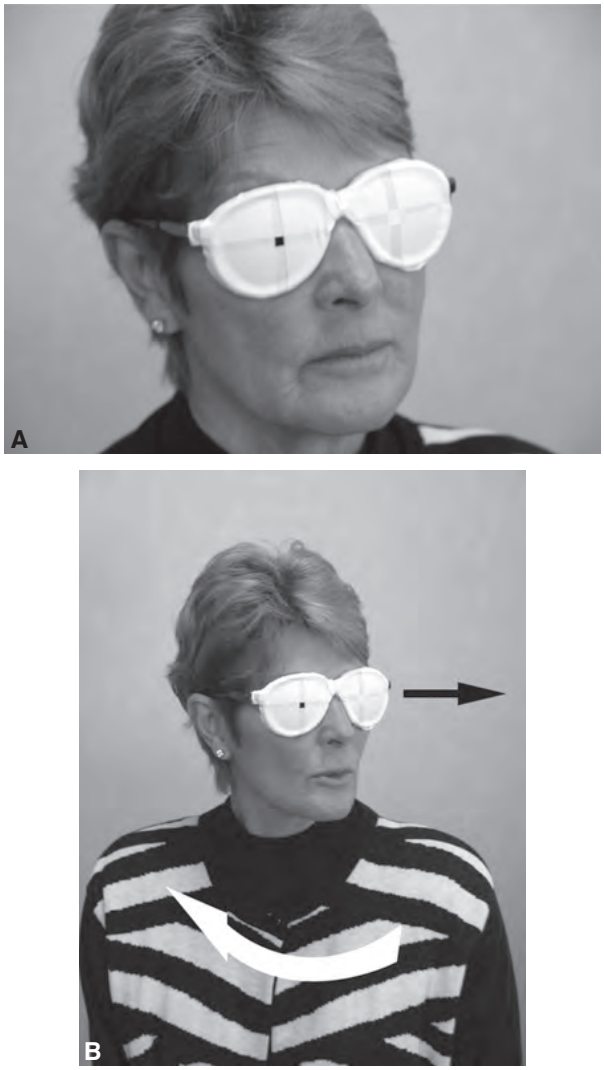

Figure 31.5 Cervical kinesthesia training exercises. **(A)** Foveal glasses. **(B)** Eyes maintain gaze on a fixed target (indicated by *straight arrow*) while the trunk is rotated to the right. The foveal glasses necessitate disassociation of head and trunk movements. Similar exercises can be performed with hip and trunk flexion and extension in the sitting position.

There is growing evidence in the literature suggesting that treatment of identified cervical dysfunctions in individuals with suspected cervicogenic dizziness may lead to resolution of the dizziness as well as the cervical signs and symptoms. For example, the effectiveness of treating the musculoskeletal findings in patients complaining of dizziness and neck pain has been shown.[59] An individualized treatment was provided based on clinical reasoning and the findings on a thorough examination. The main treatment techniques used were soft tissue treatment for muscle tenderness and tightness, local manual therapy for hypomobility, stabilization techniques for neck, trunk, and shoulders for imbalance and poor stability, and postural education. These interventions were supplemented with a home training program. The range of duration of treatment was 5 to 20 weeks, median duration 13 weeks, and the number of treatments ranged from 5 to 23, with a median of 13. The treatment resulted in reduced symptoms and increased cervicothoracic mobility. In addition, postural alignment improved, as did dynamic stabilization of the trunk, neck, and shoulders. After 6 months, 13 of 17 patients had no or reduced neck pain, and 14 had no or reduced dizziness. At a 2-year follow-up, 7 patients had no or reduced neck pain, and 11 had no or reduced dizziness.

Two systematic reviews of the literature suggest there is growing evidence to support the use of manual

Figure 31.6 Cervical repositioning using foveal glasses. **(A)** Gaze is fixed on a target, and head position is memorized. **(B)** The patient closes the eyes and fully rotates the neck in one direction while the eyes remain closed.

Figure 31.6—cont'd **(C)** The patient attempts to return the head to the starting position while the eyes remain closed. **(D)** The patient opens the eyes and, if necessary, moves the head more to reacquire the target. Because of the foveal glasses, the only way for the patient to look at the target is to move the head. The patient receives kinesthetic feedback as to direction and amplitude of error, which is incorporated into subsequent trials.

therapy, in particular spinal mobilization and manipulation, for cervicogenic dizziness.[69,70] The more recent review by Lystad et al[70] grades the level of evidence from limited (Level 3) to moderate (Level 2). This systematic review included five randomized controlled trials (RCT), and eight prospective, non-controlled, cohort studies. Six of the included studies, including two RCTs, used only spinal manipulation or mobilization, or both, as the intervention. The seven remaining investigations used a multimodal approach consisting of several different interventions. These included spinal manipulation and mobilization, soft tissue therapy, electrotherapy, medications, and home exercise programs. All but one study (of 15 articles reporting findings from 13 unique investigations) reported improvement in dizziness. Some of the studies reviewed reported improvements in postural stability, joint positioning, range of motion, muscle tenderness, neck pain, and vertebrobasilar artery blood flow velocity.

There is evidence that treatment of cervical dysfunctions can lead to decreased symptoms of dizziness and improvements in postural stability. Karlberg and colleagues [71] reported on the improvements in cervical pain and mobility as well as postural control in a controlled study of individuals undergoing physical therapy for cervical pain and dizziness. In this study, the type of treatment for the cervical dysfunction was not controlled because of the variety of symptoms and physical findings. Before treatment, the patients displayed significantly higher body sway velocities than healthy controls when the gastrocnemius muscles or posterior cervical musculature were vibrated. After treatment, which was restricted to treatment for the cervical dysfunction, there was a marked improvement in the vibration-induced sway velocity. The investigators reported that after treatment, the sway velocity during vibration of the gastrocnemius muscles returned to normal levels. They noted, however, that the patients continued to display increased sway velocities when tested with vibration applied to the posterior cervical musculature.

It is interesting to note that there are no studies that have combined the use of manual therapy and vestibular rehabilitation. A reasonable argument can be made for combining the two. As noted earlier, abnormal electronystagmographic findings are not uncommon following whiplash injuries,[33,34] so it is likely that patients with a traumatic onset to their neck pain could have coexisting vestibular deficits. Furthermore, Lystad et al[70] argue that a well-integrated system, such as one achieved through vestibular rehabilitation, would be more capable for compensating for the altered cervical sensory input in cases of cervicogenic dizziness with an associated vestibular dysfunction. It is clear that more research in this area is warranted.

CASE STUDY 31-1

Case Report

History

JB is a 63-year-old male referred by his family physician to a neurologist in the vestibular clinic. JB was evaluated by the neurologist and referred to physical therapy for treatment of possible cervicogenic dizziness. JB is initially seen in physical therapy on 11/18/96.

JB states that he has been bothered by episodic bouts of dizziness that started approximately 1 year ago. He states that the afternoon before the onset of his symptoms, he had been lifting sheets of plywood out of his pickup truck. The next morning he awoke with mild discomfort and stiffness in his neck and a strong sense of imbalance, with a tendency to lose his balance to the left. He denied having vertigo or aural symptoms in association with the onset of these symptoms. He states that since the initial episode, he has been bothered by a chronic sense of lightheadedness and disequilibrium. He states that his symptoms are exacerbated by head motion, exertion, and after sitting in one position for a prolonged period (e.g., driving or reading). Cervical and shoulder symptoms are associated with the increased symptoms of disequilibrium. He describes these symptoms as mild to moderate pain and stiffness in the cervical spine and upper trapezius muscles. He does note some difficulty walking in the dark and on uneven surfaces. He denies having oscillopsia, positioning vertigo, migraine headaches, increased symptoms in busy visual environments, extremity numbness or weakness, and incoordination.

JB states that he underwent magnetic resonance imaging and carotid ultrasound in the past, results of which were normal. Electronystagmography and caloric test showed no evidence of unilateral hypofunction or oculomotor abnormalities. Audiograms showed bilateral, mild, high-frequency sensorineural hearing loss. His medical history is unremarkable. He is taking no medications. He is a retired engineer.

Clinical Examination

Oculomotor Examination

JB's oculomotor findings are normal. There is no spontaneous or gaze-evoked nystagmus in room light or with use of Frenzel lenses. Extraocular movements are normal. His saccadic and smooth pursuit eye movements are normal. VOR cancellation and VOR gains to both slow and rapid head rotations are normal. With Frenzel lenses, there was no head shaking–induced nystagmus. Dix-Hallpike test results are negative bilaterally. Tragal compression, Valsalva maneuver, or hyperventilation does not induce nystagmus and vertigo. There is a one-line difference between static and dynamic visual acuity scores.

Balance Examination

JB's Romberg test result with eyes open is normal. With eyes closed, he performed the test for 30 seconds, but he exhibited excessive sway. Results of Romberg testing on 4-inch-thick foam with eyes open are also normal. With eyes closed, his performance was again characterized by increased sway. Sharpened Romberg test result with eyes open is normal; Sharpened Romberg test with eyes closed is abnormal because JB could perform the test for a maximum of 20 seconds. Evaluation of unrestricted gait shows normal velocity and no ataxia, but JB exhibits minimal head or trunk rotation. When asked to rotate his head while walking, he develops a mild amount of ataxia (4 deviations from a 12-inch-wide, 10-yard-long path) and notes a provocation of disequilibrium and cervical discomfort.

Upper Quarter Examination

JB's posture is characterized by a forward head. Cervical flexion and extension are mildly limited, but there is no pain or symptom provocation at end range. Cervical rotation is 75 degrees to the right and 60 degrees to the left. JB notes a provocation of his cervical symptoms and disequilibrium at the limits of his cervical rotation to the left. Trunk rotation to the right under a stable head reproduces his symptoms, but en bloc rotation does not. Palpation of the cervical spine reveals increased muscle tone and tenderness in the left suboccipital musculature, the left upper trapezius muscle, and a trigger point in the left sternocleidomastoid muscle. Segmental evaluation shows restricted rotation to the left at C1–C2. No other segmental restrictions are noted. Manual muscle testing of the upper extremities finds normal, symmetrical strength. Light touch is normal in the upper extremities. Deep tendon reflexes are also normal. Manual traction of the cervical spine in the sitting position decreases his symptoms.

CASE STUDY 31-1

Impressions

There is no evidence of unilateral or bilateral vestibular hypofunction on examination. In addition, there are no signs or history suggestive of BPPV. His history of exertion-induced symptoms may be suggestive of a perilymphatic fistula. However, there are no physical findings consistent with a fistula; specifically, there was no induction of nystagmus or vertigo with tragal compression or Valsalva maneuver. JB's balance examination shows a mild disturbance in static postural stability (increased sway on Romberg test with eyes closed and positive Sharpened Romberg test result with eyes closed). He demonstrates the ability to use vestibular cues to maintain upright stance. He has mild dynamic postural instability with head rotation while ambulating. The findings of JB's upper quarter examination are consistent with a musculoskeletal dysfunction in the upper cervical spine. The positive result with trunk under head rotation and negative result with en bloc rotation are consistent with a cervical cause of dizziness. The decreased symptoms during cervical traction are also suggestive of cervicogenic dizziness. Although "cervicogenic dizziness" is a controversial entity, this patient presents with cervical signs and symptoms that may be related to his symptoms of imbalance (specifically, the temporal correlation between the disequilibrium and cervical symptoms).

Treatment (11/18/96)

Because JB's symptoms are believed to be cervical in nature, the initial treatment focuses on his cervical problems. Treatment consists of soft tissue mobilization of the sub-occipital musculature, the left upper trapezius muscle, and the left sternocleidomastoid muscle. This is followed by mobilization of C1–C2 using a muscle energy technique (contract-relax) with active resisted isometric cervical rotation to the left at JB's end range. Following treatment, JB's rotation is symmetrical and symptom free. He is instructed in a home program to "correct" the forward head posture (dorsal glide of the head performed in prone), active resisted cervical rotation also performed in prone, and self-massage of the sub-occipital musculature.

Return Clinic Visit (12/1/96)

Upon return to the clinic, JB notes a mild decrease in his symptoms of disequilibrium and cervical discomfort. On examination, results of Romberg tests with eyes closed on both a firm surface and the 4-inch-thick foam are normal. Results of Sharpened Romberg test with eyes closed and ambulation with head rotation are unchanged. Cervical range of motion shows 70 degrees of rotation to the left and 75 degrees to the right. Palpation demonstrates increased muscle tone in the left sub-occipital musculature and left sternocleidomastoid muscle, as well as a mild limitation in rotation to left at C1–C2.

Treatment consists of soft tissue mobilization of the sub-occipital musculature and left sternocleidomastoid muscle, spray and stretch of the left sternocleidomastoid muscle, and mobilization of C1–C2. After treatment, JB has symmetrical and pain-free cervical rotation. He is instructed to continue with his home exercise program.

Return Clinic Visit (12/8/96)

JB returns to the clinic for reassessment and further treatment. He has had no symptoms of disequilibrium or cervical discomfort in the preceding week. His balance examination shows normal static balance (Sharpened Romberg with eyes closed) and no head rotation–induced symptoms or ataxia while walking. He continues to demonstrate a 5-degree restriction in cervical rotation to the left. He is treated with joint mobilization of C1–C2: distraction, muscle energy techniques, and grade 3 oscillations to increase rotation to the left. He is instructed to continue with his home exercise program for an additional 4 to 6 weeks. Because he has been symptom free for an entire week, we believe that further outpatient physical therapy is not indicated at this time. He is instructed to call in 1 month to inform us of his progress, or earlier if he becomes symptomatic.

Telephone Call (2/2/97)

JB phones to say he is doing well (no disequilibrium and no cervical symptoms). He stopped performing the home exercises 3 weeks ago, and he has noted no ill effects from stopping them. He will return to the clinic as needed.

Summary

The existence of a cervical cause of dizziness continues to be a topic of debate.Although individuals with cervical symptoms may have complaints of imbalance and lightheadedness, no clear clinical test can be used to unequivocally identify the cervical spine as the cause of the symptoms. There is some anatomical and physiological evidence to suggest that the cervical spine could influence balance and perceptions of stability, but how this influence could actually lead to symptoms of dizziness remains unclear. As stated in the introductory comments, the debate over a cervical cause of dizziness may be academic from a physical therapy standpoint. If patients present with cervical dysfunction and imbalance, we should treat them for both conditions.

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Appendix A
Questionnaire for History and Examination

Describe your major problem or the reason why you are seeing us.

Please describe in detail the circumstances and date in which the problem be gan and what were your initial symptoms and problems. Was there any stress or anxiety around the onset of the problem?

If you have spells, please describe a typical spell in as much detail as possible and describe the frequency and duration of the spells.

Duration (please check which most closely fits what you experience):
_____ less than two minutes; _____ 20 – 30 minutes; _____ 2 – 4 hours; _____ 12 – 24 hours;
_____ 1 – 3 days; _____ always present; _____ other (please write in duration)

What do you personally think your problem is due to?

1. **Please check the symptoms which characterize your problem and grade their severity from 2 (marked), 1 (moderate) to 0 (none). Put 0 if you do not have these symptoms.**
   a. **Sensation of imbalance**
      ( ) Trouble with walking. Do you use a cane, walker or wheel chair, touch walls or furniture, or use a person for balance inside buildings (If yes, please circle all that apply).
      ( ) Poor balance.
      ( ) Falls. How many falls have you had in the past 6 months, in which you une xpectedly lost your balance and landed on the floor or ground _____? Please circle all that apply: indoors, outdoors, poor lighting, due toafigue, while turning, while standing up or sitting down.
      Check ( ) if the fall(s) were due to your dizziness

b. Sense of <u>movement of the environment or of one's own body</u>
( ) Rotation (spinning, tumbling or cartwheeling);
( ) Tilt

c. Sensations <u>not associated with movement of the environment</u>
( ) Lightheadedness or impending faint;
( ) Floating;
( ) Swimming;
( ) Rocking;
( ) Fear or avoidance of being in public places

d. Associated symptoms
( ) Sweating;
( ) Nausea;
( ) Vomiting;
( ) Queasiness

e. Impaired vision
( ) Double vision;
( ) Blurred vision;
( ) Jumping of vision when walk or ride in a car

2. To what extent is your dizziness or imbalance brought on by:

Check one answer for each question.	**None**	**Some**	**Severely**
Getting out of bed on the right side ____ left side ____			
Turning over in bed, bending over or looking up			
Standing up			
Rapid head movements			
Walking in a dark room			
Walking on uneven surfaces			
Loud noises			
Cough, sneeze, strain, laugh, blowing up balloons			
Movement of objects in the environment			
Moving your eyes while your head is still			
Wide open spaces			
Tunnels, bridges, supermarkets			
Menstrual periods			

3. Other questions concerning dizziness

	YES	**NO**
Do or did you have moderate-severe motion (car or boat) sickness. If <u>yes</u> when did it start?		
Do or did you avoid situations in which you were tumbled or spun (amusement rides, merry-go-rounds)? When did this start?		
Can you sit in the back seat of a moving car and read without feeling ill?		

4. Do you have any pain? ____ yes ____ no

If yes, in what part of your body? _____

Please rate the average intensity of the pain in the last week from 0 (none) to 10 (the worst it could be): _____

5. If you have had headaches, please answer the following:

a) Approximate age they began _____;
b) Number per month _____; Pain intensity (1-10 with 10 the most severe) _____
c) Have you had sparkles, halos, or distorted vision with your headaches? _____Yes; _____No
d) Since the onset of headaches have you had at least 5 headaches that:

	YES	NO
Lasted at least 4 hours		
Started on one side of the head, if yes usually which side?		
Were throbbing or pulsatile in quality?		
Were severe enough to interfere with your schedule?		
Were aggravated by routine physical activity?		
Were associated with nausea and/or vomiting?		
Were aggravated by bright lights or loud noises?		

6. Have you had:

	Yes	Result	When
Hearing test			
Evaluation by a neurologist			
Evaluation by an ear doctor			
Evaluation by an eye doctor			
Caloric test (water or air in ear)			
MRI (was dye also given by injection?)			

7. ALLERGIES TO MEDICATIONS and please note if drug causes rash or difficulty breathing.

8. MEDICATIONS

What are your current medications, include hormones, birth control pills, special diet, etc. (Name and Amount/Day)?

1.
2.
3.
4.
5.
6.
7.
8.

9. FOR WOMEN ONLY:

Date of last menstrual period: _____

If applicable, delivery date of last birth: _____

	YES	NO
History of irregular periods now? If yes, for how long? _____		
Are you taking any hormone supplements now (e.g., Provera, Estratest, birth control pill)? If yes, how long have you been taking it? _____		
Did you stop taking hormones prior to the onset of your dizziness? If yes, which hormone(s) were you taking previously? _____		
Have you had a hysterectomy? If yes, when? _____		
Have you had your ovaries removed? If yes, when? _____ If yes, did you have <u>one</u> or <u>both</u> ovaries removed? _____		
Do you have hot flashes? If yes, when did they start? _____		
Do you think you could be going through menopause now?		

PERSONAL INFORMATION

DOB _____-_____-_____ AGE: _____
RACE ❑ Caucasian ❑ African American ❑ Asian ❑ Hispanic ❑ Other
HANDEDNESS ❑ Right Handed ❑ Left Handed Primary Language:

HOME PHONE ___-____-_____ FAMILY CONTACT _____ PHONE ___-____-____
WORK PHONE ___-____-_____ PRIMARY CAREGIVER _____ PHONE ___-____-____
CELL PHONE ___-____-_____ EMERGENCY CONTACT _____ PHONE ___-____-____
PHARMACY ___-____-_____ EMAIL if you want note sent to you _____

PRIMARY PHYSICIAN
Name: _____
Address: _____
City, ST, Zip _____
Phone: _____
Fax: _____
Check if you would like us to write to this physician ❑

WHO REFERRED YOU TO THIS CLINC?
Name: _____
Address: _____
City, ST, Zip _____
Phone: _____
Fax: _____
Check if you would like us to write to this physician ❑

Have you been in the hospital within the last 60 days? ❑ Yes ❑ No
At this visit, medical reconciliation form was given to patient: ❑ Yes ❑ No **(physicians only)**
How many times have you fallen in the last year? ___ Did you injure yourself? ❑ Yes ❑ No

SOCIAL HISTORY

Education: ❑ High School ❑ College ❑ Graduate Degree Marital Status: ❑ Single ❑ Married Currently Employed: ❑ Yes ❑ No

Present Occupation: _____ Disability Claims: ❑ Yes ❑ No

Alcohol Use: ❑ No ❑ Yes Type _____ Frequency _____ Amount _____
Tobacco Use: ❑ No ❑ Yes Type _____ Cigarette Use Packs/Day _____ Last Use _____
Illicit Drug Use: ❑ No ❑ Yes Type _____ Route _____ Frequency _____ Last Use _____
Physical Activity: Type _____ # days/week ____ # minutes/day _____

FAMILY HISTORY	Alive (age)	Deceased (age)	Cause of Death	Illnesses/Diseases
Father	___	___	_____	_____
Mother	___	___	_____	_____
Brother(s)	___	___	_____	_____
Sister(s)	___	___	_____	_____
Children	___	___	_____	_____
Other: _____	___	___	_____	_____
Other: _____	___	___	_____	_____

How many immediate relatives lived to be 65+ _____

OTHER MEDICAL PROBLEMS

Please check all that apply

Neurological
❑ Stroke or TIA
❑ Parkinson's disease
❑ Tremor
❑ Dystonia
❑ Ataxia
❑ Hydrocephalus
❑ Dementia
❑ ALS/Lou Gehrig's
❑ Myasthenia gravis
❑ Neuropathy
❑ Muscle disease
❑ Spinal cord disease
❑ Restless legs
❑ Sleep apnea
❑ Narcolepsy
❑ Multiple sclerosis
❑ Migraine
❑ Optic neuritis
❑ Epilepsy

Cardiovascular
❑ Hypertension
❑ High cholesterol
❑ Angina/heart attack
❑ Arrhythmia
❑ Syncope
❑ Peripheral vascular disease
❑ Heart failure

Endocrine
❑ Thyroid disease
❑ Osteoporosis
❑ Diabetes

Gastrointestinal
❑ Reflux/heart burn
❑ Peptic ulcer disease
❑ Crohn's disease
❑ Ulcerative colitis
❑ Gall bladder disease

Cancer
❑ Lung
❑ Breast
❑ Brain
❑ Prostate
❑ Skin
❑ Colon/Rectum
❑ Lymphoma
❑ Other

Rheumatology
❑ Rheumatoid arthritis
❑ Lupus
❑ Vasculitis
❑ Spinal stenosis
❑ Cervical spondylosis

Hematological
❑ Iron deficiency
❑ Sickle cell disease
❑ Anemia

Psychiatric
❑ Depression
❑ Bipolar disease
❑ Anxiety
❑ Panic attacks
❑ Schizophrenia

Respiratory
❑ Asthma
❑ Emphysema/COPD
❑ Tuberculosis
❑ Sarcoidosis

Genitourinary
❑ Kidney failure
❑ Kidney stones
❑ Prostatic hypertrophy
❑ STD
❑ Urinary tract infections

Past Surgeries	Type	Date	Location
1.			
2.			
3.			
4.			

REVIEW OF SYSTEMS Please check if you have had any of these symptoms **IN THE PAST MONTH**

Neurological
- ❑ Dizziness
- ❑ Headaches
- ❑ Muscle pain/cramps
- ❑ Double vision
- ❑ Droopy eyes
- ❑ Muscle twitching
- ❑ Weakness
- ❑ Numbness
- ❑ Tingling
- ❑ Loss of memory/ concentration
- ❑ Blackouts/Seizures
- ❑ Unsteadiness
- ❑ Head injury
- ❑ Slurred speech
- ❑ Trouble swallowing
- ❑ Snoring
- ❑ Daytime sleepiness
- ❑ Insomnia
- ❑ Restless legs
- ❑ Leg movements during sleep

Ocular
- ❑ Glasses
- ❑ Redness
- ❑ Dryness
- ❑ Tearing
- ❑ Pain
- ❑ Infection

Ear, Nose, Throat
- ❑ Pain or soreness
- ❑ Discharges
- ❑ Hearing loss
- ❑ Bleeding gums
- ❑ Hoarseness
- ❑ Tinnitus

Respiratory
- ❑ Cough
- ❑ Blood in cough
- ❑ Wheezing
- ❑ Pneumonia
- ❑ Bronchitis

Gastrointestinal
- ❑ GI bleed
- ❑ Nausea/Vomiting
- ❑ Hepatitis
- ❑ Hiatal hernia
- ❑ Heartbrurn
- ❑ Diarrhea
- ❑ Constipation
- ❑ Difficulty chewing
- ❑ Abdominal pain
- ❑ Bright red blood in stool

Rheumatologic
- ❑ Joint pain
- ❑ Joint swelling

Dermatologic
- ❑ Rash
- ❑ Skin color change
- ❑ Hair loss
- ❑ Skin ulcer

Constitution
- ❑ Weakness
- ❑ Tiredness
- ❑ Loss of appetite
- ❑ Increased appetite
- ❑ Weight loss
- ❑ Weight gain
- ❑ Chills
- ❑ Fever
- ❑ Night sweats

Psychiatric
- ❑ Psychosis
- ❑ Addictions
- ❑ Anxiety
- ❑ Hallucinations
- ❑ Severe stress
- ❑ Depression
- ❑ Mental illness
- ❑ Mood swings
- ❑ Phobias

Cardiovascular
- ❑ Heart attack
- ❑ Chest pain/angina
- ❑ Dizziness
- ❑ Fainting
- ❑ Aneurysm
- ❑ Palpitations
- ❑ Calf/leg pain at rest
- ❑ Calf/leg pain walking
- ❑ Swelling of ankles
- ❑ Discoloration of hands/feet
- ❑ Poor circulation
- ❑ Heart murmur
- ❑ Heart failure

Hematologic
- ❑ Easy bruising
- ❑ Bleeding disorder

Lymph
- ❑ Frequent infections
- ❑ Node enlargement

MULTIDIMENSIONAL DIZZINESS INVENTORY

In the last 6 months, what percentage of the time has dizziness interfered with your activities?

Mark Line: |————————-|————————-|————————-|————————|————————|
0% 20% 40% 60% 80% 100%

Instructions. Please answer the following questions about your dizziness and how it affects your life. Read each question and then **circle a number** on the scale under that question to indicate how that question applies to you.

1. Rate the level of your dizziness at the present moment.

1	2	3	4	5
none	slight	moderate	quite a bit	extreme

2. Since the time your dizziness began, how much has your dizziness changed your ability to work? (___Check here, if you have retired for reasons other than your dizziness.)

1	2	3	4	5
not at all	slightly	moderately	quite a bit	extremely

3. How much has your dizziness changed your ability to do household chores?

1	2	3	4	5
not at all	slightly	moderately	quite a bit	very much

4. Does your dizziness significantly restrict your participation in social activities such as going out to dinner, going to movies, dancing or to parties?

1	2	3	4	5
not at all	slightly	moderately	quite a bit	very much so

5. To what extent does dizziness prevent you from driving your car?

1	2	3	4	5
not at all	slightly	moderately	markedly	severely

MDI Section B

This scale consists of a number of words that describe different feelings and emotions. Read each item and then mark the appropriate answer in the space next to that word. Indicate to what extent you generally feel this way, that is, how you feel on the average. Use the following scale to record your answers.

1	2	3	4	5
very slightly or not at all	a little	moderately	quite a bit	extremely

_____ interested	_____irritable	_____jittery
_____ distressed	_____alert	_____active
_____ excited	_____ashamed	_____afraid
_____ upset	_____inspired	_____hostile
_____ strong	_____nervous	_____enthusiastic
_____ guilty	_____determined	_____proud
_____ scared	_____attentive	

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