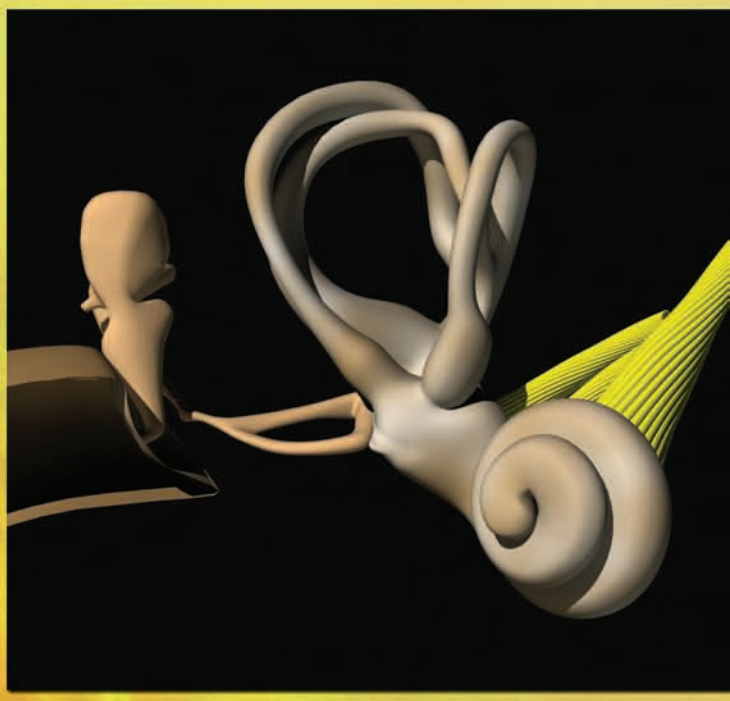
Susan J. Herdman



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# Vestibular Rehabilitation

THIRD EDITION

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## *Dedication*

***I would like to dedicate this edition of***
Vestibular Rehabilitation ***to my family.***
***I love you all very much.***

# Foreword

Baseball has always been my passion. I played the sport from the time I could walk all the way through college. I always thought one could learn a lot about life from participating and from observing the game….teamwork, responsibility, strategy and so on. In that context and for reasons far transcending the purpose of this book, I adopted the Boston Red Sox as my team. Through circumstances far more surreal than circumstantial, I was interviewed by Weekend America, a National Public Radio show the day after the Red Sox miraculous comeback against the New York Yankees to win the American League title in 2004. The interview was taped on Thursday, October 21 two days before the first game of the World Series in which my team defeated the St. Louis Cardinals in four consecutive games, and aired immediately before the first game. During the interview, Bill Radke asked if the Red Sox could win the Series (a thought that any self-respecting Sox fan would never contemplate after 86 barren years filled with countless frustrations) and win or lose, if I thought the team players would all be back the following year to start their own dynasty. To the latter question I responded that I doubted the possibility since in contemporary sports the notion of team loyalties and perpetuation of excellence amongst a cohesive unit was easily supplicated by the lure of more lucrative promises from rival baseball clubs.

So what does this stream of consciousness have to do with the third edition of *Vestibular Rehabilitation*? These thoughts about this radio interview crept into my mind as I was reviewing Susan Herdman's third edition. Here is a text already filled with contributions from clinicians and researchers acknowledged as superstars in this field. Each of these individuals is already quite busy and well in demand for other academic and intellectual opportunities. Yet rather than abandoning the "team", Dr. Herdman has strengthened it further and moreover has revised the "line up" to field an even stronger team that will have even greater appeal to the "fans". Hence at a time when star quality seeks autonomy at the sacrifice of team congruity, the exact opposite has occurred for *Vestibular Rehabilitation, Third Edition*.

Dr. Herdman has painstakingly adhered to the principles of the *Contemporary Perspectives in Rehabilitation Series* by assuring that each contributor has updated references and has, when appropriate, challenged the reader's critical thinking skills. The text is written for any specialist in vestibular rehabilitation or any student or clinician aspiring to become one. While already established as the gold standard for the assessment and management of patients with vestibular disorders, this third edition takes off from where the second edition has left. There are four new chapters and several others have been revamped considerably. The four new additions to this already comprehensive text include: a chapter (12) by Ronald Tusa on Migraine, Ménière's Disease and Motion Sickness that represents a considerable expansion from his chapter on migraine in the second edition and distinguishes these three problem areas and their medical management; a chapter (13) by Timothy Hain and Janet Helminski on Mal de Débarquement Disorder, a problem that has received increasing attention since the second edition of this book and now includes guidelines for treatment and indications that physical therapy may be inappropriate in the treatment of this disorder; a chapter (18) on Compensatory Strategies for the Treatment of Vestibulo-Ocular Hypofunction by Michael Schubert that offers new information on compensatory mechanisms used by patients undergoing vestibular rehabilitation; and a chapter (26) by Ronald Tusa on Non-vestibular Dizziness and Imbalance that uniquely addresses lesions not directly implicated in the central vestibular pathways, including disuse disequilibrium, spino-cerebellar ataxia, leukoaraiosis, and normal pressure hydrocephalus. In fact the last 5 chapters of the third edition are grouped to emphasize assessment and management of disorders either within or external to the central vestibular pathways or in the treatment of non-vestibular dizziness.

Moreover in addition to adding several new contributors including Janet Helminski, Sharon Polensek, Michael Schubert, Greg Marchetti, and Robert Landel, many chapters have undergone substantial revision. Ronald Tusa has converted what had been one presenta-

tion on quantification of vestibular function tests and clinical examination into two exciting chapters presented from the physician's perspective on History and Clinical Examination (7) and Vestibular Function Tests (8). The chapter (24) by Helen Cohen on disability now approaches the concept in a diagnosis specific manner.

Perhaps the most dominating impression created from this unique team is deriving a realization that contributions from vestibular neurorehabilitation therapists and specialty physicians blend almost seamlessly into a continuum of fact pointed toward a comprehensive understanding of the assessment and management of patients with vestibular disorders. In fact, one gets the impression that the content of this book could have easily been extracted from dialogue amongst these interdisciplinary specialists at a symposium or workshop. While students new to this topic might not appreciate the value of such a constellation of knowledgeable professionals, those clinicians familiar with many of these authors and their contributions to vestibular rehabilitation will recognize that within these hard covers lie content the sum of whose parts far exceeds the whole.

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

# Preface

The 3rd edition of Vestibular Rehabilitation! I never expected that the little black book published in 1994 would have multiple editions, much less that we would (or even could) provide a CD with the book to augment the written word with videos of patients. These videos have been chosen to provide the reader with examples of both normal and abnormal clinical tests, with visual examples of some of the exercises used in the treatment of BPPV and of vestibular hypofunction, and with examples of non-vestibular oculomotor and gait signs that should help with differential diagnosis. As I reviewed a multitude of video clips of patients we made over the past 20 years, I found that I remembered these people and their individual personalities and problems. What a wonderful experience this has been and how thankful I am for everything they taught me. If I had only one person to thank, it would be the accumulation of all these people.

Once again, we have extended the material presented to include several new chapters and have augmented the material presented in all the chapters to reflect changes in our understanding of management of vestibular disorders. The new chapters include management of persons with mal de débarquement syndrome, and persons with dizziness that is unrelated to the vestibular system such as disuse disequilibrium and central disorders. The third new chapter presents new information about the mechanisms that underlie compensation for vestibular hypofunction. In addition to these new chapters, there are a number of new topics presented within different chapters such as differential diagnosis in BPPV to identify disorders that mimic BPPV, differentiation among Ménière's, migraine, and motion sensitivity, and the role of chemical labyrinthectomy in the management of episodic vertigo.

Another shift you will find in the book, as well as in many clinical studies, is an increasing use of functional measures, rather than impairment measures, to assess outcome of rehabilitation. Of great value is the International Classification of Functioning, Disability and Health (ICF) scheme (World Health Organization 2001. *http://www.cdc.gov.hchs/about/otheract/ic99/icfhome.htm*). The ICF provides a framework for the "description of health-related states" and includes both positive experiences and negative consequences of disease. It consists of three domains that can be used to describe the effect of different disorders or diseases on a person's health, with a number of environmental and personal contextual factors that may affect each of those domains (Table below).

**HEALTH CONDITION DISORDER OR DISEASE**

| **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 | | | Co-morbidities |
| | Attitude of family | | | Social background |
| | Attitude of society | | | Education and profession |
| | Services, systems, policies | | | Past experience |
| | Products and technology | | | Coping style |

Because it provides a more comprehensive depiction of the health of an individual, the ICF model shifts the emphasis away from impairment and disability to a more balanced perspective.

Finally, we have tried, as in the other editions of Vestibular Rehabilitation, to provide you with an update on evidence that supports our practice. There is an increasing body of research that support different exercise approaches as appropriate and successful tools in the management of patients with vestibular dysfunction. The number of blinded, randomized clinical trials is growing and they provide compelling evidence that we are effectively improving outcome in these patients. Some studies offer guidance in how certain treatment can be modified to simplify treatment for the patient. Still other studies explore the extent of recovery that can be achieved. Some studies offer insight into new methods for identifying the nature of the vestibular dysfunction such as involvement of the utricle and saccule. I expect that in the next 5 to 10 years there will be another great leap in our knowledge and we will have several additional rehabilitation approaches. Researchers are exploring the use of techniques such as virtual reality, sensory substitution devices, vestibular implants, and methods to induce hair cells regeneration. These techniques are not ready yet but the next edition of this book may be filled with wonderful new ways to help people with vestibular disorders.

Susan J. Herdman, PT, PhD, FAPTA
**Editor**

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# Acknowledgments

I would like to express my gratitude to the wonderful colleagues I have worked with in the clinic. Over my years as a physical therapist, they have honed my skills, challenged my assumptions, contributed to my research and made me a better clinician. So my thanks go to Ron Tusa, Courtney Hall, Lisa Gillig, Tim Hain, John Leigh, David Zee, Doug Mattox, Rick Clendaniel, and Michael Schubert.

I also want to thank the authors of this edition of Vestibular Rehabilitation. They have contributed their considerable knowledge and perspectives so we can all learn how best to help the 'dizzy patient'. As a result, many more clinicians will become familiar with the problems and management of vestibular disorders and many, many more patients will receive appropriate treatment.

# Contents in Brief

# Contents

# O·N·E

# Fundamentals

CHAPTER 1

# Anatomy and Physiology of the Normal Vestibular System

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

## Purpose of the Vestibular System

The human vestibular system is made up of three components: a peripheral sensory apparatus, a central processor, and a mechanism for motor output (Fig. 1.1). The peripheral apparatus consists of a set of motion sensors that send information to the central nervous system—specifically, the vestibular nucleus complex and the cerebellum—about head angular velocity and linear acceleration. The central nervous system processes these signals and combines them with other sensory information to estimate head and body orientation. The output of the central vestibular system goes to the ocular muscles and spinal cord to serve three important 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 in order to maintain head and postural stability and thereby prevent falls. The performance of the VOR, VCR and VSR is monitored by the central nervous

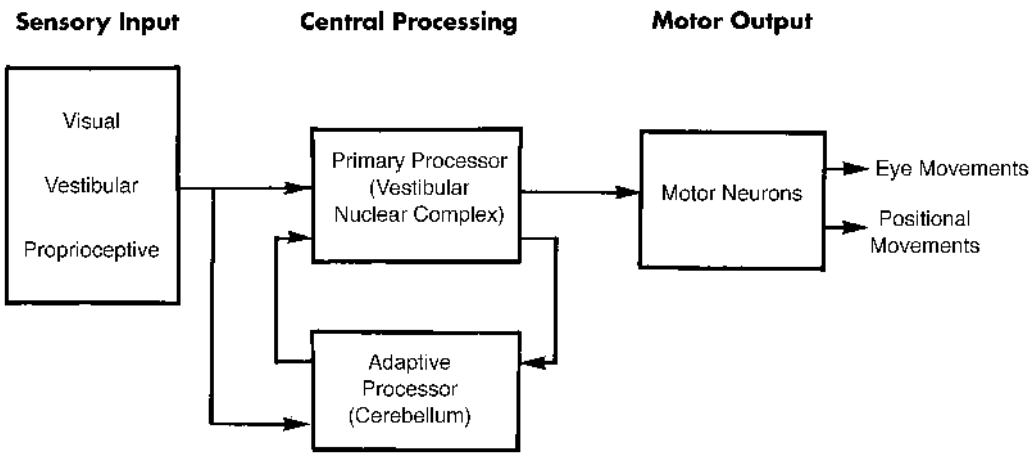


**Figure 1.1** Block diagram illustrating the organization of the vestibular system.

system, is readjusted as necessary by the cerebellum, and is supplemented by slower but more capable higher cortical processes.

From a rehabilitation perspective, it is crucial to realize that because orientation in space is a critical function, multiple fail-safe mechanisms are closely integrated into vestibular responses. The capability for repair and adaptation is remarkable! Two years after removal of half of the peripheral vestibular system, such as by a unilateral vestibular nerve section, finding clinical evidence of vestibular dysfunction is often quite difficult. The ability of central mechanisms to use vision, proprioception, auditory input, tactile input, or knowledge about an impending movement allows vestibular responses to be based on a richly textured, multimodal sensory array.

With these general philosophical considerations kept in mind, the purpose of this chapter is to describe the anatomy and the physiologic responses of the vestibular system, with particular attention to aspects relevant to rehabilitation. We proceed from the peripheral structures to central structures and conclude with a discussion of "higher-level" problems in vestibular physiology that are relevant to rehabilitation.

## The Peripheral Sensory Apparatus

Figure 1.2 illustrates the peripheral vestibular system in relation to the ear. The peripheral vestibular system consists of the membranous and bony labyrinths as well as the motion sensors of the vestibular system, the hair cells. The peripheral vestibular system lies within the inner ear. Bordered laterally by the air-filled middle ear and medially by temporal bone, it is posterior to the cochlea.[1]

### Bony Labyrinth

The *bony labyrinth* consists of three semicircular canals (SCCs), the cochlea, and a central chamber called the *vestibule* (Fig. 1.3). The bony labyrinth is filled with perilymphatic fluid, which has a chemistry similar to that of 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* is suspended within the bony labyrinth by perilymphatic fluid and supportive connective tissue. It contains five sensory organs: the membranous portions of the three SCCs and the two otolith organs, the *utricle* and *saccule*. Note that one end of each SCC is widened in diameter to form an *ampulla*. This

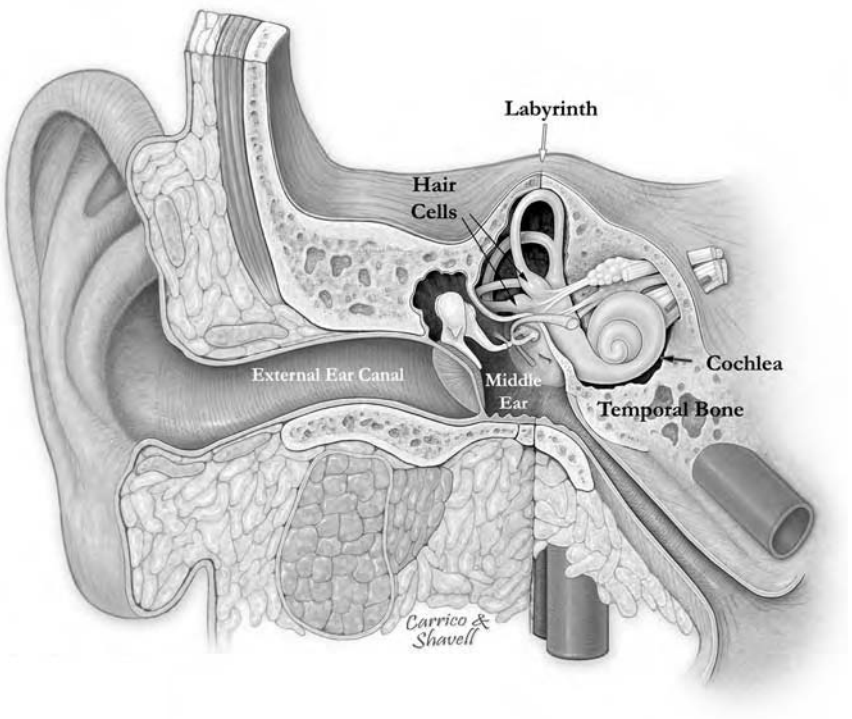


**Figure 1.2** Anatomy of the peripheral vestibular system in relation to the ear. (Illustration adapted from http://www.dizziness-and-balance.com/disorders/hearing/sensorineural.htm, with permission.)

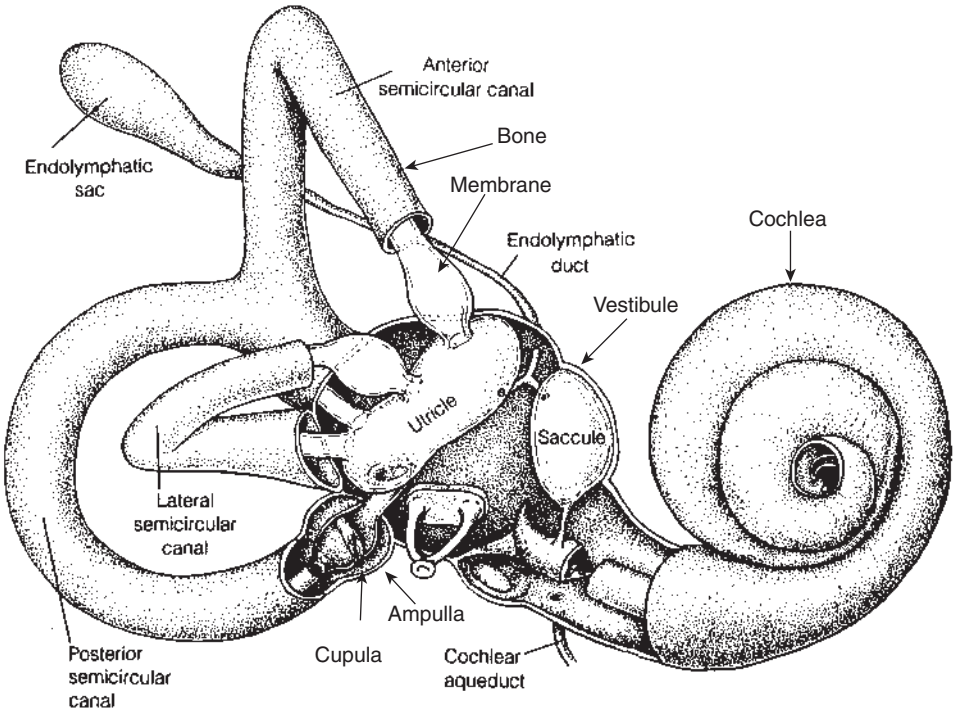


**Figure 1.3** The membranous and bony labyrinths. The *inset* illustrates the perilymphatic and endolymphatic fluid compartments. (Adapted from an illustration by Mary Dersch; originally adapted from Pender, 1992.[2])

widening is relevant to the understanding of a common vestibular condition, benign paroxysmal positional vertigo (see later).

The membranous labyrinth is filled with endolymphatic fluid (see Fig. 1.3). In contrast to perilymph, the endolymph resembles intracellular fluid in electrolyte composition (high K:Na ratio). Under normal circumstances, there is no direct communication between the endolymph and perilymph compartments.

## Hair Cells

Specialized hair cells contained in each ampulla and otolith organ are biological sensors that 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 of the saccule and the floor of the utricle. Each hair cell is innervated by an afferent neuron located 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. Associated with angular head motion, endolymphatic pressure differentials across the cupula cause the cupula to bend back and forth, stimulating the hair cells (Fig. 1.4*B*). [3]

The *otolithic membranes* are structures that are similar to the cupulae but they are also 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. [4]

## 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), 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 com-

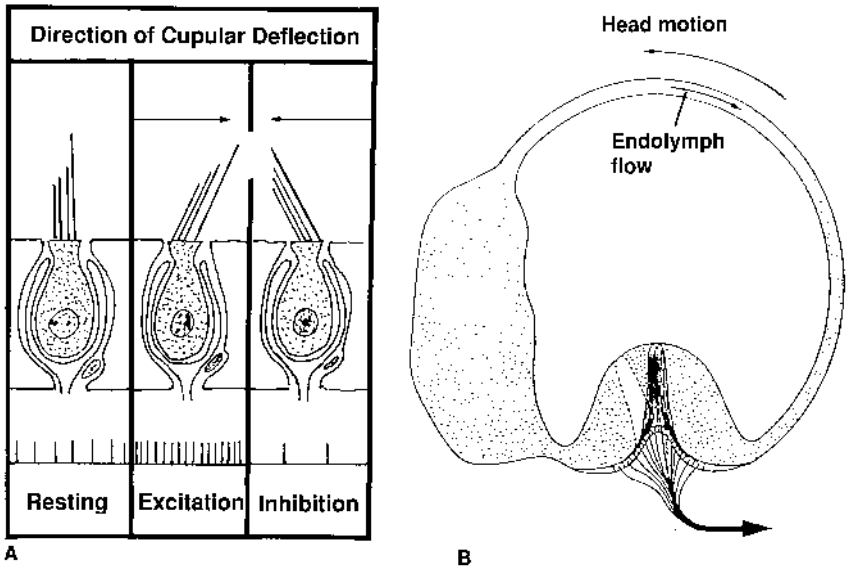


**Figure 1.4** Effects of head rotation on the canals. (*A*) The direction from which hair cells are deflected determines whether or not hair-cell discharge frequency increases or decreases. (*B*) Cross section of the membranous labyrinth illustrating endolymphatic flow and cupular deflection in response to head motion. (From Bach-Y-Rita et al, 1971.[3])

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

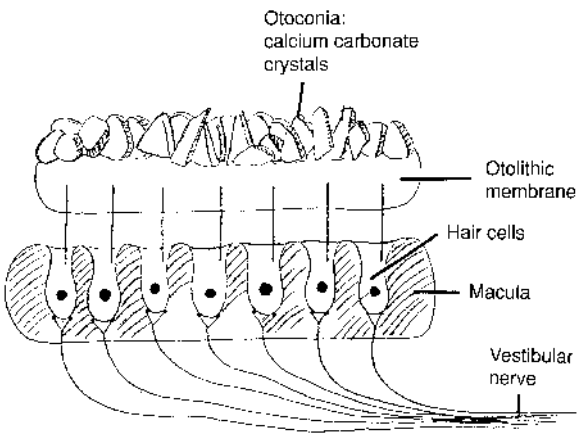


**Figure 1.5** The otolithic macula and its overlying membrane. (From Baloh et al, 1990.[4])

## 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. By virtue 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 eye remains still 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 (0.5–7 Hz). In engineering terms, the canals are "rate sensors."

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, as a perfect rate sensor should, 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 spring-like action of the cupula that tends to restore it to its resting position.[7]

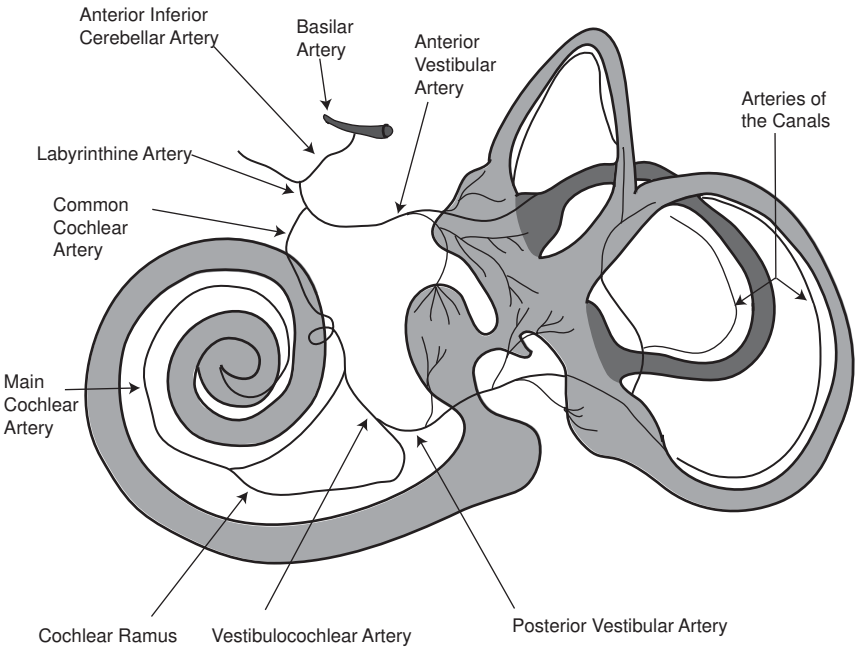


**Figure 1.6** The arterial supply of the labyrinth. (Redrawn from Schuknecht, 1993.[5])

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 SCCs become the following 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 other side. For the lateral canals, displacement of the cupula towards the ampulla (ampullopetal flow) is excitatory.

There are three advantages to the push-pull arrangement of coplanar pairing. First, pairing provides *sensory redundancy*. If disease or surgical intervention affects the SCC input from one member of a pair (e.g., as in vestibular neuritis, or canal plugging for benign paroxysmal positional vertigo), 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 due to changes in body temperature or chemistry. These changes are not related to

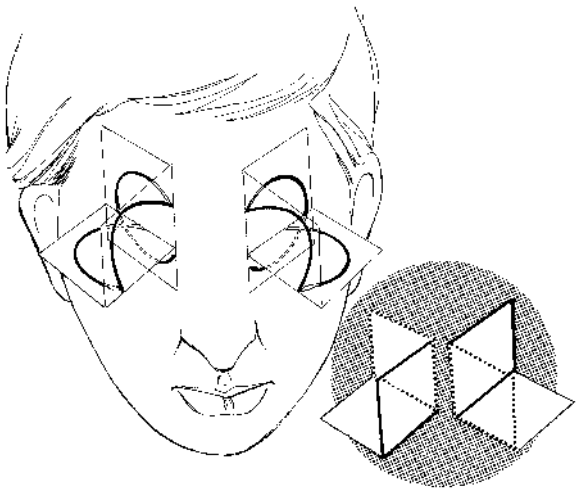

**Figure 1.7** The spatial arrangement of the semicircular canals. The canals on each side are mutually perpendicular, are paired with conjugate canals on the opposite side of the head, and also are closely aligned with the optimal pulling directions of the extraocular muscles.

head motion and are "common-mode noise." The engineering term for this desirable characteristic is "common-mode rejection." Third, as discussed in a later section, a push-pull configuration assists in compensation for sensory 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, he has no sense that he is traveling at 300 miles per hour. However, in the process of taking off and ascending to cruising altitude, he senses the change in velocity (acceleration) as well as the tilt of the plane on ascent. The otoliths therefore differ from the SCCs in two basic ways: They respond to linear motion instead of angular motion, and to acceleration rather than to velocity.[7]

The otoliths have a simpler task to perform than the canals. Unlike the canals, which must convert head velocity into displacement to properly activate the hair cells of the cristae, the otoliths need no special hydrodynamic system. Exquisite sensitivity to gravity and linear acceleration is obtained by incorporation of the mass of the otoconia into the otolithic membrane (see Fig. 1.5). Force is equal to mass multiplied by acceleration, so with incorporation of 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).

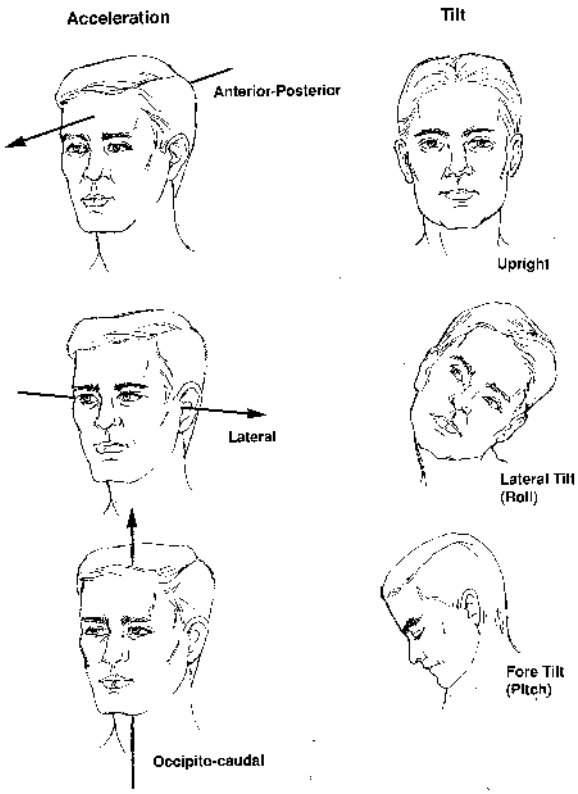


**Figure 1.8** The otoliths register linear acceleration and static tilt.

Like the canals, the otoliths are arranged to enable them to respond to motion in all three dimensions (Fig. 1.9). However, unlike the canals, which have one sensory organ per axis of angular motion, the otoliths have only 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 SCCs). In this posture, the saccule can sense linear acceleration in its plane, which includes the acceleration oriented along the occipitocaudal axis as well as linear motion along the anterior-posterior axis. The utricle senses acceleration in its plane, which includes lateral accelerations along the interaural axis as well as anterior-posterior motion.[8]

The earth's gravitational field is a linear acceleration field, so in a person on the earth, the otoliths register tilt. For example, as the head is tilted laterally (which is also called *roll*; see Fig. 1.8), shear force is exerted upon the utricle, causing excitation, but shear force is lessened upon the saccule. Similar changes occur when the head is tilted forwards or backwards (called *pitch*). Because linear acceleration can come from two sources—earth's gravitational field as well as linear motion—there is a sensor ambiguity problem. We discuss strategies that the central nervous system might use to solve this problem later, in the section on higher-level vestibular processing.

In the otoliths, as in the canals, there is redundancy, with similar sensors 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 tilt increases afferent discharge from one part of a macula while reducing the afferent discharge from another portion of the same macula. This extra level of redundancy in comparison with the SCCs 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

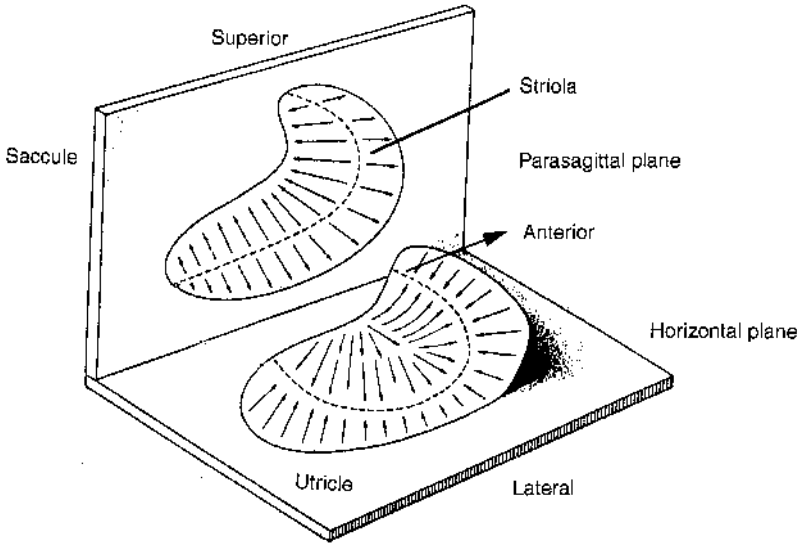


**Figure 1.9** Geometry of the otoliths. (From Barber and Stockwell, 1976.) [8]

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 sensation), and the labyrinthine artery. The IAC travels through the petrous portion of the temporal bone to open into the posterior fossa at the level of the pons. 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, because in experimental animals irregular afferents can be ablated without much change in the VOR. However, 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 a sensitivity to head velocity of about 0.5 spike per degree per second.[10,11] We can speculate about what happens immediately after a sudden change in head velocity. Humans can easily move their heads at velocities exceeding 300 degrees per second (deg/sec). As noted previously, the SCCS are connected in a push-pull arrangement, so that one side is always being inhibited while the other is being excited. Given the sensitivity and tonic rate noted previously, the vestibular nerve, which is being inhibited, should be driven to a firing rate of 0 spikes per second, for head velocities of only 180 deg/sec! In other words, head velocities greater than 180 deg/sec 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 those to rotations that inhibit a canal.[12,13] Cutoff behavior explains why a patient with unilateral vestibular loss avoids head motion toward the side of the lesion. More is said about this issue in the later discussion of 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 vestibular input and implements direct, fast connections between incoming afferent information and motor output neurons. The cerebellum is the adaptive processor; it monitors vestibular performance and readjusts central vestibular 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 VSRs and coordinates head and eye movements that occur together. The lateral vestibular nucleus is the principal nucleus for the VSR. The descending nucleus is connected to all of the other nuclei and the cerebellum but 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 that 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.[14]

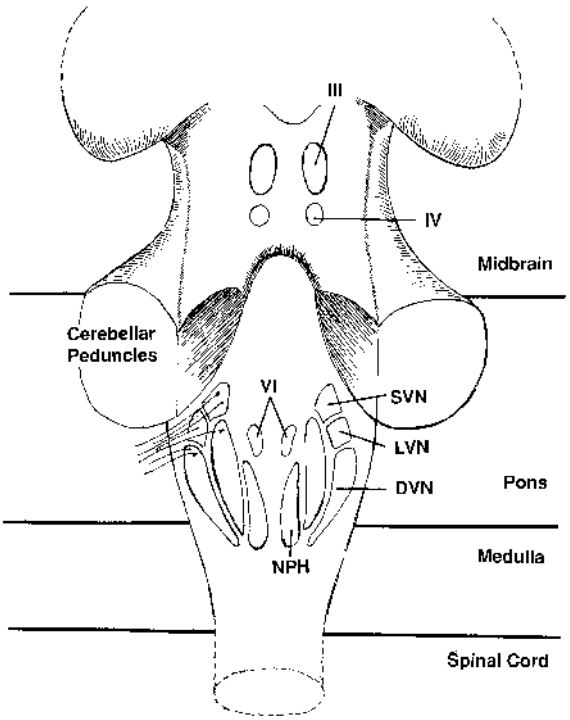


**Figure 1.10** The vestibular nuclear complex. This section shows the brainstem with the cerebellum removed. DVN = descending vestibular nucleus; LVN = lateral vestibular nucleus; NPH = nucleus prepositus hypoglossi; III = oculomotor nucleus (inferior oblique muscle and medial, superior, and inferior rectus muscles); IV = trochlear nucleus (superior oblique muscle); VI = abducens nucleus (lateral rectus muscle). The medial vestibular nucleus (not labeled) lies between the NPH and the DVN. (From Brodal, 1981.[14])

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

## Vascular Supply

The vertebral-basilar arterial system supplies blood to the peripheral and central vestibular system (Fig. 1.11). The

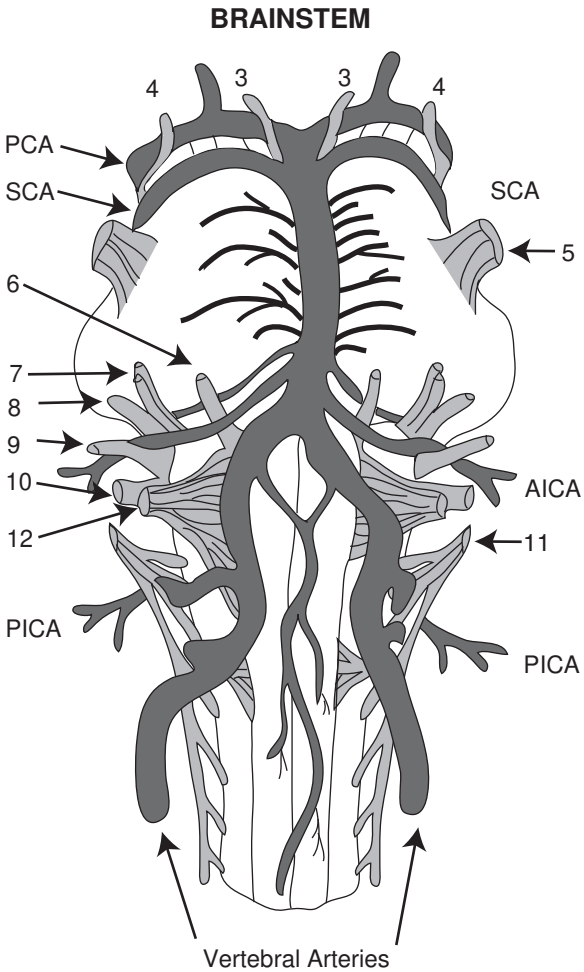


**Figure 1.11** The vertebral-basilar system. AICA = anterior inferior cerebellar artery; PCA = posterior cerebellar artery; PICA = posterior inferior cerebellar artery; SCA = superior cerebellar artery. *Numerals* indicate individual cranial nerve roots (all nerves are paired, but for clarity, both sides are not always labeled here). (© Northwestern University, with permission.)

posterior-inferior cerebellar arteries (PICAs) branch off 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 vestibular system via the labyrinthine artery. The AICA also supplies blood to the ventrolateral cerebellum and the lateral tegmentum of the lower two-thirds of the pons. Recognizable clinical syndromes with vestibular components may appear after occlusions of the basilar artery, labyrinthine artery, AICA, and PICA.

### *Cerebellum*

The cerebellum, a major recipient of outflow from the vestibular nucleus complex, is also a major source of input itself. Although the cerebellum is not required for vestibular reflexes, vestibular reflexes become uncalibrated and ineffective when this structure is removed. Originally, the "vestibulocerebellum" was defined as the portions of the cerebellum receiving direct input from the primary vestibular afferents. We now understand that most parts of the *cerebellar vermis* (midline) respond to vestibular stimulation. The cerebellar projections to the vestibular nuclear complex have an inhibitory influence on the vestibular nuclear complex.

The *cerebellar flocculus* adjusts and maintains the gain of the VOR.[15] Lesions of the flocculus reduce the ability of experimental animals to adapt to disorders that reduce or increase the gain of the VOR. Patients with cerebellar degenerations or the Arnold-Chiari malformation typically have floccular 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 those 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.

Lesions of the anterior-superior vermis of the cerebellum affect the VSR and cause a profound gait ataxia with truncal instability. Patients with such lesions are unable to use sensory input from their lower extremities to stabilize their posture. The lesions are commonly 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.[15] Although a similar structure must exist for the vestibulospinal system, the location of the VSR neural integrator is currently 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. 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.

Two white matter tracts 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) (Fig. 1.12). Because the median longitudinal fasciculus is often injured in multiple sclerosis, this connection may account for central vestibular symptoms in patients with this disorder.[14]

### *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 compli-

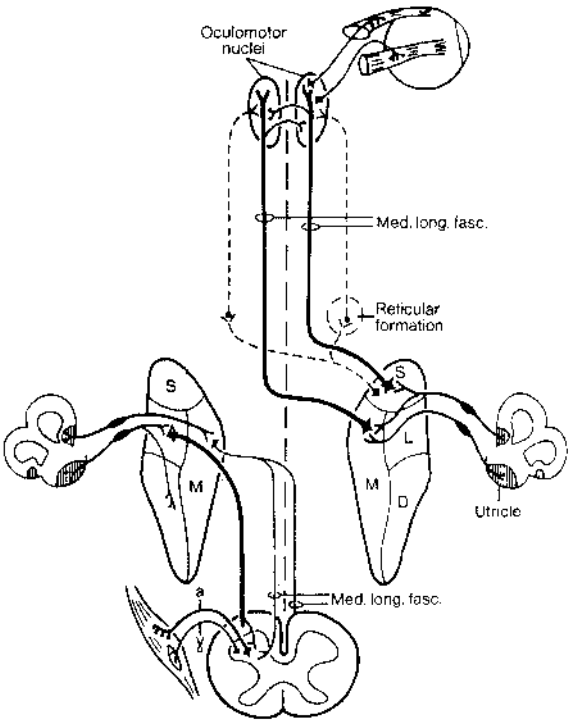


**Figure 1.12** The vestibulo-ocular reflex (VOR) and vestibulospinal reflex (VSR) arcs. S, L, M, and D indicate the superior, lateral, medial, and descending vestibular nuclei, respectively. The lateral vestibulospinal and medial vestibulospinal tracts are shown as *heavy lines* and *light lines*, beginning in the lateral vestibular nucleus and medial vestibular nucleus, respectively. (From Brodal, 1981.[14])

cated than that for the VOR. The VSR has a much more difficult task than the VOR, because multiple strategies involving entirely different motor synergies can be used to prevent falls. For example, when one is shoved from behind, one's center of gravity might become displaced anteriorly. In order to restore "balance," one might (1) plantar-flex 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 later 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.

Three major white matter pathways 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. 1.12). This pathway generates antigravity postural motor activity or protective extension, primarily in the lower extremities, in response to the head position changes that occur with respect to gravity. The *medial vestibulospinal tract* originates from the contralateral medial, superior, and descending vestibular nuclei (see Fig. 1.12) and mediates ongoing postural changes or head righting in response to SCC 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 of the vestibular nuclei as well as from all of 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 it 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 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. This reflex has two components. The angular VOR, mediated by the SCCs, 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 when 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 follows:

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, whereas 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. Simultaneously, contraction of the left lateral rectus and right medial rectus muscles and relaxation of the left medial rectus and right lateral rectus muscles 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 more than 2 deg/sec, the cerebellar projection to the vestibular nuclei 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); these reflexes are discussed in more detail in Chapter 2. As an example of a VSR, let us examine the sequence of events involved in generating a labyrinthine reflex, as follows:

1. When the head is tilted to one side, both the canals and the 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 sensed by the motion sensors.

### *The Vestibulocollic Reflex*

The VCR acts on the neck musculature to stabilize the head. The reflex head movement produced counters the movement sensed by the otolithic or SCC 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.[16] The COR is facilitated when the vestibular apparatus is injured.[17,18] It is rare, however, for the COR to have any clinical significance.

### *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 chin is pointed and flexion of the limb on the contralateral side. Vestibular receptors influence both of these systems by modulating the firing of medullary neurons in a pattern opposite to that elicited by neck receptors. With their interaction, the effects on the body of vestibular and neck inputs tend to cancel one another when the head moves freely on the body, so that posture remains stable.[19]

### *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. The reflex is thought to be primarily a monosynaptic one.[16] The extent to which the CCR contributes to head stabilization in normal humans is cur-

rently uncertain, but it seems likely that the CCR is useful primarily to stabilize head movement in the vertical plane, and it may also be facilitated after labyrinthine loss.

## Visual Reflexes

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

## Somatosensory Reflexes

Somatosensory mechanisms appear to be involved in postural stability as well. Bles and associates documented somatosensory-induced nystagmus ("stepping-around nystagmus").[20] Interestingly, the subjects in their study with bilateral vestibular loss developed a more pronounced nystagmus than normal subjects. This finding implies that subjects with bilateral vestibular loss use somatosensory information to a greater extent than normal subjects.

## *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 now been made in our understanding of BPPV.

Figure 1.14, from Squires and colleagues,[21] 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.[21]

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

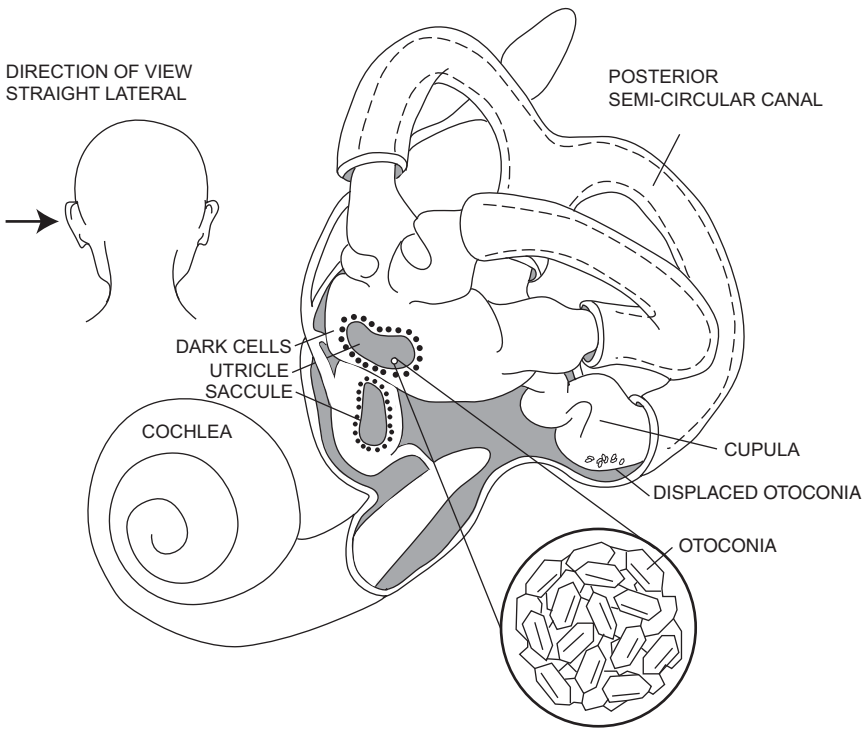


**Figure 1.13** Physiology of benign paroxysmal positional vertigo. Otoconia become displaced from the utricle and relocate to the bottom of the posterior semicircular canal, which is the lowest part of the inner ear. (© Northwestern University, with permission.)

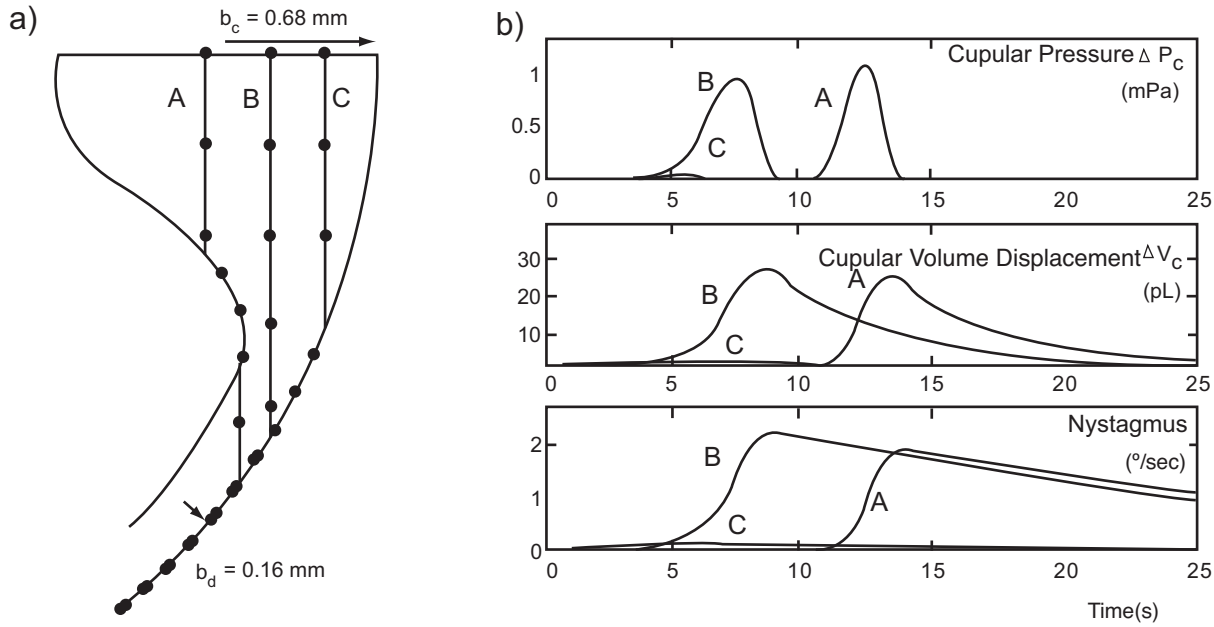


**Figure 1.14** Fluid mechanics of benign paroxysmal positional vertigo. (*A*) Trajectories of three otoconia after a sudden change of head position that makes the posterior canal vertical. Otoconia begin close to the cupula, fall through the ampulla with radius $b_c$, and then enter the duct with radius $b_d$. (*B*) Simulated pressure, displacement, and nystagmus due to otoconia falling with the trajectories of *A*. (From Squires et al, 2004.) [21]

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.[22]

## Higher-Level Vestibular Processing

In this section we identify 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? In order to keep the eye still in space while the head is moving, the velocity of the eyes should be exactly opposite to that of head movement. When this happens, the ratio of eye movement to head movement velocity, called the *gain*, equals –1.0. In order to maintain normal vision, retinal image motion must be less than 2 deg/sec. In other words, for a head velocity of 100 deg/sec, 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 not for low-frequency head motion. This fact can be most easily seen if one considers the response of the SCCs 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; 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, replacing 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*.[23]

The velocity storage mechanism is used as a repository for information about head velocity derived from several kinds of motion sensors. 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.[24] The example discussed here shows how the vestibular system resolves multiple, partially redundant sensory inputs.

## Estimation: Going Beyond Reflexes

*Reflexes* are by definition 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 noisy (such as when a sensor has been damaged)? A mechanism that combines sensory inputs, weights them according to their relevance and reliability, 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. The shuttle has dozens of sensors and motors. Some sensors respond quickly, and some slowly. They may differ in accuracy, scaling, coordinate frame, timing, and noise characteristics. No single sensor can provide a complete picture of the shuttle's state. A mechanism is needed to integrate sensor output and to develop an internal estimate of the state of the system (i.e., position, velocity, acceleration) in order to keep the shuttle on the desired course and heading.

The engineering solution to this problem developed out of work performed by Kalman and is often called a *Kalman filter*. It is also commonly called an "optimal estimator" or an "internal model." The essentials of the Kalman filter are shown in Figure 1.15. There is considerable evidence that mechanisms similar to Kalman filters are used for human sensorimotor processing.[25]

The Kalman filter is far more powerful than a simple reflex. Several key concepts must be considered before one can understand how it is superior. First, internal models of sensors and motor output are used to develop an estimate of the current sensory and motor state. These internal models are adjusted according to experience and

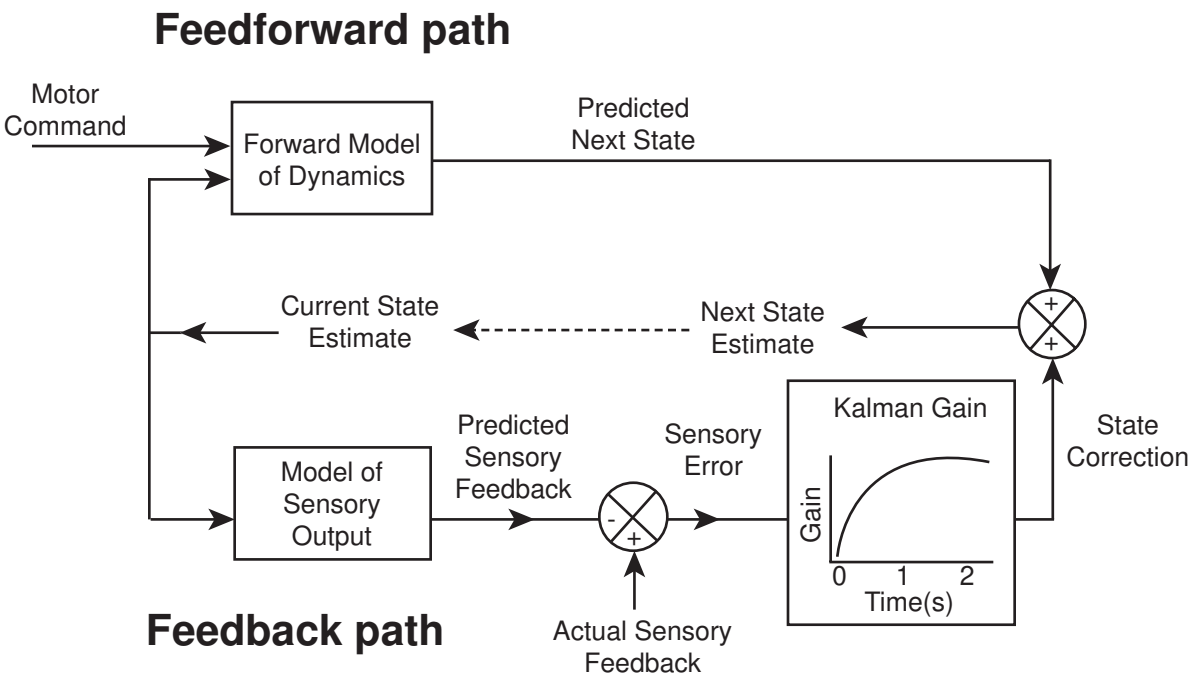


**Figure 1.15** Block diagram showing a Kalman filter such as may be used by the body for sensorimotor integration. Sensory inflow and motor outflow are used to estimate the current state. (From Wolpert and Miall, 1997.) [25]

must track changes in bodily function. It seems likely that vestibular rehabilitation affects internal models.

Second, sensory input is not used to directly compute body state, but rather, the difference between sensory input and predicted sensory input is used to correct the current estimate of body state. This design allows the Kalman filter to easily combine multiple sensor inputs—from eyes, ears, and somatosensors. The Kalman filter continues to work even in the absence of a sensory input, because it uses its estimate when the sensor is missing. Both of these highly desirable features make the Kalman filter far superior to a simple assemblage of reflexes.

The *Kalman gain* weights the extent to which a sensory input affects the ongoing state estimate. This weighting provides a method of adjusting for the salience and reliability of sensory streams. It seems highly likely that vestibular rehabilitation adjusts the Kalman gain.

Overall, this sort of mechanism is clearly far superior to vestibular reflexes: Although not as fast, it can be far more accurate, it functions even in the absence of sensory input, and it is modifiable by experience and rehabilitation.

## Higher-Level Problems of the Vestibular System

### Compensation for Overload

Humans can easily move their heads at velocities exceeding 300 deg/sec. Consider, for example, driving a car. When one hears a horn to the side, one's head may rapidly rotate to visualize the problem and, potentially, 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 high-velocity signals. The reason is the cutoff behavior discussed in the earlier section on 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 arranged in "push-pull" configuration with the inhibited side. Whereas the inhibited side can be driven only 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 in patients with unilateral vestibular loss, this mechanism is not available to deal with the overload problem, and they are commonly disturbed by rapid head motion toward the side of the lesion.

### Sensor 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 whether 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 graphically 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 through the use of a Kalman filter as discussed previously. As long as one does not have to make a quick decision, these senses may be perfectly adequate. However, visual input takes 80 msec to get to the vestibular nucleus and tactile input must be considered in the context of joint position and of the 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 but linear accelerations are brief. Neural filters that pass low or high frequencies can be used to tell 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.[26] 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

The phenomenon of motion sickness illustrates how the brain routinely processes multiple channels of sensory information simultaneously. The *motion sickness syndrome* consists of dizziness, nausea or emesis, and malaise after motion. It is thought to be caused by a conflict between movement information in related sensory channels, such as visual-vestibular conflict 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, allowing 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."[27] 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 the force 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 one is outside earth's gravitational field, like 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 now untrue, causing the motion sickness syndrome.[27,28]

Structures that appear to be essential for the production of motion sickness are (1) intact labyrinth and central vestibular connections, (2) cerebellar nodulus and uvula that coordinate labyrinthine stimuli, (3) the chemoreceptive trigger zone located in the area postrema, and (4) the medullary vomiting center.[29] Why some people 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 sensory apparatus and the constraints of physics. In normal individuals, these problems can be satisfactorily resolved by relying on redundancy of sensory input and central signal processing. In addition to these intrinsic problems, there are extrinsic problems 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 regarded as an integral part of its physiology; for this reason, it is our final topic.

Adaptive plasticity for peripheral vestibular lesions is dealt with elsewhere in this volume. Suffice it to say here that repair is amazingly competent, even enabling the vestibular system to adapt to peculiar sensory situations requiring a reversal of the VOR.[30] Adjustments of internal models and weighting of sensory inputs (e.g., Kalman gain) are probably at least as important as readjustment of reflexes, because internal models provide many important features that reflexes cannot provide (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, there is a high degree of *context dependency* to 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.[31] Similarly, when the VOR of cats is trained with the use of head movements of low frequency, no training effect is seen at high frequencies.[32]

Another type of context dependency relates to the VSRs 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 in which the head is turned 90 degrees 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 degrees. 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 in which the head assumes other postures, such as looking down at the 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 television breaks down you can take it to the repair shop and get it fixed. If, however, your television is broken and the repair shop is out of business, 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 that 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 difficult as well as critical to survival. Not surprisingly, the body uses multiple, partially redundant sensory inputs and motor outputs in combination with central state estimators and competent central repair. The system as a whole can withstand and adapt to major amounts of peripheral vestibular dysfunction. The weakness of the vestibular system is a relative inability to repair central vestibular dysfunction.

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CHAPTER 2

# Vestibular Adaptation

*David S. Zee, MD*

A robust and versatile capability for adaptive control of vestibular motor behavior is essential if an organism is to maintain optimal visual function and stable balance throughout life. Changes associated with normal development and aging, as well as with disease and trauma, demand mechanisms both to detect errors in performance and to correct them. An understanding of such mechanisms is crucial in the design and evaluation of programs of physical therapy that are used to rehabilitate patients with vestibular disorders. The purpose of this chapter is to review information about adaptive mechanisms that bears on the management of patients with disorders of the vestibular system. By necessity, this discussion emphasizes studies of the vestibulo-ocular reflex (VOR), because more is known about its adaptive control. When possible, however, vestibulospinal (VSR), vestibulocollic (VCR), and cervico-ocular (COR) reflexes are also discussed.

## Recalibration, Substitution, and Alternative Strategies

Adaptive control of vestibular reflexes must be regarded in the larger context of overall compensation for vestibular deficits. Restoring adequate motor behavior by readjusting the input-output relationships (e.g., gain, timing, or direction) of the VOR or VSR may be impossible, especially when deficits are large. Other mechanisms of compensation must then be invoked (Box 2-1). Examples are as follows:

- Substitution of another sensory input to drive the same motor response (e.g., substitution of a spared otolith-driven response in the absence of canal-driven responses, or the substitution of the COR for an absent angular VOR)
- Substitution of an alternative motor response in lieu of the usual compensatory motor response (e.g., use of saccades instead of slow phases to help stabilize gaze during head motion)
- The use of strategies based on prediction or anticipation of intended motor behavior (e.g., preprogramming compensatory slow phases *in anticipation of* a combined eye-head movement toward a new target)
- Furthermore, there is considerable variability among subjects as to which particular mechanisms are primarily used for compensation.[1] This heterogeneity dictates a need for quantitative evaluation of vestibular function in patients before and during rehabilitation. Any plan of therapy must focus on what is likely to work best and what is working best. The promotion of different goals may require a different therapeutic emphasis, and different therapeutic programs may potentially work at cross purposes. Therefore, it is essential to plan the program of

Box 2-1

**COMPENSATORY MECHANISMS**

Adaptation: changing the gain, phase, or direction of the vestibular response Substitution of

- Other sensory input mechanisms (e.g., cervico-ocular reflex for absent VOR, otolith-ocular response for absent canal-ocular response)
- Alternative motor responses (e.g., saccades or pursuit)
- Strategies based on prediction or anticipation

therapy on the basis of (1) what is most likely to succeed in the context of the type and the degree of deficit and (2) the patient's inherent potential for compensation.[2] To illustrate these points, I first discuss two archetypal paradigms requiring vestibular compensation: unilateral loss and bilateral loss of labyrinthine function.

## *Compensation after Unilateral Labyrinthectomy*

Unilateral labyrinthectomy (UL) has been used as an experimental model to study motor learning and compensation for more than 100 years. Yet, until recently, little has been known about the error signals that drive the compensatory process, the precise mechanisms—both physiological and molecular—that underlie recovery from both static and dynamic disturbances created by the loss of labyrinthine input from one side, and the additional strategies that are available to assist in the overall goal of gaze and postural stability during movement.[3–5]

Let us use the VOR as an example. UL creates two general types of deficits for which a correction is required: static imbalance, related to the differences in the levels of tonic discharge within the vestibular nuclei, and dynamic disturbances, related to the loss of one half of the afferent input that normally contributes, in a push-pull fashion, to the generation of compensatory responses during head movement. Spontaneous nystagmus with the head still and sustained torsion (ocular counter-roll) with the head still are examples of the static type of disturbances that occur from an imbalance in inputs from the semicircular canal and from the otoliths, respectively. Decreased amplitude (gain), abnormally directed slow phases (i.e., misalignment between the axes of rotation of the eye and head), and a left-right asymmetry of eye rotation during yaw axis head rotation are examples of the dynamic type of disturbance.

### The Error Signals that Drive Vestibulo-ocular Reflex Adaptation

What might be the error signals that drive VOR adaptation? Image motion or "slip" on the retina, when associated with head movements, is the obvious candidate; the raison d'être of the VOR is to keep images stable on the retina during head movements. My colleagues and I investigated the role of vision, and especially of visual information mediated by geniculostriate pathways, on both the acquisition and the maintenance of adaptation to UL in monkeys.[6,7] VOR function was recorded in three groups of animals. One group had undergone bilateral occipital lobectomy many months before the labyrinthectomy. These "cortically blind" animals were allowed normal light exposure after UL. The other animals had experienced no prior lesions and were divided into two groups: those that were kept in complete darkness for 4 days after UL and those that were allowed normal exposure to light after the UL.

### Restoration of Static Vestibulo-ocular Reflex Balance after Unilateral Labyrinthectomy

The results from this experiment were clear-cut. Restoration of static balance, as reflected in the disappearance of spontaneous nystagmus, proceeded independently of whether or not the animals had undergone a prior occipital lobectomy and whether or not they were kept in the dark after UL. Furthermore, a bilateral occipital lobectomy performed nearly a year after the labyrinthectomy, when compensation to UL had taken place, did not result in the reappearance of spontaneous nystagmus. Thus, both the acquisition and the maintenance of static balance in semicircular canal–mediated VORs after UL are independent of visual inputs. Another example of restoration of static imbalance after UL is the elimination of cyclotorsion [ocular counter-roll] that follows UL.[8] This bias arises from imbalance in otolith-ocular reflexes. Whether this readjustment also requires visual inputs is not known.

### Restoration of Dynamic Vestibulo-ocular Reflex Function after Unilateral Labyrinthectomy

On the other hand, the restoration of dynamic performance after UL depended critically on visual experience in these experiments.[6,7] There was no increase in the amplitude (gain) of the VOR until exposure to light. Similar results occur in monkeys after unilateral plugging of a lateral semicircular canal, which causes a nearly 50%

decrease in VOR gain.[9] Presumably, the critical stimulus for recalibration of dynamic VOR responses is not light per se but, rather, the presence of motion of images on the retina during head movements. A similar dependence on vision has been shown for recovery of balance in cats after vestibular neurectomy.[10]

Furthermore, my colleagues and I found that the compensatory changes in the gain of the VOR that occurred after UL were lost, over the course of several weeks, after a bilateral occipital lobectomy.[6,7] The occipital lobectomy itself was unlikely to have been responsible for the decrease in the gain of the VOR, because only small alterations in the gain and in the symmetry of the VOR are produced by occipital lobectomy in animals that have not undergone any prior vestibular lesions.

Taken together, these findings suggest that adaptation of VOR gain is a dynamic process that requires visual experience for its acquisition and depends on the posterior cerebral hemispheres for its maintenance. Presumably, the contribution of the occipital lobes is that they transmit information about image slip on the retina during head movements to more caudal structures, which in turn use this error information to readjust the dynamic performance of the VOR. The nucleus of the optic tract (NOT) in the pretectal region of the midbrain is one such structure shown to be important in VOR adaptation. The NOT receives visual information from the cerebral cortex as well as directly from the retina. It has a strong projection to the inferior olive and to other brainstem nuclei that also project to the cerebellum. Lesions in the NOT interfere with compensation to UL[11] and VOR adaptation in general.[12]

The NOT does more, however, than simply relay visual information for adaptive changes in the VOR, because lesions in the NOT also lead to change in the baseline VOR gain. Whether the NOT is also important for maintaining long-term adaptive changes is not known, but presumably it is. The cerebellum must be constantly apprised of the need for adaptive recalibration, and visual inputs are critical. Even in a patient who can "see" in the usual sense, lesions in the brainstem interfere with the transmission of visual information to the cerebellum, so the VOR can still become uncalibrated.

Other studies have shown that recovery after unilateral labyrinthine lesions is less complete when stimuli are composed of high accelerations or high frequencies.[5] Different strategies are needed, especially preprogramming of movements similar to those movements shown by patients who have lost labyrinthine function bilaterally. Indeed, a major implication of much recent research on recovery from unilateral vestibular loss is how important the saccade contribution really is. These issues are discussed later along with saccade strategies.

## Restoration of Spontaneous Activity in the Deafferentiated Nuclei

One might ask why visual experience is necessary for recalibration of dynamic VOR function but not for restoration of static balance. Without motion of images on the retina during head movements, adaptive mechanisms do not have a reliable error signal that they can use to recalibrate the dynamic VOR. With respect to static imbalance, however, deafferentation of the vestibular nucleus might (1) initiate alterations in the intrinsic properties of the vestibular neurons themselves, (2) lead to denervation hypersensitivity to remaining sensory inputs, or (3) stimulate sprouting of extralabyrinthine afferents.[3,13,14] Each process might result in a rise in the level of the spontaneous activity of neurons on the lesioned side. One should recall that restoring vestibulospinal balance also depends on rebalancing of the vestibular nuclei and that many neurons within the vestibular nuclei have axons that bifurcate and project both rostrally, to the ocular motor nuclei, and caudally, to the spinal cord.[15] Vestibulospinal compensation would not be expected to rely solely on visual inputs to provide the necessary error signals for central readjustment of balance.[16] Proprioceptive and somatosensory cues are probably more important and could provide the requisite error signals leading to the static rebalancing of the vestibular nuclei that would affect both vestibulo-ocular and vestibulospinal responses.

## A "Critical" Period for Vestibulo-ocular Reflex Adaptation

Another finding, potentially of important clinical relevance, emerged from our study in labyrinthectomized monkeys. Monkeys that had been kept in the dark for 4 days after UL and then allowed normal visual experience generally showed a recovery of dynamic VOR performance at about the same rate, and to the same level, as monkeys allowed visual experience immediately after UL. There was, however, one important exception: For high-velocity rotations directed to the lesioned side, recovery was markedly delayed in the monkeys initially deprived of vision, although the final level reached was close to that of monkeys that had not been deprived of vision immediately after UL. This finding suggests that there may be a "critical period" during which, if error signals are not provided to the adaptive mechanism and recalibration does not get under way, the rate and, perhaps, the ultimate extent of recovery may decrease. Restoration of postural control after UL also appears to be subject to a critical period.[17] If animals are immobilized for a period of seven days after UL, their recovery of locomotor func-

tion, even after normal activity is reinstituted, is delayed and limited.

The obvious implication of these findings is that when patients incur enduring vestibular damage, they should be encouraged to move about in the light and to try to engage their VORs and VSRs as much as possible. In this way, they will generate and experience the sensorimotor mismatches that make the central nervous system aware of a need for adaptive readjustment of its motor reflexes. As a further caveat, heavy sedation, immobilization, and restriction of activity of patients with a recent loss of vestibular function should be discouraged. Such practices may potentially retard or even limit the ultimate level of compensation after a vestibular lesion.

In the same vein, physical exercise promotes recovery of balance function in monkeys that have undergone either unilateral or bilateral experimental vestibular ablations.[18] Although carefully controlled studies of the effects of different types of activity on vestibular compensation are sparse in human beings, physical therapy most likely also promotes recovery in patients. Most evidence is based on evaluations of balance,[19–26] but there is some evidence that vestibulo-ocular function, too, may improve with physical therapy.[26,27]

## Substitution of Saccades for Inadequate Compensatory Slow Phases

Although we usually think of the main function of the VOR as stabilizing images on the retina, getting the image near the fovea and keeping it relatively close there, by whatever strategy, can be the optimal strategy for preserving vision during head motion. This is especially true when the brain is confronted with trying to compensate for a severe loss of function, either bilateral or unilateral. Because of the inherent limitations with respect to maximum frequency, velocity, and acceleration, the ability of subjects with just one labyrinth to generate slow phases of the correct magnitude when the head is rotated toward the side of the lesion may be limited (Ewald's second law).[28–30] Patients may learn to generate catch-up saccades (elicited, automatically, even in complete darkness) in the same direction as their inadequate compensatory slow phases. A number of factors influence which saccade strategies are invoked. Perhaps most important are whether the movements are active or passive and the ability to predict and invoke the gaze stabilizing strategy that is best suited for the upcoming pattern of head motion.[31–34] The severity of deficit, whether it was acquired suddenly or gradually, and the duration of the deficit are likely key factors as well.

## Recovery Nystagmus and Related Phenomena

A practical clinical implication of these ideas about recovery from unilateral labyrinthine lesions relates to the phenomenon of *recovery nystagmus*. As indicated previously, the static imbalance created by a unilateral peripheral lesion leads to a spontaneous nystagmus that is compensated by a rebalancing of the level of activity between the vestibular nuclei. If peripheral function should suddenly recover, central adaptation becomes inappropriate, and a nystagmus would appear with slow phases directed away from the paretic side. Recovery nystagmus may confuse the clinician or therapist, who may think that the reappearance of symptoms and of a spontaneous nystagmus are due to a loss of function on the previously healthy side rather than to a recovery of function on the previously diseased side. In either case, a spontaneous nystagmus ensues that must be eliminated, adaptively, by a further readjustment of levels of activity within the vestibular nuclei.

An analogous circumstance—although rather than recovery, there is further damage after compensation—occurs if the other labyrinth is destroyed after recovery from an initial unilateral labyrinthine lesion. In this case, a deficit occurs as if the original damaged labyrinth were intact. This *Bechterew's phenomenon* reflects the rebalancing of central vestibular tone after the first lesion. The second lesion then creates a new imbalance.[35,36] These clinical examples illustrate the importance of taking into account the actions of adaptive mechanisms in interpretations of patterns of nystagmus that do not seem to reflect the effects of the sudden onset of a new lesion.

A similar mechanism may account for some instances in which nystagmus is provoked by hyperventilation.[37] In these cases, the induced alkalosis alters the amount of calcium available for generating action potentials along the vestibular nerve and, so, may lead to a restoration of function on partially demyelinated fibers (for example, due to multiple sclerosis, chronic microvascular compression, petrous bone tumors such as cholesteatoma, or cerebellopontine angle tumors such as acoustic neuroma). Hyperventilation, however, may also produce nystagmus in patients with a perilymphatic fistula, by an alteriation in intracranial pressure that is then transmitted to the labyrinth. Other mechanisms for hyperventilation-induced nystagmus are possible, including alterations in compensatory mechanisms.[38,39]

There may even be a dynamic equivalent of recovery nystagmus. After sustained head shaking, patients who have a unilateral peripheral vestibular loss often demonstrate a transient eye nystagmus with slow phases

directed toward the impaired ear, so-called head-shaking nystagmus.[40] The initial phase of head-shaking nystagmus may be followed by a reversal phase in which nystagmus is directed opposite to the initial phase. The primary phase of head-shaking nystagmus is related to a dynamic asymmetry in the VOR such that excitation (rotation toward the intact side) elicits a larger response than inhibition (rotation toward the impaired side). Head-shaking nystagmus is another manifestation of Ewald's second law. The secondary phase is often attributed to short-term (with a time constant of about 1 minute) vestibular adaptation, probably reflecting adaptive processes in both the peripheral nerve and central structures.

If recovery occurs peripherally, however, any prior central VOR gain adaptation (which would increase the dynamic response during rotation toward the impaired side) could lead to a head-shaking–induced nystagmus with the slow phases of the primary phase directed away from the impaired ear. Thus, one can appreciate the pivotal role that adaptation plays in determining the particular pattern and direction of any spontaneous or induced nystagmus that may appear during the process of recovery.

## Bilateral Vestibular Loss

An even more challenging problem for the central nervous system is to compensate for a complete bilateral loss of labyrinthine function (BL). In the case of a truly complete loss, there is no labyrinthine-driven VOR to recalibrate, so alternative mechanisms, such as sensory substitution, motor substitution, and predictive and anticipatory strategies, must be invoked.

Whether or not labyrinthine function is completely absent helps determine what type of program of physical therapy should be prescribed. Because the response of the VOR to high frequencies of head rotation is usually spared until the labyrinthine function loss is complete, caloric responses (which primarily simulate a low-frequency rotational stimulus) may be absent even when the patient has a relatively functional VOR. One should recall that response to the high-frequency components of head rotation requires the fast-acting labyrinthine-driven VOR; visual stabilizing reflexes are adequate only to ensure stabilization for the low-frequency components of head rotation. Accordingly, the results of a rotational test can help determine the level of preservation of labyrinthine function.[41] Such information may help the therapist decide whether to prescribe a program of rehabilitation that stresses recalibration of a deficient but present VOR or a program that emphasizes sensory substitution and alternative strategies.

## Experimental Results in Nonhuman Primates

Dichgans and colleagues[42] first described compensatory mechanisms of eye-head coordination in monkeys that had undergone bilateral labyrinthine destruction. These researchers identified the following three major adaptive strategies used to improve gaze stability during head movements: (1) potentiation of the COR (neck-eye loop), as reflected in slow-phase eye movements elicited in response to body-on-head (head stable in space) rotation, (2) preprogramming of compensatory slow phases in anticipation of intended head motion, and (3) a decrease in the saccadic amplitude–retinal error relationship, selectively during combined eye-head movements, to prevent gaze overshoot. With respect to the last phenomenon, saccades made with head movements would normally cause gaze to overshoot the target if there were no functioning VOR to compensate for the contribution of the movement of the head to the change in gaze. Dichgans and colleagues[42] also showed that saccades were programmed to be smaller when an accompanying head movement was also anticipated. When the head was persistently immobilized, however, saccades were programmed to be their usual size.

### Additional Strategies in Human Patients with Bilateral Loss of Labyrinthine Dysfunction

The compensatory mechanisms described for monkeys without labyrinthine function have also been identified in human beings who have a complete BL.[43–47] How much one or another mechanism is adopted, however, varies from patient to patient. In addition, several other strategies have been identified in humans that are also quite idiosyncratic from patient to patient (Box 2–2).[48–51] These strategies are as follows:

1. Substitution of small saccades in the direction opposite to head rotation, in order to augment inadequate compensatory slow phases.
2. Enhanced visual following reflexes.
3. Predictive strategies to improve gaze stability during tracking of targets jumping periodically or during self-paced tracking between two stationary targets.
4. Effort of spatial localization, as judged by a much better compensatory response to head rotation when the patient imagines the location of stationary targets, as opposed to the response while the patient performs mental arithmetic.

Box 2-2

**COMPENSATORY MECHANISMS AFTER BILATERAL VESTIBULAR DEFICITS**

- Potentiation of cervico-ocular reflex
- Central preprogramming of slow phases or saccades
- Decreased saccade amplitude
- Enhanced visual following
- Effort of spatial localization
- Suppression of perception of oscillopsia

As a corollary to these last two mechanisms, responses during active (self-generated) head rotations occur at a shorter latency, and usually with a larger gain, than those during passive head rotations.[33,34,52] Even somatosensory cues from the feet can be used to augment inadequate compensatory slow phases of the eyes—producing so-called stepping-around nystagmus.[53] Finally, perceptual mechanisms—suppression of oscillopsia (illusory movement of the environment) in spite of persistent retinal image motion—may also be part of the "compensatory" response to vestibular loss.[54,55]

Another example of potentiation of a reflex that is normally vestigial in human beings can be shown in the ocular motor response to static lateral tilt of the head. This presumably otolith-mediated reflex is composed of a static torsion of the eyes opposite to the direction of lateral head tilt—so-called ocular counter-rolling. In labyrinthine-defective human beings, but not in normal subjects, ocular counter-rolling can be produced by lateral tilt of the trunk with respect to the (stationary, upright) head.[56] This response probably reflects a potentiation of a static COR in lieu of the missing otolith signals. Patients with BL also show a greater sensitivity to visually induced tilt. With the loss of the inertial frame of reference normally provided by the dominating labyrinthine inputs, visual inputs become more potent stimuli to vestibular reflexes.

A new reliance on visual inputs is a necessary and useful adaptation to labyrinthine loss, although it may become a liability if visual inputs should become incongruent with the actual motion or position of the head. For example, when one is reading a newspaper in a moving elevator or escalator, visual inputs (from the motion of the image of the newspaper on the retina) provide misleading information about the position or the movement of the head that, if relied upon (as may occur in patients with BL), would lead to an inappropriate (or lack of) compensatory response and, possibly, a fall.

## Studies of Vestibulo-ocular Reflex Adaptation in Normal Subjects

Adaptive control of the VOR has been investigated in normal subjects through artificial creation of motion of images on the retina, by optical or other means, during head rotation. For example, in the pioneering experiments by Gonshor and Melvill Jones[57,58] subjects wore right-left reversing prisms, which required and led to a reversal in the direction of the slow phase of nystagmus with respect to the direction of head motion. Likewise, magnifying and minifying spectacle lenses (used to correct for farsightedness and nearsightedness, respectively) require and produce adaptive increases and decreases, respectively, in VOR gain. A practical consequence of this phenomenon is that normal subjects wearing a spectacle correction undergo an adaptive change in VOR gain to meet the needs of the new visual circumstances created by their optical correction. Such gain changes must be considered in the evaluation of the results of vestibular function tests in individuals who habitually wear spectacles.[59] Furthermore, if a patient does show a change in VOR gain in response to wearing spectacles, one can infer that the patient has at least some capability to undergo adaptive VOR recalibration.

VOR adaptation can also be studied via prolonged rotation of subjects during artificial manipulation of the visual surround. One can use an optokinetic drum that surrounds the subject and rotate it either in phase—in the same direction as chair rotation—to produce a decrease in VOR gain, or out of phase—opposite the direction of chair rotation—to produce an increase in VOR gain. If the amplitude of drum rotation is exactly equal to that of the chair, the required VOR gain would be 0.0 for in-phase viewing (so-called x0 viewing) and 2.0 for out-of-phase viewing (so-called x2 viewing). The usual duration for VOR training in these types of paradigms is an hour or two, although VOR adaptation can probably be detected within minutes of the onset of the change in the relationship between the visual and vestibular stimuli.[60]

A potentially important practical application of vestibular rehabilitation comes from a recent novel finding related to "consolidation" in a short-term learning paradigm used to stimulate adaptation of saccade amplitude. Frequent but short (a minute or so) rest periods during about 20 or 30 minutes of training were found to

considerably enhance the rate and degree of adaptation.[61] Potentially, then, a similar strategy might be attempted with vestibular rehabilitation exercises.

## Imagination and Effort of Spatial Localization in Vestibular Adaptation

Finally, we note that one's imagination can be a potent substitute for the real stimulus to VOR adaptation—motion of images on the retina during head rotation. Melvill Jones and colleagues[62] have shown that the VOR of normal subjects can be adaptively modified (as measured in darkness and tested under the same mental set) with just a few hours of imagining a visual stimulus moving in such a way that it would normally create slip of images on the retina if it were actually visible. Indeed, simply imagining body rotation can lead to compensatory slow-phase eye movements.[63] Along these lines, Eggers and associates[64] have shown that just paying attention to, without even looking at, the new location of a target after a head rotation (i.e., using a position rather than a retinal image motion error) leads to adaptive modification of the VOR.

Thus, what are usually called psychological factors—motivation, attention, effort, and interest—may actually play a more specific role in promoting adaptive recovery than originally believed. The habit of a professional athletes—downhill skiers or ice skaters, for example—of going through their routes or routines in their minds as they prepare for actual events is probably an example of using this "cognitive" capability to create an internal model of the external environment (and the sensory consequences of moving within it) in which to rehearse and improve motor performance.

Similar types of paradigms, in which the motion of the visual surround is artificially manipulated with respect to the motion of the head, have been used to induce an alteration in the phase (timing) of the angular or translational VOR[65,66] or a change in the direction of the VOR, so-called cross-axis plasticity.[67–69] In the latter paradigm, the visual surround is rotated around an axis orthogonal to that of rotation of the head. This cross-axis plasticity accords with electrophysiological evidence that secondary neurons in the vestibular nucleus receive inputs from one, two, or all three pairs of semicircular canals.[70]

Similar considerations account for the recovery in VOR function when just one single semicircular canal is plugged. The spatial tuning of information from an intact canal (as a function of the plane of rotation) is readjusted centrally so that it can provide a better signal of rotation in a plane close to that of the plugged canal. Furthermore, during cross-axis training, neurons in the vestibular nuclei that are normally maximally sensitive to pitch axis (vertical) stimulation increase their sensitivity to yaw axis (horizontal) rotation,[71] providing a neurophysiological substrate for the change in direction of the VOR. The torsional VOR undergoes similar adaptive adjustments, as do the vertical and horizontal VOR, although the mechanisms may be somewhat distinct.[72–75]

A clinical consequence of a disturbance of cross-axis VOR plasticity is the occurrence of "perverted" nystagmus—nystagmus in which the slow phases are in the wrong direction relative to that of the stimulus. A strong vertical nystagmus induced during either caloric stimulation or vibration of the mastoid bone is such an example, and it usually occurs with central vestibular lesions.

The linear (translational) VOR, and other otolith-ocular reflexes, are also under adaptive control.[66,76,77] In the absence of canal-driven responses, otolith-ocular responses to a changing gravity vector (which can occur when rotation is around an axis tilted away from the vertical) are potentiated, leading to improved stabilization of gaze during off-axis rotation of the head.[78] The vestibular responses to translation, and their potential for rehabilitation in vestibular patients, have been largely neglected in clinical practice.

These short-term adaptation experiments probably test only one particular type of vestibular adaptation, because the learned response is not sustained in the absence of continued stimulation. There is also a long-term adaptive process, taking days to weeks rather than just minutes to hours, that gradually supervenes and leads to a more enduring, resilient adaptive change. Thus, one must be cautious when extrapolating results from these short-term experiments to the long-term problems of patients adapting to chronic vestibular deficits.

Adaptive capabilities have been investigated in elderly subjects[79]; at higher frequencies (above about 0.75 Hz), the ability of these subjects to increase VOR gain adaptively is significantly diminished. Because the angular VOR is most needed to compensate for the high-frequency components of head rotation, a loss of adaptive capability in elderly patients could account for the more devastating and persistent symptoms in such patients after a vestibular loss. Furthermore, with aging, patients with deficient vestibular responses lose some ability to substitute corrective saccades for hypometric slow phases.[80,81]

Potentiation of the cervico-ocular reflex (COR) can be well demonstrated in patients with bilateral loss of labyrinthine function.[44,45,47] Even normal individuals can be shown to undergo modification of the COR in short-term VOR adaptation paradigms.[54,82] The role of the

COR in compensation for unilateral vestibular loss is not clear, although in an occasional patient the reflex may be potentiated and may even be subject to improvement with vestibular exercises.[83]

### *Context Specificity*

One finding that may have important clinical implications is the demonstration that VOR gain adaptation is context-specific.[69] Baker and associates[84] showed that cats can be trained to adaptively change the gain of their canal-induced (rotational) VOR in one way when the head is oriented with respect to gravity in one particular position (e.g., right ear down), and in another way when the head is oriented in the opposite position with respect to gravity (e.g., left ear down). Thus, even though the pattern of canal activation is the same, the pattern of static otolith inputs determines or "gates" different central responses to an identical input from the semicircular canals.

VOR adaptation in humans also depends on the static orientation of the head in which the training of the canal-induced VOR took place.[85,86] Similar results have been shown for the translational VOR.[87] Shelhamer and coworkers[88] have also shown that adaptation of the gain of the VOR can be made to depend on the position of the eye in the orbit in which the training took place. In their paradigm, the horizontal VOR gain was trained to depend on the vertical position of the eye in the orbit. Such a capability would be particularly useful for individuals who wear a bifocal spectacle correction (although clearly, context-specific adaptation of the VOR did not evolve to meet the needs of the spectacle-wearing human, because glasses are a relatively new arrival to the human experience). When viewing through the lower part of the lenses, which have the stronger prescription needed to overcome the effects of presbyopia, subjects require a higher VOR gain during head rotation than when viewing through the top part of the spectacles.

Potentially, then, simply putting one's glasses on (or perhaps just the frames, or putting on the glasses in complete darkness) might be enough of a cue to generate a different vestibular response. Just such an effect of spectacles has been shown for another type of ocular motor adaptation in which the eyes are required to rotate by different amounts when the two eyes have different spectacle corrections.[89]

Thus, context specificity of vestibular learning, which potentially can be derived from a variety of cues, both vestibular and non-vestibular, must be considered in the design of programs of physical therapy: Will the particular training paradigm that is being used to promote vestibular compensation "transfer" to the more natural circumstances in which the patient usually becomes symptomatic?

## Neurophysiologic Substrate of Vestibulo-ocular Reflex Adaptation

Where might the structures be that elaborate various types of vestibular plasticity within the central nervous system? First, one should remember that one must distinguish between static and dynamic VOR adaptation. The cerebellum, especially the flocculus, seems to play an important role in the acquisition of adaptive changes in VOR gain.[90–96] The flocculus also plays a role in recovery of function after unilateral labyrinthine loss. Although restoration of relatively small degrees of imbalance between the vestibular nuclei can probably take place independently of the flocculus,[97] large amounts of spontaneous nystagmus and the recovery of amplitude and symmetry of gain during head movement probably require the flocculus.[98–101] Some of this effect may occur through the vestibular commissural pathway, which likely mediates some aspects of recovery of static balance and dynamic function.[102] Furthermore, potentiation of the COR as an adaptive strategy during head rotation is lost in patients with bilateral vestibular loss who also have cerebellar atrophy, implying a role for the cerebellum in this aspect of vestibular adaptation.[45]

The exact sites and specific mechanisms underlying these types of vestibular learning are still unsettled; evidence exists for multiple mechanisms and both a cerebellar cortex and a brainstem (vestibular nuclei) locus.[94,95,101,103–106] The VOR pathway has been parsed into linear and nonlinear (velocity-dependent) components, and depending on the pattern of head motion during learning, one or both are engaged.[107,108] There may be differences in mechanisms that underlie adaptive increases versus decreases in the amplitude of the VOR and context-specific versus generalized VOR adaptation.[95] Long-term, more "hard-wired" changes in the VOR may take place in the vestibular nuclei themselves[94] but again, are subject to higher-level control, which can gate particular responses according to prevailing circumstances and needs. As emphasized earlier, there certainly are multiple mechanisms and multiple time frames even for something as simple and fundamental as adaptation of the gain of the VOR.

Many neurotransmitters and neuropeptides have been implicated in the process of vestibular adaptation.[3,109–118] In the vestibulocerebellum, nitric oxide, NMDA receptors, acetylcholine, and catecholamines appear to be important.[3] Long-term depression (LTD) has

been implicated in the cerebellar contribution to VOR adaptation, and long-term potentiation (LTP) may also be important.[104,119–122] Specific transmitters may relate to the direction (increase versus decrease) or the frequency spectra (high versus low) of a particular adaptive response.[123,124] The exact role of these processes and of specific neurotransmitters in vestibular compensation has not yet been settled. The challenge remains as to how to apply the results of the many studies of vestibular compensation in experimental animals to human patients with vestibular disturbances.[125]

Still unclear are the substrates for the variety of strategies and cognitive influences (e.g., context and imagination, and effort of spatial localization) that are incorporated as part of the compensatory response to vestibular damage. A possible anatomical substrate for such higher-level influences may reside in the extensive reciprocal connections between the vestibular nuclei and the cerebral cortex.[126] Indeed, a number of areas in the cerebral hemispheres are activated with vestibular stimulation.[127–131] On the other hand, it has been shown that when a rabbit is exposed to sustained sinusoidal oscillation of the head, some climbing fibers in the nodulus of the rabbit discharge in a sinusoidal fashion after the animal stops rotating.[132] This finding is compatible with the idea that the cerebellum can learn patterns of vestibular stimulation and generate them even after the actual stimulus has ceased.

## Summary

This discussion has emphasized that consideration and knowledge of the adaptive capabilities of the brain are essential to the diagnosis and management of patients with vestibular disorders. The proper interpretation of the symptoms and signs shown by patients with vestibular dysfunction, the design of an optimal plan of physical therapy to promote recovery from vestibular dysfunction, and an objective analysis of any salutary effects of physical therapy cannot be accomplished unless constant attention is paid to the actions of the variety of compensatory mechanisms that are used to cope with abnormal vestibular function. Furthermore, the discussion has reemphasized that compensation is far more than a simple readjustment of low-level, largely subconscious reflexes. The roles of anticipation and prediction, altered motor strategies, sensory substitution, and cognitive factors related to mental set, psychological effort, imagination, and context are all important in the adaptive process and must be considered in the planning of physical therapy.[2] We are only now beginning to identify the wide repertoire of compensatory mechanisms available. Indeed, the remarkable adaptive capabilities of the vestibular system may lend itself to a successful response to a vestibular prosthesis, much like the success story of the cochlear prosthesis.[133] The challenge now is for us to learn to marshal these adaptive mechanisms to best promote recovery in our patients.

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CHAPTER 3

# Role of the Vestibular System in Postural Control

*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, the vestibular system is one of the nervous system's most important tools to control posture. The vestibular system is both a sensory system and a motor system. As a sensory system, the vestibular system provides the central nervous system (CNS) with information about the position and motion of the head and the direction of gravity. The CNS uses this information, together with information from other sensory systems, to construct a picture (sometimes called a "model" or "schema" or an "internal representation" or "map") of the position and movement of the entire body and the surrounding environment. In addition to providing sensory information, the vestibular system also contributes directly to motor control. The CNS uses descending motor pathways, which receive vestibular and other types of information, to control static head and body positions and to coordinate postural movements.

Because the vestibular system is both a sensory system and a motor system, it plays many different roles in postural control. This chapter explores the four most important roles (Fig. 3.1). First is a discussion of the role of the vestibular system in the sensation and perception of position and motion. Second, its role in orienting the head and body to vertical, including the static alignment of the head and body and the selection of appropriate sensory cues for postural orientation in different sensory environments, is dealt with. These first two roles are primarily sensory in nature; the vestibular system provides the sensory information about head motion and position and the direction of gravity that the CNS needs to carry out these functions. Third is a discussion of the role of the vestibular system in controlling the position of the body's center of mass, for both static positions and dynamic movements. Fourth is a description of the vestibular system's role in stabilizing the head during postural movements. These latter two roles involve motor aspects of the vestibular system.

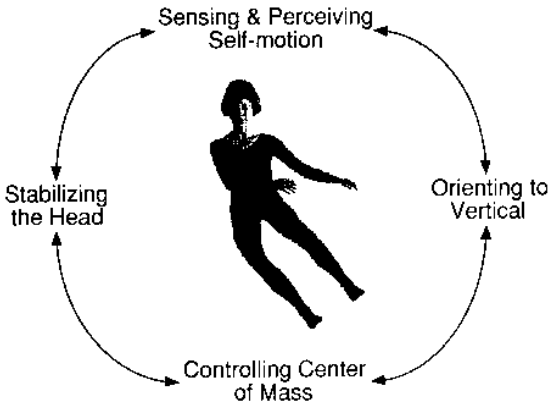


**Figure 3.1** Four important roles of the vestibular system in postural control interact with other sensory and motor systems to accomplish tasks like maintaining equilibrium and body alignment on an unstable surface.

## Sensing and Perceiving Position and 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 as a whole. The vestibular system consists of two types of motion sensors, the semicircular canals (SCCs) and the otoliths. The SCCs sense rotational movement of the head. Rotational movements in the sagittal and frontal plane are detected by the vertical (anterior and posterior) SCCs. The horizontal canals are sensitive to motions in the horizontal plane. The largest head motions during quiet stance and walking or running occur in the sagittal (anterior-posterior) plane. Frontal plane (side-to-side) and horizontal plane (as if to shake the head "no") movements also occur, but they are smaller.[1] Information from all three sets of SCCs contributes directly to the perception of self-motion.

In contrast to the SCCs, which sense rotational motion, the otoliths sense linear accelerations. Vertical linear accelerations of the head, like the head translations generated during deep knee bends, are sensed by the saccular otoliths. Horizontal linear accelerations, like the translations of the head generated during walking forward, 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 alignment with respect to gravitational vertical.

As important as good vestibular function is for determining the position and motion of the body, vestibular information by itself is not enough. First, the vestibular system can provide information only about head movements and not about the position or movement of any of the other body segments. Second, vestibular information about head movements can be ambiguous. A signal from the vertical canals indicating anterior head rotation can be produced by the head flexing on the neck or by the body flexing at the waist, but the vestibular system alone cannot distinguish between the two. In addition to these problems, the vestibular system is not equally sensitive to the entire range of possible head movements. The SCCs are most sensitive to faster head movements, such as those that occur at heel strike during gait or as a result of a sudden trip or slip.[2–5] The canals respond poorly to slower head movements, such as the slow drifting movements that occur during quiet stance. The otolith organs can signal tilts with respect to gravity and slow, drifting movements, but only when these movements are linear, rather than rotational.[6–9]

In order to clarify the ambiguities inherent in vestibular information and to get good sensory information about the entire range of possible head and body movements, the CNS relies on information from all available sensory systems. Each sensory system contributes a different kind of important information about body position and motion to the CNS, and each sensory system is most sensitive to particular types of motion.[10–15] 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 door frames 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.[12,16,17]

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 one another. For example, somatosensory information can help the CNS distinguish whether a head rotation signal from the vertical canals is due to motion of the head on the neck or due to flexion of the body at the waist. The somatosensory system can also provide information about how body segments are aligned with respect to each other and the support surface, through sending information about muscle stretch and joint position at the ankle or more proximal joints. The somatosensory system is particularly sensitive to fast movements, like those generated by sudden perturbations of joint positions.[18]

The contribution of each sensory system to the sensation of self-motion has been demonstrated experimentally through stimulation of the individual sensory systems. Electrical stimulation of the vestibular nerve by means of current passed through electrodes placed on the skin over the mastoid bone produces sensations of self-motion or tilt in humans by mimicking the vestibular signals that would be generated by an actual head movement.[19,20] Similar results can be achieved by presenting subjects with large moving visual scenes (Fig. 3.2).[12,17,21–23] When the head is moved, the image of the entire visual scene moves in the opposite direction. When subjects watch a large moving visual scene, their CNSs often misinterpret the visual stimulus, and the observers feel self-motion in the opposite direction. Vibrating ten-

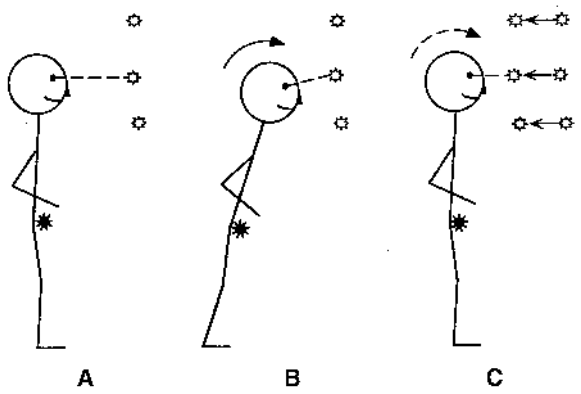


**Figure 3.2** Visual information can be used to determine how the body is moving. In this and all subsequent stick figures, the *asterisk* corresponds to the position of the body's center of mass, which is located about 2 cm in front of the spinal column at the level of the pelvis (*A*). As the body sways forward (*B*), visual objects placed in front of the subject loom toward the subject. If visual objects are moved toward a stationary subject (*C*), the subject can experience an illusion of forward sway (*dotted arrow*), especially in cases of vestibular loss.

dons in the neck and legs, which stimulate somatosensory motion detectors, can also give rise to sensations of body motion.[10,24,25]

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") can contribute powerfully to how sensory signals are interpreted.[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 his vestibular system and somatosensory system indicate that he has not moved. This phenomenon 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.[27] This observation is particularly interesting because these illusions of motion occur despite the fact that in either case the vestibular system is signaling lack of motion.

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 or rocking or that the room appears to spin around them. These sensations may be associated with particular head positions, depending on the disorder. Patients may adopt leaning postures while insisting that they feel themselves aligned vertically, indicating that self-motion perception and automatic postural responses may be independent of each other to some extent. Patients with profound losses of vestibular function have difficulty determining how they are moving in environments that lack good visual and somatosensory orientation cues, such as while walking at night on a sandy beach or while swimming in muddy 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 motion of the whole body; each sensory system contributes different and necessary information. The next section explores 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.

## Orienting the Body to Vertical

Keeping the body properly aligned parallel to gravity and directly over the feet 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 one another and with respect to vertical, other sensory systems contribute to body orientation as well. This section discusses the role of the vestibular system in the orientation 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 radiographs and fluoroscopy have revealed that most vertebrates hold the vertical spine parallel to gravitational vertical.[28] The vestibular system, which signals the direction of gravity, plays an important, but not exclusive, role in the alignment of head and trunk in animals. A unilateral vestibular lesion in animals results in head and body tilts toward the lesioned side.[28–30] The amount of asymmetrical posturing gradually diminishes over time, and the return to normal postural alignment has

been considered a sign of vestibular compensation.[31] 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 more short-lived than in lower species.[29,30,32] 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.[31,32] However, within 6 months to 1 year of total unilateral vestibular loss in humans, postural alignment and control can be indistinguishable from that of normals.[33] Bilateral loss of vestibular function may be associated with a forward head position.[34] Altered postural alignment, sometimes associated with excessive muscle tension and pain, especially in the neck, is a familiar problem for patients with vestibular dysfunction.[35]

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 the lesion and then regain normal weight distribution over the course of several weeks.[36] Fukuda[37] developed a stepping-in-place test to document the asymmetry and gradual compensation that follow unilateral vestibular loss. In this test, subjects attempt to step in place with eyes closed; patients with unilateral loss typically rotate slowly toward the side of the lesion. Patients with bilateral vestibular loss have also been reported to shift their weight forward or backward.[38]

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.[19,20,39–46] Cathodal currents increase the tonic firing in the vestibular nerve, and anodal currents decrease the firing rate.[47] 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 that would result if the body were tilted toward the side of the cathode. Subjects sway toward the anode to correct the apparent tilt induced by the galvanic stimulation. Galvanic stimulation reliably results in tonic head tilts and weight shifts in normal humans.[19,20,39–46] These responses are typically absent or abnormal, however, in patients with vestibular nerve sections, although they can also be normal in patients with loss of peripheral hair cell receptors, a finding that confirms that the galvanic current directly stimulates the vestibular nerve.[48,49] Postural responses to electrical stimulation can be enhanced when subjects stand on a sway-referenced surface that provides poor somatosensory feedback for orientation or when galvanic current is delivered during responses to a platform movement.[46,48] These findings suggest that the role of the vestibular system in automatic postural alignment is larger when somatosensory information for postural control is unreliable.[19]

Studies of postural responses to electrical stimulation of the vestibular system also show that the CNS 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. If the head is turned on the trunk so that the ear with the anode is turned forward, the galvanically induced sway is forward.[19] This observation suggests that information about head position from neck receptors is combined with vestibular information to trigger an appropriate postural response regardless of head position with respect to the body.[19] When the head and lower limbs are aligned forward, but the trunk is aligned differently, postural responses to galvanic stimulation are appropriate to alignment of the head and feet regardless of trunk alignment.[50] 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 and appropriate postural responses.[51]

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[13] have suggested that the CNS constructs a model or internal map of the direction of gravity on the basis of vestibular and other sensory information, and then aligns the body according to this map. This hypothetical explanation could also account for the body realignments that occur with galvanic stimulation. The galvanic current, which simulates the pattern of vestibular nerve firing that results when the body is tilted toward the cathode, may lead to a change of the CNS's estimate of the position of gravity—that is, that it has shifted toward the anode. 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 a patient with vestibular disorders may also occur from a faulty internal map based on abnormal information from the malfunctioning vestibular system.

Vestibular information also contributes to another important internal map, the map of stability limits, and vestibular pathology may lead to defects in this internal

map as well. A human standing with feet planted on the ground may sway forward or backward a small amount (about 4 degrees backward and about 8 degrees forward) without losing balance and taking a step. The boundaries of the area over which an individual may safely sway are called the *stability limits*.[52] The actual stability limits for any individual in any situation are determined by biomechanical constraints, such as the firmness and size of the base of support, and by neuromuscular constraints, such as strength and swiftness of muscle responses.[53] Vestibular disorder 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. Figure 3.3 shows a patient with bilateral loss of vestibular function, peripheral neuropathy, and cataracts, who aligns herself near her backward limits of stability in stance.

**Figure 3.3** Elderly woman with loss of vestibular function, peripheral neuropathy, and cataracts who aligns herself near her backward limits of stability. In this photograph, she is standing on a compliant foam pad, which reduces her ability to use somatosensory cues for orientation.

## Weighting Sensory Information

The studies of abnormal body alignment resulting from vestibular lesions show that the vestibular system plays an important role in the orientation of the body in space. The fact that normal alignment can be recovered with time even after bilateral vestibular loss, however, also argues that vestibular information is not the only source of sensory information that can be used for orientation. Visual and proprioceptive information also contribute to body alignment, as experiments with large, moving visual surrounds and tendon vibrations have shown.[17,21–24] Whether and how vestibular or other sensory information is used for 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), 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 upon vestibular, visual, and somatosensory inputs for postural orientation, studies now suggest that they rely primarily on somatosensory information from the support surface in these conditions.[54,55]

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 joint and other somatosensory and proprioceptive information from the feet and legs may bear little relationship to the orientation of the rest of the body; that is, the body's center of mass could be aligned well within the stability limits despite large amounts of ankle motion (Fig. 3.4). Under such circumstances, it is critical that the nervous system be able to extract from the available sensory information the actual orientation of the body with respect to gravity and the base of support, because failure to align the body's center of mass properly in a gravity environment will almost certainly lead to a fall.

The relative dependence on, or weighting of, each sensory system changes with changes in environmental conditions.[56] Peterka[54] showed experimentally that sensory weighting of somatosensory. vestibular, and visual information changes with postural orientation (Fig. 3.5). For example, when blindfolded subjects were exposed to

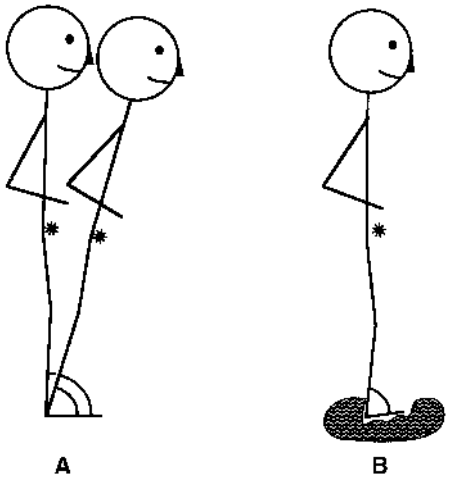


**Figure 3.4** When subjects stand on a flat, firm support surface (*A*), ankle flexion corresponds to a forward body center of mass (CoM) position. The position of the CoM is indicated with an *asterisk*. When the support surface is compliant or tilted (*B*), however, ankle flexion can correspond to an upright or even backward body CoM position. Thus, vestibular information is more important for postural control on the unstable surface in *B* than on the firm surface in *A*.

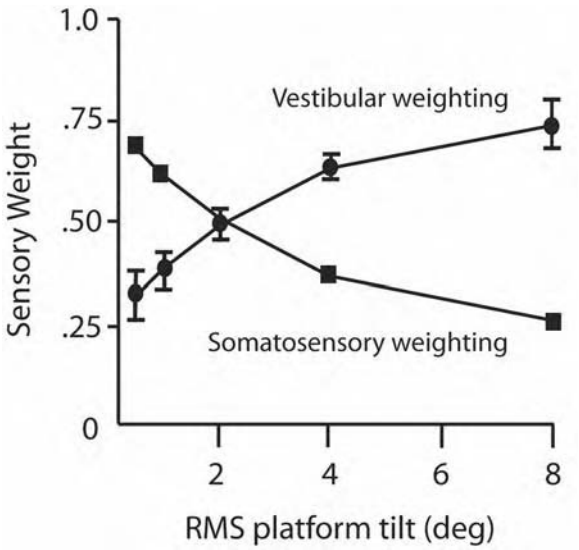


**Figure 3.5** As the magnitude (RMS [Root Mean Squared]) of tilt of surface rotation increased, blindfolded healthy subjects increased their relative weighting of, or dependence on, vestibular inputs and decreased their dependence on somatosensory input. (Adapted from Peterka, 2003.[54])

surface oscillations of various amplitudes, healthy subjects tended to align themselves to the surface for very small amplitudes of rotation but to orient their posture more with respect to gravity with larger amplitudes of rotation, as if relying more on vestibular information. Peterka[54] 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. As surface oscillations increased from 1 to 8 degrees, however, he found a change in sensory weighting so that subjects relied 70% on vestibular information, 20% on vision, and only 10% on somatosensory information (Fig. 3.5). Because the human nervous system normally depends primarily on somatosensory information for postural orientation, it can be difficult to identify postural problems in patients with vestibular disorders when they are standing on a firm support surface, even though they may have severe balance problems on an unstable surface.

The way in which the nervous system weights sensory information for body orientation in different environments can be measured using a Sensory Organization Testing paradigm developed by Nashner and colleagues[57–59] and adapted to use in the clinic by Shumway-Cook and Horak.[60] In this paradigm, the subject is asked to stand quietly in each of six different sensory environments, and the subject's postural sway is measured each time. 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 used in clinical evaluations of standing balance.

In the remaining four environments, either 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* (Fig. 3.6). Sway-referencing the support surface and/or the visual surround can disrupt the normal sensory feedback relationships between the different sensory systems. For example, when the support surface is sway-referenced, somatosensory information from the ankle joints correlates poorly with the position of the body. 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.[60] 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 3.7 shows how normal subjects react when exposed to such altered sensory environments. Normal subjects sway a small amount even under normal cir-

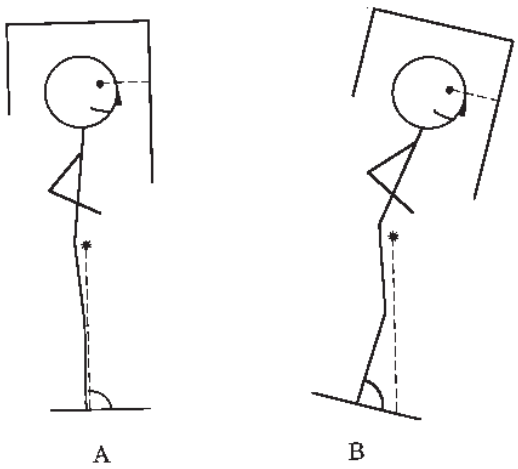


**Figure 3.6** This figure shows how "sway-referencing" the subject's support surface and visual surround interferes with the ability to use somatosensory and visual information to orient the body to vertical. (*A*) The subject stands aligned to vertical, and the center of mass projects directly over the foot. (*B*) The subject has swayed forward, resulting in a toe-down rotation of the platform and a forward rotation of the visual surround. Although the ankle angle and the position of the head with respect to the visual surround have not changed, the center of mass is now in front of the foot, and the subject is in imminent danger of a fall. Normal subjects can detect this forward sway using vestibular information and avoid a fall, but patients with vestibular losses have difficulty doing so.

cumstances (condition 1), and this sway may or may not increase slightly when visual information is removed by eye closure (condition 2).[61] Although sway may increase slightly when either the visual surround alone (condition 3) or the support surface (condition 4) alone is sway-referenced, subjects are still able to select useful sensory information from the sources available and maintain body orientation with respect to gravity. Even when both the support surface is sway-referenced and visual information is eliminated by eye closure (condition 5) or altered by sway-referencing (condition 6), normal subjects are able to use vestibular cues to orient the body, albeit somewhat less efficiently. This is not surprising; conditions 5 and 6 are similar to walking on uneven surfaces in poorly lit environments (Fig. 3.8), swimming in murky water, or standing in the cabin of a ship—all tasks that normal subjects can perform without great difficulty.

Patients with clinically diagnosed vestibular disorders have also been tested with use of the same paradigm.[57–60] One of the most important findings of these studies is that not all patients with vestibular disorders respond in the same way. Patients identified in Figure 3.6 as having a *vestibular loss pattern* lose all sense of orientation and fall in conditions 5 and 6, in which orientation information from both the surface and vision have been altered and the subject is forced to rely primarily on vestibular information. One should note, however, that these patients can perform as well as normal subjects in conditions 1 through 4, in which at least one unaltered source of sensory information was available to them. In conditions 5 and 6, however, patients with vestibular loss lose balance when forced to rely on vestibular information, as if 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 he or she may have some 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 (equivalent to conditions 1 and 2) as a test of vestibular function.[62]

In contrast to the patients with vestibular loss, patients who have uncompensated vestibular disorders may show increased sway in conditions 3 through 6 (see the sensory selection pattern in Fig. 3.6). That is, they have difficulty with orientation whenever somatosensory *or* visual information is altered or is not available. Why some patients with vestibular disorders have difficulty orienting in condition 3 or 4—in which has normal somatosensory or visual information, respectively, is normal—is not clear. Their nervous systems appear to have oriented their bodies to visual and support surface references that correspond poorly to gravity when these are sway-referenced. Investigators have hypothesized that even healthy subjects vary in how much they "weigh" (rely more heavily on) sensory information from a particular source, such as vision or somatosensation,[15,27] particularly when vestibular cues to orientation are unreliable. Incomplete CNS adaptation to a vestibular lesion may also be a factor. This pattern of results is typical of patients in the acute stages of vestibular lesions.[63] 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 vestibular loss eventually return to normal sensory orientation for postural control.[33] A *surface dependent pattern*, in which patients show postural instability whenever the surface is sway-referenced, is common in patients with central cerebellar disorders. [64,65]

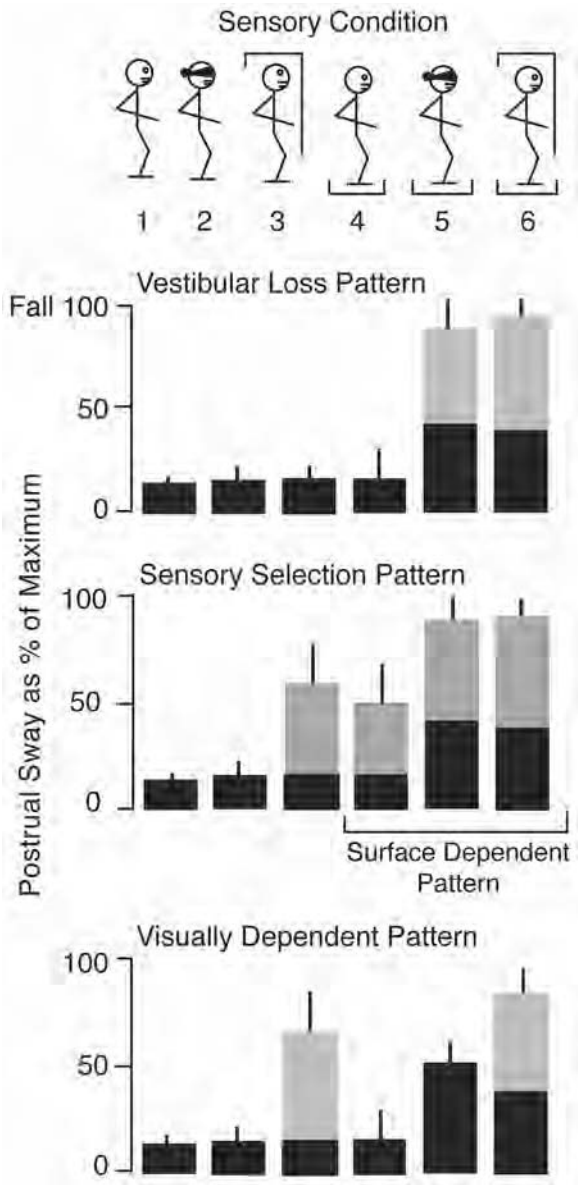


**Figure 3.7** Postural sway of healthy subjects (*black bars*) and of patients with vestibular loss (*gray bars*) under six sensory conditions. Peak anterior-posterior postural sway (as a percentage of the maximum sway possible without a fall) and standard errors are shown for patients with vestibular loss who demonstrate three types of sensory orientation patterns: vestibular loss, sensory selection, and visually dependent. Patients who show excessive sway only in conditions 4, 5, and 6 are called *surface dependent*, but data for that group are not shown. *Stick figures* along the abscissa show the sensory information disrupted in each condition: *Blindfold* indicates eyes closed, *box* around the head indicates a sway-referenced visual surround, and *box* around the feet indicates that the platform was sway-referenced.

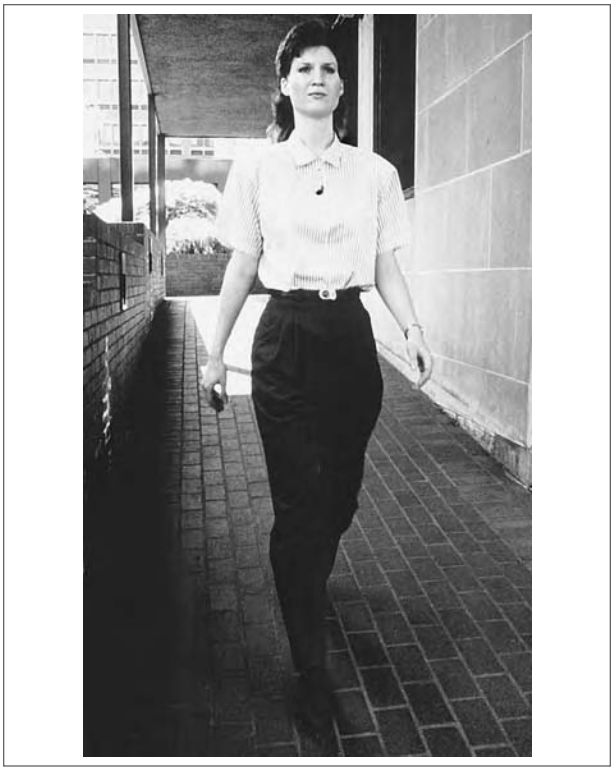

**Figure 3.8** Walking in a poorly lighted environment on an uneven surface like a ramp puts demands on vestibular information for postural control.

Some patients who have distorted but not absent vestibular function such as acute hydrops (see Chapter 6), which alters but does not eliminate vestibular input, show a *visually dependent pattern* of sensory organization.[58] These patients demonstrate excessive sway whenever the visual surround is sway-referenced but normal sway when their eyes are closed. It is as if the nervous system relies on visual information whenever the eyes are open, even when vision is not providing accurate information about body sway. However, when their eyes are closed, such patients are able to rely on surface information on a firm surface and upon vestibular information when standing on a sway-referenced surface.

In summary, these studies show that vestibular information for body orientation is most important in environments that lack good somatosensory 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, regardless of its usefulness, when vestibular information is bad or missing. Figure 3.9*A* shows a healthy child who

maintains a normal orientation with respect to gravitational vertical despite standing on a tilted surface. Figure 3.9*B* shows a child with abnormal vestibular function who appears to rely most heavily on surface information for orientation; he maintains a perpendicular orientation to his tilted support surface. If the support surface is orthogonal with respect to gravity, this strategy may work fairly well. However, when the support surface is tilted, as it is in Figure 3.9, this strategy works poorly.

Although the subject shown in Figure 3.9*B* chooses somatosensory information for orientation, other patients with vestibular disorders behave as if they rely most heavily on vision for orientation and sway or fall when attempting to stand near large moving objects (like buses or cars). These 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, patients with vestibular disorders may align themselves either with a faulty vestibular estimate of the direction of gravity or with an estimate of the direction of gravity from another sensory system.

## Controlling Center of Body Mass

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 describes how motor output from the vestibular system contributes to static body positions and dynamic postural movements, which help subserve the postural goal of maintaining equilibrium or controlling the center of body mass within its limits of stability.

We know from 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 neck, trunk, and limb muscles. 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. In order 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 flexions 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

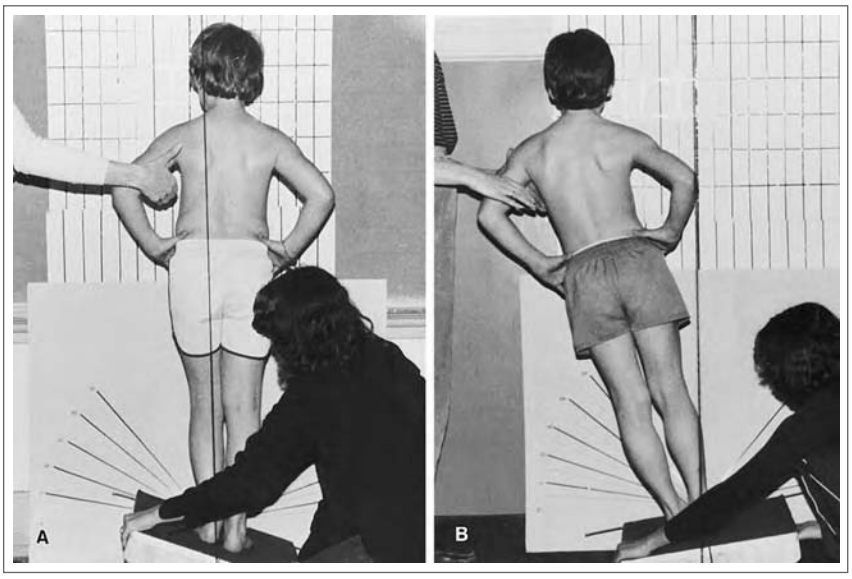


**Figure 3.9** (*A*) Child with normal vestibular function orienting the head and body with respect to gravity while the surface is tipped and the eyes are closed. (*B*) Child with absence of vestibular function orienting the head and body with respect to the tipped surface while the eyes are closed.

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.

## *Role in Automatic Postural Responses*

### 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 (50–100 msec in cats; 80–200 msec 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 absence of vestibular function; the responses remain, however, after 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.[61,66–68]

Quick displacements directly to the head during stance also result in muscle activations in the neck (45 msec), trunk (85 msec), thigh (90 msec), and ankle (70 msec).[63,69,70] These responses are absent in patients with adult-onset vestibular loss, indicating a vestibular origin. However, adults who lose 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.[69,70]

Direct stimulation of the eighth cranial nerve by galvanic stimulation behind the ears can result in asymmetrical tonic activation of the vestibular system that is associated with body tilt toward the side of vestibular inhibition.[71,72] Subjects tilt toward the side of the vestibular inhibition regardless of the direction their heads are facing (sideways if the head is facing forward, and forward or backward if the head is rotated to the right or left). This observation is consistent with an integration of vestibular with somatosensory information so that the vestibular signal in the head is interpreted with respect to the position of the base of support. In fact, the muscles activated in response to galvanic vestibular stimulation also depend greatly on the somatosensory conditions.[73,74] In subjects who are freely standing, ankle muscles are activated first, but in those who are sitting, trunk muscles are activated, and in those holding onto a stable support with the hands, arm muscles are activated. The shortest latency responses to direct vestibular stimulation are very small and require very large currents and averaging across many trials, whereas the medium-latency electromyographic (EMG) responses that result in measurable body sway are very sensitive to somatosensory conditions. Thus, direct vestibulospinal reflexes are very weak and do not provide the functional effect on postural sway like the longer latency vestibulospinal influences that are interpreted via a whole-body somatosensory map.[72]

Because limb muscles are activated in response to sudden drops, perturbations in head position, and galvanic stimulation,[44–46] investigators have hypothesized that vestibulospinal mechanisms may also play a role in automatic postural responses to stance perturbations induced at the feet by movable platforms.[75,76] Two types of platform perturbations have been used to test this hypothesis: backward or forward translations, which induce forward or backward sway, respectively, and platform rotations, which induce ankle dorsiflexion or plantar flexion (Fig. 3.10). Platform translations result in activation of the stretched ankle muscles, which occur 80 to 100 msec after the onset of platform movement and act to restore the body to initial position. Vestibular inputs do not appear, however, to 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.[34,57,77–79] Proprioceptive information from stretched muscles appears to be sufficient for recovery from platform translations. This conclusion is supported by the finding that automatic postural responses to surface translations are delayed when proprioceptive inputs are disrupted by sway-referencing of the surface or eliminated with pressure cuffs at the thigh or in peripheral neuropathy.[80–82] 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.[73,69,70] Therefore vestibulospinal responses probably do not play a large role in the recovery of equilibrium following platform translations.

In contrast to responses to translations, responses to platform rotations may rely much more heavily on vestibular mechanisms. Platform rotations produce ankle dorsiflexion or plantar flexion, stretching muscles in the

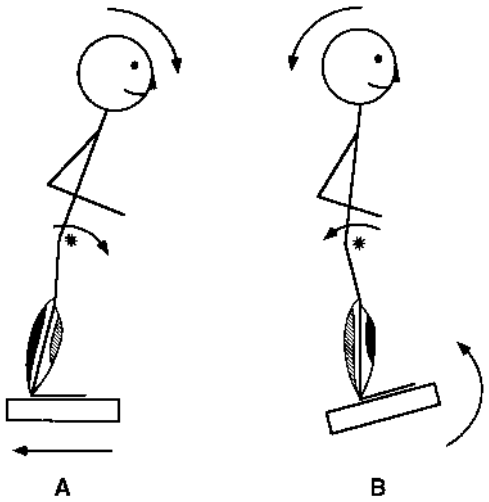


**Figure 3.10** Platform translations and rotations. (*A*) Backward horizontal translation of the platform results in stretch of ankle plantar flexors (*solid*) and forward sway of the body. Medium latency activation of ankle plantar flexors and other muscles restores equilibrium in response to backward platform translations. (*B*) However, toe-up rotation of the platform, which also results in stretch of ankle plantar flexors, results in backward sway of the body. For toe-up platform rotations, medium latency activation of the shortened ankle dorsiflexors (*hatched*) restores equilibrium. The CNS relies more heavily on vestibular information for postural control in platform rotations.

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 msec, which is probably triggered by proprioceptive inputs[69,75,76,82–84] and which could, if unopposed, actually destabilize the body. Slightly later, a stabilizing response occurs in the shortened ankle muscle (at 100–120 msec), and this response is probably more dependent on vestibular and visual inputs. In patients with bilateral or unilateral loss of the vestibular function, this stabilizing response is reduced in magnitude and delayed in latency.[76] The ability to adaptively reduce the magnitude of the destabilizing response to repeated surface tilts is impaired in some patients with loss of vestibular function.[57] The cause of this failure to adapt could be either that vestibular information is required to trigger the adaptive process or that patients with absence of vestibular function become hypersensitive to proprioceptive information.[85]

These studies, like many others, suggest that (1) the role of the vestibular system automatic postural responses enlarges when proprioceptive information about body sway is lacking or inaccurate and (2) accurate, efficient recovery of equilibrium after stance perturbation requires a close interaction of vestibular and somatosensory information.[86] This hypothesis is supported by studies showing that the magnitude of responses to head perturbations increases when subjects stand on compliant or moving surfaces.[70,83] Further, responses to peripheral vestibular stimuli are stronger in cats suspended by hammocks than in cats who support themselves on their legs and are stronger in patients with somatosensory loss due to peripheral neuropathy than in healthy subjects.[72] Cats suspended in hammocks and patients with neuropathy have less access to proprioceptive information about body position and are thus more affected by vestibular stimulation.[87]

## Selection of Postural Strategies

Not all postural tasks require the same type of movement for the recovery of equilibrium, and different automatic postural responses are triggered in different situations. These different responses, which differ in muscle activation patterns, body movements, and joint forces, are called *postural control synergies* or *postural control strategies*.[84] Two very different strategies for moving the body's center of mass without moving the feet have been identified in previous studies: ankle strategy and hip strategy (Fig. 3.11).[88] An *ankle strategy* is used by most subjects recovering from a postural disturbance such as a horizontal translation of the support surface while standing on a firm, flat support surface. The body sways roughly like 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), on compliant or tilting support surfaces (foam, tiltboards), when stance is narrow (one-foot standing or tandem stance), or when center of mass position must be corrected quickly (i.e., when the perturbation velocity is high). There is evidence that these postural control strategies are centrally programmed and can be combined according to biomechanical conditions, subject expectations, and prior experience. For example, normal subjects typically show a mixed ankle and hip strategy when responding to a translation of a 10-cm beam for the first time to translations of a flat surface for the first time after responding to a series of beam trials, or to intermediate or fast translations of a flat support surface.[90–92]

Vestibular information does not seem to be essential for initiation or execution of a normal ankle strategy, because subjects with complete absence of vestibular function show normal kinematic and EMG patterns associated with an ankle strategy.[34,79,91,92] This finding suggests that proprioceptive information from the body is sufficient to produce normal ankle strategy responses. In

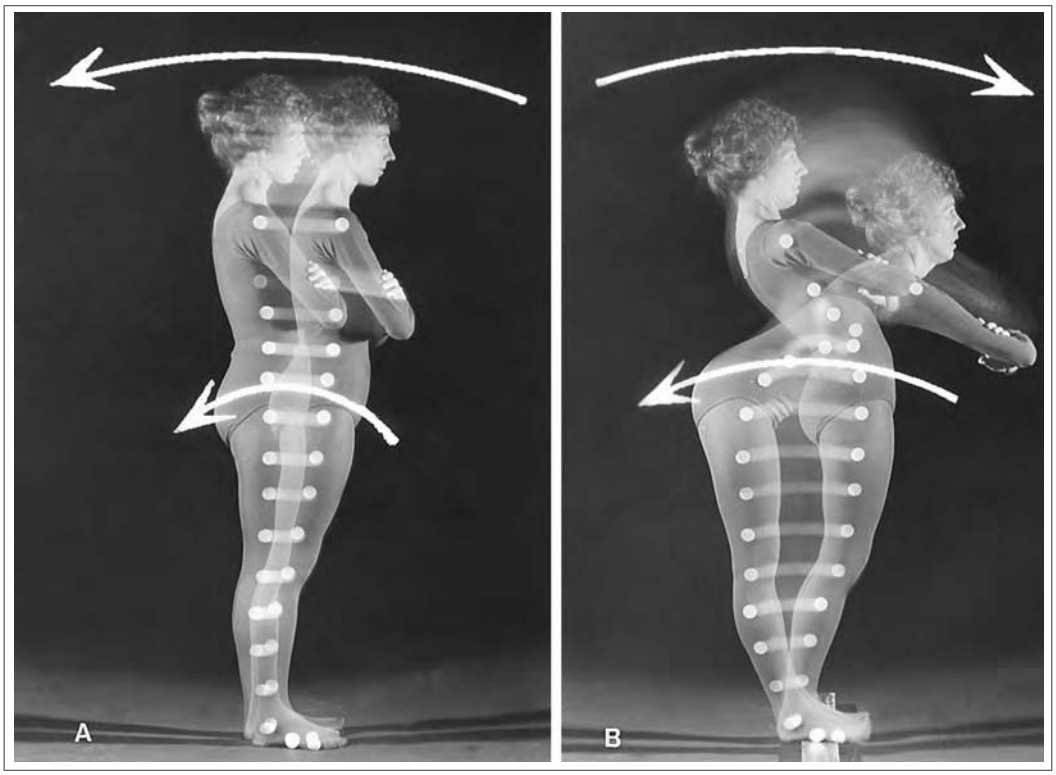


**Figure 3.11** Normal subject using an ankle strategy (*A*) and a hip strategy (*B*) to control postural sway. *Arrows* at the hips show direction of corrective center of mass movement; *arrows* at the head show direction of corrective head movement. The relationship between vestibular information and somatosensory information is different for ankle and hip strategies. In *A*, recovery from forward sway using ankle strategy, ankle extension corresponds to backward movement of the center of mass and backward pitch of the head. In *B*, recovery using hip strategy, ankle extension and backward movement of the center of mass now correspond to forward movement of the head and trunk. (From Horak, 1987.[89])

contrast, some evidence suggests that patients with vestibular loss have difficulty using 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.[34,77,79,93–95] There is also experimental evidence that signals from the vestibular system could act as a trigger for some postural responses. Subjects undergoing support surface translations show horizontal and vertical head acceleration peaks that are above threshold for the vestibular system at 40 to 60 msec after the onset of a platform translation.[91,92] Also, subjects with loss of somatosensory information from the feet due to ankle ischemia, who presumably must increase their reliance on the vestibular system, use a hip strategy when an ankle strategy would be more efficient.[77] All of these findings suggest a critical link between the vestibular system and the use of hip strategy to control posture.

However, evidence also suggests that the relationship between vestibular information and the use of hip strategy is not simple. In a study reported by Runge and associates,[92] patients with bilateral vestibular loss were tested using fast platform translations, and many of them showed bursts of abdominal muscle activity and early hip flexion torques similar to those in normal subjects, indicating that normal vestibular function is not necessary to trigger hip strategy responses when subjects are standing on a firm, flat surface. Why does vestibular information seem to be critical for a hip strategy in some circumstances, but not others?

The answers to this question may lie both in the way sensory information from the vestibular and somatosensory systems is interpreted and in the way motor activity produces body movements in the different postural tasks. The tasks requiring hip strategy in which subjects with vestibular loss perform poorly all involve changes in the support surface—that is, in its compliance (for foam or

tiltboards), in its size (beams), or in the way the body contacts the surface (beams, tandem stance, or stance on one foot). Further, these situations involve changes in the way forces generated at the ankle result in body movements; torques generated at the ankle cannot be used effectively to move the center of mass on short or compliant support surfaces. In these situations, body movements (either self-generated or due to a perturbation) do not result in the same somatosensory feedback as during normal bipedal stance on a firm surface, nor will an ankle strategy, which relies on generation of torque at the ankle joints, be very effective in correcting balance after a perturbation. It may be that in these special situations, normal subjects use vestibular information to interpret the changes in somatosensory information in order to produce an appropriately changed postural response. Patients with bilateral vestibular loss, who lack vestibular information, may not be able to accomplish the reinterpretation in time to prevent losses of balance when standing on compliant or short support surfaces. Responses to fast translations on a large, firm support surface, however, may not require reinterpretations of somatosensory information or changes in the way ankle torque can be used, possibly explaining why patients with bilateral vestibular loss do not show obvious abnormalities in this task.

Although patients with vestibular loss appear to be able to initiate hip strategy responses to fast platform translations, they take compensatory steps to maintain balance in response to these faster translations more frequently than normal subjects.[92] This finding suggests that such patients do have some difficulty with fast translations despite their ability to initiate a hip strategy. The fact that patients with bilateral vestibular loss tend to step more often to correct balance when responding to fast platform perturbations also suggests a further connection between a failure of hip strategy and vestibular loss. Although they may be able to initiate hip torques, patients with vestibular loss may be unable to execute hip strategy efficiently, and so may resort to stepping, because hip strategy may require efficient control of the trunk and good stabilization of the head with respect to gravity, which is likely to be faulty in patients lacking vestibular information.

Whereas vestibular loss is associated with a failure to use hip strategy in some conditions, some patients with disorders of the vestibular system habitually use hip movements to control center of mass position.[34,58,96] These patients typically report vertigo and ataxia consistent with vestibular dysfunction and have histories consistent with vestibular disorder but often show normal horizontal vestibulo-ocular reflex function during rotation testing; these findings suggest that such patients have sustained damage to the vestibular system, but have not lost a significant amount of vestibular function.[34,35,58,96,97] Preliminary results suggest that some patients with vestibular loss who show hip sway use a coordinated pattern of active hip motions similar to the hip strategy seen in normal subjects. Other patients appear to generate large ankle forces, which result in hip sway because the patients exert no control over their trunks. Coordination of head and trunk motions are abnormal in both types of patients.

Several possible explanations for an overreliance on hip sway in some patients with vestibular disorders are as follows:

1. As a result of disease or damage, the vestibular system in these patients may become hyperresponsive. The hyperresponsive vestibular system may overestimate the velocity of head motion signals during body sway, and the CNS may respond to small perturbations as though they were much bigger.
2. The vestibular dysfunction may impair the patients' ability to interpret somatosensory input from feet, so they may perform similarly to normal subjects with acute somatosensory loss due to ankle ischemia, and therefore use hip strategy.
3. The abnormal vestibular information may contribute to abnormal internal representation of stability limits, so patients may behave as if small disturbances in stance push them beyond their stability limits.

Patients who perceive themselves to be in a different relation to their stability limits from where they actually are may show inappropriate postural movement strategies in response to destabilization.[35,97] 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 in turn affect body alignment and recovery from postural disturbances. The information provided by the vestibular system and its relationship to information from the other senses changes according to the movement strategy used in controlling

equilibrium. This discussion has hypothesized that postural strategies are specific prescriptions for mapping interactions among sensory and motor elements of postural system; these maps are, in essence, a method for solving a sensorimotor problem. Individuals need a variety of different movement strategies to choose from, depending on current, past, and expected environmental conditions and task constraints. 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.

## Development of Motor Coordination

Consistent with the concept that the vestibular system does not shape the details of coordinated postural patterns, intact vestibular function does not appear to be crucial for the normal development of many aspects of motor coordination. Deaf children who sustain complete or partial loss of vestibular function within the first year of life score within or above normal limits in tests of interlimb coordination, such as kicking, walking, running, skipping, hopping, and fine coordination of the hands, although they often start walking a few months later than normal.[94,98,99] Later in childhood, clinical measures of balance function, such as duration of one-foot standing and ability to walk on a balance beam, are affected by loss of vestibular function. These tasks are difficult because one-foot stance and balance beams compromise a person's ability to use somatosensory information to control posture. However, these tasks also require hip strategy for center of mass control, which is usually abnormal in patients with loss of vestibular function.[34,73,75] Thus, these children may perform nearly normally in situations with normal support conditions but may show abnormalities in situations in which hip strategy is unavoidable.

In general, adults who have lost vestibular function as infants show much better postural compensation than those who have lost vestibular function as adults. For example, patients with infant-onset vestibular loss are more likely to show the vestibular loss pattern rather than the sensory selection pattern in sensory organization testing, indicating that they can use visual and/or somatosensory inputs to compensate for missing vestibular inputs. In response to surface translations, those with infant-onset loss of vestibular function show much more normal trunk and head stability than subjects with adult-onset vestibular loss. Figure 3.12 shows the excessive upper body and head displacement associated with excessive neck EMG activity in response to a backward surface translation in a subject with adult-onset vestibular loss and that responses in the infant-onset vestibular loss subject are much more similar to those of the healthy control

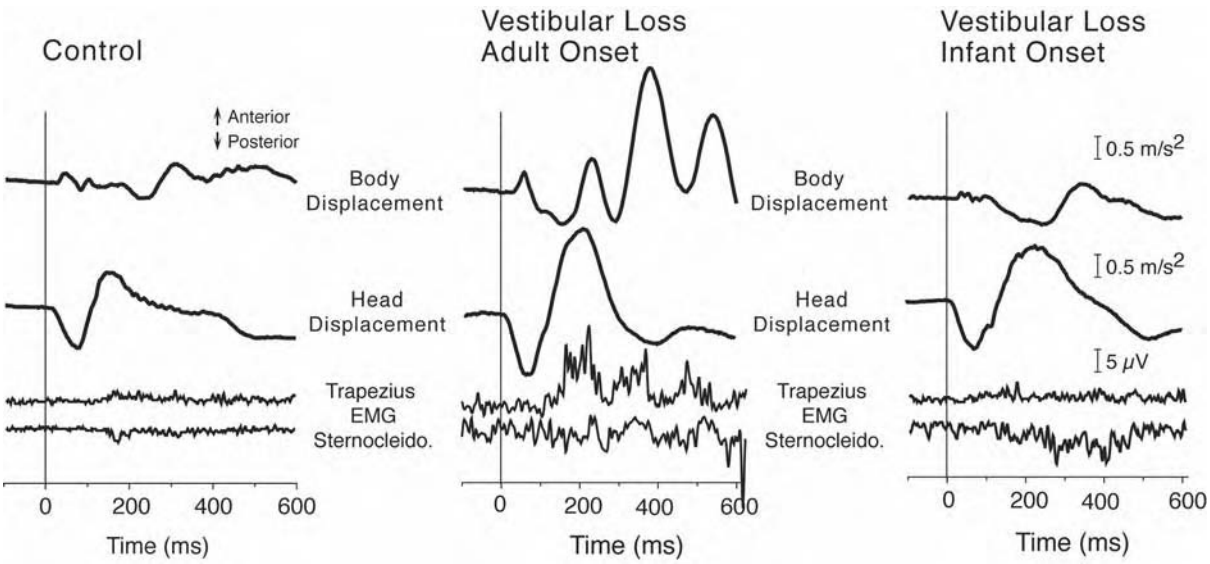


**Figure 3.12** Comparison of body and head displacement (angular velocity) and neck muscle electromyographic (EMG) activity in response to a backward surface translation in a representative healthy control subject and subjects with adult-onset bilateral vestibular loss and infant-onset vestibular loss. The subject with adult-onset vestibular loss shows the most trunk and head instability and excessive neck EMG activity in response to postural perturbations. Sternocleido. = sternocleidomastoid muscle. (Adapted from Horak et al, 1994.[69])

subject.[79] In addition, patients who have lost vestibular function as young children complain of oscillopsia or of balance or spatial orientation problems much less often than patients who have lost vestibular function as adults. Thus, the increased central nervous plasticity in young children facilitates maximal compensation for loss of vestibular function.

## Stabilizing the Head and 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 results 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 what can be compensated for by the VOR, blurred vision could result. For these reasons, investigators have suggested that the nervous system might stabilize the head with respect to gravity during postural control, either to simplify the interpretation of vestibular information or to facilitate gaze stabilization (Fig. 3.13*A*).[100,101] In the absence of good information about gravity from the vestibular system, or in an attempt to simplify the coordination of head and trunk movements, the nervous system might stabilize the head with respect to the trunk (Fig. 3.13*B*).[100]

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 activities like hopping and running.[101–103] For example, neck muscle activations are observed in normal subjects using a hip strategy.[34,79,91,92] In these studies, the neck muscle activations occurred prior to 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.

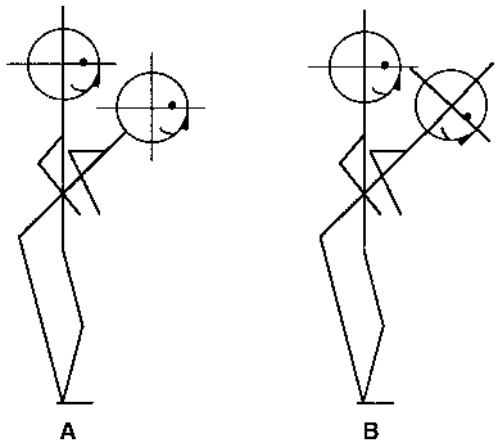


**Figure 3.13** Alternative strategies for control of head position during postural movements. Example is given for a subject using hip strategy to control the center of mass. Head is stabilized with respect to gravity in *A*, and with respect to trunk in *B*.

To determine the role of the vestibular system in maintaining head position with respect to gravity, the control of head position has also been studied in patients with vestibular loss.[34,79,104,105] During activities like walking and running, patients with profound vestibular loss show greater variability in head position with respect to gravity. When standing on an oscillating surface with the eyes closed, patients with vestibular loss show large oscillations of their 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.[106] Figure 3.14 compares the stable

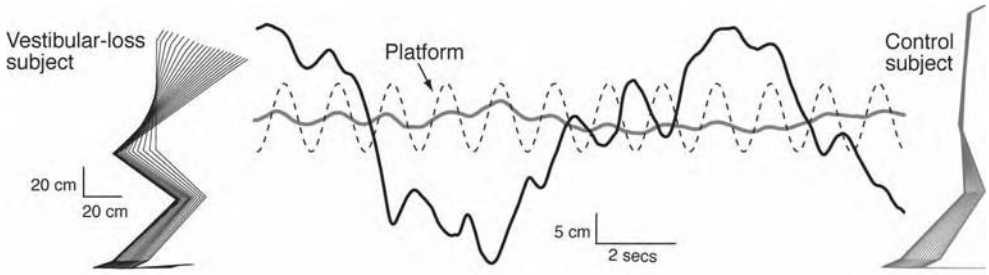


**Figure 3.14** Comparison of stable trunk in space in healthy control versus large trunk instability in a subject with bilateral vestibular loss during fast, anterior-posterior surface translations. *Stick figures* show lateral view of trunk, thigh, and legs during surface oscillations. Anterior (up) and posterior (down) trunk displacement in a representative control subject (*gray line*) and subject with vestibular loss (*black line*). *Dashed line* represents platform surface oscillation.

control of the trunk in a control subject with the large forward-and-backward drift of the trunk in a patient with vestibular loss during forward and backward translations of the support surface. Some patients with well-compensated vestibular loss, however, can stabilize their trunks in space when their eyes are open, whereas those with poorly compensated vestibular loss cannot use vision to compensate for the loss of vestibular function.

Figure 3.15 shows that three subjects with well-compensated vestibular loss demonstrated very little vertical, medial-lateral or anterior-posterior variation in head position, like contol subjects; three subjects with poorly compensated vestibular loss, however, showed very large variations in head position during surface oscillations.[106] Despite abnormal top-down coordination of the head and trunk, subjects with vestibular loss showed normal coordination of the legs at all frequencies of surface oscillation, even with eyes closed, consistent with a bottom-up somatosensory control of postural coordination of the legs.

Subjects with vestibular loss are very successful in substituting light fingertip touch on a stable surface for the lost vestibular function to stabilize posture during tandem stance and during surface rotations.[107,108] They can also use audiobiofeedback about trunk motion to control posture when standing on a compliant foam surface with the eyes closed.[109] These findings suggest that patients with vestibular loss who complain of balance problems would benefit from use of a cane or other biofeedback device 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, 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 center of mass or postural coordination of the legs, especially when good somatosensory cues about body position are available. These observations are consistent with the hypothesis that the distal-to-proximal muscle activations that control the center of mass are triggered by somatosensory information from the feet and legs and that the neck and trunk muscle activations that control head position are in turn controlled by vestibular mechanisms.[3–5]

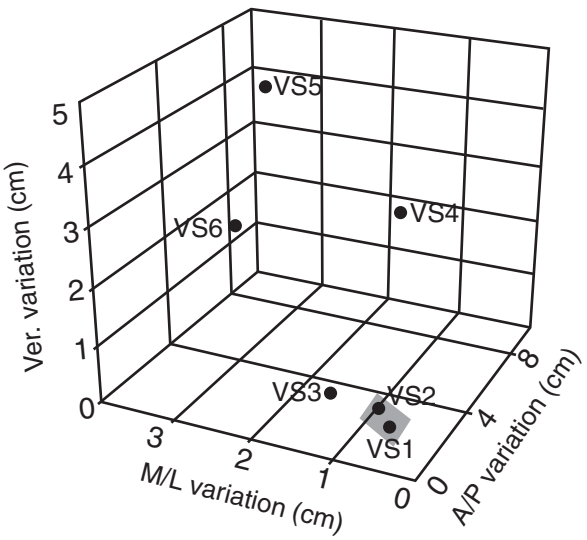


**Figure 3.15** Three dimensional graph of head acceleration in the sagittal, frontal, and horizontal planes in 6 patients with vestibular loss. Three patients with well-compensated bilateral vestibular loss (VS1, VS2, and VS3) show very little head instability during surface oscillations with the eyes open, like normal subjects (*shaded area*). Three patients with poorly compensated bilateral vestibular loss (VS4, VS5, and VS6), however, show large variation in vertical (Ver.), medial-lateral (M/L), and anterior-posterior (A/P) head displacements.

## Summary

The vestibular system plays many potential 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. When the stabilization of the head or trunk is critical for good performance, the vestibular system is very important. Likewise, when somatosensory (and, to a lesser degree, visual) information is not available, vestibular information for postural control has a dominant role. Table 3-1 lists some tasks and conditions in which vestibular information for postural control is important as well as 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 center of mass, and stabilizing the head result in impairments in gait as well as in standing balance.

Consider, for example, a task and a set of environmental conditions that occur frequently in clinical examinations of postural control: The patient sits, with eyes closed, on a board, which is tilted (see Fig. 3.1). 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

## CASE STUDY 1

A 60-year old woman required high doses of intravenous gentamicin for a bone infection after a fracture repair. After several days of treatment, she noticed a partial loss of hearing and imbalance when walking in a dark room on a soft carpet, such as in a movie theater.

### Assessment

**Perception of Motion.** Impaired perception of limits of stability and of verticality in sitting and standing. She had complaints of unstable vision with head movements and during walking and riding in a car, but no dizziness.

**Motor Strategies.** The patient was unable to balance in tasks requiring use of the hip strategy, such as standing across a beam, standing on one foot, and heel-toe walking. Her head was actively fixed on the trunk during gait and turns, and she complained of neck stiffness.

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

### Discussion

The history of gentamicin treatment followed by hearing loss and vestibular symptoms is consistent with ototoxicity affecting both motor and sensory strategies for postural control. Loss of vestibular function is associated with loss of a hip strategy in tasks with compromised surface somatosensory information as well as fixing of the head on the trunk, which can lead to cervical symptoms. The oscillopsia associated with head movements was likely associated with increased imbalance. The patient showed context-dependent instability when vestibular information was required for postural orientation in environments when both visual and surface somatosensory information were inadequate for stability.

Physical therapy goals focused on reduction of oscillopsia, increasing use of remaining vestibular function, and increasing use of a hip strategy when tasks required 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.

lost vestibular function demonstrate abnormalities in the use of vestibular information in each of its four roles. First, they have difficulty perceiving and reporting whether their bodies or heads are properly oriented to gravity; that is, they have difficulty identifying when they are upright. Second, 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. Fourth, head position with respect to gravity varies a great deal as such patients attempt to recover from the perturbation.

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, patients with vestibular loss often have stiffness of 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 of 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 result from 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 orien-

## CASE STUDY 2

A 34-year-old, physically fit, mounted policeman was kicked in the head by his horse. Afterwards, he reported severe disorientation when riding his horse, especially near moving traffic. Any busy visual environment, such as crowds, stores, patterns, and windshield wipers, was aversive, and he fell when trying to walk on the beach near the ocean waves. He also reports spinning vertigo when he pitches his head backwards.

### Assessment

**Motor Strategies.** Excessive hip strategy with head instability when standing on a firm or foam surface when wearing a visual dome that stabilized vision and when attempting to stop suddenly while walking.

**Sensory Weighting.** The patient had normal sway when standing on a firm or foam surface with eyes open and closed. Sway was excessive, dizziness increased, and he could not stand independently for 30 seconds when wearing a visual dome that stabilized vision.

### Discussion

The visually 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 of 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 that 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. The patient also practiced use of the ankle strategy by swaying forward and backward slowly at the ankles in a variety of sensory conditions and when stopping during gait.

■ 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 | Trunk is oriented to support surface instead of to gravity |
| | Patient stands on foam with eyes closed | Patient falls or sway increases markedly |
| Controlling center of mass | Patient stands or walks on a beam | Patient is not able to use hip strategy to control center of mass, and so falls |
| Stabilizing the head | Patient leans or is tilted | Head is not stabilized with respect to vertical |

tation and balance, the vestibular system also interacts with the parts of the CNS responsible for expectation and learning. Although it is automatic and rapid, postural control is also flexible, capable of adapting to many different sensory environments and musculoskeletal constraints. The role of the vestibular system in postural control cannot be fully appreciated until we better understand the complex and multifaceted nature of postural control itself.

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CHAPTER 4

# Postural Abnormalities in Vestibular Disorders

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When this chapter was written for the first edition of this book, the primary role of the vestibular system was considered to be the control of posture and balance. In recent years, however, 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 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, and another might complain of imbalance and falling, yet both could have the same vestibular disorder.[9] Because the process of central nervous system compensation proceeds over a lengthy time, patients can also have different symptoms when they finally arrive at a clinic, although suffering a similar deficit. Both clinical and experimental observations have shown that along with symptoms of vertigo, past pointing, and nystagmus, equilibrium disturbances are one of 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 is more efficient or effective for the population of patients with a vestibular deficit. If one strategy were better, then a systematic approach to treatment could be followed. But to determine the effectiveness of the compensation, we must first determine how to reliably indicate whether the patient is suffering from postural dyscontrol, and whether vestibular dysfunction is responsible for the symptoms. Although the standard clinical tests of the vestibular system have not altered, new technologies, such as virtual reality (VR), have arisen that allow us to test vestibular function when combined with other sensory signals.[11] In this chapter, some of the methods available for testing postural disorders that are associated with vestibular disease are briefly discussed. Then, postural behaviors that have been quantified and associated with specific vestibular disorders 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, is 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 methods cannot fully describe all disorders of the vestibular system. One problem is that tests of vestibulo-ocular integrity and 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 that are intimately involved in the control of posture and balance. Second, tests of the VOR are commonly performed in the plane of the horizontal semicircular canals, whereas vestibulospinal reflexes depend 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). The VEMP is a sound-evoked muscle response that is believed to be generated from acoustical stimulation of the saccule (one of the otoliths that transduces linear accelerations and decelerations). Thus, the presence or absence of this muscle response provides diagnostic information about the function of the saccule or the inferior vestibular nerve.[12–14] The measured variable is a short-latency response from surface electrodes over the tonically contracted sternocleidomastoid muscle. The VEMP can be recorded from patients who have no hearing but have intact vestibular system function. The diagnostic utility of the VEMP has been examined for various audiovestibular and neurological disorders, including vestibular labyrinthitis, Ménière's disease, benign paroxysmal positional vertigo (BPPV), superior canal dehiscence, Tullio phenomenon, vestibular schwannoma, multiple sclerosis, and spinocerebellar degeneration.

Although dynamic posturography does not directly assess peripheral or central vestibular function, it is a useful tool for identifying disorders of the vestibulospinal system.[15–17] Dynamic posturography assesses balance rather than specific vestibular function. But response patterns specific to individuals with vestibular dysfunction are elicited with dynamic posturography, making it a useful adjunct to more traditional methods of testing vestibular function. Diagnostic tests of the vestibulospinal system that are more easily and inexpensively available to the clinician than dynamic posturography are also discussed in this chapter, as are some of the problems inherent in each method of testing. Table 4-1 presents a summary of the advantages and disadvantages of the tests discussed here.

## *Dynamic Posturography*

### Automatic Postural Responses

In the 1970s, Nashner[18,19] 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. 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.[18–23] Subjects stand on a platform that can be translated in an anterior and posterior direction, or rotated so that the ankles are moved into plantar flexion or dorsiflexion (see Fig. 4.1). The expected response to anterior motion of the platform is backward 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 flexion or dorsiflexion would produce equivalent changes at the ankle (see Fig. 4.1), 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 latencies longer than the stretch reflex but shorter than voluntary reactions in order to bring the body back over the base of support. These restabilizing ankle muscle responses (at latencies of 90 to 120 msec) were followed within 10 to 20 msec by the muscle in the upper leg on the same side of the body (i.e., soleus muscle followed by the hamstrings; tibialis anterior muscle 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.[18,19]

Nashner's original conclusion that the body acts as a rigid, inverted pendulum, reliant primarily on ankle proprioceptive inputs to initiate the restabilizing actions,

**■ Table 4-1 ADVANTAGES AND DISADVANTAGES OF CLINICAL TESTS OF POSTURAL INSTABILITY**

| | Advantages | Disadvantages |
|---|---|---|
| *Static tests:* | | |
| Romberg | Easily performed in clinic | Qualitative<br>Does not test adaptive responses |
| Stabilometry | Quantitative<br>Can manipulate sensory inputs | Requires a force platform<br>Intersegmental shifts confound results<br>Does not test adaptive responses |
| Vestibular evoked myogenic potential | Specific test of the saccule and inferior vestibular nerve | Requires electromyography of the sternocleidomastoid muscle |
| *Dynamic tests:* | | |
| Stepping tests | Easily performed in clinic<br>Can be quantified | Does not test adaptive responses<br>Has not been shown to be reliable |
| Tiltboards | Easily performed in clinic<br>Requires adaptation to external forces | Qualitative<br>Amplitude and application of force are not controlled |
| Posturography | Quantitative<br>Requires adaptation to external forces<br>Can manipulate sensory signals | Requires a posture platform |
| 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 physiological measurements |

does not accurately describe the complex, multisegmental actions that occur during postural restabilization.[21,23–26] 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 with a *hip strategy*.[27] Allum and colleagues[28] concluded that (1) all balance corrections result from one of two basic synergies defined by the timing between segmental responses and (2) the selected synergy was amplitude-modulated according to the initial movement velocities at all joint segments, thereby producing a continuous repertoire of movement strategies for balance. With more realistic stimuli (i.e., 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.[29,30] Other studies have demonstrated that the initiation of the balance reactions depends greatly both on the ability to predict the occurrence of the perturbation[31–34] and on the location of the disturbance on the body.[35–38]

## 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.[39–43] Studies in which the labyrinthine receptors were directly stimulated by vertically dropping human and animal subjects, thereby producing linear acceleratory stimuli,[40–43] demonstrated that direct labyrinthine stimulation can produce automatic or "triggered" postural reactions in the lower limb. Nashner[20] hypothesized, however, that the vestibular system contributes to the control of lower limb balance reactions only when proprioceptive signals are absent or unreliable. With use of a servomechanism, the posture platform was made to match the sway at the hip, thereby maintaining a neutral position at the ankle and, one assumes, eliminating any change in the proprioceptive feedback from the ankle during normal quiet standing. In subjects thus tested, automatic postural responses were significantly delayed when

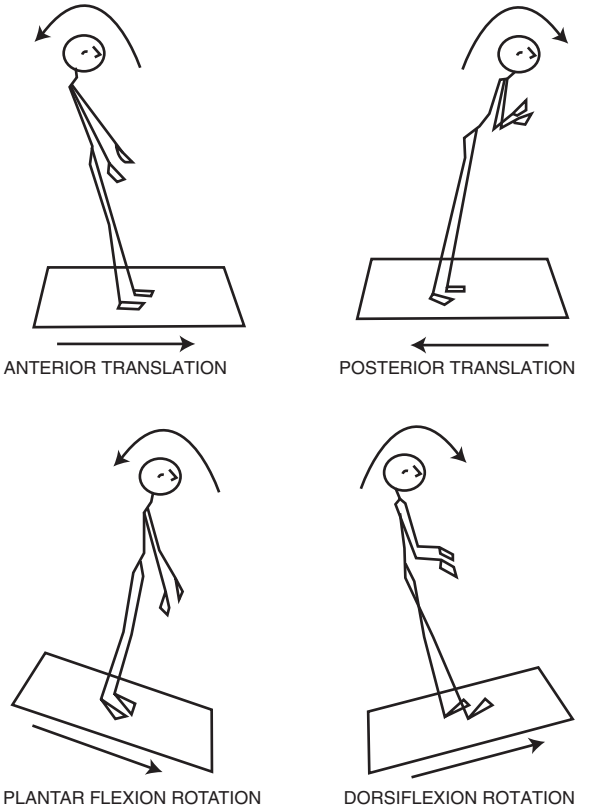


**Figure 4.1** Four directions of perturbation on the dynamic posture platform. Note that in anterior translations and plantarflexion rotations, ankle angles increase as the person compensates for the motion of the platform. In posterior translations and dorsiflexion rotations, ankle angles decrease.

the subjects had to rely on vestibular signals in the absence of proprioceptive feedback from the ankles.

Most probably, the servomechanism did not fully remove the ankle proprioceptor feedback but, rather, produced distorted or modified signals that altered the automatic postural reactions; alternatively, responses to vestibular inputs during quiet stance may not be transferable to the responses observed during dynamic gait and a loss of balance. A novel experiment reported by Allum and colleagues[44,45] appears to resolve the issue of labyrinthine involvement in the generation of postural reactions to support surface displacements. In this experiment, angular displacement of the ankle was kept equal for both platform translations and rotations, thereby producing equivalent proprioceptive signals from the ankle even though the labyrinthine inputs differed. When head acceleration and neck and lower limb muscle EMG responses recorded during both perturbations did not exhibit the same response patterns, these investigators concluded that labyrinthine signals must be directly involved in the generation of lower limb postural reactions. The differences between the subject groups were greatest and most consistent during platform rotations,[45] leading the researchers to conclude that vestibular loss is best diagnosed through a rotation of the support surface. Using a range of platform translation velocities (5–35 cm/sec), Runge and colleagues[46] found that equivalent head accelerations produced very different torques at the hip and knee in healthy subjects and in patients with bilateral vestibular deficit. These findings suggest that the magnitude and force pattern of the muscles depends on 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 and addition of a layer of dense foam to the base of support to make somatosensory inputs less effective.[47] Visual conditions have been controlled by either stabilizing or rotating the visual field in an anterior-posterior plane.[47–49] The emergence of VR technology has now enhanced the ability of scientists and clinicians to explore the impact of visual perturbations on postural responses (see Table 4-1).[11,50]

Although normal subjects, elderly individuals, and people with vestibular deficit exhibit a greater tendency to fall under conditions of altered sensory input,[47,48,51–53] 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 upon 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 patients with labyrinthine loss when visual[48] or somatosensory[54,55] 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.[56–60]

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 using a servomechanism to match the angle of the platform to 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, which alter the transmission of forces from the lower limb to the head and make it difficult to compare vestibular influences on balance in different 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,[23,46,61–63] 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 to obtain 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.[64] 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, and its results are highly variable even for testing the same subject repeatedly.[65] 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.[66]

Tests of quiet stance may indicate the severity of a balance problem, because patients with vestibular system damage demonstrate increased sway and falling when the base of support is constrained during quiet standing. Conclusions about the neural processes contributing to postural imbalance are severely limited, however. 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,[66] but simple removal of vision does not alter the temporal or spatial organization of the automatic postural reactions.[39] Furthermore, behavioral measures as to how often a subject falls, or to which direction he or she deviates, do not convey information about the motor and sensory mechanisms that may be involved in postural control.[67,68] 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.[69,70] Despite these limitations, the concept of deviation from the vertical during quiet standing continues to underlie clinical testing of vestibular dysfunction.

## 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.[71–73] During that time, the subject stands quietly on a force plate, and the excursion of the center of gravity is measured during 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 through alteration of signals from other sensory pathways—for example, adding a layer of foam rubber to the base of support to make somatosensory inputs less effective[47] or placing the subject within a visually controlled environment to modify visual feedback.[48] This attempt to quantify the classic 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.[71] In general, because the sensory apparatus of the vestibular system is most responsive to changes in acceleration and orientation in space,[74] and because patients with vestibular deficits tend to have normal Romberg responses, 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] When the base of support was tilted, the subject reacted to regain a stable equilibrium 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 ($<$ 150 msec after) 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 the 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 test[75] or stepping test of Fukuda[76] 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.[77] A battery of tests developed by Graybiel and Fregly[78] (Ataxia Test Battery) examine subjects standing upright, on one leg, and with feet aligned in tandem position with eyes open or closed as well as while 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 lower than normal subjects on measures of deviation from the straight line or of the number of steps made prior to 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. In order 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.[79] VR technology offers the sensory complexities found in the physical world in the controlled environment of the laboratory (Fig. 4.2).[80] Individuals become immersed in the training environment so that they feel they are part of the scene[81] and perceive that the world is moving about them.[82] Thus, VR allows researchers and clinicians to create a synthetic environment with precise control over a large number of physical variables that influence behavior while recording physiological and kinematic responses.[83]

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.[84] In this case, the visual information had no parameters in common with the physical disturbance and, thus, could have been treated as irrelevant to the postural perturbation and ignored. Instead, in both normal subjects and subjects with labyrinthine deficit, the postural responses became more complexly organized: an additional frequency was incorporated into the intersegmental response, which was also further increased in magnitude. 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 they were unable to produce responses of any greater magnitude in the absence of labyrinthine inputs[39] or that modulating the response magnitude depended on an ability to calculate the visual-vestibular conflict.

## 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.[9,85,86] The focus here is 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.[87,88] A *deficiency* in the

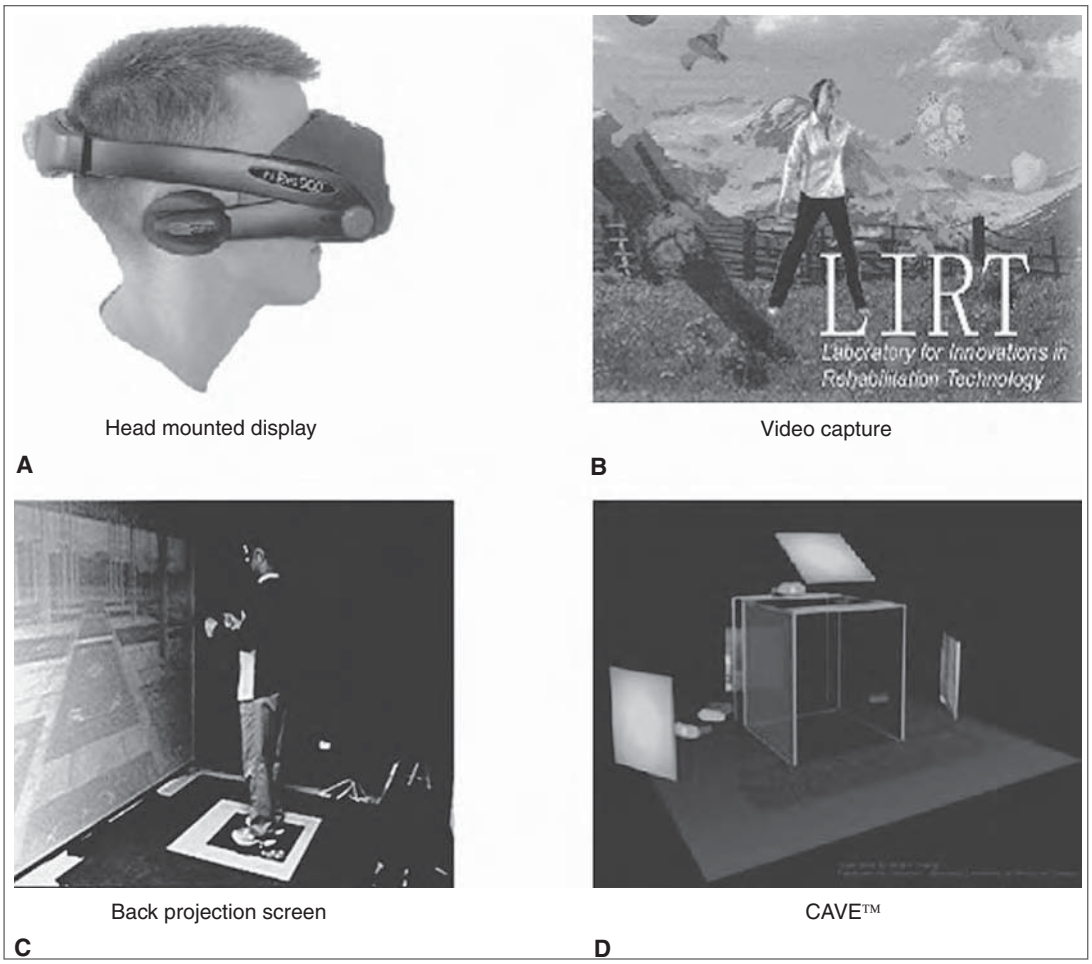


**Figure 4.2** Some examples of virtual reality technology in which the performer can freely move while interacting with the visual images. (*A*) The performer can walk about while wearing the head-mounted display (Hi-Res 900, available from http://www.vrealities.com/hi-res900.html). (*B*) Video capture permits performers to observe themselves interacting with virtual objects at the Laboratory for Innovations in Rehabilitation Technology (http://hw.haifa.ac.il/occupa/LIRT). (*C*) In the Virtual Environment and Postural Orientation Laboratory at the Rehabilitation Institute of Chicago, a dynamic posture platform (NeuroCom International, Inc., http://www.onbalance.com/) has been placed in front of a full field-of-view, back-projection screen in order to simultaneously perturb the base of support and the visual system of the performer (http://www.smpp.northwestern.edu/index.php?option-=com_content&task=view&id=66&Itemid=85). (*D*) The visual image is projected on three walls and the floor of the CAVE Virtual Reality System, and the performer is free to walk about within the virtual environment (http://www.evl.uic.edu/pape/CAVE).

system usually implies that the sensory (i.e., labyrinthine) inputs have been reduced or abolished, resulting mostly in complaints of unsteadiness and instability. *Distortion* means 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 4-2 for the disorders discussed in this chapter.

## Deficient Labyrinthine Inputs

With damage along the VIIIth nerve or within the vestibular labyrinth, signals from the peripheral vestibular apparatus are lost or diminished.[74] Central disturbances originate at the vestibular nuclei or in the higher central pathways that communicate with the vestibular nuclei. In both cases, patients can experience disequilibrium, imbalance, and ataxia. With unilateral lesions of the peripheral

**■ Table 4-2 POSTURAL DISTURBANCES OBSERVED WITH VESTIBULAR DISORDERS**

| | |
|---|---|
| Peripheral vestibular disorders: | |
| Deficient inputs | Need more energy to maintain the upright position<br>Instability increases in the presence of inappropriate sensory signals<br>Amplitudes of EMG and torque are inversely related to severity of deficit |
| Distorted inputs | Still able to process vestibular inputs<br>Falls increase in the presence of inappropriate sensory signals |
| Central vestibular disorders | Impaired perception and location of the gravitational vertical<br>Direction-specific ataxia<br>Falling tends to occur in the direction of quick-phase nystagmus |
| Aging | Longer response latencies and delayed reaction times<br>Diminished sensory acuity and impaired signal detection<br>Temporally disordered postural response patterns |

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 activation of extensor muscles in the extremities of both monkeys[89] and humans[90] with unilateral deficit was found to be 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 while subjects maintain an upright position.[91] Both in the study using this approach and in others using force plates to record sway during quiet stance,[10,73] intersubject variability and overlap between normal and clinical populations reduced the strength of the findings. Results suggest, however, that more energy needs to 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 colleagues,[52,87,91] 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 ("servoed"). 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 in whom labyrinthine inputs were reduced or absent were more unstable than normal controls only when ankle proprioceptive references were proportional to body sway and visual references were either removed or inappropriate (condition 2 or 3). It was believed that when the only reliable source of feedback was the vestibular inputs, patients with vestibular deficiencies would fall because they depended on the somatosensory and visual reference to correctly organize their postural responses. From their studies, the researchers suggested that vestibular deficits could be quantified by systematically altering the sensory information provided by the support surface and the visual surround.

Several problems limit our reliance on these results for clinical diagnosis and measurement. First, patients with unilateral or partial bilateral deficits were sometimes as unstable as patients with total loss of vestibular function,[67] rendering this approach a poor test of graduated

function in the vestibular system. Second, the researchers tested only patients with well-compensated deficits. As discussed later, compensation could occur as a central reorganization in the system. Thus, these experiments may be testing not 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 (T. Hain, S.J. Herdman, personal communication, [1999]).

Allum and colleagues[21,39,44,45,68,92,93] examined both the latencies and the amplitudes of muscle EMG responses on a platform that dorsiflexed the ankle. Areas under the EMG bursts in the ankle muscles as well as soleus and tibialis anterior muscles, and ankle torque recordings of patients with complete bilateral labyrinthine deficit were significantly diminished compared with those in normal subjects with eyes both open and closed.

Using these data, Allum and associates[69] explored the presence of a linear correlation between EMG amplitudes and the extent of the peripheral vestibular deficit. The population measured included subjects with intact labyrinths (normal subjects) as well as those with acute unilateral labyrinthine deficit, chronic unilateral labyrinthine deficit, and bilateral labyrinthine deficit, 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.[45,55] Although latencies of the postural reactions were produced without significant time delays, segmental organization of the postural response were disordered. Allum and colleagues[93] 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 of the postural response to platform rotations. These investigators suggested that the selection of movement strategy depended on the initial direction of trunk and head acceleration (which is oppositely directed in platform rotations and translations) and that the strategy was executed as if preprogrammed from the beginning. Although Horak and coworkers[55] 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 dynamic posturography is used as a diagnostic tool. Runge and associates[46] found that the joint torques at the hip and knee were abnormal over a range of velocities during rearward translations in patients with bilateral deficit. Because head accelerations were the same in the patients and healthy subjects, these investigators' findings agreed with others[21,39] that the magnitude and force pattern of the muscles depends on vestibular inputs from early head movements.

One 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 to deviate toward the affected side. Severe postural disturbances occur when these individuals must rely on vestibular inputs, but the deficit is compensated within 2 weeks after the lesion.[94] Individuals with bilateral deficit exhibit normal postural sway in quiet stance.[55] Removal of vision via eye closure increases this sway by only a small amount.[87] 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.[95]

## Ménière's Syndrome

*Ménière's syndrome*, or endolymphatic hydrops, is considered a vestibular deficiency even though it manifests 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 normal Romberg response during remission,[96] but symptoms of dizziness and instability can occur for several days after intermittent episodes of vertigo. These episodes appear at irregular intervals for years, and about one third of the patients eventually have bilateral involvement.[9]

Objective diagnosis of Ménière's syndrome has depended on long-term documentation of the fluctuating hearing loss. Quantitative posturography measures have been able to identify consistent changes in the postural response of affected patients, even during periods of remission. In one study, the movement pattern of the center of gravity was measured during a stepping test after the researchers observed that patients deviated toward the affected side even during the remission period.[96] The stepping test was performed in the dark with eyes open and closed, and patients were required to perform at a frequency that both was optimal for normal walking and elicited a smooth rhythmical pattern (i.e., 1.2 Hz). When their 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 that 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.[97,98] High-frequency components of standing sway were observed during acute phases of Ménière's syndrome, but not during remission periods.[98] Patients with Ménière's syndrome in whom vestibular hypofunction had not developed, as determined from VOR gains, were also tested under conditions of sensory conflict during quiet standing (see earlier description in section "Indicators of Vestibulospinal Deficiency").[91] These patients responded very much like patients with well-compensated 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 Peripheral Positional Vertigo

In order to study the effects of distorted labyrinthine inputs on posture, patients with BPPV have been examined.[91] 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; they include dizziness and nausea lasting for hours or days.[9] Degeneration of the utricular macula, releasing otoconia that settle on the cupula of the posterior semicircular canal, is strongly implicated as the cause of BPPV and could have 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.[99]

Because of the positional component of this syndrome, postural changes are easily recorded during quiet standing by having a patient with BPPV alter the position of the head in space. After tilting of the head, large amplitudes of anterior-posterior sway and sway ipsilateral to the direction of head tilt were observed in one study.[99] Instability gradually decreased as the vertigo diminished, but with eyes closed, the sway could not be compensated by other inputs, and the patients fell.

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 upon 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.[67,87,91] 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.[74] Convergence of vestibular and somatosensory inputs on the vestibulospinal and reticulospinal[100] neurons can take place at the level of the vestibular nuclear complex, at the adjacent reticular formation, and upon spinal interneurons[101,102] and motoneurons.[100] But growing evidence[103,104] suggests that correct alignment of the head with the trunk and with the gravitoinertial vertical[105] 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.[106] Inputs from either of these modalities are not necessarily redundant, because each represents different parameters and is effective within a particular frequency domain.[107–109] In fact, the frequency of stimulation is important to control in patients with compensated loss, because motor output of the visual system as well as the vestibular system has been found to be frequency dependent.[63,110–113] Thus, it is unlikely that normal postural responses are reflective of the isolated labyrinthine and neck reflexes observed in the decerebrate animal.[114–116] 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.[117,118] A convergence of inputs from more than one sensory modality would then be necessary to create this estimated postural orientation. Studies by Keshner and Kenyon[119] in a virtual environment 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 largely out of phase with the upper body. These researchers inferred, as in a previous study on a posture platform, that the upper body responded to visual-vestibular signals but the ankle responded to proprioception and changes in ground reaction forces.[24] Buchanan and Horak[120] 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 due to a disorder of head and trunk spatial orientation rather than lower limb instability.[121]

Several clinical findings have been suggested to differentiate between a peripheral disturbance and a 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.[122] Balance disorders due to abnormalities of the vestibular nuclear complex have been observed[123,124] but are poorly documented. The majority of the literature about central vestibular brainstem lesions reports only oculomotor abnormalities, but Brandt and colleagues[124] 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 therefore may affect balance.

*Downbeat nystagmus* is specific for a lesion of the paramedian craniocervical junction (30% of cases due to 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 affected patients do not complain of vertigo or balance problems. Brandt and colleagues[124] suggest that the backward sway is vestibulospinal compensation for the forward vertigo resulting from the downbeat nystagmus. *Ocular tilt reaction* is actually a triad of responses, consisting of ipsilateral head-trunk tilt, ocular torsion, and ocular deviation. This condition has been observed in patients with brainstem abscess, multiple sclerosis, and acute Wallenberg's syndrome. Patients seem to have a readjustment in their perception of the vertical that matches tilt deviation of the eye, head, and trunk.

*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 that causes the body to deviate toward the side of the lesion and, consequently, fall. *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 VIIIth nerve can mimic a peripheral labyrinthine disorder, and *drop attacks* (sudden, unpredictable forward falling) can occur with basilar insufficiency. Thus, the evidence from clinical reports suggests that a central vestibular dysfunction results in impairment of perception and location of the gravitational vertical exhibited throughout the whole body postural system. But with all of these syndromes, other motor structures are affected as well and may contribute to the impairment.

## Postural Dysfunction with Disorder of Other Sensory-Motor Centers

The vestibular nuclear complex communicates with motor as well as sensory centers.[122] In fact, extensive

reciprocal connections between the vestibular nuclei and the *cerebellum*[125] 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 depend on the site of the lesion.[126] 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 after dorsiflexion rotations and backward translations on a platform, but amplitudes and durations of response were two to three times greater than normal,[127,128] and habituation to the stimulus was absent.[129] Postural response magnitudes in the patients with anterior lobe damage were scaled correctly when they relied on current somatosensory feedback, but the patients were unable to scale their responses when relying upon 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.[128]

A characteristic sway frequency of 3 Hz has been recorded in this population.[130] Intersegmental counterbalancing actions were enhanced in these patients, so that falling was not commonly observed, but they tended to exhibit a stiff-legged gait. Stance ataxia was found to improve with visual feedback, unlike that appearing with vestibulospinal lesions.[131,132] 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 they cannot walk. Ataxia of the head and trunk is observed during 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 responses with eyes open and closed are compared. These patients tend to fall even when sitting down, which may be due to diminished intersegmental movement for counterbalancing or to truncal ataxia.[126,131] *Neocerebellum* lesions produce little postural instability or disturbance of stance even with eye closure. Control of position of the body's center of mass seems to be disturbed, because patients with these lesions exhibit ataxia during a limb and trunk pursuit task.[126] Reports of head and trunk deviation to the side of the lesion have also appeared.[131]

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 patients with such disorders,[133] although long-latency responses to direct stretch of a muscle have been observed to be enhanced in the Parkinson population in both the arm[134] and leg.[135] Response latencies to sudden platform displacements were found to be within the normal range,[136,137] although the ability to suppress long-latency muscle reactions to a perturbation was impaired in these patients even under supported conditions.[138] 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.[139] 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.[140] The inability to generate adequate force in the stabilizing muscles appears, therefore, to be hampering successful postural reactions in these patients. Inferring that 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 *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.[141] 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.[142] With augmented feedback, such as a warning tone and knowledge of perturbation direction, however, timing of the postural responses improved.[143] When balancing on a seesaw apparatus, patients with spastic hemiparesis minimized the high-frequency, anterior-posterior sway on the affected side with a corresponding reduction of the EMG response in the tibialis anterior muscle.[144,145] Electrical stimulation of the tibial nerve in patients on the seesaw revealed a delayed and diminished EMG response of the tibialis anterior muscle 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.[144,145]

Children with cerebral palsy have also been studied with a posture platform.[146] 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 but 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 "Indicators of Vestibulospinal Deficiency"). Thus, postural abnormalities of children with cerebral palsy were due either to muscle incoordination or to 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.[141]

## 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 (Box 4-1). Changes in the postural response organization with loss of labyrinthine inputs have been described in detail earlier in 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.[92] Thus, the response does not reach an appropriate gain to maintain stability, and restabilizing torques at the ankle are inadequate to prevent falls.

Second, the patient with peripheral vestibular deficits tends 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 their heads in the same direction as their bodies (head locked to trunk) in association with absence of neck torques,[147] whereas normal subjects exhibit a counter-rotation of the head and body in the sagittal plane.[61,93] This finding correlates with clinical observations that patients with vestibular deficits increase the 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.[39] 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[63,111] and is typical of natural head movements during locomotion.[148,149] 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.

Box 4-1

**MODIFICATIONS OF POSTURAL STABILITY AFTER LOSS OF SPECIFIC SENSORY INPUTS**

Labyrinthine deficits:
- Stiffening between body segments
- Increased sway at high frequencies

Somatosensory deficits:
- Low-frequency sway during quiet stance
- Delayed restabilization
- Increased lateral sway

Visual deficits:
- Increased sway at low frequencies
- Increased sway at high frequencies when labyrinthine inputs are also absent

Somatosensory inputs provide powerful feedback about motion of the limbs and stabilize body sway at the lower frequencies (<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 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.[138,150] When proprioceptive feedback from the ankle is excluded or suppressed in normal subjects during perturbations on a posture platform,[18,151] a characteristic low-frequency (1-Hz) sway emerges. 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.[152] In the absence of feedback from these pathways to the cerebellum, a significant delay of the restabilizing response of the tibialis anterior muscle after dorsiflexing ankle rotations on a posture platform has been observed.[127] Affected patients exhibit large lateral sway deviations in the low-frequency range (>1 Hz) with eyes closed, as do patients with tabes dorsalis.[151] Patients with sensory polyneuropathy of the lower extremities demonstrate ataxia and instability during quiet stance. Falls tend to occur when the eyes are closed,[152] 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.[153,154] The neck proprioceptors have intimate connections with the vestibular system and are probably used both as feedforward and feedback to the vestibulocollic reflexes.[155] Cervical ataxia has been a controversial diagnosis, however, because there are no hard signs to identify the neck as the source of the dizziness.[154] Karlberg and colleagues[156–158] 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 towards 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 that in patients with vestibular neuritis, suggesting that disorders of the neck should be considered in the assessment of patients complaining of dizziness, vertigo, and balance disturbances.

Visual signals are used to accurately detect and reduce motion relative to the surround.[159,160] 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 does not produce significant changes in the postural response organization, although greater sway amplitudes may appear.[39,49,51] Instead, visual information is thought to be redundant unless both vestibular and somatosensory inputs are lost.[160] 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.[48] 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 normals. At higher frequencies (0.2 Hz), the patients' sway increased beyond normal limits, indicating that patients with vestibular deficit could rely upon visual inputs at lower frequencies, but suffered for the loss of vestibular signals at higher frequencies.[66,161] Patients with labyrinthine deficit tested with a stabilometer were better able to stabilize sway when fixating on a stationary light.[162] When an optokinetic stimulus was introduced, the patients became unstable, suggesting that velocity information received through peripheral vision was the cause of the increased sway.

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 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 domain.[66,107,108] Thus, the falls observed in patients with vestibular deficits, particularly after a platform perturbation or in the absence of other sensory signals, may be due to uncontrolled or poorly compensated oscillations of intersegmental structures at particular frequencies of sway.

## Compensatory Processes

Compensation for vestibular disorder is a gradual process of functional recovery that is probably of central origin.[163,164] Although adaptation (i.e., long-term changes in gain) of lower limb postural reactions has been observed in patients with labyrinthine deficit,[90,165] 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.[164] 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 among subjects or in a particular subject for different functions.[164,166] 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.[167–169] Subjects asked to co-contract their neck muscles to counteract random sum-of-sines rotations exhibited neural control over a greater bandwidth than when distracted or relaxed,[170] suggesting that feedforward signals could alter the reactive components of the response. Horak and Nashner[27] 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.[60]

Central control over postural responses can be measured in studies examining predictive processes. For example, Guitton and associates[110] 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. Patients with vestibular deficits 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 msec) 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 $>$ 150 msec) 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.[169,171] 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,[19,39] suggesting central habituation of these responses at the cortical or spinal level. Selection of postural strategies on a translating platform is influenced by prior experience as well as current feedback information.[27] 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 patients with chronic labyrinthine deficits eventually demonstrate normal sway,[73] 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.[172] With testing 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, revealing 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 greater instability of the elderly, because anatomic studies have revealed a gradually decreasing density of labyrinthine hair cell receptors beginning at age 30 years, and a steeper decline in the number of vestibular receptor ganglion cells beginning around age 55 to 60 years .[173–175] Although caloric measures of the peripheral vestibular system have demonstrated declining function with age,[174] 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.[173–177] Thus, their compensatory approach to postur-

al instability may not be the same as that in patients who have experienced an acute but sustained loss of a single input (see Table 4-2).

Age-related trends in the VOR and optokinetic reflexes have been shown to correlate well with anatomic changes found in the peripheral vestibular system.[176,177] Declining gains and greater 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. These findings would suggest that elderly subjects rely more on the visual tracking reflexes, and that longer visuomotor processing time could contribute to feedback delays and poor performance.[176] Thus, elderly individuals 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.

If conflicting sensory information is not suppressed, but instead can influence and modify the weighting of all other sensory inputs, older individuals may be particularly disturbed in the presence of conflicting visual and somatosensory information. Postural responses of older subjects have been observed to be poorly organized during combined support surface perturbations and mental distraction tasks[178,179] and the predictive components of posture were noted to be absent.[180] Results from a number of studies[181–183] 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 balance, 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 subjects in several studies.[184–186]

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.[187,188] The dynamic process of maintaining an upright posture is compromised in the elderly, as evidenced by this higher incidence of falling. Lengthened response latencies, increased static sway, and muscle weakness have been cited as contributing to falls in the elderly,[189,190] but none has been identified as a causal factor. Attentional demands and disorganized postural strategies are more global parameters that have been targeted as potentially generative causes of falls.[95,178,191–193] Increased instability on a moving platform[194,195] and during locomotion[196–198] has also been attributed to an increasingly rigid trunk with aging. It has been observed that older adults stiffen the trunk in order to decrease the controlled degrees of freedom in an effort to make themselves more stable,[192,199] yet they still produce reduced head stabilization in space.[200,201]

A longitudinal study of elderly people who have fallen has found significant changes in the mean frequency of postural sway in the medial-lateral direction.[202] A low-frequency component was identified in this plane, suggesting a slow postural drift during quiet standing.[203] 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.[204] Latencies of EMG responses and of reaction times are increased in the elderly population,[191,204,205] as is quiet sway.[206,207] Although measures of both sway induced by platform perturbation and quiet sway demonstrated significant aging-related decreases in stability,[208] 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."[208]

A study of elderly individuals on a rotating posture platform[95] 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,[39] lead to decreased stabilizing ankle torques. Weakness of the tibialis anterior muscle has been described in the elderly[190] and could be a major contributor to this diminished torque response.

Greater stiffening between the head and trunk has also been reported in elderly subjects attempting to keep the head stationary in space when the trunk was moving[199,200,209] and during whole-body postural control.[36] 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,[36,200,210,211] which could be due to a stiffer mechanical system. If elderly individuals lock the head to the trunk in order to decrease the controlled degrees of freedom, these greater torques would be transmitted to the head. These data imply that elderly subjects rely upon 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 propriospinal reflex responses and a postural strategy to increase stiffness of the aging musculoskeletal system.

## Summary

The following conclusions can be drawn 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.

Thus, clinicians and researchers should identify the preplanned and automatic components of a postural response to determine how best to influence the postural response organization. Recognizing the multiple factors that contribute to the outcome of a postural response should help clinicians determine the approach and effectiveness of their intervention strategies for retraining and restoration of postural function in patients.

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CHAPTER 5

# Vestibular Compensation: Clinical Changes in Vestibular Function with Time after Unilateral Vestibular Loss

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Sudden, complete unilateral loss of vestibular function in normal, healthy individuals results in a dramatic series of symptoms: rapid eye movements, postural instability, inadequate compensatory responses to head movement, as well as changes in the perception of body orientation and movement. We use the term *uVD* (unilateral vestibular deafferentation) *syndrome* to refer to these symptoms, which are discussed in more detail later. Within a few days or weeks, some of these symptoms diminish and some do not; the term used to describe the general recovery from this syndrome is *vestibular compensation*. The symptoms of the uVD syndrome are complex and varied, and many factors can affect their occurrence and diminution. Simply listing the symptoms and their change over time does not help the understanding of the uVD syndrome and the recovery from it. To clarify what happens, we will examine the neural basis of the symptoms and their diminution.

Why does a uVD cause these symptoms? Why are these symptoms not permanent? The answers lie in the neural basis of the uVD syndrome and the changes in the pattern of neural activity over time, mainly in the vestibular nuclei of the brainstem. Immediately upon loss of one labyrinth, there are major changes in neural activity in each of 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.

The following 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 and relay output to many destinations, most importantly to oculomotor (vestibulo-ocular), postural (vestibulo-spinal), and also 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 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 their activity 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 their activity (Fig. 5.1).

The multiple-input character of neurons in the vestibular nuclei is important for understanding why so many seemingly unrelated sensory inputs can affect the uVD syndrome and recovery from it (see Fig. 5.1). The transmission of vestibular activity can be affected by the cerebellum. 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!

In this chapter, we review the behavioral, clinical, and experimental evidence concerning neural transmission of vestibular information, how it is modified by unilateral loss, and what happens over time as recovery takes place. For other reviews of vestibular compensation, see references 1 through 18.

## Overview

Similar patterns of responses after uVD occur in different species, which 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. But in some of these species (rats and guinea pigs), this spontaneous nystagmus disappears in about 1 day, whereas in other species (such as humans), the disappearance takes longer. Given this similarity among species, one can reasonably assume that similar neural processes underlie the recovery in these different species; that assumption is the basis for using results from experimental studies on animals to understand human vestibular compensation. Although vestibular compensation appears to be a single recovery process, there is overwhelming evidence that many different processes are changing during vestibular compensation and that the various processes recover at different rates or, in some cases, do not recover at all.

One conundrum is that, although most patients recover from uVD and lose their acute symptoms quickly, others experience incomplete recovery. The term used to describe such partial recovery is "poorly compensated". In other patients who have recovered completely and appear to be fully functional (who are "well-compensated" ), there can be a near total relapse—called *decompensation*—under particular conditions, such as severe stress.

## Causes

Patients with uVD are not a homogeneous group but the very opposite: They constitute a very heterogeneous group, and that heterogeneity is a major reason for the heterogeneous results of vestibular compensation. The cause and extent of the loss of vestibular input are factors that must be considered in individual patient cases. There are many possible causes of unilateral loss of peripheral vestibular function, including disease, trauma, surgery

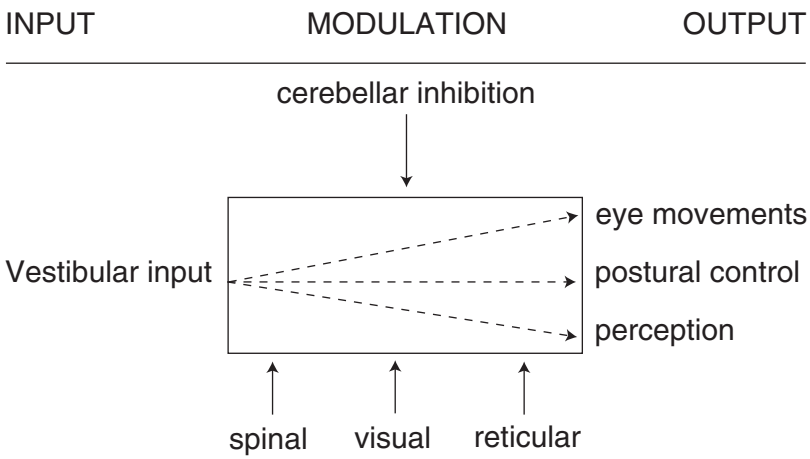


**Figure 5.1** A simple general schematic illustration to represent the responses that can be controlled by vestibular input and how transmission of the vestibular input to govern those responses can be modulated by various sources—visual, spinal, reticular, and cerebellar. This scheme should not be taken too literally; for example, there is evidence that visual input differentially affects eye movements and posture.

(e.g., for removal of acoustic neuroma), and therapeutic intratympanic injection of gentamicin for the treatment of peripheral vestibular disorders.

The reason for the uVD can be a determinant of the severity of the symptoms and the rate of recovery. A normal healthy person who suddenly loses all peripheral vestibular input from one ear experiences dramatic symptoms (nystagmus, postural instability, and perceptual phenomena). At the other extreme, someone who has had a slowly growing acoustic neuroma on one vestibular nerve and then has undergone surgical removal of the neuroma and the entire VIIIth nerve may exhibit almost no uVD symptoms. The reason for lack of symptoms in such a patient is presumably a progressive vestibular loss for which the patient has been progressively compensating as the neuroma grew. So by the time the surgery takes place, the patient has no effective vestibular function in the affected ear. Therefore, he or she has no uVD syndrome and so no vestibular compensation after surgery. Without knowledge of the preoperative level of vestibular function and an understanding of the mechanism of vestibular compensation, these totally different uVD syndromes would be inexplicable.

The anatomy of the peripheral vestibular nerve is also important for understanding the symptoms experienced by the patients, their recovery, and the variability among patients. Although most vestibular afferent neurons course in the vestibular divisions of the VIIIth cranial nerve, some vestibular neurons course in the cochlear division of this nerve. If a patient still has hearing after undergoing a procedure such as surgical vestibular neurectomy, it is highly likely that residual vestibular neurons still project from the affected labyrinth to the brainstem, where 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 VIIIth nerve, whereas vestibular neurectomy involves sectioning only one division of the nerve (e.g., the superior vestibular nerve). The various vestibular sensory regions project their information to the vestibular nucleus in different bundles of fibers within the vestibular nerve, so neurectomy will spare some vestibular neurons. 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 other branch (the inferior vestibular nerve).

One procedure now very widely used to treat unilateral peripheral vestibular dysfunction, largely because of its safety, is intratympanic injection of a solution of the ototoxic antibiotic gentamicin via a fine needle inserted through the eardrum into the middle ear (see Carey[19] for a review). The gentamicin is taken up into the inner ear through the round window of the cochlea and selectively attacks vestibular receptors in preference to cochlear receptors, even though both auditory and vestibular receptors are hair cells.

At low doses, gentamicin attacks the cilia of vestibular receptors, disabling their response to natural vestibular stimuli but having little effect on cochlear receptors, so hearing is minimally affected. Interestingly, 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.[20] Physiological evidence shows that these receptors and the afferent neurons terminating on them convey information primarily about *changes* in vestibular stimulation; one would expect that if they are lost, information about changes in vestibular stimulation would be affected early. Evidence now shows that behavioral changes correspond to that progressive loss.[21] It is worth noting that the cell bodies of the receptors may be preserved during the course of the gentamicin treatment, and the receptor cilia on these vestibular receptor cells may regrow afterwards, so that vestibular function (and possibly vestibular symptoms) may reappear following gentamicin treatment.[22]

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

One of the themes of this chapter is 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. Obtaining such data requires a very modest investment of time in relation to the possible outcome in a patient who compensates poorly for the loss due to the procedure because the supposedly healthy other ear was, in fact, dysfunctional. Such a patient may spend many years in therapy and rehabilitation in an attempt to overcome the poor vestibular compensation that was due to 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 few, some long-term quantitative data on the precise sensory and motor consequences of uVD in humans are available.[23–25] In particular, some patients with Ménière's disease probably have normal or near-normal labyrinthine function between attacks of vertigo, and some of these patients have been studied before and after unilateral vestibular neurectomy.

## The uVD Syndrome

What are the symptoms after uVD? Humans have intense disequilibrium with 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 become apparent during movement. We discuss each category in turn.

### *Static Symptoms*

Immediately after uVD, there is a spontaneous, mainly horizontal, ocular nystagmus. Both eyes show rapid eye movements, so that they appear to be beating away from the affected (lesioned) side. In fact, recordings of eye position show there are slow eye deviations (called slow phases) toward the affected side, followed by rapid eye movements (called quick phases) away from the affected side (Fig. 5.2). Those slow deviations are not apparent to an observer and can be detected only by careful recording procedures. To the observer, therefore, both eyes appear to be beating away from the affected ear. Because this nystagmus can be reduced or entirely suppressed by visual stimuli, a way of viewing eye movements in darkness or without any visual fixation stimuli present is needed in order to observe it. Two ways of doing so are the use of Frenzel glasses and video recordings of eye movements in total darkness with the eye illuminated by (invisible) infrared light. In the week after the occurrence of uVD, the strength of the spontaneous nystagmus in darkness declines, but for some patients, a very small spontaneous nystagmus in darkness remains as a permanent "legacy" of the uVD.

The horizontal component of the spontaneous nystagmus occurs because the uVD removes peripheral vestibular input to one vestibular nucleus, creating an imbalance in average neural activity between the two vestibular nuclei.[26,27] This imbalance in neural activity is very similar to the imbalance produced by a large maintained horizontal angular acceleration to the intact side. In both cases the nystagmus is the vestibulo-ocular response generated by that vestibular neural imbalance.

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 eye movements (anterior canal causes upward movements, and posterior canal causes downward movements). The loss of both of these sensory regions will result in a net cancellation of vertical components.

#### Ocular Tilt Reaction

In addition to spontaneous nystagmus, there is a static otolith component called the *ocular tilt reaction*.[28] It seems that these static otolithic symptoms are also due to the neural imbalance between the vestibular nuclei. Removing all otolithic input from one vestibular nucleus results in an imbalance in neural activity between the two vestibular nuclei similar to that occurring during a large maintained roll head tilt toward one side.[29] The otolithic responses to unilateral vestibular loss can be seen as compensatory responses for such a head tilt –

- Head tilt to the lesioned side
- Skew deviation to the lesioned side
- Conjugate ocular torsion to the lesioned side

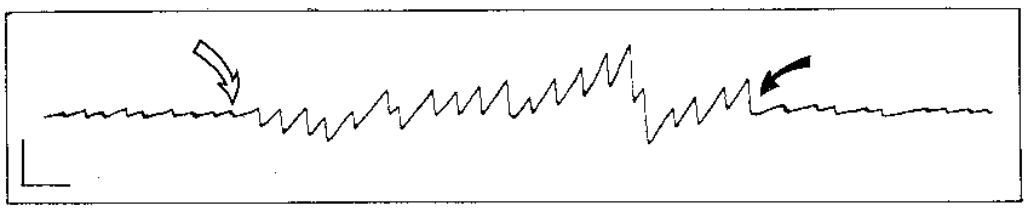

**Figure 5.2** Peripheral vestibular nystagmus. Oculographic recording showing a left-beating primary-position nystagmus that is obvious only when visual fixation is removed (*open arrow*) and that is quickly suppressed again when visual fixation is permitted (*solid arrow*). One can detect peripheral vestibular nystagmus clinically by viewing the fundus of one eye while occluding the other. This patient had undergone a right vestibular neurectomy the previous day. Upward deflections indicate rightward eye movements, and downward deflections indicate leftward eye movements. *Bars* represent 10 degrees and 1 second.

In *skew deviation*, the ipsilesional eye is positioned lower in its orbit relative to the 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 resolves rapidly.[28,30,31] In *ocular torsion*, both eyes roll with the upper pole toward the lesioned ear and take up a maintained ocular torsional position, which can be up to 15 degrees of ocular torsion relative to the preoperative torsional position (Fig. 5.3).[25,28,30,32,33]

This aberrant ocular torsion position resolves slowly over time, but some residual ocular torsion seems to be a permanent legacy of a uVD.[25] How can it be measured? Examination of the fundus of the eye of affected patients shows that the torsional position of the eye has changed: The upper pole of both eyes has rolled to the lesioned side.[25] However, when the torsion is small, it is difficult to detect with fundus observation. A more sensitive way is to use the change in perception of the subjective

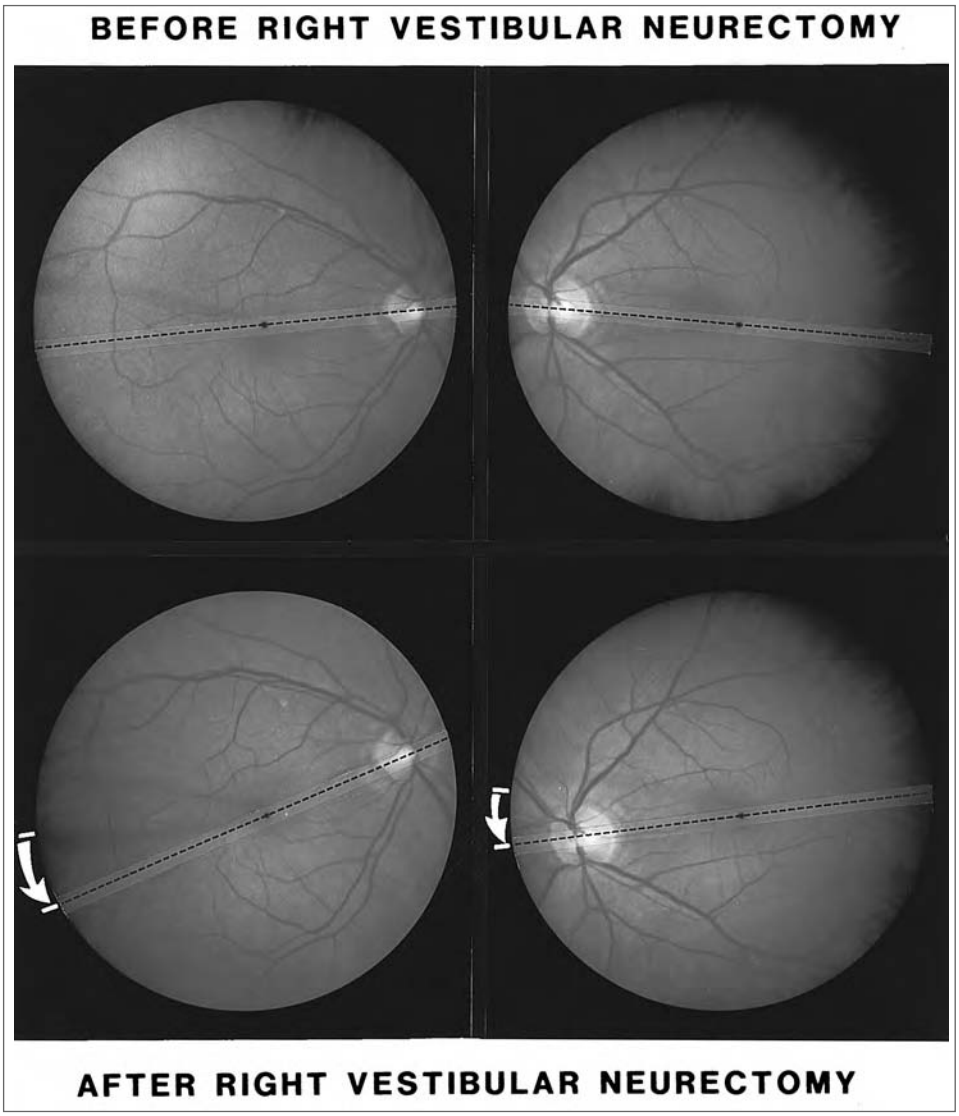


**Figure 5.3** Fundus photographs of the left and right eyes of a patient before (*top*) and 1 week after (*bottom*) right vestibular neurectomy. After unilateral vestibular deafferentation (uVD) there is tonic torsion of the 12 o'clock meridian of each eye toward the patient's right side. The change 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.[25])

visual horizontal, which closely corresponds to the change in ocular torsional position.[25,34] The clinician can so measure visual perception 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.[25,31,35–37] Comparison of the ocular torsional position and the subjective visual horizontal shows that patients rotate this visual line so that its image lies along their (torted) retinal meridian.

We have studied patients before and after unilateral vestibular neurectomy and demonstrated that the size of the change in the perception of the subjective visual horizontal corresponds almost exactly to the change in ocular torsional position and that the torsion position and perception recover in parallel over time (see Chapter 9). Our findings have been confirmed by others. The change in visual perception of the horizontal (or vertical) is a useful indicator of the recovery of otolithic function over time.[38–40]

The clinical significance of these findings is that careful standardized measurement of the subjective visual horizontal 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 Chapter 9).[25,40] A significant tilting of the subjective visual horizontal (more than 3 degrees from the true horizontal) indicates vestibular, probably mainly otolithic, hypofunction on the side to which the patient tilts the bar. It is reasoned to be mainly otolithic, because semicircular canal activation or loss in alert, drug-free humans generates not a tonically maintained eye position but, rather, a changing eye position (nystagmus). Although the tilting of the visual horizontal appears to be due to ocular torsion, the mechanism by which the unilateral otolithic loss causes this ocular torsion itself is speculative. It is most likely similar to the mechanism of the spontaneous nystagmus that occurs after uVD: It probably reflects an imbalance in average neural activity in the two lateral vestibular nuclei (LVNs) due to decreased resting activity in otolithic secondary vestibular neurons in the ipsilesional vestibular nucleus, which itself results from the loss of input from primary otolithic neurons.[29]

### Lateropulsion

After uVD, there is an offset in posture toward the affected side that can be demonstrated by a variety of simple tasks showing 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, called *lateropulsion,* decreases and disappears within about a month.[42]

### Recovery from 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 following a uVD do not change very much over time, although they may appear to resolve. It seems that these dynamic symptoms are progressively masked by other behaviors as time goes by. Careful testing with posture platform tests shows a permanent deficit in postural stability after uVD.[43]

## *Dynamic Symptoms*

The dynamic vestibulo-ocular reflex (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.[44–46] 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.[24,47] Because of the failure to exclude extraneous sources of oculomotor control, there may *appear* to be vestibular recovery, but careful measures with specific vestibular tests show that there is no such recovery. 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 that patients with uVD complain about have high acceleration (2000–3000 degrees per second per second [deg/sec/sec]) and high frequency (5–12 Hz).

In order to measure dynamic vestibular function *specifically,* it is necessary to use high accelerations. One test that does so is the head impulse test, which uses short-duration angular accelerations in the natural range (2000–3000 deg/sec/sec). A simple version of this test can be conducted anywhere. The clinician faces the patient, holds the patient's head at arm's length, and then turns the patient's head in an abrupt, unpredictable horizontal head rotation of about 20 degrees in less than 1 second while asking the patient to stare at the tip of the clinician's nose and to not blink. Although it is very brief, this kind of abrupt head rotation has a peak angular velocity of around 200 deg/sec and a peak angular acceleration of 2500

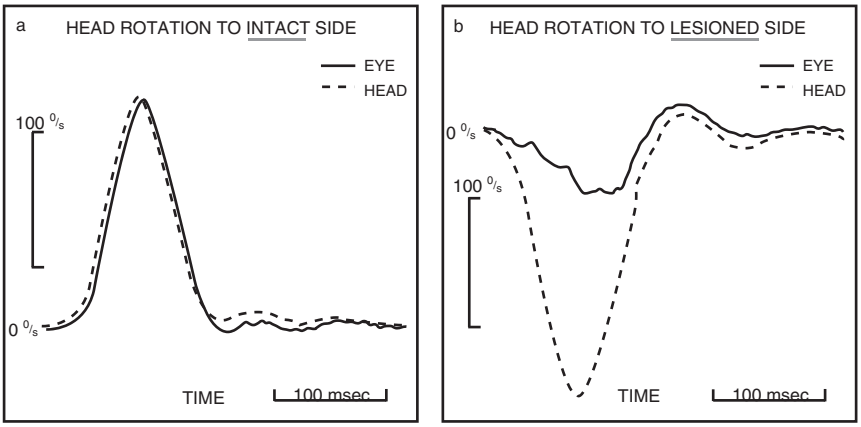


**Figure 5.4** Time series of the eye velocity and head velocity response during a single horizontal passive head impulse from a patient who had undergone unilateral vestibular neurectomy 3 years previously. Head velocity is represented by *dashed lines*, and eye velocity by *continuous lines*. For rotation to the intact side, eye velocity mirrors head velocity. 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.[24])

deg/sec/sec. In normal subjects this head rotation results in a short-latency (about 10 msec) smooth compensatory eye movement, so that the gaze remains fixed on the clinician's nose irrespective of whether the rotation is to the left or right. This means that in normal subjects, *VOR gain* (the ratio of eye velocity to head velocity) is close to 1.0. For a patient with a unilateral vestibular 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 such so-called ipsilesional head rotations, whereas the VOR gain for head rotations to the healthy side (contralesional rotations) is only modestly reduced (Fig 5.4).

The inadequate eye movement response during the ipsilesional head rotation means that during the head rotation, the patient's eyes are dragged with the head and so accumulate a position error, which is usually corrected by a saccade at the end of the head movement that takes the patient's gaze back to the clinician's nose. It is that corrective saccade that the clinician can (with a little training and practice) detect—the telltale sign of an inadequate VOR (Fig. 5.5).[24,48]

Testing over time shows that the ipsilesional VOR gain during the first 100 msec, before other sources of oculomotor control can affect the response, remains at around 25% of normal, whereas the contralesional VOR gain remains at around 80% of normal values.[24,49,50] This VOR asymmetry also applies to animals.[51–54] That permanent deficit of dynamic VOR function after uVD shows that vestibular rehabilitation of uVD patients should be aimed at developing new behaviors that overcome or substitute for the dynamic vestibular loss, rather than trying to restore something that cannot be restored.

If patients with unilateral vestibular loss are asked to maintain gaze in a comparable fashion to the passive test just described while *actively* turning the head abruptly to left or right, most patients learn to pre-program a small saccade to correct the inadequate VOR after some trials, but they learn to insert this saccade *during* the head movement (Fig. 5.6).[43] This is an important observation, because it shows that there is a substitution of a saccade for the deficient vestibular slow phase, and this saccade

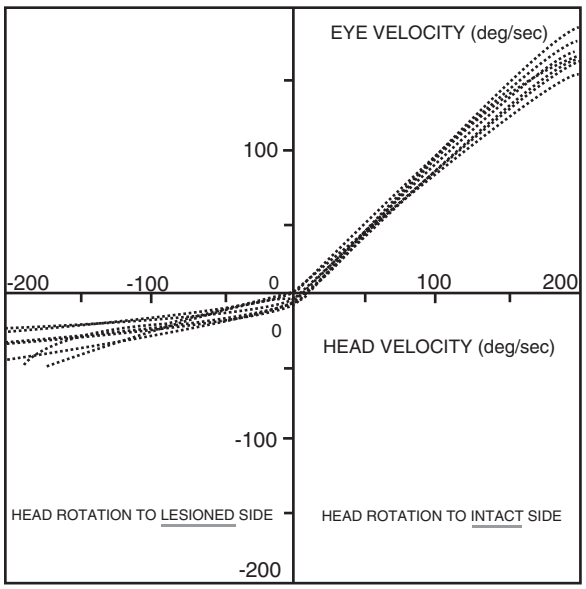


**Figure 5.5** Plots showing the relationship between horizontal eye velocity and the corresponding horizontal head velocity during a head impulse for repeated horizontal head impulses in a patient who had undergone a left vestibular neurectomy 3 years previously. There is a profound vestibulo-ocular reflex (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.[24])

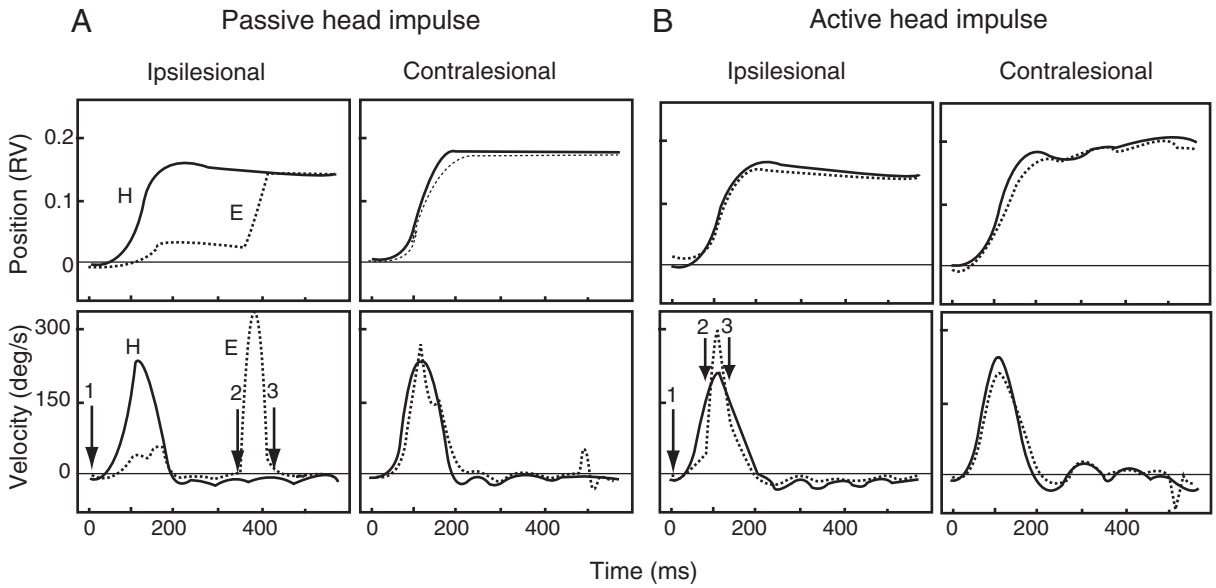


**Figure 5.6** Time series of eye and head velocity during typical passive (*A*) and active (*B*) head impulses for both ipsilesional and contralesional head rotations. The eye velocity response has been inverted to allow close comparison with the head velocity. The start of the head rotation is indicated by the *arrow* (*1*), the start of the compensatory saccadic eye movement by *2* and the end of that compensatory saccade by *3*. (*A*) During passive ipsilesional head rotations, a large gaze error develops during the head rotation, persists until the saccade occurs after the end of the head rotation, then acts to return the patient's eyes to the target. (*B*) With a comparable active head rotation to the ipsilesional side, this compensatory saccade occurs *during* the head rotation, so the gaze error is small and no corrective saccade is needed after the end of the head rotation. Only close inspection of this low-noise search-coil record shows the presence of this small but very effective saccade during the active head rotation. (From Black et al., 2005.[43])

acts to minimize the effect of the uVD on the patients' permanent dynamic VOR deficit; in this way the saccade minimizes retinal smear.[55–58] Another way of concealing a VOR inadequacy is with 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 patients with uVD. Ironically, most tests of VOR (including the head impulse test) require the patient to keep the eyes open during testing. In other words, the testing procedure itself may require the patient to suppress one of the strategies (blinks) that they may have learned to assist vestibular recovery.

After uVD there is a small symmetrical loss of VOR gain for pitch head rotations for both pitch nose-down and pitch nose-up. [59–63] But there is a larger loss for roll rotations. 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.[63]

## Otolithic Dynamic Responses

In normal healthy people, during dynamic otolithic stimulation, as the head is rolled at constant velocity the magnitude of the otolithic stimulation on the receptors in the inner ear changes, and the result is that both eyes show a small compensatory ocular torsion response called *ocular counter-rolling* (OCR). This small ocular torsion is in the direction opposite to roll tilt. OCR is one of the few accepted measures of otolith function.[64,65] After uVD there are deficits in otolithic dynamic responses. Schmid-Priscoveanu and colleagues,[66] testing patients before and after uVD, found that shortly after uVD, there was an asymmetry in the OCR response, with smaller OCR for roll-tilts of the head toward the affected side. However, they found this otolithic asymmetry

recovers fairly quickly; it is not like the permanent asymmetry of the angular VOR.

Impulses of linear acceleration (so-called head heaves) — whereby the patient's head is moved laterally toward or away from the affected ear[67,68]—also show an asymmetrical response.[69–71] This lateral or horizontal head movement causes a horizontal eye rotation in normal subjects that appears to be due to the otoliths because there are no angular head rotations. These head heave responses after uVD show an asymmetry, with a smaller horizontal eye velocity for linear accelerations directed to the operated ear (again, 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]

## Sensory Components

After uVD, patients exhibit two different false spatial sensations. Both illusions occur while patients are stationary, and both 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, patients judge themselves to be rotating around the long axis of the body, toward the side of the uVD (i.e., ipsilesional rotation). However, if their eyes are open, the patients perceive that they are stationary and that the world is rotating around them in the opposite direction, toward the intact side. This false sensation of rotation, called *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. Patients with uVD underestimate the magnitude of both angular and linear accelerations directed to the ipsilesional side.[23] The linear acceleration perception has been studied with the use of roll-tilt stimuli; Dai and associates[23] found that patients with uVD underestimate roll-tilts to the affected ear, compared with the same patients' accurate performance before uVD. Other researchers have confirmed asymmetries of roll-tilt perception.[35,37,38,73] This perceptual deficit is reduced with time, but the recovery is never total.

Box 5-1 lists the permanent legacies of unilateral vestibular loss. Table 5-1 summarizes the uVD symptoms and the general extent of recovery from them. Most symptoms are at their maximum shortly after the uVD procedure (when effects of anesthesia, etc., have worn off).

## Clinical Evidence Concerning Factors Affecting the uVD Syndrome and Vestibular Compensation

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.

Most human 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, these patients present with symptoms similar to those of the patients with good compensation and the uVD procedures were similar, but they do not recover as well as others. "Recovery" has a large subjective component, and it is probable that some patients had expected a much better outcome. Detailed comparison of the vestibular performance of such patients has not been able to identify any clear differences after uVD procedures between those with good compensation and those with poor compensation (see Box 5-1).

Patients with poor compensation exhibit a syndrome called *chronic vestibular insufficiency,* which consists of sensations of dysequilibrium, gait ataxia (especially with restricted vision on unstable surfaces), and oscillopsia. The ataxia is particularly evident when vision is disrupted and proprioception is challenged (e.g., by asking

Box 5-1

**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.

## ■ Table 5-1 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<br>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<br>Reduced VOR gain (80% of normal values) for high frequency high angular acceleration stimulation (natural values) in a contralesional direction<br>Net result—asymmetrical VOR | Little change over 1 year and apparently a permanent loss<br>Little change over 1 year and apparently a permanent loss<br>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<br>Reduced VOR gain for high angular accelerations in a contralesional direction | Little change over 1 year<br>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<br>Reduced VOR gain (80% of normal values) for high frequency high angular acceleration stimulation (natural values) in a contralesional direction<br>Net result – asymmetrical VOR | Little change over 1 year |

*(continued on following page)*

## ■ Table 5-1 uVD SYNDROME: SYMPTOMS AND EXTENT OF RECOVERY *(continued)*

| uVD Syndrome | Description/Type | Extent of Recovery |
|---|---|---|
| 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 |
| Vestibular evoked myogenic potential (VEMP) | Absent short latency inhibition of ipsilesional sternocleidomastoid response to intense air-conducted clicks | 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<br>*OR*<br>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 |

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 it appears to slide or smear during a rapid horizontal head movement during driving). Oscillopsia is evident when the person's head is moved abruptly (high accelerations) either passively or actively—such as while running, while looking from side-to-side to cross 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 determinants of 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.[74,75] Certainly there is neural and behavioral evidence from animal studies that underpins this distinction between the initiation and maintenance of vestibular compensation.[76] 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 of these patients with poorly compensated vestibular loss may have had inadequate vestibular function or even central (i.e., cerebellar) deficits before the uVD procedure. Thus, the uVD procedure is potentially dangerous. That is why careful testing is needed to ensure an adequate level of function of the remaining labyrinth and an absence of central deficits before the procedure is undertaken. Patients may suffer postural disequilibrium and gait ataxia for virtually the rest of their lives if the uVD procedure is carried out inappropriately. Rehabilitation procedures (see later) are of great potential value to patients with poorly compensated vestibular loss, 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 common for both ears to be affected by the disease, but one ear more than the other. If a uVD procedure is carried out on the more affected ear, the patient is relying on a partly dysfunctional ear to provide full vestibular input.[77] It is not too surprising that in such cases the eventual result is poor compensation or chronic vestibular insufficiency.

A simple principle for a uVD procedure is that the sole remaining labyrinth will provide all the vestibular input for oculomotor, postural, and perceptual control for the rest of the patient's life. Is the level of function in that remaining, supposedly healthy, labyrinth adequate to carry that burden? How can the level of functioning in the supposedly healthy labyrinth be established? Horizontal canal function can be assessed clinically with the caloric test and the head impulse test. Another important test of a different vestibular sensory region is the vestibular evoked myogenic potential (VEMP) to air-conducted sound delivered by headphone to the ear under test.[78,79] The VEMP is a short-latency inhibitory potential detected in the ipsilateral sternocleidomastoid (SCM) muscle about 12 msec after the onset of the auditory click or brief tone burst (see Chapter 9).

## Decompensation

An apparently related phenomenon is decompensation. *Decompensatio*n refers to situations in which vestibular compensation is nullified and the patient experiences a partial or complete relapse. The behavioral evidence of compensation disappears, 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,[80,81] but even changing the vestibular environment may also do so: Reber and associates[82] reported that when rats with well-compensated vestibular loss were exposed to a brief interval of microgravity during parabolic flight, they experienced decompensation, showing spontaneous nystagmus and other symptoms upon removal of 1 g. That decompensation disappeared within seconds of a return to 1 g. This finding indicates that changing the otolithic stimulation of the intact ear was sufficient to elicit this brief decompensation. This study and others[17,83] suggest that otolithic input from the remaining ear plays an important role in compensation. One assumption that also must be considered is that central neural function, especially cerebellar function, is normal. The patient may be put at risk of poor compensation or chronic vestibular insufficiency if that assumption is incorrect.

## Psychological Factors

Of course, psychological factors play a major role in compensation,[84] but it is difficult to determine their precise role. Are psychological factors causal in determining the outcome of uVD, or are they caused by the procedure

itself? Bowman,[85] measuring the personality characteristics of patients with well-compensated and poorly compensated vestibular loss, found that those with poor compensation showed higher scores on tests of somatic awareness (attention to bodily sensations) than control subjects. It is not clear, however, 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. That people with a predisposition to hypochondriasis would find the symptoms after uVD distressing is not surprising. Patients who have undergone a uVD procedure for treatment of a life-threatening neuroma may be expected to respond rather differently from patients who have chosen to have such a procedure to alleviate a much less threatening condition.

### *Medication*

At the start of this chapter, we discussed the variety of influences on transmission of neural information in the vestibular nucleus. Vestibular neurons not only use a variety of different neurotransmitters themselves but also receive input from many other brain regions that can modulate the activity of these neurons as well as the transmission and processing of vestibular information. Because of the multiple-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 5, 9, and 86 through 88.

## Evidence from Animal Studies

Despite the similar time course of vestibular compensation in humans and animals, the data from animal studies of vestibular compensation does not transfer directly to human studies. The reason is that 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 procedure. In most patients, who tend to be middle-aged, 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.

Referring again to the simple schematic diagram of information transmission in the vestibular system (see Fig. 5.1), one can understand 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[89] and the static component of roll head tilt,[90,91] but seems to have little effect on the reduction of spontaneous nystagmus after uVD.[92] Visual inputs do augment the diminished motor responses to linear acceleration[93] and the deficient righting reflexes[94] that occur after uVD. Visual motion deprivation delays recovery of locomotor equilibrium.[95–98]

Similarly, stimulation of proprioceptive receptors appears to facilitate the recovery of dynamic postural equilibrium,[99] 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.[100] Somatosensory proprioceptive deprivation appears to retard static compensation,[93] whereas somatosensory proprioceptive stimulation appears to facilitate the restoration of dynamic postural equilibrium.[99] Acute spinal lesions can produce a temporary decompensation of static postural symptoms.[80,81]

There are only scant data on the effects of vestibular stimulation or deprivation on static or dynamic compensation. In frogs, otolithic stimulation hastens but otolithic deprivation delays static compensation of roll-tilt of the head.[101] In cats, low-frequency combined visual-vestibular stimulation helps reverse the deficit in VOR gain that occurs in response to low-frequency stimulation after uVD[102] but has no effect on the asymmetry of the VOR at high frequencies.

One idea prevalent in this literature is that the relative importance or salience of other non-vestibular sensory inputs may change during the course of vestibular compensation.[103] Support for this idea comes from the results of neck vibration. Vibration of neck muscles usually causes no detectable nystagmus or changes in postural perception in normal, healthy subjects. However, after uVD, the same vibration causes horizontal nystagmus and ocular torsion and changes in the visual perception of a horizontal line even in well compensated patients.[104] Interestingly, this result occurs for vibration on either the ipsilesional or contralesional neck muscles[105]: the nystagmus has quick phases directed away from the affected side, and such vibration has been proposed as a means of supplementing vestibular diagnostic testing. This nystagmus may be an example of a stimulus producing a temporary decompensation.

### Plasticity of the Vestibulo-Ocular Reflex

There is a long history of studies of vestibulo-ocular responses showing that the gain of the VOR is modifiable in normal, healthy subjects. Gonshor and Melvill Jones[106,107] required subjects to wear reversing spectacles during the course of their usual daily activities. In order 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 (references 55 and 108 list many papers on this topic). It appeared that patients with uVD may use mechanisms responsible for this VOR plasticity to assist their recovery.

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. But closer consideration shows that such evidence is not really relevant for the situation confronting the patient with uVD who is undergoing vestibular compensation. Some of the procedures that have been developed for the study of VOR plasticity do help patients improve their normal everyday functioning. We suggest, however, that these improvements are not due to 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 VOR is very modest at natural head angular accelerations. We think that for the patient with uVD, 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. Of course, this achieves the sought-after goal of returning the patient to normal lifestyle, but for us, it is important to identify the mechanism by which it occurs.

Patients with uVD differ in many important ways from 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, possibly disrupting the neural substrate needed for vestibular modification.

### Rehabilitation

Many years ago, Cawthorne[109] and Cooksey[110] suggested a number of exercises to assist in the rehabilitation of patients with vestibular disorders. Those exercises are similar to exercises in use today. Doubts about the efficacy of such exercises have now been largely dispelled.[111–114] 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 the patient has active control of the response, and we suggest that the Cawthorne-Cooksey exercises and others are acting to teach patients how to substitute these other responses to conceal and thus overcome the sensory deficit. The results of others[115] agree with our own.[43] 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 the inadequate VOR during that ipsilesional head rotation.[43]

We suggest that the process of vestibular rehabilitation should be thought of as an opportunity for other nonvestibular sensory inputs and cognitive-behavioral strategies to assume larger roles in controlling the patient's equilibrium.

## Neural Evidence Concerning Recovery after Unilateral Vestibular Deafferentation

The neural evidence about recovery after uVD is a huge area; in this chapter we refer only to the major features.

For reviews of neural processing of vestibular sensory input, see references 4 and 116 through 119.

One simple principle helps us understand the neural basis of vestibular responses and vestibular compensation: 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 has the head still, the neural activity in the pair of vestibular nuclei (one on each side of the brainstem) is approximately equal. A horizontal (yaw) angular acceleration in one direction (e.g., to the person's left) 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, whereas many neurons in the right vestibular nucleus concurrently decrease their rate of firing, and 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, nystagmus occurs.

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 the self 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 in guinea pigs 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, the behavioral and perceptual symptoms decline.[26,27]

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 process happens in the space of about 1 day, and by 52 hours after uVD, the average activities on the two sides are about equal.[26,27] The spontaneous nystagmus disappears, although the dynamic deficits remain. As the balanced activity returns, the behavioral symptoms disappear. Concurrently, other sources of control of neural transmission through the vestibular nuclei (e.g., cerebellar input) are changing. The following discussion describes this process in more detail (Fig. 5.7), summarizing material from references 26, 27, and 120 through 126.

Within the medial vestibular nucleus are two types of neurons responding to horizontal (yaw) rotation. Both types discharge spontaneously when the head is stationary. During yaw angular head accelerations in one direction, type I neurons in the vestibular nucleus on the side to which the head turns increase their firing; these neurons decrease their firing for yaw head accelerations in the contralateral direction. These neurons receive direct input from the ipsilateral peripheral vestibular labyrinth. Other neurons within that vestibular nucleus (type II neurons) show precisely the opposite pattern of response; they increase their firing for contralaterally directed head accelerations and decrease it for ipsilateral head accelerations. The reason these type II neurons respond in such a mirror-image fashion is that 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*. Type II neurons are inhibitory 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. These synergistic mechanisms mean that there is enhanced sensitivity of type I neurons during acceleration compared with peripheral vestibular neurons. This arrangement is called a *push-pull system*, and it functions to generate a fast, sensitive response.

Immediately after uVD, changes occur 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 that of ipsilesional type II neurons is substantially increased.[26] 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.

As 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, which 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 receive progressively more inhibition, further enhancing the restoration of balanced neural activity. Therefore, 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.

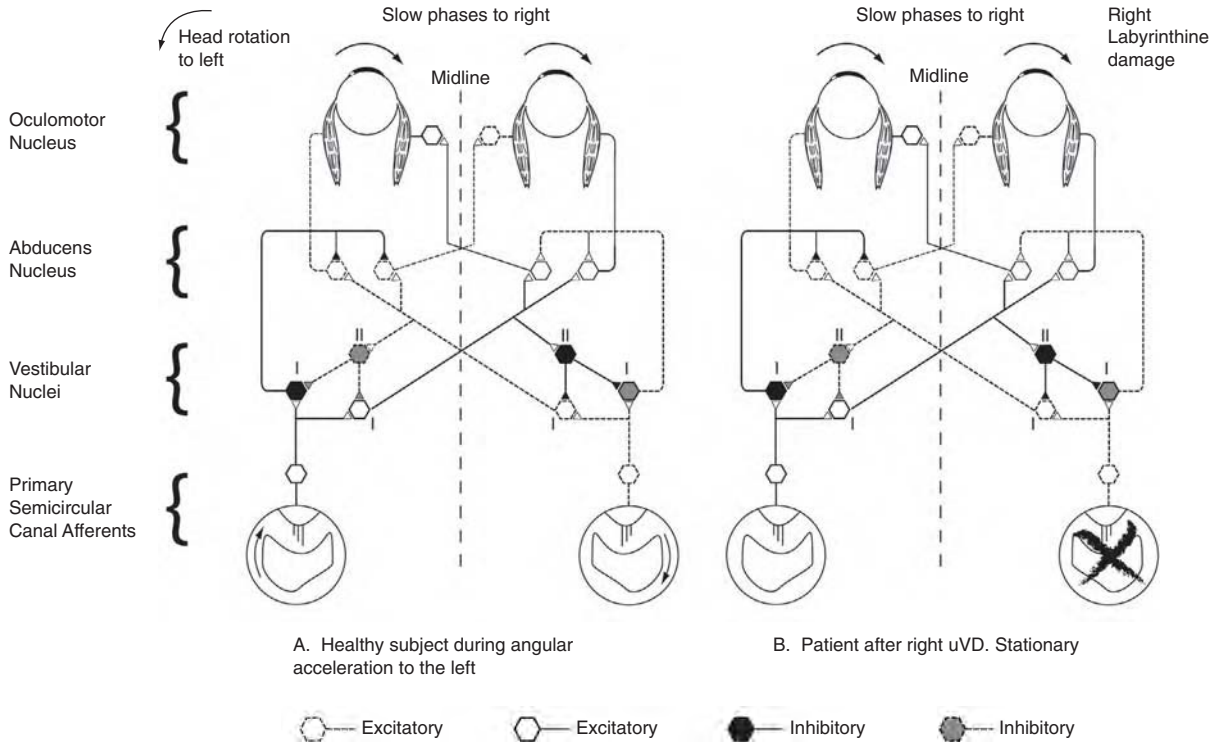


**Figure 5.7** To show the neural basis of the nystagmus after unilateral vestibular deafferentation (uVD) and how a patient with uVD at rest (*B*) has vestibular activity closely similar to that of a person undergoing a rotation in the direction of the intact ear (*A*). The *left panel* shows the responses in some of the identified neural connections of the vestibulo-ocular pathways during a leftward yaw head acceleration. *Open hexagons* = excitatory neurons; *solid hexagons* = inhibitory neurons; *solid lines* = activated neurons; *dashed lines* = disfacilitated neurons. . The sequence occurs as follows: during ipsilateral acceleration the primary neurons from the left semicircular canal are activated while those from the right are inhibited. 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.

In the hours, days, and weeks that follow, the activity of vestibular nucleus neurons changes remarkably. 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 due merely to a neural process such as adaptation, or is there an "error signal" that triggers them? Does the removal of peripheral vestibular input trigger synaptic or membrane changes that act to restore the balance between the two nuclei? There is evidence for changes both at the synaptic level and the cell membrane level during compensation.[127–129]

How could the balance be restored? Adaptation is one possibility. 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 gamma-aminobutyric acid (GABA).[128,130–136] A reduction in the efficacy of the neurotransmitter receptor for GABA would allow for greater activity of 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, referred to as *downregulation of GABA sensitivity*, may be partly 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.[137,138] After uVD, the response of ipsilesional type I neurons shows a decreased sensitivity, as would be expected from the preceding analysis.[121] Over time, there are significant anatomical changes in the vestibular nuclei,[139–142] although it seems that the behavioral effects of compensation occur before sprouting.[143]

Evidence from Campos-Torres and colleagues[144] indicates that the effects of silencing peripheral neurons by chemical means (*tetrodotoxin*) and those of silencing by surgical sectioning of the nerve are not identical. Tetrodotoxin does not cause degeneration, and Campos-Torres and colleagues [144] found that it causes no glial changes in the vestibular nuclei, as opposed to the dramatic increase in glia after sectioning or damage to the vestibular nerve. Results such as these suggest that the "error signal" may well be a chemical factor released by the injured neurons.

Studies have also recorded the characteristics of neurons in slices of brainstem tissue taken from animals with compensation at 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, and type B neurons tend to be phasic. Beraneck and associates[145,146] have reported that after uVD, the membrane characteristics of type B neurons appear to change. These previously phasic neurons appear to take on much more tonic characteristics. Other studies have reported very long-lasting changes in intrinsic membrane characteristics after prolonged inhibition.[147,148] It is likely that membrane changes are responsible in part for the changes in global neural activity after uVD previously described.

## 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 during ipsilateral roll-tilts show that most neurons show an increase in firing for such stimuli.[29] After a uVD, there is a decrease in the proportion of ipsilesional LVN neurons and a decrease in their average resting activity.[149,150] So a similar imbalance occurs to natural linear accelerations as described previously for natural angular accelerations. That imbalance is mimicked by a removal of all the peripheral vestibular neural input from one side.

## Cerebellum

Finally, the cerebellum may play an important role in vestibular compensation that has not been fully recognized because so much focus has been put 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,[151] others show no effect.[152] 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.[92] Lesions of the brainstem[153] or transcerebellar vestibular commissures[154] do not impede the vestibulospinal symptoms of static compensation, at least not in mammals. This finding suggests that input from the contralesional (intact) vestibular nucleus is not essential for static compensation. Kitahara and associates[155,156] have shown that neurons in the flocculus of the cerebellum play an important role in the early stages of vestibular compensation for static symptoms. 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, a reduction that would in turn act to relieve inhibition on the ipsilesional type I neurons.[157–159] Human patients with cerebellar lesions show slow compensation.[160]

## 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.[161–164] The physiologically realistic neural network model developed by Cartwright and associates,[162,163,165] which was trained on guinea pig eye movement responses before and after uVD, showed that changes in activity in various stages through the neural network determine the responses. The model pointed to neurons on the intact side as being especially important because their gain changes most.

## Summary

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 due to 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 due to the decreased dynamic sensitivity of vestibular nucleus neurons as documented by Markham and colleagues.[121]

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.

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CHAPTER 6

# Vestibular System Disorders

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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. Bilateral involvement can be found in 10% of the spontaneous cases and 20% of the traumatic cases. Spontaneous remissions are common, but recurrences can occur, and the condition may trouble the patient intermittently for years.

Evaluation should include a careful otoneurologic examination, the most important part being the history. A key diagnostic maneuver is the Dix-Hallpike positioning test[1] while the examiner observes the patient's eyes with a pair of Frenzel lenses or in combination with electronystagmography (ENG) monitoring. 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.[2] The nystagmus changes with the direction of gaze, becoming more torsional as the patient looks toward the dependent ear and more vertical as the patient looks toward the higher ear.

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 decreases 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, and (3) the nystagmus may occur as downbeat nystagmus only in the head-hanging position. BPPV must be differentiated from positional nystagmus in Ménière's disease, perilymphatic fistulas, and alcohol intoxication.

A few patients do not display the typical torsional upbeat nystagmus but, for example, show a strong horizontal nystagmus, which nevertheless follows a similar pattern of buildup and decline but often over a longer period. This horizontal nystagmus may indicate a lateral canal variant of BPPV.

The classic explanation of the underlying pathophysiology (cupulolithiasis) was first described by Schuknecht[3] in 1969. His study of the temporal bones of two patients afflicted with this disorder showed deposition of otoconial material in 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 is typically seen.

Brandt and Steddin[4] emphasized a second theory, canalithiasis, which better explains the typical features of BPPV. It suggests that the debris of a higher density than the endolymph is free-floating in the long arm of 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, further investigation, such as magnetic resonance imaging (MRI), should be made to assess for unusual causes of positional vertigo, such as acoustic neuroma and tumors of the fourth ventricle.

BPPV is usually a self-limiting disorder and commonly resolves spontaneously within 6 to 12 months. Simple vestibular exercises or maneuvers aimed at dispersing the otolithic debris from the cupula can speed recovery; antivertiginous drugs are not helpful. One approach is to instruct the patient to assume repeatedly the positions that bring on the symptoms.[5] In 1988, Semont and colleagues[6] introduced a single liberatory maneuver, and Epley,[7] in 1992, proposed a variation, later modified by Herdman and associates[8] (see also Chapter 17). With these newer liberatory maneuvers, most patients can be treated successfully. The period of recovery varies from immediate after one positioning maneuver (physical displacement) to usually 6 weeks to 6 months. Only in a few patients, usually the more elderly, do the symptoms persist 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 cause is

never proved, evidence to support a viral etiology (similar to that for Bell's palsy or sudden hearing loss) comes from histopathologic changes of branches of the vestibular nerve in patients who have suffered such an illness[9] and the sometimes epidemic occurrence of the condition. Onset is often preceded by the presence of a viral infection of the upper respiratory or gastrointestinal tract. 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, and nausea. Hearing loss is not usually present, but when it is, mumps, measles, and infectious mononucleosis, among other infections, have been implicated. The presence of hearing loss together with acute onset rotational vertigo should alert the physician to consider other diagnoses (e.g., ischemia of labyrinth artery, Ménière's disease, acoustic neuroma, herpes zoster, Lyme disease, neurosyphilis). The condition mainly affects those aged between 30 and 60 years, with a peak for women in the fourth decade and for 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 of an acute unilateral vestibulopathy and a benign paroxysmal positioning vertigo simultaneously in the same ear of an affected patient.[10] 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), which left part of the saccule and the posterior semicircular canal afferents intact.[11] 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, which is 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 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.

Initial treatment is accomplished with the use of vestibular suppressants, such as the antihistamine dimenhydrinate or the anticholinergic scopolamine. In addition, bedrest is very helpful early in the course of the disease. After the most severe vertigo and nausea have passed (after 24 to 72 hours), the patient may resume ambulation 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 20.)[12]

Animal experiments have shown that alcohol, phenobarbital, chlorpromazine, diazepam, and adrenocorticotropic hormone (ACTH) antagonists retard compensation; caffeine, amphetamines, and ACTH accelerate compensation.[13] In 141 patients with vestibular neuritis, Strupp and coworkers[14] performed a prospective randomized trial of methylprednisolone, valacyclovir, and the two in combination within 3 days after the onset of symptoms. The researchers 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.

## 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) persists anywhere from approximately 30 minutes to 24 hours. Gradually, the severe symptoms abate, and the patient is generally ambulatory within 72 hours. Some sensation of postural unsteadiness persists for days or weeks, and then normal balance returns. During this recuperation time, hearing gradually returns. Hearing may return to the pre-attack 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 also usually diminishes as hearing returns. As the disease progresses, hearing fails to return after the attacks, 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. In vestibular Ménière's disease, only vestibular symptoms and aural pressure are present, and in cochlear Ménière's disease, only cochlear symptoms and aural pressure are encountered.[15]

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 with classic Ménière's disease.[16] About 15% of the patients have blood relatives with the same disease, suggesting genetic factors. The incidence of bilaterality of involvement ranges between 33% and 50%.[17]

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 is a pathologic 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 of 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 routinely results in endolymphatic hydrops in many animal models.[18] 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 of 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 cause tamponade of the cochlear duct, contributing to fluctuating progressive sensorineural hearing loss and other cochlear symptoms, whereas distention 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 for Ménière's diesase include an 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.

A brainstem-evoked acoustic response (BEAR) procedure must be performed in patients with findings of retrocochlear disease on routine audiometry, to screen for cochlear nerve or brainstem pathology. If the BEAR is found to be positive, MRI with the use of intravenous gadolinium should be obtained to assess for central nervous system pathology or VIIIth 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 and avoidance of exposure to low temperatures. Stellate ganglion blocks, diuretics, vasoactive agents, tranquilizers, neuroleptics, and lithium have been employed under the mistaken assumption that it is possible to diminish endolymphatic hydrops 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.[19] 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 should be used primarily during the acute episodes of vertigo and should be discouraged as long-term daily medications.

In addition to pharmacologic therapies, many patients with Ménière's disease require psychological support to help them cope with the frustrations and changes brought about by their medical condition. Those patients in whom the vertigo becomes disabling by virtue of greater 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.[20]

Sacculotomy has been proposed by a variety of authorities 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 performed with 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 selectively damage the secretory epithelium (and thereby improve endolymphatic hydrops) before significantly affecting vestibular and cochlear function.[21]

The current treatment that is 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 investigators. 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 they have 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 experience a loss of vestibular function at the end stages of the disease, and for these patients, vestibular rehabilitation may be appropriate. Vestibular exercises are also beneficial in patients with surgical destruction of the inner ear.

## Perilymphatic Fistula

Perilymphatic fistula may lead to episodic vertigo and sensorineural hearing loss because of the pathologic 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 due to perilymphatic fistula, in most cases from superior canal dehiscence, but subluxation of the stapes foot plate and other ear disease may be responsible. The symptoms often sub-

side while the patient is at rest, only to resume with activity. Sneezing, straining, nose blowing, and other such maneuvers can elicit the symptoms after the initial event.

Perilymphatic fistulas probably account for a considerable proportion of patients presenting with vertigo of unknown origin. Diagnosing perilymphatic 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.[22]

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 the application of manual pressure over the tragus or to the tympanic membrane with the pneumatic otoscope; a positive response 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 may 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 patients with a suggestive history and symptoms, treatment is indicated. Often the diagnosis is made definitively only at the time of surgical exploration by tympanoscopy as the patient performs Valsalva maneuvers.

Medical treatment consists of absolute bedrest 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 are important for reduction of further explosive and implosive forces that may activate perilymph leakage.[23]

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 perilymphatic fistulas are found at surgery. Surgical management consists of middle ear exploration and packing of the oval and round window areas with fat, absorbable gelatin sponge (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%, likely reflecting some element of variable patient selection.

A variant of perilymphatic fistula causing episodic vertigo has been described by Minor and coworkers.[24] The disease is due to dehiscence of the superior semicircular canal. It is probably the most common form of a fistula and probably the most often overlooked. Hallmarks are vertigo spells induced by pressure increase such as happens with 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. In more than half of the cases, symptoms start after even mild head trauma or barotrauma. The diagnosis can be made with high-resolution computed tomography (CT) of the temporal bone, which shows a dehiscence of the apical part of the superior semicircular canal.

## Vestibular Paroxysmia (Disabling Positional Vertigo)

Neurovascular cross-compression of the root entry zone of the vestibular nerve can elicit disabling positional vertigo.[25] The term describes a heterogeneous collection of signs and symptoms rather than a reliable diagnosable disease entity. Brandt and Dieterich[26] 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 of the 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).

Neurovascular cross-compression can cause local demyelinization of the root entry zone of the VIIIth nerve. Ephaptic transmission between bare axons and central hyperactivity initiated and maintained by the peripheral compression are the suggested mechanisms. As in 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. Autoimmune conditions affecting the inner ear are rare but distinct clinical entities,[27] 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 are 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 via 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 performed 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.[28] Other agents that may produce vertigo are 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 therapy with such agents.

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.[29]

Alport's syndrome (inherited sensorineural deafness associated with interstitial nephritis), Usher's syndrome (inherited sensorineural deafness associated with retinitis pigmentosa), and Waardenburg's syndrome (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), and anoxia.

Controlled physical exercises can improve the condition in patients with permanent bilateral vestibulopathy by recruiting non-vestibular sensory capacities such as the cervico-ocular reflex and proprioceptive and visual control of stance and gait (see Chapter 21).

## Summary

This chapter describes the clinical presentations of the more common peripheral vestibular disorders and their differential diagnosis from 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. Therefore, a thorough evaluation should always include, in addition to the specific otoneurological investigation, a detailed history and a general physical examination. For a quick review, Table 6–1 summarizes the hallmarks of the peripheral vestibular disorders treated in this chapter.

## ■ Table 6-1 SUMMARY OF VESTIBULAR SYSTEM DISORDERS

| | Benign Paroxysmal Positional Vertigo | 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 | 30 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 phenomenon, Hennebert's sign | Frequent attacks, tinnitus, hypacusis | Gait ataxia |
| Precipitating action | Positioning, turning in bed | – | – | Head trauma, ear surgery, sneezing, straining, nose blowing | Changing of head position | – |

– = absent, + = present; –/(+) = variable; ++ = very strong.

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# T·W·O

# Medical Assessment and Vestibular Function Tests

CHAPTER 7

# History and Clinical Examination

*Ronald J. Tusa, MD, PhD*

Management of the dizzy patient depends on history, bedside clinical examination, and laboratory testing. The first two portions of this evaluation are covered in this chapter. History is key to determining 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 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 8) 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 start can be extremely tedious, because the patient's complaints are often vague and are frequently filled with anxiety-provoked symptoms. The history can be divided 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 complaint are the three key items in the history (Table 7-1).

#### 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 dizziness. 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 upon. *Dizziness* is an imprecise term used to describe a variety of symptoms, each of which has a different pathophysiological mechanism and significance (Table 7-2). If the patient cannot describe the symptoms, the clinician should ask whether the dizziness causes problems primarily in the head or with balance. If the patient has

## ■ Table 7-1 KEY ITEMS IN THE HISTORY OF THE DIZZY PATIENT

| Disorder | Tempo | Symptoms | Circumstances |
|---|---|---|---|
| Vestibular neuritis | Acute dizziness | Vertigo, dysequilibrium, nausea and vomiting, oscillopsia | Spontaneous, exacerbated by head movements |
| Labyrinthitis | Acute dizziness | Vertigo, dysequilibrium, nausea and vomiting, oscillopsia, hearing loss and tinnitus | Spontaneous, exacerbated by head movements |
| Wallenberg's infarct | Acute dizziness | Vertigo, dysequilibrium, 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 | Dizzy, dysequilibrium, 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<br>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 7-2 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<br>Anxiety |
| Motion sickness | Visuovestibular mismatch |
| Nausea and vomiting | Stimulation of medulla |
| Oscillopsia: illusion of visual motion | Spontaneous: acquired nystagmus<br>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 |

spells, have him or her describe in detail the initial spell and the last severe spell.

### *Disequilibrium*

*Disequilibrium* is an 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.

### *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 is called *mal de débarquement*. Most likely it is due to a vestibular adaptive process to the continuous, passive motion during travel. Once individuals are back in a stable environment, this symptom persists until readaptation occurs. Most patients respond to reassurance. Some individuals continue to have this symptom for weeks to years after a voyage, and others complain of the same symptom arising spontaneously.

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] It may be due to a problem in the otoliths, which is not immediately accessible to clinical testing. It may also be due to or exacerbated by anxiety or a somatoform disorder. 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 Chapter 13).

### *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 due to a sensory mismatch between visual and vestibular cues.[3] 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.[4,5] The cause for this relationship is not clear. Symptoms of increased motion sensitivity are often reproduced when the patient is exposed to moving full-field visual target.

### *Nausea and Vomiting*

Nausea and/or vomiting is due to stimulation of the solitary and vagus centers in the medulla. In peripheral vestibular lesions, these symptoms are usually mild 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 for central lesions. 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.[6] 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. Spontaneous oscillopsia is caused by acquired nystagmus and is due to apparent motion of the visual scene from movement of the retina (*retinal slip*). Patients with congenital nystagmus usually do not report oscillopsia because of feedback of the involuntary eye movement to the central nervous system (*efference copy*).

Head-induced oscillopsia occurs in patients with severe, bilateral loss of the vestibular-ocular reflex (VOR), which is frequently experienced after ototoxicity due to 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.

### *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 due to a skew eye deviation from peripheral or central otolith dysfunction.

### *Vertigo*

*Vertigo* is the illusion of movement of the self or the environment due to sudden imbalance of tonic neural activity in the vestibulocortical pathway (labyrinth–VIIIth nerve–vestibular nucleus–vestibular thalamus–vestibular cortex). It is due either to normal head movements (physiologic) or to 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 due to horizontal semicircular canal (SCC) dysfunction, which commonly occurs from labyrinthine (e.g., labyrinthitis or Ménière's) or VIIIth nerve dysfunction (vestibular neuritis). Rotational vertigo in the torsional plane (clockwise or counterclockwise direction) is due to anterior and posterior SCC dysfunction on one side from a central lesion in the dorsal medulla. Tilt— lateral translation or lateropulsion—is due to utricle dysfunction, which can be caused by lesions in the labyrinth or VIIIth 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 of the head cause dizziness and there is no eye movement disorder (such as ocular misalignment or an internuclear ophthalmoparesis), the symptom is not likely to be due to a vestibular or neurological problem. When dizziness occurs without provocation (spontaneous) and it is vestibular in origin, it is commonly exacerbated by head movements.

## 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 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 cause dizziness, some of which are used to treat dizziness (see Chapter 11). 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 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*

## Sensations

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 the Appendix). The second is to use a *Visual Analogue Scale* (VAS) to quantify the intensity of specific symptoms. An example would be the head movement VAS shown in Figure 7.1. 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 (Fig. 7.1), 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.

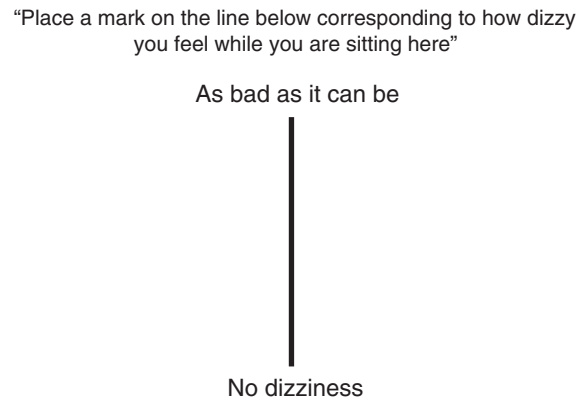


**Figure 7.1** Head movement Visual Analogue Scale (VAS). The patient is instructed to place a mark on the line corresponding to how dizzy he or she feels while sitting and then while performing a task. For this scale, test-retest reliability is $r = 0.59$, based on a separate sample of patients with 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, page 483).

## Perceived Disability

The perceived disability caused by a patient's dizziness can be determined from the Disability Scale, shown in Box 7-1.[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 7-1

**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, i.e., that patient's condition is unlikely to change with rehabilitation. 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 correlation ($r=-.64$) 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); anxiety or depression, 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 7-3 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 7-4).

### *Spontaneous Nystagmus*

#### Peripheral Vestibular Disorders

Selective lesions in the peripheral vestibular pathways result in spontaneous nystagmus due to the unopposed higher spontaneous neural activity in the intact vestibular pathways (Fig. 7.2). 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 peripheral vestibular loss resolve spontaneously in 1 to 2 weeks, without any intervention. Simultaneous bilateral vestibular loss does not cause spontaneous nystagmus because there is no asymmetry between the two sides.

Three features of spontaneous nystagmus can be used to separate peripheral (inner ear or VIIIth cranial nerve) vestibular orders from central vestibular disorders (Table 7-5). First, nystagmus that is caused by peripheral disturbances can be decreased with fixation, but nystag-

## ■ Table 7-3 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 only during maneuvers | Usually, inner ear debris from benign paroxysmal positional vertigo<br>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 | Any listed in table |

mus resulting from brainstem and cerebellum 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

## ■ Table 7-4 WHEN TO USE FRENZEL LENSES OR VIDEO INFRARED 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 thrust 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 |

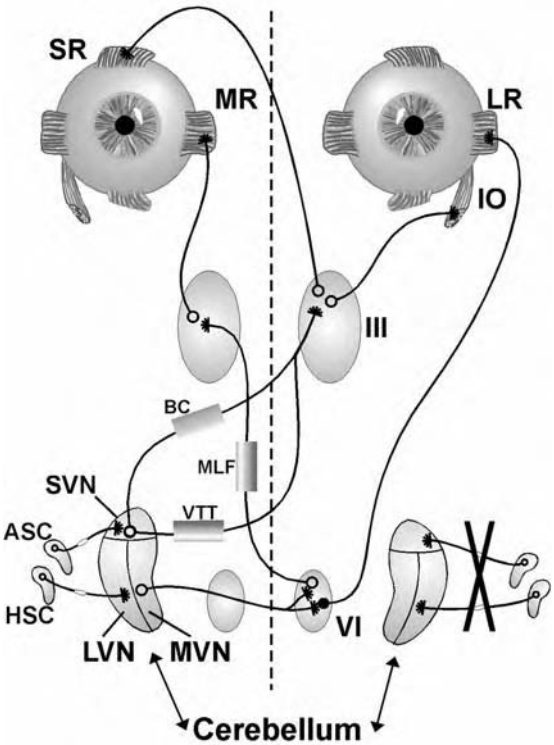


**Figure 7.2** Peripheral vestibular loss on the left side from vestibular neuritis. This disorder disrupts the portion of the vestibular nerve from the horizontal semicircular canal (HSC) and anterior semicircular canal (ASC) (X). The ASC and HSC project to the superior vestibular nucleus (SVN) and 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 vial 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 7-5 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 |

in Table 7-6. 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. 7.3). 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).

## ■ Table 7-6 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 |

### Central Vestibular Disorders

Figure 7.4 illustrates the structures and pathways that mediate central vestibular nystagmus, and Table 7-7 describes the neural mechanisms.

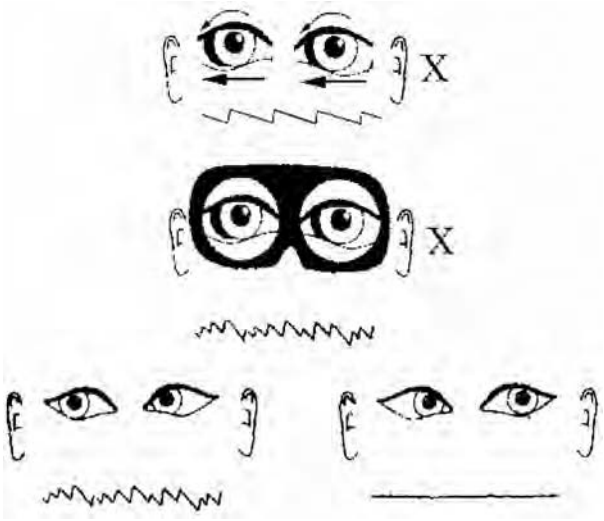


**Figure 7.3** Peripheral vestibular nystagmus. This figure depicts disruption of the left superior division of the VIIIth nerve from vestibular neuritis (X). *Top*, Mild right-beating and right torsional nystagmus when fixation is present. *Middle,* Vigorous nystagmus when fixation is suppressed in 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])

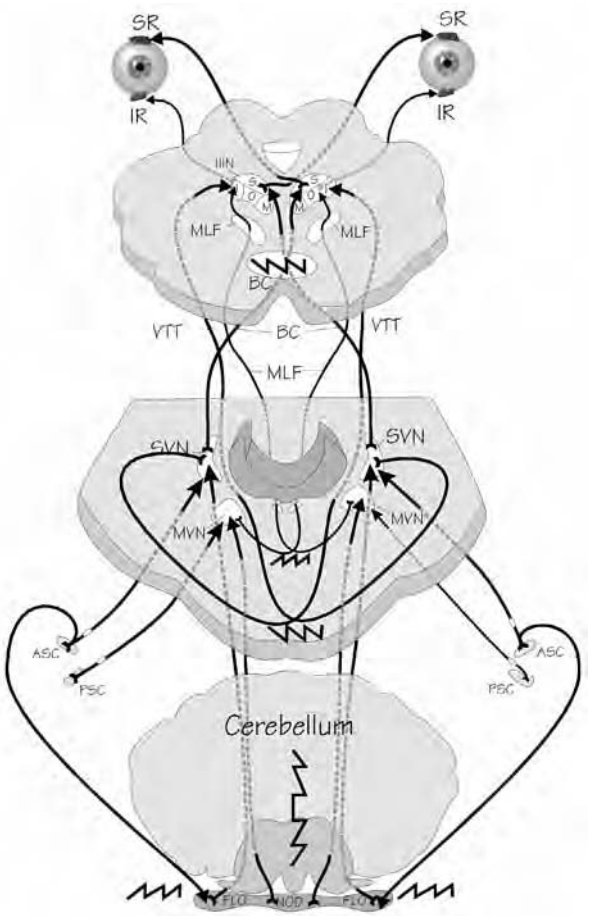


**Figure 7.4** Central vestibular pathways and lesion location for central vestibular nystagmus. Downbeat nystagmus occurs whenever the tone within the central pathways from the anterior semicircular canals (ASCs) is relatively higher than the tone within the posterior semicircular canals (PSCs). This can occur from lesions of the cerebellar flocculus (FLO) on both sides due to disinhibition of the superior vestibular nucleus (SVN). It can also occur from bilateral lesions of the medial longitudinal fasciculus (MLF), which caries input from the PSC to the IIIrd nerve (IIIN) nuclei. The IIIN nuclei 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 IIIrd 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 1–2 minutes and then nystagmus to the left for 1–2 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])

## Skew Eye Deviation

*Skew eye deviation* is a vertical misalignment of the eyes due to 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 7.5.[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 VIIIth nerve section or vestibular neuritis causes a pathologic ocular tilt response because of the unopposed excitation of the intact utricle.[16]

Figure 7.6 depicts the findings in a left-sided lesion (leftward pathologic 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 pathologic OTR can be due to 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 is a *static imbalance* due to a difference in the tonic discharge rate between the vestibular nuclei on the two sides of the brainstem. The tonic resting firing rates of most or all of primary afferents on the lesioned side are lost.[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 owing to 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

## ■ Table 7-7 VESTIBULAR NYSTAGMUS DUE TO 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 of the visual input into the MVN) |

INC = interstitial nucleus of Cajal; MVN = medial vestibular nucleus; NOT = nucleus optic tract; SCC = semicircular canal.

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 7-8). 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-thrust tests are strongly positive in patients with bilateral vestibular loss, but the head-shaking nystagmus test result is not positive in this group.

### 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 thrusts, observing the eyes during the thrusts. After a head thrust, the observation of a refixation saccade indicates decreased VOR.[18] It is important that the patients be tested while they are wearing their usual glasses, because the VOR is calibrated for visual inputs through those glasses. Table 7-9 shows the sensitivity and specificity of the head-thrust test with respect to the caloric test for UVL. For complete UVL due to nerve section, the sensitivity and specificity are 100%; for a variety of types of UVL, the overall sensitivity is 36%, but the specificity is 97%. 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 thrust unpredictable.[19] Table 7-10 shows how the head thrust test results vary with the severity of UVL.[23] For example, for moderate paresis of the caloric test (50–75% weakness), 19 patients had a negative head thrust 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 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.[24] This sign may persist indefinitely after a peripheral or central unilateral vestibular lesion. Table 7-11 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 7-12

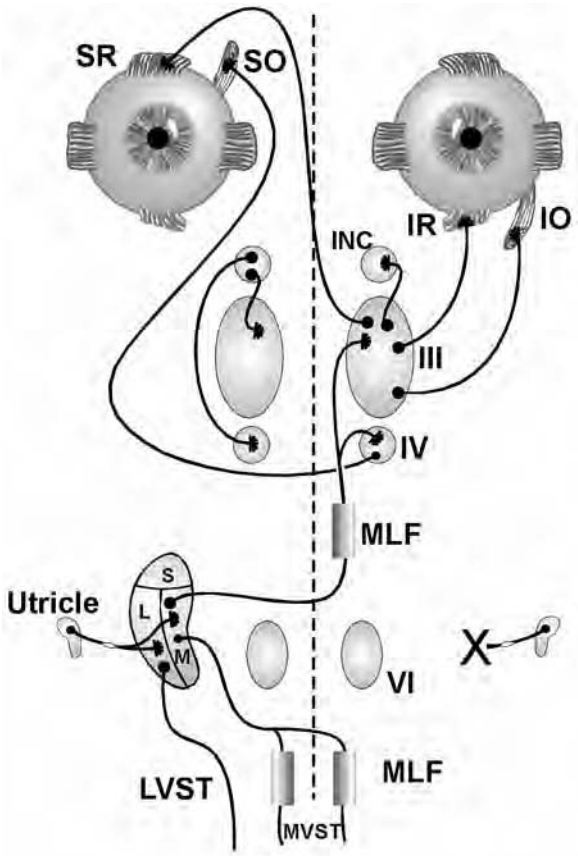


**Figure 7.5** Otolith pathway from the left utricle. This figure depicts disruption of the left utricular division of the VIIIth nerve from vestibular neuritis. The utricle projects to the lateral (L) and medial (M) divisions of the vestibular nucleus. These portions of the vestibular nucleus project 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])

shows how the sensitivity and specificity vary with the severity of UVL.[28]

## 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


**Figure 7.6** Pathologic 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.

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 their 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. 7.7). After vestibular adaptation exercises, dynamic visual acuity improves, possibly because of the development of preprogrammed or anticipatory eye movements.[29,30]

## *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 7-13).

### Position Testing

The *Hallpike-Dix* test result is positive in patients with BPPV (Fig. 7.8). 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 due to a central disorder (central positional vertigo). The only exception to this statement is BPPV due to cupulolithiasis. In this condition, otoconia are attached to the cupula of the SCC, so the Hallpike-Dix test will result in

## ■ Table 7-8 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<br>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 thrust | The patient fixates a distant visual target, and eye position is observed immediately after a small thrust of the head to the left and right | A refixation saccade after the head thrust indicates decreased VOR<br>If the head thrust 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 |

persistent nystagmus and vertigo (see Chapter 17 for further details). Positional nystagmus may also be seen in patients with a variety of other mechanical problems of the inner ear, but the characteristics of the nystagmus are usually not classic for BPPV. 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.

## ■ Table 7-9 HEAD-THRUST TEST COMPARED WITH CALORIC TEST FOR UNILATERAL VESTIBULAR LOSS (UVL)*

| Type or Cause of UVL | Sensitivity (%)* | Specificity (%)* | No. of Patients | Study |
|---|---|---|---|---|
| Complete UVL due to nerve section | 100 | 100 | 20 | Halmagyi & Curthoys (1988)[18] |
| Complete UVL due to nerve section | 100 | 100 | 12 | Foster et al (1994)[25] |
| Various | 39 | 97 | 112 | Harvey & Wood (1996)[26] |
| Various | 35 | 95 | 105 | Harvey et al (1997)[27a] |
| Various | 34<br>*Average:* 36 | 100<br>*Average:* 97 | 150 | Beynon et al (1998)[23] |
| Various | 70 | 81 | 77 | Grine et al (2000)[19] |

*Caloric test used as the standard.

**■ Table 7-10 HEAD-THRUST 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 7-11 HEAD-SHAKING NYSTAGMUS TEST COMPARED WITH CALORIC TEST***

| Sensitivity (%)* | Specificity (%)* | No. of Patients | Study |
|---|---|---|---|
| 40 | 60 | 108 | Wei et al (1989)[27b] |
| 95 | 62 | 85 | Takahashi et al (1990)[28] |
| 27 | 85 | 116 | Jacobson et al (1990)[29] |
| 44 | 65 | 105 | Burgio et al (1991)[30] |
| 42 | 85 | 197 | Goebel and Garcia (1992)[31] |
| 35 | 92 | 105 | Harvey et al (1997)[27a] |
| 38 | 79 | 290 | Asawavichianginda et al (1997)[28] |
| *Average:* 46 | *Average:* 75 | | |

*Caloric test used as the standard.

**■ Table 7-12 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.[28]

## *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

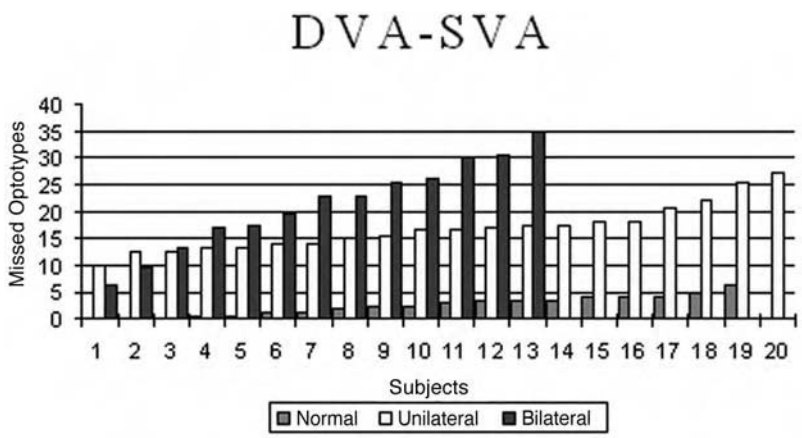


**Figure 7.7** Dynamic visual acuity (DVA) scores in patients with vestibular hypofunction and normal controls. On the ordinate is the number of letters (optotypes) missed on a standardized visual acuity (SVA) chart called the ETDRS chart (used in the Early Treatment Diabetic Retinopathy Study). This chart has 5 letters on each line. On the abscissa are the rankings of individual subjects from the best visual acuity (to the left) to the more 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 4 irrigations) and rotary chair test (gain < 0.1).

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 7.9. In patients with cerebellar degeneration or other bilateral disorders, smooth pursuit and VOR cancellation are impaired in both directions.

■ Table 7-13 **MANEUVER-INDUCED VERTIGO AND EYE MOVEMENTS FROM MECHANICAL PROBLEMS IN THE INNER EAR**

| Positive Result of: | Disorder(s) |
|---|---|
| Position testing | Benign paroxysmal positional vertigo<br>Central positional nystagmus<br>Central positional vertigo<br>Dehiscence of superior semicircular canal<br>Perilymphatic fistula |
| Pressure testing, tragus movement, or Tullio phenomenon | Dehiscence of superior semicircular canal<br>Ménière's disease<br>Perilymphatic fistula |

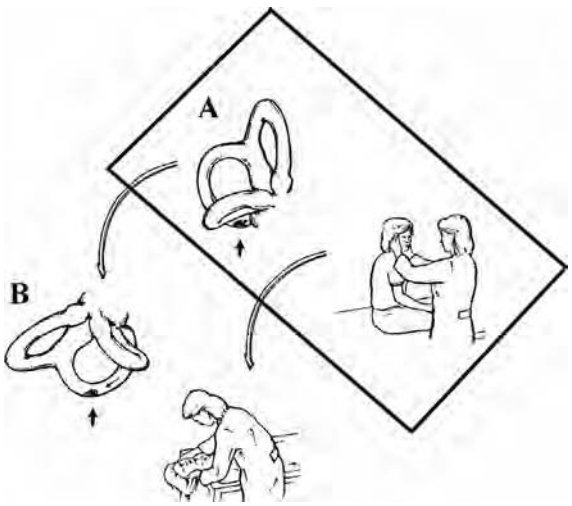


**Figure 7.8** Hallpike-Dix test for benign paroxysmal positional vertigo (BPPV). (*A*) In this test, the patient sits on the examination table and the examiner turns the head 45 degrees horizontally. (*B*) The examiner then quickly brings the head and trunk straight back "en bloc" so that the head is 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 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. (Modified from Tusa and Herdman 1998 [see Chapter 17 for reference].)

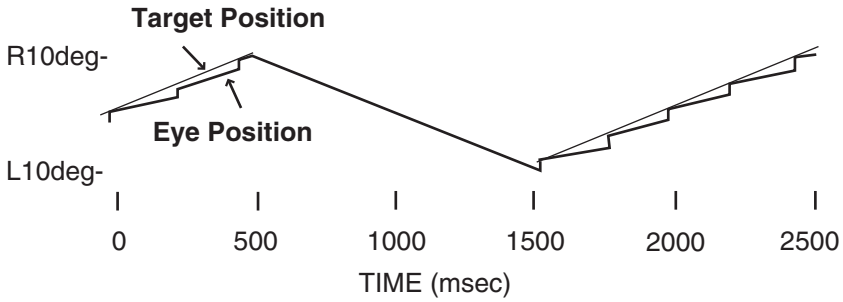


**Figure 7.9** Impaired smooth pursuit to the right. The *light line* represents target trajectory. The *dark line* represents eye position. R10deg and L10deg represent eye positions (right and left 10 degrees, respectively) within the orbit.

## Saccadic Eye Movements

Saccadic eye movements are very fast 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 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 (Table 7-14). 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.

Cerebellar vermal lesions result in hypometric saccades to the right and to the left. Slow saccades can be due to 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 be due also to an internuclear ophthalmoparesis (INO). An INO occurs when the pathway between the VIth and IIIrd 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, and

■ Table 7-14 **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 |

retropulsion test should be performed to evaluate stance, and gait and tandem gait should be examined.

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 *positiv*e Romberg test result is one in which the patient is stable with eyes open but loses balance with eyes closed. A positive Romberg result 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 as well as in those with chronic vestibular defects and in some normal individuals older than 65 years.

For the Fukuda's Stepping Test, the subject steps in place for 50 steps with arms extended and eyes closed.[20] Progressive turning toward one side of more than 30 degrees is abnormal. A *positive* Fukuda's Stepping Test result is frequently found in patients with a unilateral vestibular defect but it is also seen in patients with a leg-length discrepancy or other structural abnormalities of the legs.

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).

Normal 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.[21,22]

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CHAPTER 8

# Vestibular Function Tests

*Ronald J. Tusa, MD, PhD*

Laboratory testing serves several purposes. It (1) confirms what one already suspects from the history and clinical examination (i.e., unilateral or bilateral vestibular loss versus no vestibular loss), (2) quantifies the extent of loss (complete labyrinthine loss innervated by superior and inferior vestibular loss versus incomplete loss), (3) shows evidence of central compensation after physical therapy, and (4) shows evidence of an aphysiological component. Each of these points is discussed in this chapter. The discussion is divided into tests that are specific for different portions of the labyrinthine and vestibular nerve (Fig. 8.1) and tests that assess function but are not specific to labyrinth and vestibular nerve (Table 8-1).

## Tests that Specifically Assess Labyrinth or Vestibular Nerve

### *Caloric Test*

The caloric test is the best method of determining whether a unilateral vestibular defect is peripheral or central and also of identifying the side of the defect. This test is usually not well tolerated in children younger than 8 years. The caloric test is usually part of a battery of tests included in electronystagmography (ENG) (or electrooculography [EOG]), or videography (VOG). The battery of tests consists of spontaneous eye movements, position testing,

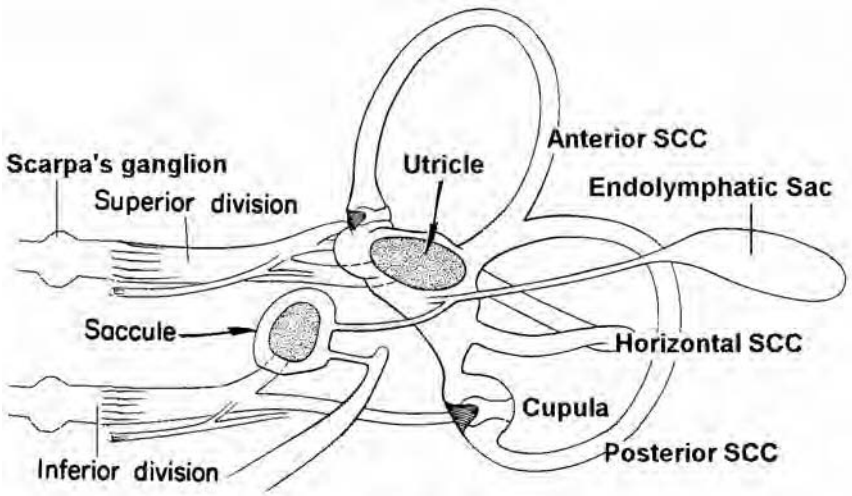


**Figure 8.1** Labyrinth and vestibular nerve. The superior vestibular nerve innervates the anterior semicircular canal (SCC), horizontal SCC, and utricle. The inferior vestibular nerve innervates the posterior SCC and the saccule (Adapted from Schuknecht, 1974.[1])

**■ Table 8-1 TYPES OF TESTS USED TO ASSESS PATIENTS WITH DIZZINESS**

| Tests that assess the function of the labyrinth and vestibular nerve | Functional tests that do not evaluate peripheral vestibular function |
|---|---|
| Caloric test — appropriate for horizontal SCC and portion of the superior vestibular nerve. | Spontaneous and positional tracking, smooth pursuit, OKN and saccadic eye movements |
| Rotary chair test— appropriate for horizontal SCC and portion of the superior vestibular nerve | Computerized dynamic posturography |
| Quantified DVA test— appropriate for horizontal SCC and portion of the superior vestibular nerve | |
| VEMP test—appropriate for saccule and portion of the inferior vestibular nerve | |
| Subjective visual vertical test— appropriate for utricle and portion of the superior vestibular nerve | |

visual tracking, and vestibular testing (caloric testing or rotary chair tests). These tests are objective and quantitative and ideally are performed in all patients in whom a vestibular defect is detected by clinical examination. ENG and EOG record eye movements by means of small electroencephalography (EEG) electrodes placed on either side of the eye to record small voltage differences between the cornea and retina of the eye. VOG records eye movements by means of a video camera and software that track the pupil of the eye.

Before the caloric test is performed, the internal auditory canal is examined with an otoscope, and any wax or debris that blocks the canal is removed. If there is a tympanic membrane perforation, different temperatures of air instead of water should be used as the stimulus. The caloric test uses a nonphysiological stimulus (water) to induce endolymphatic flow in the semicircular canal (SCC) by creating a temperature gradient within it.

Each ear canal is irrigated for 40 seconds, with a constant flow rate of water at two temperatures (30°C and 44°C). Eye movements are recorded for 2 minutes after each irrigation. During the test, the patient is supine, but the head is tilted up 30 degrees, placing the horizontal SCC in a vertical plane. When cold water (30°C) or iced water is flowing into external auditory canal, endolymph fluid in the horizontal SCC sinks. This movement of endolymph causes the cupula to depress away from the utricle (Fig. 8-2). Movement of the cupula away from the utricle inhibits the spontaneous activity in the vestibular nerve (Fig. 8.3), resulting in nystagmus with slow phases directed toward the side of irrigation (Fig. 8.4). When

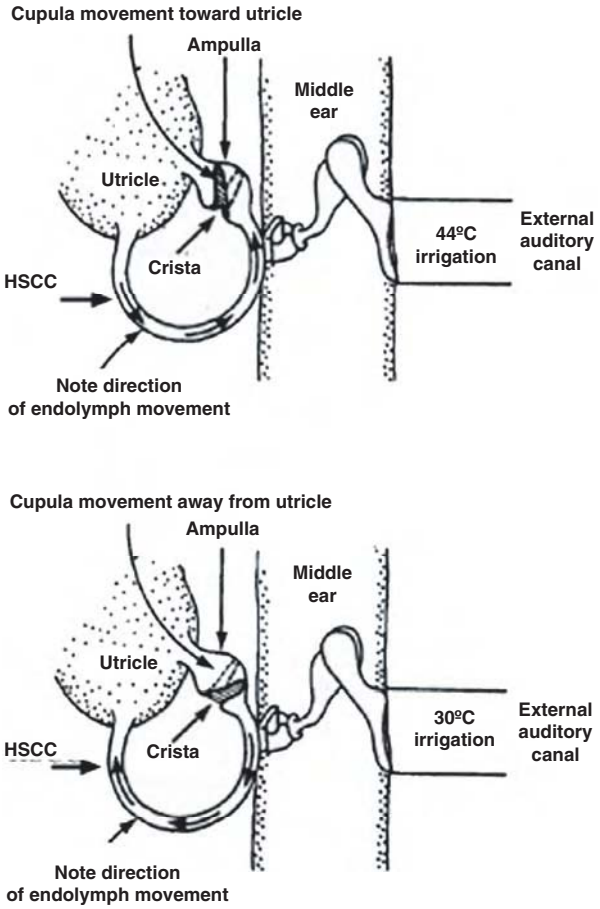


**Figure 8.2** Diagram illustrating how irrigation of external auditory canal with water of different temperatures sets up convection currents in different directions in the endolymph of the horizontal semicircular canal. This results in deflection of the cupula in different directions, with an end result of either excitation or inhibition of the vestibular neurons. In this illustration, the subject has been positioned supine with the head elevated 30 degrees, placing the horizontal semicircular canal (HSCC) in a vertical alignment. (Modified from Baloh and Honrubia,1990[2])

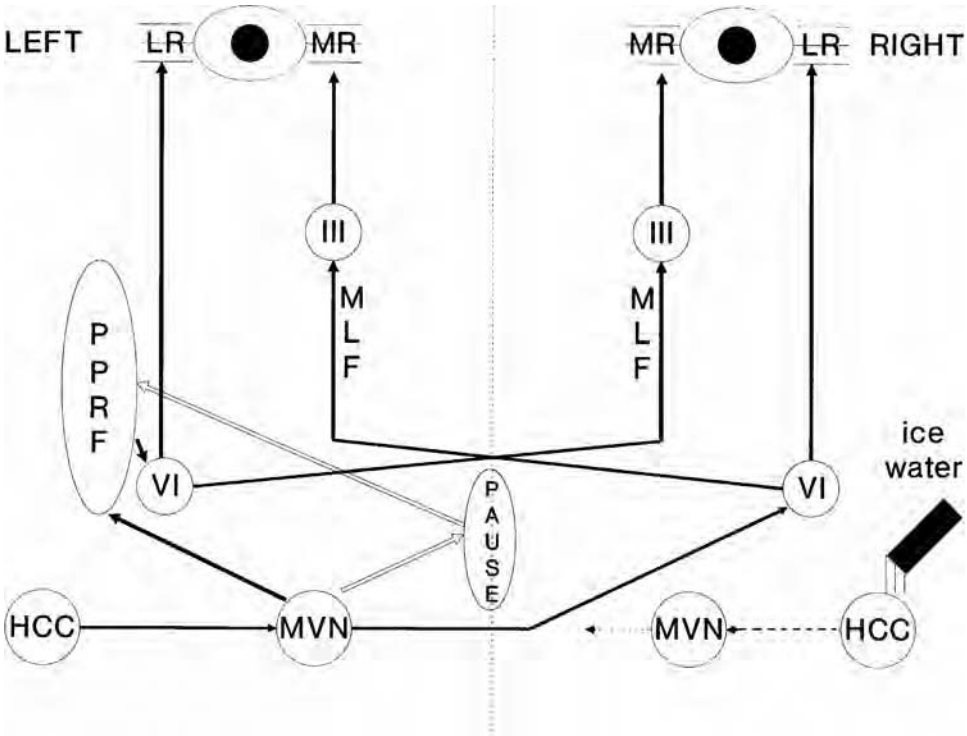


**Figure 8.3** Schematic diagram of caloric test in right ear. Iced water irrigation will cause endolymph to sink within the horizontal semicircular canal (HCC), resulting in decreased spontaneous neural activity in the medial vestibular nucleus (MVN). Asymmetric firing of the MVN on each side leads to nystagmus. In this figure, a left-beating nystagmus would occur (slow phases to the right and quick phases to the left), mediated by the lateral rectus muscle (LR), medial rectus muscle (MR), VIth nerve nucleus (VI), oculomotor nucleus (III) and connection between VI and III, called the medial longitudinal fasciculus (MLF). Quick phases are generated by the parapontine reticular formation (PPRF). PAUSE cells suppress cells in the PPRF.

warm water (44°C) is flowing into external auditory canal, the opposite response occurs. At the end of this period, the ear is emptied of any remaining water, and sufficient time is allowed for the nystagmus to stop before the next irrigation is begun (usually 5 minutes).

The results of caloric testing, and the determination whether there is a meaningful asymmetry in the responses, can be quantified using Jongkees formula, as follows:

$$\frac{(LC + LW) - (RC + RW)}{(LC + LW) + (RC + RW)} \times 100$$

where the values are the velocity of the slow-component eye moments to cool-water irrigation of the left ear *(LC)*, warm-water irrigation of the left ear (*LW*), cool-water irrigation of the right ear (*RC*), and warm-water irrigation of the right ear (*RW*).

The comparison of responses from the irrigation of the right and left ears also can be depicted graphically (Fig. 8.5).

### Strengths of Test

The caloric test is the best test for determining whether a vestibular defect is peripheral or central and for identifying the side of the defect. It is inexpensive relative to rotary chair testing. See the discussion of rotary chair testing for sensitivity and specificity information.

### Weaknesses of Test

The caloric test assesses only the horizontal SCC or the portion of the superior vestibular nerve that innervates this canal. Because it compares the response from one ear with that from the other ear, it cannot be used to assess bilateral vestibular loss. The test also cannot be used to determine central compensation.

## *Rotary Chair Testing*

Rotary chair testing is a physiological test that can be used to assess the horizontal SCC or the superior vestibu-

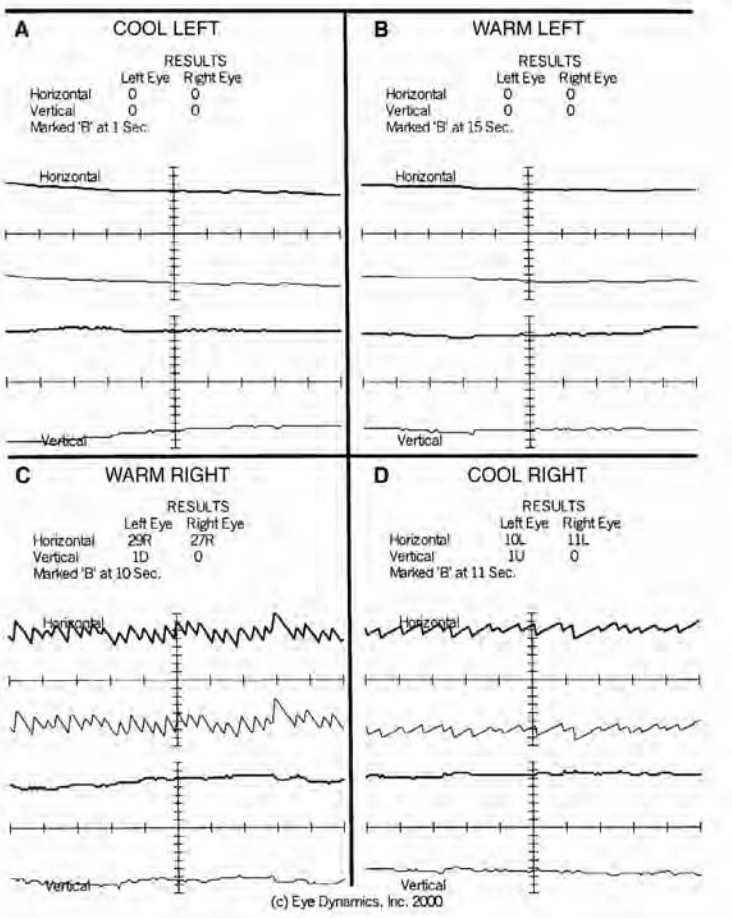


**Figure 8.4** Caloric nystagmus in a patient with left vestibular hypofunction. Note that irrigation of the right ear with warm water (*C*) results in a right-beating nystagmus and that irrigation with cool water results in a left-beating nystagmus (*D*). In contrast, there is no response from the left ear to either cool water (*A*) or warm water (*B*).

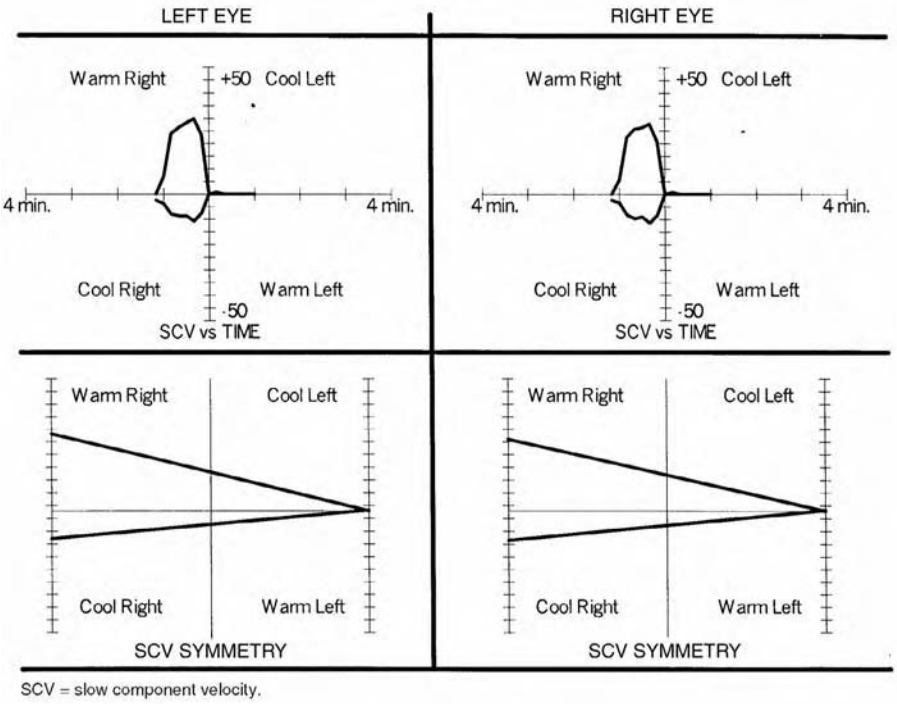


**Figure 8.5** Graphic illustration of the caloric asymmetry in the patient represented in Figure 8.4.

**Figure 8.6** Rotary chair testing: Rotation of the patient generates a response that reflects stimulation of both inner ears simultaneously. Eye movements are recorded using electrooculography (EOG) or infra-red video-oculography.

lar nerve that innervates it (Fig. 8.6). Eye movements are measured by EOG or VOG. After calibration, peak slow-phase eye velocity is measured during either sinusoidal chair rotations or constant-velocity step rotations in the dark, to assess vestibulo-ocular reflex (VOR) gain (eye velocity ÷ chair velocity), time constant during constant velocity steps (rate of decay of slow-phase eye velocity), and phase during sinusoidal rotation (offset between time of peak eye velocity and time of peak chair velocity). VOR gain is normally near 1.0 at birth and decreases in adults.[3] In my laboratory, gain for VOR step rotation less than 0.3 is abnormal and indicates vestibular hypofunction.

If head rotation is sustained in one direction (e.g., spinning around at a constant speed), the slow-phase eye movements of the VOR drive the eyes into extreme contraversive deviation in the orbit. Moving the eyes in this extreme position during the VOR triggers a quick phase (type of saccade) in the opposite direction, which brings the eyes back into the working ocular motor range. A sustained sequence of slow and quick phases is called *nystagmus* (Fig. 8.7).

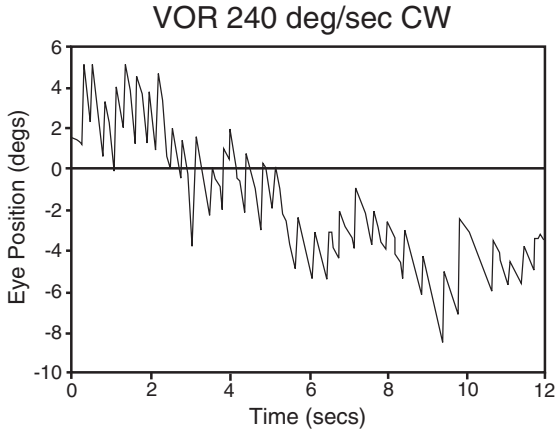


**Figure 8.7** Vestibular nystagmus from rotary chair test (constant velocity chair rotation at 240 deg/sec). With rotation in a clockwise direction (patient's right), a slow-component eye velocity is generated to the left. In the depiction here, eye movements to the right are positive or up, and eye movements to the left are negative or down. Note the gradual decrease in the slope of the slow-component eye movement trace with time, which indicates that the eye movement velocity is slowing.

The sensory stimulus for the VOR is head acceleration, and yet, during head rotation at a constant velocity, slow-phase eye velocity still matches head velocity fairly well, at least for the first several seconds after head acceleration has stopped. This function is mediated by the velocity storage system located in the brainstem, a system capable of storing the head velocity signal for a limited portion of the time. During a sustained, constant velocity rotation, slow-phase eye velocity decreases in an exponential manner (Fig. 8.7). The time constant of the decay (time to decay to 37% of initial value) is 10 to 30 seconds. This decay can be graphically illustrated by plotting the velocity of each slow-component eye movement against time (Fig. 8.8). Another form of rotary chair testing uses sinusoidal chair movements rather than step rotations at a constant velocity. In sinusoidal rotary chair testing, the chair rotations are performed at 0.0125, 0.05, 0.2, 0.4, and 0.8 Hz. The gain and phase of the slow-component eye velocities are plotted for each frequency of rotation (Fig. 8.9).

## Strengths of Test

Rotary chair testing is one of the most physiological tests to assess vestibular function and can be performed even in children of any age, although calibrations are usually obtainable only in infants 6 months or older (by using a "happy face" or similar stimulus). Children younger than 4 years can sit in a parent's lap. The test is not susceptible to mechanical problems of the ear, so it can be done in individuals with such problems, includ-

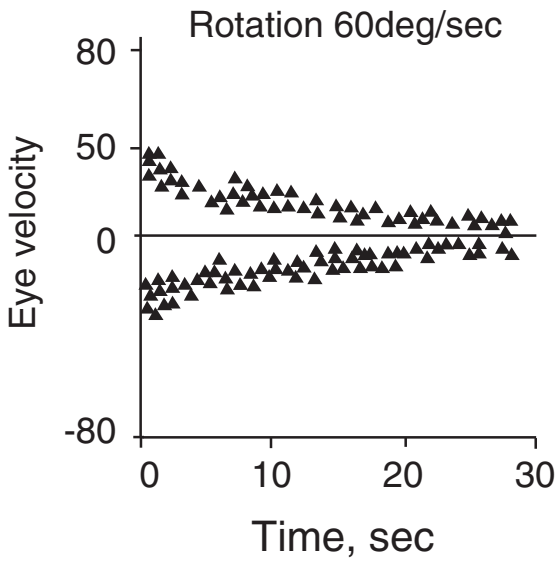


**Figure 8.8** Slow-phase eye velocity decay during 60 deg/sec, constant velocity rotary chair test. Vestibulo-ocular reflex (VOR) gain (peak slow-phase eye velocity ÷ peak chair [or head] velocity) can be measured; in this example, it is 30 ÷ 60 = 0.5. VOR time constant (time it takes for eye velocity to decrease to 37% of peak value) can be measured; in this example it is 14 seconds.

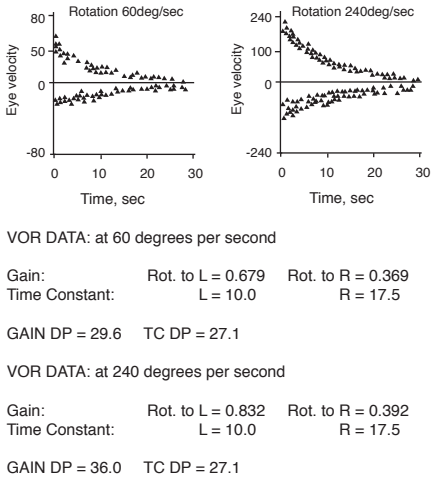


**Figure 8.10** In individuals with uncompensated unilateral vestibular hypofunction, gain is reduced for rotation to the side of the lesion in both 60 deg/sec and 240 deg/sec chair rotations. The patient represented by this figure has an uncompensated defect on the right side. In individuals with compensated defects, the vestibulo-ocular reflex (VOR) gain at low chair velocities (60 deg/sec) would begin to return to normal.

ing perforated tympanic membrane. It is superior to caloric test in patients with bilateral vestibular weakness. Some patients with central vestibular defects have increased VOR gain (some forms of spinocerebellar atrophy), which is not detected with caloric tests but is with rotary chair testing. This test can be used to help determine central compensation from vestibular rehabilitation (Fig. 8.10).

The sensitivity of bithermal caloric test for identifying vestibular hypofunction ranges from 31% to 90%, and its specificity is 86%.[4,5] The sensitivity of rotary chair testing for identifying vestibular hypofunction ranges from 66% to 71%, and its specificity is 54%.[4,5] One reason for this large range in sensitivity is that there is no "gold standard" for identifying vestibular hypofunction other than when it occurs surgically (vestibular nerve section).

## Weaknesses of Test

Rotary chair testing can assess the horizontal only SCC or the portion of the superior vestibular nerve that innervates it. Because rotation affects vestibular

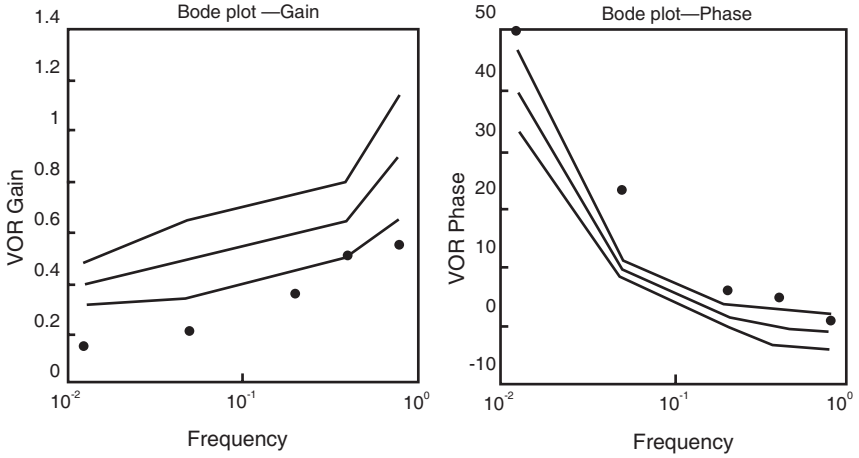


**Figure 8.9** Bode plot of gain and phase of slow-component eye movements during sinusoidal rotation in the dark. Phase is analogous to the time constant obtained during step rotations. The three *solid lines* in each plot indicate the mean and standard deviation for a normal response. The responses depicted in this figure are for the patient represented in Figures 8.4 and 8.5, who has a left vestibular hypofunction.


**Figure 8.11** Setup for quantified dynamic visual acuity test. See text for explanation.

function on both sides, it can be more difficult to identify a small lesion on one side with this test than with the caloric test. The rotary chair test equipment is very expensive and requires yearly calibration and maintenance.

## *Quantified Dynamic Visual Acuity*

Quantified dynamic visual acuity (DVA) is one of the best functional tests in patients with unilateral and bilateral vestibular hypofunction.[6] It measures visual acuity while the head is moving. In this test, the static, distant visual acuity is first determined with the head still. Visual acuity is then determined while the patient's head is oscillated manually or actively. During the test, the subject is asked to tell the operator the direction that the optotype is pointing (letter E in the computerized test my colleagues and I use[6]). The optotype comes on only during head velocities of 120 to 180 degrees per second (deg/sec) as determined by a velocity rate sensor (Fig. 8.11). The results, DVA, are expressed as the difference between acuity measured with the head stationary and acuity measured with the head moving. We usually measure visual dynamic visual acuity as the logarithm of the minimal angle of resolution (LogMAR), one of the best ways to measure change in visual acuity.

Dynamic visual acuity decreases as vestibular loss increases. Figure 8.12 ranks patients according to the

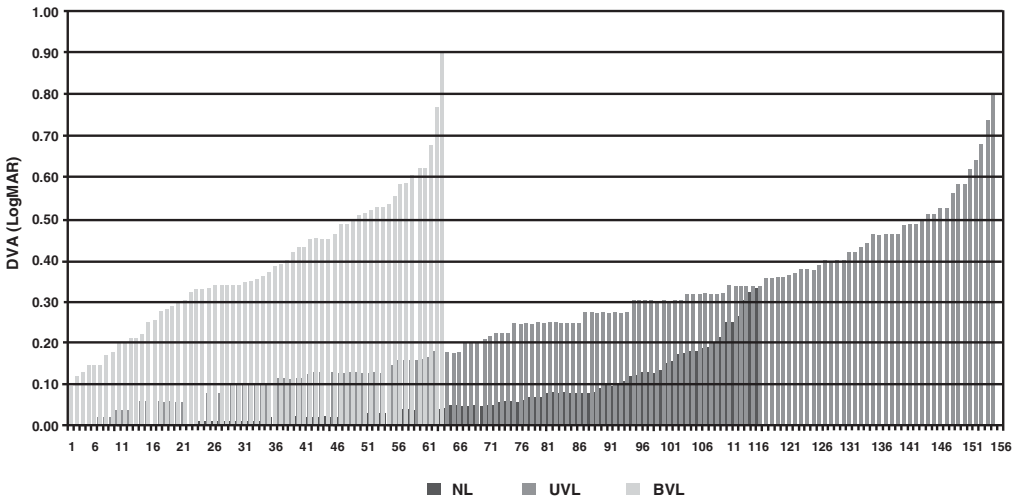


**Figure 8.12** Variability in dynamic visual acuity (DVA) among patients with dizziness without vestibular loss (NL), with unilateral vestibular loss (UVL), and with bilateral vestibular loss (BVL). DVA is measured as the logarithm of the minimal angle of resolution (LogMAR), where LogMAR 0.000 is equivalent to a visual acuity of 20/20, and LogMAR 1.00 is equivalent to one of 20/200.

degree of vestibular loss based on caloric test result and LogMAR score. There is good reliability in any single patient, but great variability among patients with specific vestibular defects. One reason for this variability is that the test is age-dependent, older patients having higher (worse) LogMAR scores. Therefore, the test should not be used to diagnose unilateral or bilateral vestibular loss.

DVA is best used to identify change in a patient undergoing a 5-week course of vestibular adaptation exercises (see Chapter 20) (Fig. 8.13). Note that not all patients show an improvement in DVA after such a course of exercises.

## Strengths of Test

DVA is one of the most useful tests for following functional VOR in patients with unilateral and bilateral vestibular deficits. It can be used to help identify central compensation. DVA is a highly reliable test in patients with vestibular hypofunction ($r = 0.84$), and normal values by age are available.[6]

## Weaknesses of Test

This test can assess only the horizontal SCC or the portion of the superior vestibular nerve that innervates it. Because rotation affects vestibular function on both sides, DVA may not identify a problem in patients with small lesions on one side. Ocular motor defects (eye muscle weakness, strabismus, central nystagmus) and glasses with high refractive changes (bifocals and progressive lenses) can decrease DVA; therefore, the test should not be used to diagnose unilateral or bilateral vestibular loss.

## Vestibular Evoked Myogenic Potential Test

The vestibular evoked myogenic potential (VEMP) test assesses the saccule and its central projections, including the inferior vestibular nerve.[8,9] VEMPs evaluate the saccule–medial vestibular spinal projection–XIth central nerve circuit. The test assesses the function of the saccule by presenting tones in the ears while recording the evoked responses from the sternocleidomastoid muscle (SCM). Subjects are positioned supine in a dimly lit room. Surface electrodes are placed over the belly of each SCM with a ground electrode placed on the forehead. The subject is asked to flex the neck while a series of tones is presented in each ear individually. The stimulus can be tones or clicks. The intensity of the stimulus ranges between 60 and 94 dB, normalized hearing level (nHL). The rate of the stimulus is typically 5.0 per second, and 128 trials are averaged. The response to the stimulus is an inhibitory response of SCM tone detected by a surface

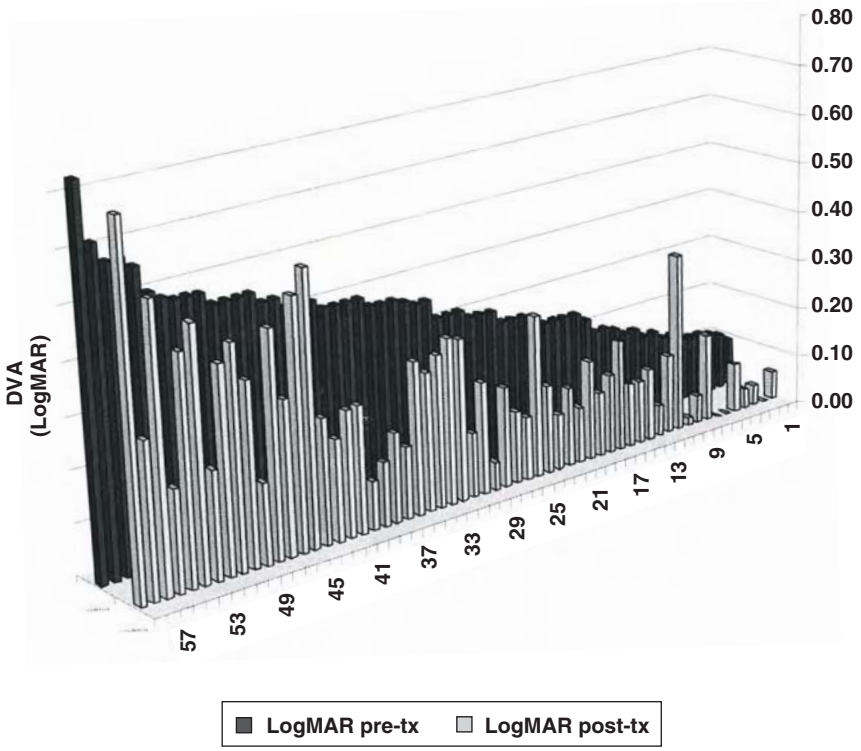


**Figure 8.13** Change in dynamic visual acuity (DVA) in individual patients with unilateral vestibular hypofunction after 4 to 6 weeks of vestibular rehabilitation. Approximately 22% of the patients did not show an improvement in DVA, defined as a change greater than the mean plus 2 SD of the test-retest variability. (From Herdman et al, 2003.[7])

electromyography (EMG) recording. Amplitude of the response depends on tone intensity and baseline muscle contraction. The response is a positive and negative wave (Fig. 8.14). The evoked responses in the SCMs are recorded (total of 750 trials for each ear) and summed for each ear. Absence of the evoked myogenic response reflects damage to the saccule on that side.

### Strength of Test

The VEMP test is the only test for assessment of the saccule and inferior vestibular nerve.

### Weaknesses of Test

It is difficult to determine partial defects on the basis of the amplitude of the response, because the amplitude varies according to the muscle tone of the SCM. One way around this problem is to record from both muscles simultaneously with the subject lifting the head up while supine. This test cannot be used to determine central compensation. It can be done in individuals with sensorineural hearing loss, but not in individuals with conduction hearing defects (e.g., otosclerosis). There is some evidence that the VEMP can reappear in approximately 17% of patients in whom it previously was absent.[10]

## *Subjective Visual Vertical Test*

The subjective visual vertical (SVV) test assesses utricular function and its central connections, including the superior vestibular nerve.[11] It is a subjective test of the degree of ocular torsion present after unilateral lesions in these pathways. Subjects sit in a dark room and orient a projected image of a tilted line until it looks vertical to them. They are given at least 10 trials before the mean and standard deviation of the offset from true vertical are determined. Usually, 2 degrees of tilt or less is considered a normal response. Ocular torsion for peripheral problems is ipsilateral; that is, with a right utricular or superior vestibular nerve defect, the offset is to the right.

The degree of offset depends on the completeness of the lesion and the age of the defect. Studies show that people with unilateral vestibular loss in which the utricle is damaged shift their perception of vertical orientation in a way that reflects the asymmetry in utricular input.[12,13] This phenomenon occurs only when there are no visual cues to aid in orientation, which is why the test is performed in complete darkness (a light-tight room). In a study of patients who had undergone unilateral vestibular nerve section, the mean offset was 12 degrees at 1 week after surgery, 6 degrees at 1 month, and 4.5 degrees at 1 year.[11] The mean deviation of the line (to the right or left) is compared with values for healthy individuals that we have established in the literature[11] (deviation from true vertical $>$ 2.0 degrees is the criterion for abnormal response).

### Strength of Test

The SVV test is the easiest means available for assessing utricular function, the other means of directly assessing degree of ocular torsion being with retinal photographs.

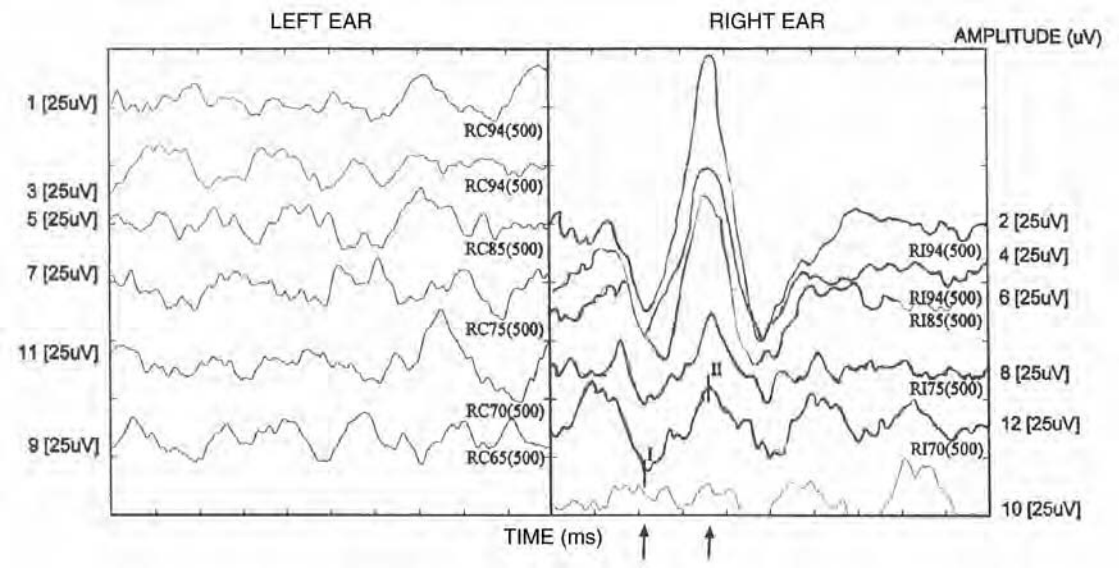


**Figure 8.14** Vestibular evoked myogenic potential (VEMP) test results. Normal *P13, N 23* VEMP values in the right ear with absence of response from the left ear.

Because the SVV response changes with time, it can be used to assess central compensation of a static defect, but only by looking at changes in the same individual.

### Weaknesses of Test

The SVV test must be done in a completely dark room. It will not detect bilateral utricular defects.

## Tests That Do Not Specifically Assess Labyrinth or Vestibular Nerve

### *Visual Tracking*

#### Smooth Pursuit Eye Movements and Vestibulo-Ocular Reflex Cancellation

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. With quantitative testing, unlike the clinical exam, the patient is asked to track a small, slowly moving target (20 degrees/second) and the eye movement is recorded. Thus the gain (eye velocity/target velocity) of smooth pursuit can be calculated. This is especially important because smooth pursuit eye movements degrade with age and the decision as to whether or not the eye movements are normal can be made more accurately if the eye movements are quantified. See Chapter 7 for further discussion.

#### Saccadic Eye Movements

Saccadic eye movements are very rapid changes in eye position. The advantage of measuring saccadic eye movements using a tracking system is that the gain, amplitude, accuracy and latency to onset of the saccades can be measured and compared to normal values. See Chapter 7 for further discussion.

### *Computerized Dynamic Posturography*

Postural sway is quantified with dynamic posturography, which measures sway in conditions in which visual and somatosensory cues are absent or altered (Fig. 8.15). Automatic postural responses can also be measured in response to perturbations of the support platform. Deficits in a variety of different neural systems can be identified, including cerebral cortex, anterior cerebellum, and the spinal cord. The test is not specific for vestibular disorders, although patients with uncompensated or severe vestibular deficits typically have difficulty maintaining their balance when both visual and somatosensory cues are altered. This test is also very

**Figure 8.15** Computerized dynamic posturography. The patient stands on a force platform. Sensors are oriented to detect vertical pressure changes and shear forces. In some equipment, as shown here, both the support surface (force platform) and the visual surround can move. (With permission from NeuroCom International Inc, Clackamas, OR).

useful in demonstrating objective signs of a functional component.

The advent of computerized posturography as part of balance assessment has extended the ability of clinicians and researchers to identify subtleties in postural control, specific system impairments, and functional limitations. *Postural control* involves the interplay among the sensory, central nervous, and musculoskeletal systems. To meet the constantly changing demands of maintaining balance, body position and movement must be detected by the visual, somatosensory, and vestibular systems. The central nervous system then must integrate this information and select an appropriate response. Finally, the response must be performed by the musculoskeletal system within the appropriate timeframe and with adequate force. Research has demonstrated that interventions targeted at improving specific deficits are successful in improving postural control and in reducing the risk for falls.[14]

Computerized posturography can be used to quantify many aspects of postural control, such as the ability to maintain quiet stance and to shift weight voluntarily, as well as to assess automatic postural reactions. Computerized posturography has now been expanded to include the assessment of activities such as moving from sitting to standing and turning while walking.

## Balance under Different Sensory Conditions

Postural sway can be measured under conditions in which visual and somatosensory feedback is altered (Table 8-2). Such manipulation of feedback is called the *sensory organization test* (SOT). In one commercially available apparatus (NeuroCom International, Inc., Clackamas, OR), the change in sway angle is used to move either a visual surround or the support surface. At low frequencies of sway (<0.3 Hz), the visual surround or support surface moves in synchrony with the individual's sway, and visual and somatosensory cues are minimized.

At higher frequencies of sway, the mechanics of the system cannot move the surround or surface at the same frequencies as body sway, so the sensory feedback is simply altered in a way that is novel to the subject. Movement of the visual surround alters the visual cues normally used for postural stability. For example, in quiet stance, humans normally have a small amount of anterior and posterior (AP) sway. As humans sway, a number of visual cues influence postural stability, including stereopsis, contrast sensitivity, and retinal disparity.[15–17] If the visual world around a person (visual surround) moves in parallel with the body as he or she sways, the usual visu-

■ Table 8-2 **SENSORY ORGANIZATION TESTS IN COMPUTERIZED DYNAMIC POSTUROGRAPHY**

| Sensory Organization Test Conditions | Sensory Cues | Cues Available for Stablity |
|---|---|---|
| Condition 1 (C1) | Eyes open<br>Visual surround and platform stable | Normal visual, somatosensory, and vestibular cues |
| Condition 2 (C2) | Eyes closed<br>Visual surround and platform stable | No visual cues<br>Normal somatosensory and vestibular cues |
| Condition 3 (C3) | Eyes open<br>Moving visual surround<br>Platform stable | Normal somatosensory and vestibular cues<br>Altered visual feedback<br>Normal visual and vestibular cues |
| Condition 4 (C4) | Eyes open<br>Visual surround stable<br>Platform moving | Altered somatosensory feedback<br>Normal vestibular cues<br>No visual cues |
| Condition 5 (C5) | Eyes closed<br>Platform moving | Altered somatosensory feedback<br>Normal vestibular cues |
| Condition 6 (C6) | Eyes open<br>Visual surround and platform moving | Altered visual and somatosensory feedback |

al cues are reduced or inaccurate and so cannot be used effectively to maintain postural stability. Similarly, when the support surface moves in parallel with body sway, the alteration in somatosensory feedback is less effective as a signal in the maintenance of upright posture. Other systems (e.g., Balance System, Biodex Medical Systems, Shirley, NY; Balance Quest, Micromedical Technologies, Chatham, IL; the Modified-Clinical Test of Sensory Interaction in Balance [M-CTSIB] test, NeuroCom International, Inc.) alter vision by having the subject close the eyes or by projecting an optokinetic stimulus and alter the somatosensory feedback by having the subject stand on dense foam or an unstable surface.

## Automatic Postural Responses

Tests of reactive balance involve sudden, brief displacement of the support surface in order to assess the automatic postural responses used in the recovery of balance. These tests are important because people must be able to react to external disturbances in balance if they are to move safely in a challenging environment, such as when a train suddenly starts moving. Tests of the automatic postural responses typically use forward and backward translations of the support surface or pitch movements of the support surface (toes-up or toes-down). During the translational perturbations of the support surface, the relative distribution of weight on each leg, the latency of onset of force development used to regain postural stability, and the amplitude or strength of the response are recorded. The larger the perturbation of the support surface, the greater the response needed to regain balance. Normal subjects and patients with bilateral vestibular deficits exhibit a relative increase in the response (initial rate of change of torque developed) as the amplitude of surface displacement increases. In contrast, patients with cerebellar disorders fail to modify their automatic postural responses as measured by surface reactive force.[18]

The shift in the center of gravity alignment that occurs with pitch perturbations of the support surface destabilizes the individual. Measurements of the magnitudes of response to five identical 'toes-up' or 'toes-down' perturbations are used to assess motor learning or adaptation. As equal perturbations are repeated, there should be a decrease in the amplitude of the force developed to maintain postural stability. Normal individuals quickly learn to maintain their balance during this paradigm, and loss of balance is unusual except during the initial trial in younger subjects.[19] In older subjects ($>$ 70 years), there is an increased likelihood of loss of balance on all trials of "toes-up" perturbations even though adaptation still occurs.

The addition of surface electrode EMG during toes-up perturbations allows the clinician to measure muscle activity—the postural evoked response. The "toes-up" movement of the platform elicits both a short-latency response and a middle-latency response in the gastrocnemius-soleus muscle group (Fig. 8.16*B* and *D*). The short-latency response is equivalent to the monosynaptic stretch reflex, and the middle-latency response is a multisegmental spinal reflex.[20] This is followed by a long-latency response in the anterior tibialis muscle, which acts to restore postural stability by shifting the center of gravity alignment forward (Fig. 8.16*C* and *E*). The long-latency response is believed to encompass a transcortical pathway.[20] The presence, absence, or delay of any of the reflex responses has been correlated with site of neurological lesion (Table 8-3). Note, however, that the measurement of postural evoked responses with surface electrode EMG is not a typical component of the evaluation of patients with vestibulopathy.

## Application to Patient Management

It is important to recognize that dynamic posturography is not a diagnostic test and cannot be used to identify vestibular deficits. Dynamic posturography measures the output of multiple balance systems, not just the vestibulospinal system. Thus, during posturography testing, vestibular deficits may be "masked" by other, functioning balance systems.

Although dynamic posturography should not be used as a diagnostic tool, the results of studies in patients with different problems show performance patterns that are useful as guides to expectations for certain diagnoses (Table 8-4). The overlap in findings among diagnoses can be clearly seen; therefore, difficulty on test conditions 5 and 6 (see Table 8-2) cannot be interpreted as indicating a vestibular deficit. For instance, it is well documented that patients with severe bilateral vestibular loss are unable to maintain balance under conditions in which both visual and somatosensory cues are altered.[21] However, patients with other balance problems, as diverse as Huntington's disease and fear of falling, also may lose their balance under these same conditions.[22,23]

It is probably most appropriate to interpret the results of the SOT in terms of the functional implications of the test performance. Several common patterns in test performance have been identified and ascribed to difficulties using different sensory cues (Table 8-5) rather than diagnoses. It is important to remember that although this test provides reliable data, test performance can be strongly influenced by subjective factors, including patient effort, fear, and cognition.

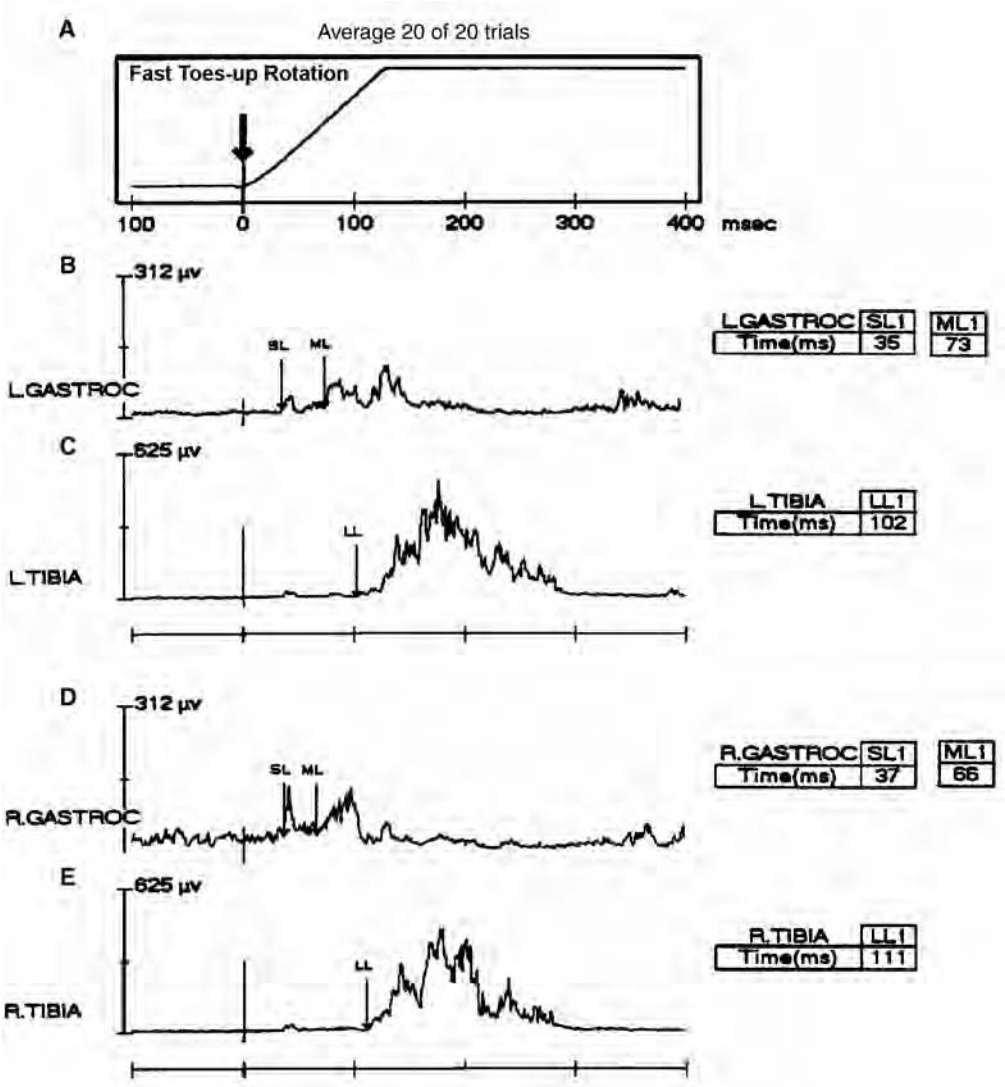


**Figure 8.16** Postural evoked responses as measured by surface electrode electromyography (EMG) in a normal subject. (*A*) Tracings are averaged from a series of 20 pitch (toes-up) perturbations of the support surface. The *ramped line* indicates the perturbation of the support surface, with the *thick arrow* indicating the initiation of the perturbation. (*B* and *D*) Short-latency (SL) and middle-latency (ML) responses elicited in the left (L) and right (R) gastrocnemius/soleus muscles. (*C* and *E*) The long-latency response (LL) occurs in the left (L) and right (R) anterior tibialis muscles and acts to correct for the sudden posterior shift in the center of mass.

## Identification of Aphysiological Test Performance

An aphysiological or nonorganic basis of balance test performance can be characterized by specific patterns of performance on the SOT and motor test (Table 8-6).[37] Cevette and colleagues[35] examined patients who had symptoms unrelated to organic findings. They found that these patients had erratic performances within each condition (i.e., greater intertrial variability) and relatively better performance on the more difficult conditions than on the easier conditions of the SOT (Fig. 8.17). Additionally, they noted a discrepancy between the patients' abnormal performance on dynamic posturography and their normal or near-normal ability to walk.

Goebel and colleagues[34] identified two additional criteria from the motor control tests—exaggerated

■ Table 8-3 **SHORT-, MIDDLE-, AND LONG-LATENCY POSTURAL RESPONSES AND SITE OF LESION**

| Condition | Short-Latency Response | Middle-Latency Response | Long-Latency Response |
|---|---|---|---|
| Normal (mean ± 1 SD) | 43.5 ± 4.2 msec | 89.5 ± 10 msec; absent in 25% of subjects | 125.3 ± 17.8 msec; duration 76 ± 19 msec |
| Normal (mean ± 2 SD) | 51.9 msec | Latency = 109.5 msec | Latency = 160.9 msec; duration = 114 |
| Peripheral neuropathy | Delayed | Delayed | Delayed |
| Anterior lobe cerebellar atrophy; diffuse cerebellar disease | Delayed due to concomitant peripheral neuropathy | Normal | Increased duration in 66% of subjects |
| Friedreich's ataxia | Absent in 66% of subjects | Absent in 66% of subjects; when present, is delayed | Prolonged rate of rise (> 50 msec); delayed (>225 msec) |
| Cerebellar hemispheric lesion | Normal | Normal | Delayed (146 msec) |
| Parkinson's disease | Normal latency; normal integral | Normal latency; greatly increased integral | Normal latency; normal integral |
| Spinal cord lesion | Normal | Absent in 50% of subjects | Delayed (>164 ± 36.5 msec) |
| Intracranial lesions | Normal latency | Normal latency | Delayed latency |

From Diener and Dichgans, 1986.[20]

responses to small translational perturbations and inconsistent responses to small and large perturbations of the support surface—that had a high specificity for identifying persons who were deliberately feigning instability. These researchers also found that a combination of SOT and motor control test criteria reduced to zero the likelihood of identifying a problem where there is no organic cause.[34] The criteria for identifying an aphysiological test performance were validated in a study by Gianoli and associates,[36] who found that 76% of patients who would benefit from an abnormal test (secondary gain) had exaggerated test results, compared with only 8% of patients without secondary gain.

One concern about the use of computerized posturography in identifying malingerers or patients with other nonorganic problems is that individuals may learn to manipulate their results. That is, people who deliberately malingering may learn how to produce results that mimic real disorders rather than producing results that are clearly identifiable as aphysiological patterns. Fortunately, Morgan and colleagues[38] showed that an individual's ability to manipulate the results does not improve when he or she has additional information about dynamic posturography.

## Treatment

Dynamic posturography is useful in establishing treatment and in monitoring patient recovery, especially as part of the rehabilitation process. For example, patients may use different sensory cues to maintain postural stability. Figure 8.18 illustrates the results of the sensory organization test in three patients with bilateral vestibular loss secondary to aminoglycoside ototoxicity. None of the patients can maintain balance when both visual and somatosensory cues are altered (conditions 5 and 6), a finding consistent with the findings of numerous studies in patients with bilateral vestibular loss.[24,25,27] The first

## ■ Table 8-4 PATTERNS SEEN ON POSTUROGRAPHY WITH VESTIBULAR DEFECTS

| Diagnosis | Posturography Performance Pattern* |
|---|---|
| Peripheral vestibular deficits[24,25] | Abnormal sensory organization test results<br>Rare motor test abnormality (< 2%) |
| Severe bilateral vestibular loss[24,25,27] | Loss of balance on C5, C6; may or may not have increased sway on C3, C4<br>Correlation of sensory organization test results with severity of deficit (VOR Tc) |
| Incomplete bilateral vestibular deficit | Increased # of falls on C3, C5, and C6, but not necessarily on all trials |
| Acute unilateral vestibular deficit[28] | Increased sway or loss of balance especially on C5 and C6<br>Can be normal within few days after onset |
| Compensated unilateral vestibular deficit[29,30] | Normal sensory organization test results but evidence of decrement in performance several months out |
| Benign paroxysmal positional vertigo[31–33] | Increased sway on C3 and C6 in BPPV plus head injury<br>Increased sway on C5 and C6 in BPPV with no head injury<br>Increased sway on all tests pre-treatment; improved with remission of symptoms post treatment |
| Central deficits[26] | >90% of subjects have abnormal results of motor tests (especially latencies) as well as sensory organization tests |

*C1 through C6 are sensory conditions; for definitions, see Table 8-2.

## ■ Table 8-5 INTERPRETATION OF FUNCTIONAL DEFECTS SEEN ON POSTUROGRAPHY

| Functional Defect(s) | Possible Interpretations(s) |
|---|---|
| Loss of balance or increased sway: | |
| On C2, C3, C5, and C6 | Visual dependency |
| On C4, C5, and C6 | Somatosensory dependency |
| On C5 and C6 | Reflects increased difficulty of these conditions, vestibular deficit, or severe bilateral vestibular loss |
| Generally increased sway<br>Stopping test by putting hands out to touch wall<br>Verbalized expression of concern or fear | Fear of falling |
| Increased sway or loss of balance occurring on later trials only, with normal performance on initial trials of same condition | Fatigue |
| Loss of balance or increased sway on initial trial of each sensory condition | Inability to handle novel postural challenges |

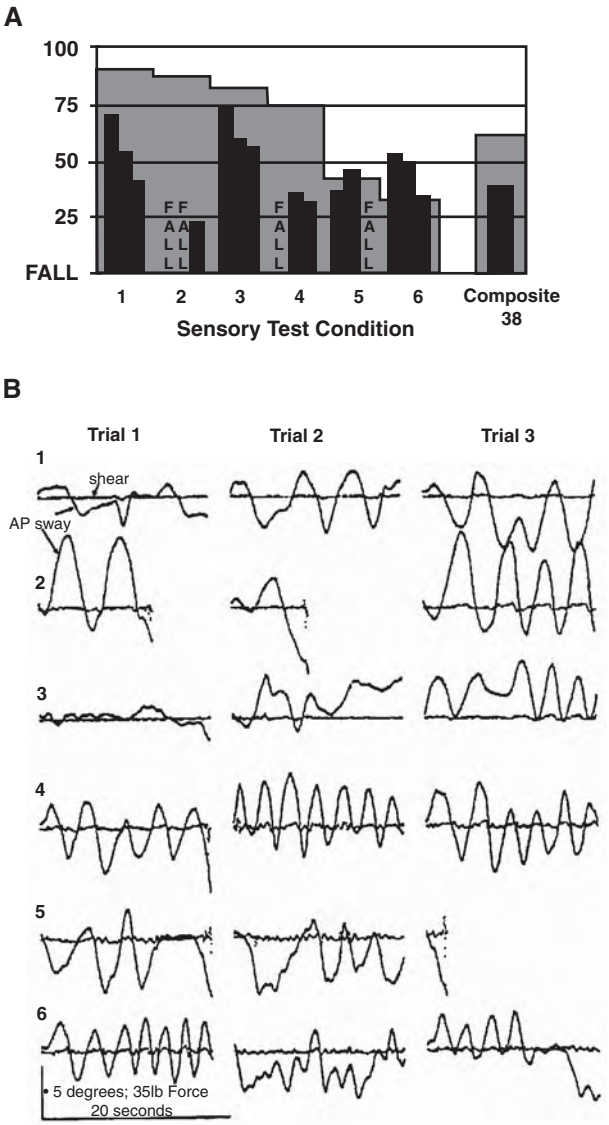


**Figure 8.17** (*A*) Performance on the sensory organization test by patients with aphysiological balance problems is characterized by inconsistent performance on different trials of the same condition and by better performance on more difficult conditions than on easier conditions (compare condition C3 with C1 and C5 with C2). (B) Inspection of the anterior-posterior sway trace often reveals a regular periodicity to the sway.

patient (Fig. 8.18*A*), however, has no difficulty with the other tests. In contrast, the second patient may rely on somatosensory cues for stability and therefore lose his balance when somatosensory feedback is altered (test condition 4) but not when visual cues are altered (test conditions 2 and 3) (Fig. 8.18*B*). Similarly, other patients may be reliant on visual cues and so have difficulty in test conditions 2 and 3 but not test condition 4 (Fig. 8.18*C*). Identification of reliance on a particular sensory

■ Table 8-6 **CRITERIA USED FOR APHYSIOLOGICAL PERFORMANCE ON POSTUROGRAPHY TESTS**

| Study | Criteria for Aphysiological Performance |
|---|---|
| Goebel et al, 1997[34] | Substandard performance on C1<br>Score = No. of points below norm for the best trial of C1<br>Exaggerated motor responses to small translations<br>Score = average number of degrees of sway in all trials for small forward and backward translations (should be < 2 degrees)<br>Inconsistent motor responses to small and large translations<br>Score = No. of tests with at least 2 of 3 concordant trials per test (max = 4) |
| Cevette et al, 1995[35] | Lower scores on C1 and 2, higher scores on C5 and 6<br>Score = [(C1 – norm1) + (C2 – norm2)] – [(C5 – norm5) + (C6 – norm6)]<br>Highest score on the following 3 equations:<br>Aphysiological = –158.2 + (1.94 × C1) + (1.09 × C2) + (1.37 × C4) – (0.15 × C6)<br>Normal = – 238.14 + (2.24 × C1) + (1.45 × C2) + (1.70 × C4) – (0.13 × C6)<br>Vestibular = –251.21 + (2.31 × C1) + 1.54 × (C2) + (1.89 × C4) – (0.58 × C6) |
| Gianoli et al, 2000[36] | Substandard performance on C1 and C2<br>Large-amplitude anterior-posterior (AP) sway without falls<br>Score = average number of AP sways > 5 degrees on C4, C5, and C6 without falls<br>Large amplitude lateral sway<br>Score = average number of lateral sways > 1.25 degrees on C4, C5, and C6 without falls<br>Excessive intertrial variability (no score calculated)<br>Circular sway (no score calculated) |

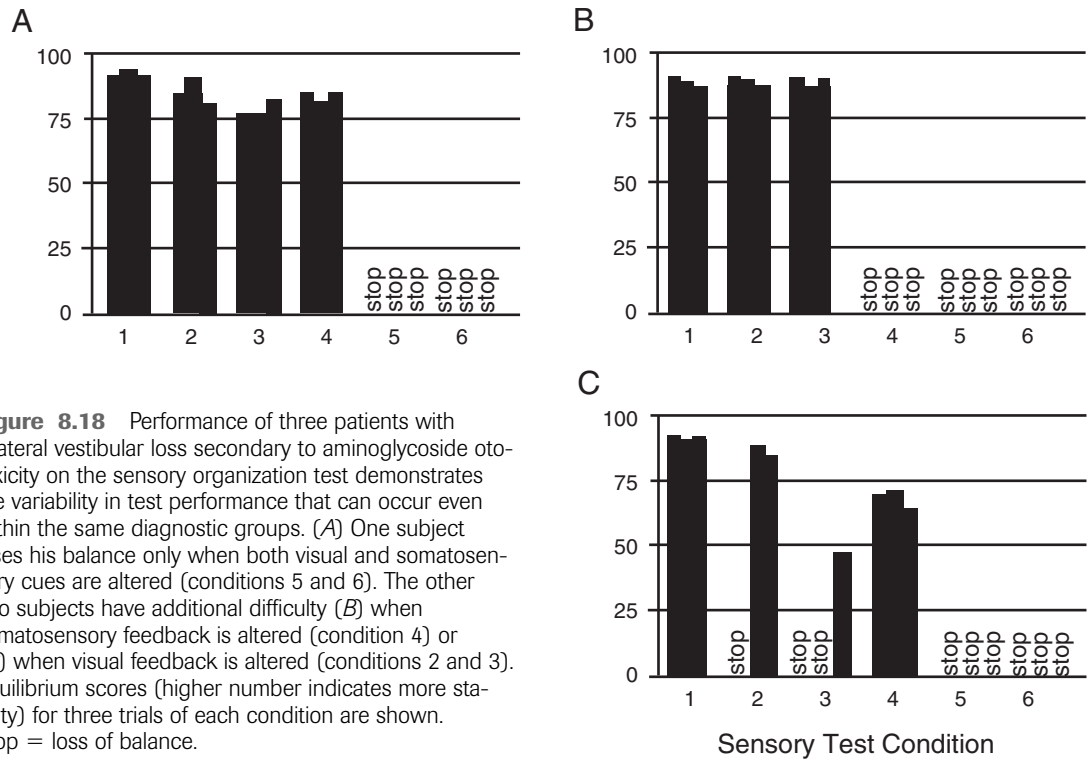


**Figure 8.18** Performance of three patients with bilateral vestibular loss secondary to aminoglycoside ototoxicity on the sensory organization test demonstrates the variability in test performance that can occur even within the same diagnostic groups. (*A*) One subject loses his balance only when both visual and somatosensory cues are altered (conditions 5 and 6). The other two subjects have additional difficulty (*B*) when somatosensory feedback is altered (condition 4) or (*C*) when visual feedback is altered (conditions 2 and 3). Equilibrium scores (higher number indicates more stability) for three trials of each condition are shown. Stop = loss of balance.

cue can then be used to establish specific exercises for the patient.

Computerized dynamic posturography results can be used as an outcome measure for vestibular rehabilitation.[25,39–41] Improved postural stability has been documented in patients with acute and chronic unilateral vestibular hypofunction,[39,42] benign paroxysmal positional vertigo,[31–33] and bilateral vestibular loss,[43] although patients may continue to demonstrate significant physical impairments.

## ■ Table 8-7 OVERVIEW OF TESTS FOR VESTIBULAR FUNCTION

| Problem | Helpful Bedside Test(s) | Helpful Laboratory Test(s) |
|---|---|---|
| Acute unilateral vestibular loss (vestibular neuritis, labyrinthitis, vestibular nerve section) | Spontaneous nystagmus<br>Head-thrust<br>Skew eye deviation | Electronystagmography (ENG) (spontaneous nystagmus and caloric)<br>Rotary chair |
| Chronic unilateral vestibular loss (same causes as above) | Head-thrust<br>Head-shaking nystagmus<br>Dynamic visual acuity | ENG (caloric)<br>Rotary chair |
| Mechanical problems (benign paroxysmal positional vertigo) | Position testing | ENG (position testing) |
| Central problems (stroke, multiple sclerosis, trauma) | Spontaneous nystagmus<br>Smooth pursuit<br>Saccades | ENG (spontaneous nystagmus, visual tracking) |

There is also some evidence that dynamic posturography can be used to predict the outcome of vestibular rehabilitation. Poor pre-rehabilitation disability, including spontaneous and/or continuous symptoms in conjunction with motion sensitivity and abnormalities in four or more of the SOT test conditions, predicts poorer outcomes of a course of rehabilitation.[30,44,45] Although computerized posturography is reliable test and therefore is appropriate as an outcome measure, the relationship between posturography results and functional activities is still not clear.

## Summary

Laboratory testing should be used in conjunction with the history and clinical examination to identify the cause of a patient's dizziness and extent of the defect. Table 8-7 lists the bedside tests that best correlate with laboratory tests for particular problems.

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CHAPTER 9

# Otolith Function Tests

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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 tests—the subjective visual horizontal and the vestibular evoked myogenic potentials—come near to fulfilling these requirements. Both have been regularly used in our clinical laboratories for several years, and what follows is in part a distillation of our own experience with these tests and of their scientific basis. Before considering these tests, however, one must have some familiarity with the structure and function of the otoliths.

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. These biological gyros detect forces: the forces imposed by gravity and the forces generated when we move. They 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 could have consequences for the overall integration of all of 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.

## Otolith Structure

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. 9.1). The SCCs and the otoliths detect these two different forces, not so much because of any differences in the intrinsic properties of the hair cells themselves but because of the way the structures that surround the hair cells are affected by the stimulating force. Each receptor cell has several 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

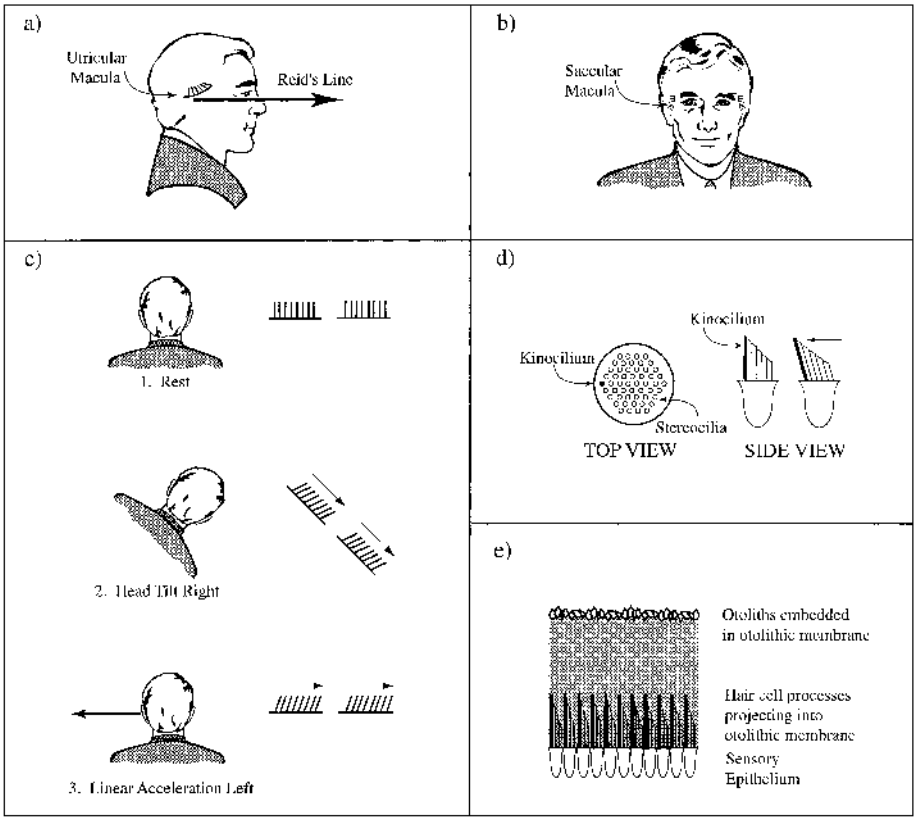



**Figure 9.1** The structure and function of the otolithic system. (*a* and *b*) Schematic representation of the approximate orientation of the utricular and saccular maculae in the head. Reid's line is a standard reference line joining the center of the external ear and the lower edge of the bony orbit. (*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.

structure. The remaining cilia, the *stereocilia,* are of uniform cross-sectional structure and are arranged in a series of increasing height as they approach the kinocilium. Forces cause the otolithic membrane to slide so that the cilia of the receptor hair cells are bent.

Each labyrinth consists of endolymph-filled tubes and sacs containing receptor structures. The two specialized sacs are the *utricular sac* and the *saccular sac.* Contained within each of these sacs is a plate of specialized receptor hair cells and connective tissue, the *macula.* Each macula takes its name from the sac in which it is located, so that in each inner ear there is an 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. Embedded on the outer (free) surface of the otolithic membrane are dense crystals of calcium carbonate, the *otoconia.* 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 in the order of one micron.

The utricular and saccular maculae are almost 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 6000 primary afferent nerve fibers supply the utricle, and only about 4000 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; 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, with the result that there is a highly ordered arrangement of polarization vectors across each macula.

Primary afferent neurons synapse on a number of adjacent receptor hair cells so that some of the precision of 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, presumably 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 frequency declines. Utricular afferents in general show a preference for forces directed 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.

## Primary Otolithic Afferents

Recordings from primary otolithic afferent neurons in the vestibular nerve of experimental animals show a number of important characteristics (for a review see Goldberg & Fernandez[6]). 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. They 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 force 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. 3. The afferents show a marked asymmetry in bidirectional sensitivity. This characteristic results in an increase in firing rate for forces in the excitatory direction that is larger than the decrease in firing rate for the same magnitude of force in the opposite, disfacilitatory direction. 4. 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 forces to contralaterally directed forces. There is 3:1 preponderance of utricular afferents preferring ipsilateral force, whereas there is about a 1:1 distribution of saccular afferents preferring up-and-down force. 5. At rest, some otolithic afferents have a regular interspike interval, and others show an irregular pattern of firing. The dynamic response characteristics of these two groups of neurons are quite different. Regular neurons show little adaptation to maintained forces, whereas the irregular neurons show a transient mode of responding: They fire vigorously during the change in the stimulus but adapt rapidly for a maintained stimulus. This means that even at the level of the afferent neurons leaving the macula, functional specialization has already taken place, in that different aspects of the force stimulus are being signaled by these different parallel pathways.

## Central Projections

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. 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.

Some cells in each vestibular nucleus can be activated both by SCC stimulation and by otolithic stimulation. These convergent neurons integrate linear and angular acceleration input and thus help maintain posture and equilibrium. 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.[7] Complementing that finding are studies that have shown that unilateral section of the vestibular nerve causes a torsion of both eyes toward the operated side.[3,8] In natural 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.[4]

## 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.[3] Friedmann[9,10] 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. We studied the ability of patients with Ménière's disease to set such a light-bar to the visual horizontal before and after uVD.[3,8] 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, so that a slight ipsilesional tilt or *offset* of the subjective visual horizontal (SVH) was a permanent legacy of a uVD procedure for treatment of Ménière's disease.

These findings have been confirmed by others.[11–16] After uVD for vestibular schwannoma, some patients have permanent tilt or offset of the SVH of 15 degrees or more, values similar to that found acutely after uVD. In other words, the SVH appears not to be compensated for, even though in other respects compensation has occurred normally.[17] The explanation for this difference in the SVH long after uVD for Ménière's disease and vestibular schwannoma is not clear but might have to do with involvement of the brainstem or cerebellum during removal of large schwannomas. The ipsilesional offset of the SVH after uVD is enhanced by vibration over the ipsilesional mastoid bone or sternomastoid muscle.[18]

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 in order 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,[19,20] and others just a partial one with ocular torsion and skew deviation.[20–23] We measured torsional ocular position as well as SVH before and after uVD[8] 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. 9.2). One week after uVD, there was up to 15 degrees of ipsilesional ocular torsion and a

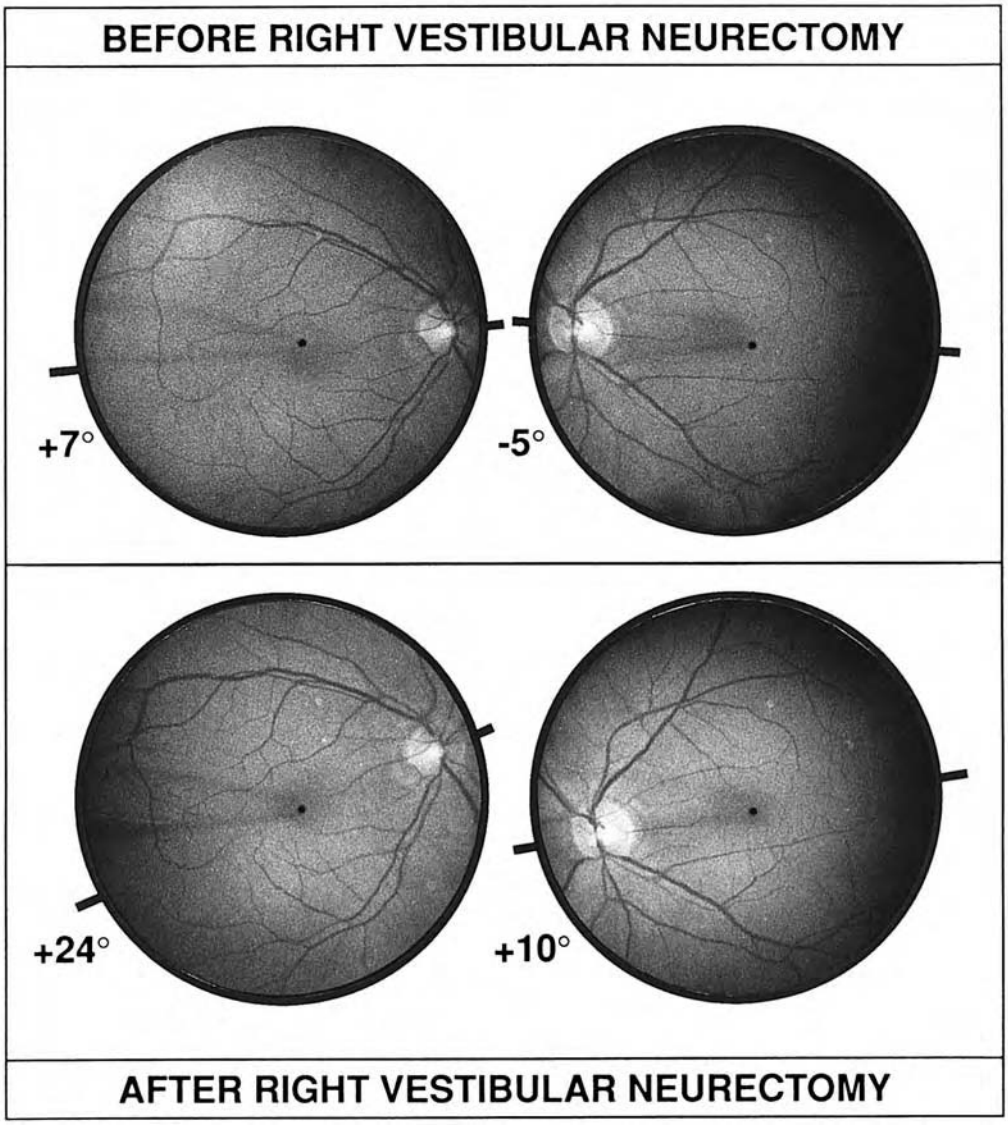



**Figure 9.2** Ocular torsional position before and after unilateral vestibular deafferentation (uVD). Fundus photographs of the left and right eyes of a patient before *(top row)* and one week after *(bottom row) right* vestibular neurectomy. After operation, there is tonic rightward torsion of the 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.

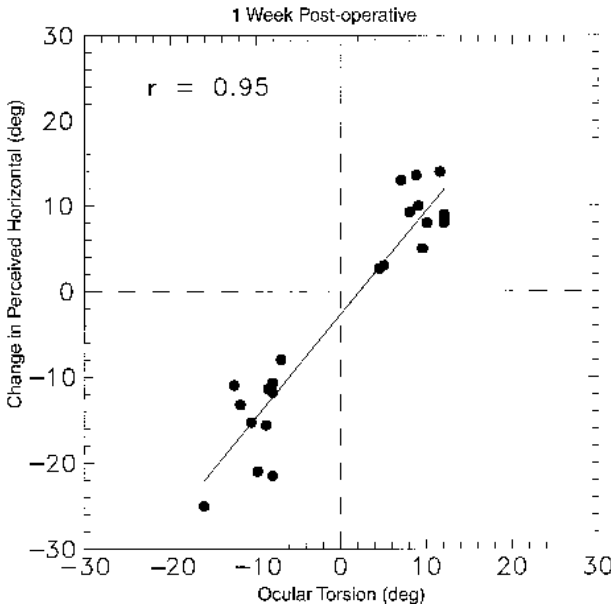



**Figure 9.3** The relationship between ocular torsional position and the subjective visual horizontal. The average value 1 week after unilateral vestibular deafferentation of the change in ocular torsional position was calculated for each patient and correlated with that patient's average change in the visual horizontal. The correlation (0.95) is statistically significant. (From Dai et al, 1989.[3])

close correlation ($r = 0.95$) between the magnitude of the ocular torsion and the offset of the SVH (Fig. 9.3). 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.[8] It is of interest that after acute uVD in frogs there is head torsion, which follows the same time course as conjugate eye torsion in humans and has been used to monitor drug effects on vestibular compensation.[24]

The offset of the SVH appears to be due to ocular torsion, but the mechanism of the ocular torsion itself is speculative. The conjugate ocular torsion is part of the ocular tilt reaction, which could be considered the equivalent in the otolithic system of the spontaneous nystagmus in the SCC 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.[8] 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.[25,26] Nonetheless, ocular torsion in the absence of spontaneous nystagmus is likely to be otolithic, a conclusion based on the proposition that ocular torsion represents a tonic offset of the dynamic ocular counter-rolling mechanism, which appears to be under utricular control (Fig. 9.4).[4,27]

## Central Vestibular Lesions and Settings of the Subjective Visual Vertical

Patients with acute brainstem[28–31] or cerebellar[32,33] lesions can show offsets of torsional eye position and of the SVH or subjective visual vertical (SVV). 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 (paramedian thalamic infarcts) or with cerebellar lesions involving the nodulus 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. 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 eye can be much larger than the incyclotorsion of the contralesional eye.

## Clinical Significance

Standardized measurement of the SVH, with use of 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.[34] 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, to exclude all visual cues.[29]

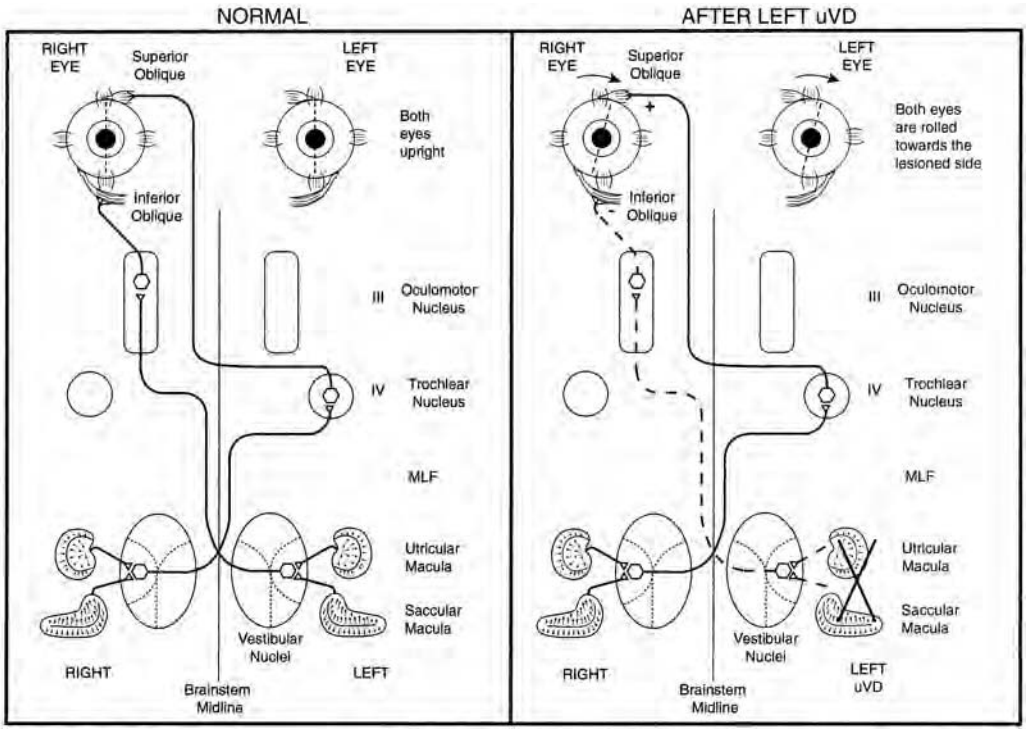



**Figure 9.4** Explanation of the changes in torsional eye position after unilateral vestibular deafferentation (uVD). Normally, second-order afferents from the vestibular nucleus send excitatory projections to the contralateral inferior oblique and ipsilateral superior oblique muscles, as well as to 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, and reduced activity in the ipsilateral superior oblique (and superior oblique, *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.)

A significant offset of the SVH or the SVV indicates acute vestibular hypofunction, possibly otolithic. It indicates a lesion either at the level of the end-organ, vestibular nerve, or vestibular nucleus on the side to which the patient offsets the bar or at a level above the vestibular nucleus on the side opposite to which the patient rotates the bar. Although the greater the deviation of the SVH or SVV, the more acute as well as the more extensive the lesion, a small permanent deviation of the SVV might be a permanent legacy of both central and peripheral vestibular lesions. The SVH test is the single most useful investigation in the acute phase of suspected vestibular neuritis: There is a deviation of the SVH, sometimes by more than 20 degrees, always toward the side of the lesion.[14,15] SVH testing can also be used to follow the progress of vestibular loss and compensation after intratympanic gentamicin treatment for Ménière's disease.[13]

## Vestibular Evoked Myogenic Potential Testing of Otolith Function

### *Physiological Background*

Brief (0.1 msec) loud (>95 db above normal hearing level [nHL]) monaural clicks[35,36] or short tone-bursts[37,38] produce a large (60–300 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). Unlike a neural evoked potential such as the brainstem auditory evoked potential, which is generated by the synchronous discharge of nerve cells, the VEMP is generated by synchronous discharges of muscle cells or, rather, motor units. Being a myogenic potential, the VEMP can be 500 to 1000 times larger than a brainstem potential,[39] 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 VEMP.[40]

The amplitude of the VEMP is linearly related to the intensity of the click and to the intensity of sternomastoid muscle activation during the period of averaging, as measured by the mean rectified electromyography (EMG) value.[36,41] Inadequate sternomastoid contraction produces spurious results by reducing the amplitude of the VEMP (see, for example, Ferber-Viart et al.[42]). A conductive hearing loss abolishes the response by attenuating the intensity of the stimulus (see Halmagyi et al.,[43] Fig. 9). In such cases, the VEMP can be elicited by a tap to the forehead,[44] by a bone vibrator,[45–47] or by a direct current applied to the mastoid bone.[48]

There are two main reasons to suppose that the VEMP arises from stimulation of the saccule. First, the saccule is the most sound-sensitive of the vestibular end-organs,[49,50] possibly because it lies just under the stapes footplate,[51,52] in an ideal position to receive the full impact of a loud click delivered to the tympanic membrane. Second, not only do click-sensitive neurons in the vestibular nerve respond to tilts,[53,54] most originate in the saccular macula[54,55] and project to the lateral and descending vestibular nuclei as well as to other structures (Fig. 9.5).[56,57] The VEMP measures vestibular function through what appears to be a disynaptic vestibulocollic reflex, originating in the saccule and transmitted via the ipsilateral medial vestibulospinal tract to sternomastoid motoneurons.[58]

## Method

Any equipment suitable for recording brainstem auditory potentials will also record VEMPs. Because the amplitude of the VEMP is linearly related to the intensity of both the

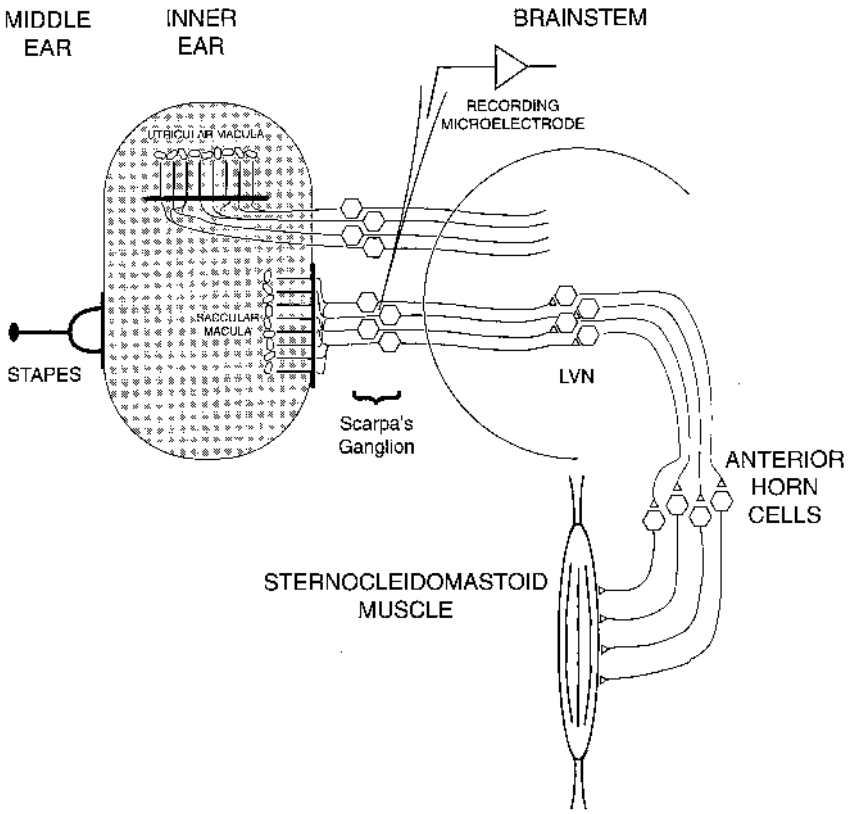



**Figure 9.5** Explanation of the pathways for the click-evoked vestibular myogenic potential (VEMP). The loud click moves the stapes to activate receptors in the underlying saccular macula, which thereby causes, via the vestibulospinal tract, disynaptic activation of ipsilateral C2 segment anterior horn cells supplying the sternomastoid muscle. LVN = lateral vestibular nucleus.

click and of sternomastoid 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 sternomastoid EMG activation is measured. Two reasons why the VEMPs could be absent or less than 50 μV in amplitude are a conductive hearing loss and inadequate contraction of the sternomastoid muscles.

For clinical testing, three superimposed runs of 128 averages 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 sternomastoid 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 sternomastoid 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.[36] One ear is best evaluated through comparison of the amplitude of its VEMP with that of the VEMP 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.[59,60] 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.[61]

## *Clinical Applications*

### Superior Semicircular Dehiscence

A third window into the bony labyrinth allows sound to activate the vestibular system in animals[62,63] and in humans.[64–66] Patients with a bony opening or dehiscence from the superior SCC to the middle cranial fossa (Fig. 9.6) not only have sound- and pressure-induced vestibular nystagmus but also have abnormally large, low-threshold VEMPs.[65–68] In normal subjects the VEMP, just like the acoustic reflex, has a threshold, usually 90 to 95 dB nHL. In patients with the superior SCC dehiscence, the VEMP threshold is about 20 dB lower than in normals (Fig. 9.6) and the VEMP amplitude at the usual 100- to 105-dB stimulus level can be abnormally large (> 300 μV). If a VEMP can be consistently elicited at 70 dB nHL, the patient has a superior SCC dehiscence. Patients with superior SCC dehiscence also have an abnormal large, low-threshold, click-evoked vestibulo-ocular reflex.[69,70]

### Ménière's Disease

In Ménière's disease as well as in delayed endolymphatic hydrops,[71,72] and unlike in acute low-tone hearing

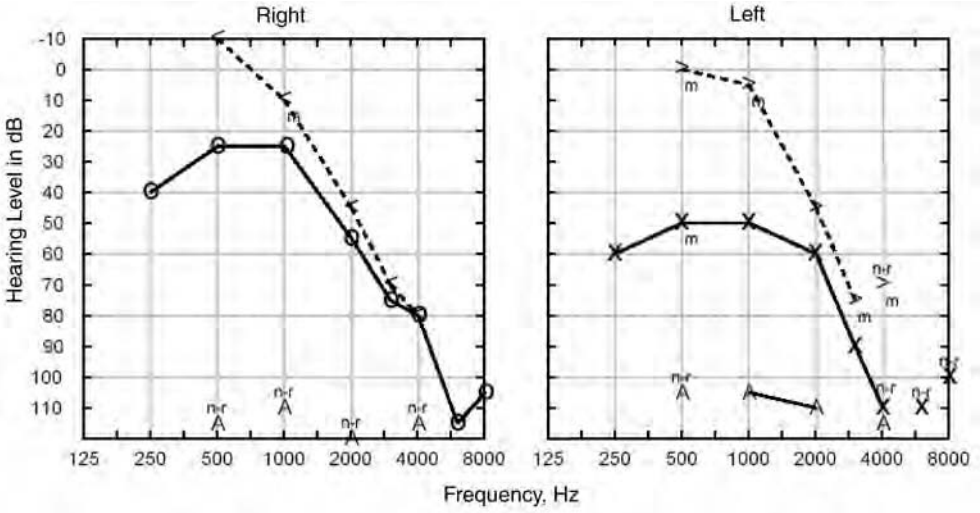



**Figure 9.6** (*A*) Audiogram. Following the left stapedectomy and two revisions, there is still a large air-bone gap at 500 and 1000 Hz on the left as well as on the unoperated right side. The contralateral acoustic reflex, the sound stimulus in the operated left ear and the volume probe in the unoperated right ear, is present at 1kHz 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 due to noise damage. Speech comprehension at 65 dB was 95% on the right and 85% on the left; m = masked; n-r = no response.

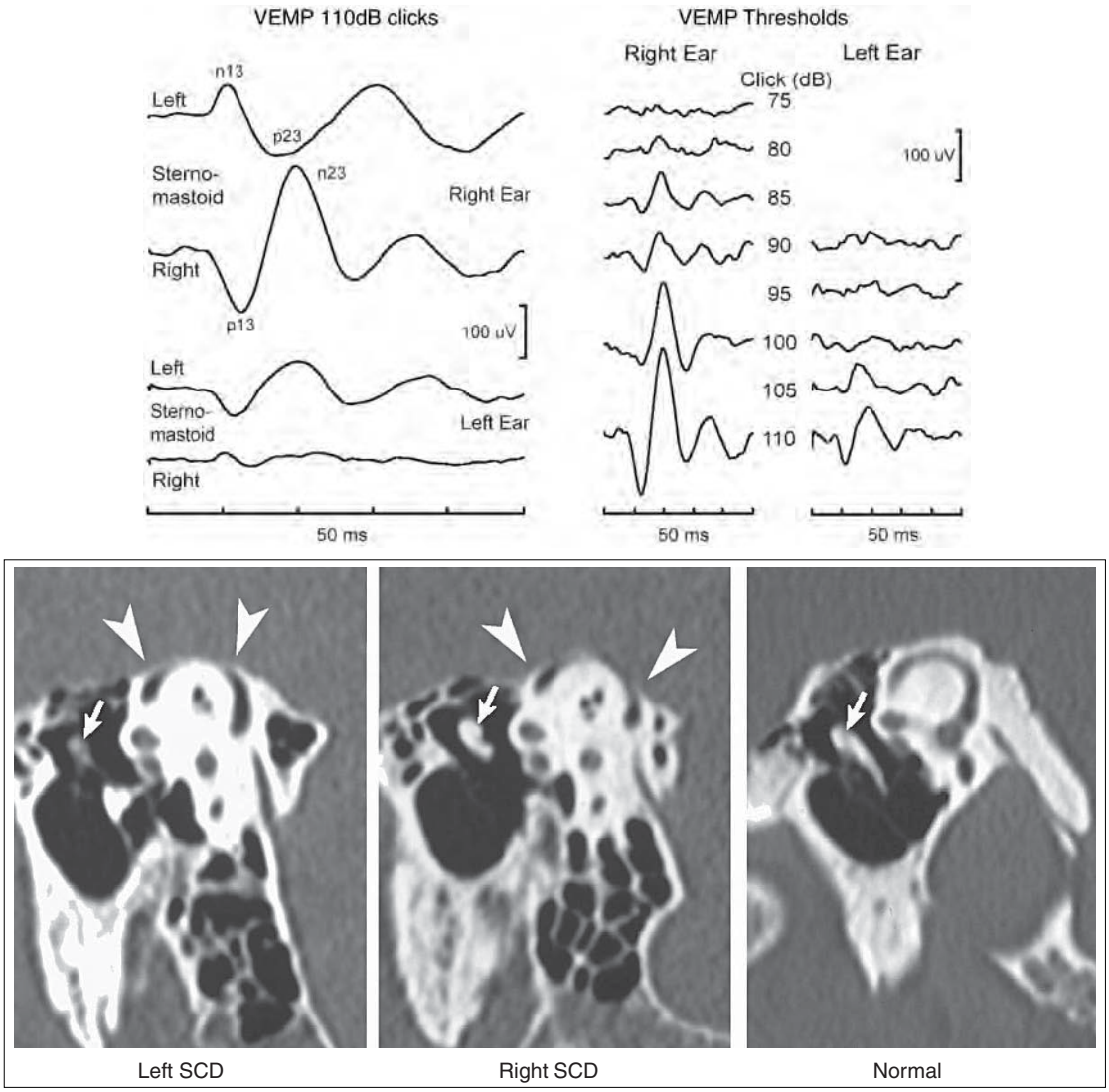



**Figure 9.6** ***(continued)*** (*B)* Vestibular evoked myogenic potentials (VEMPS). *Left,* Despite the significant air-bone gap, p13-n23 VEMP responses to 110 dB clicks are still present. The VEMP from the left ear is normal in amplitude but smaller (115 μV) than from the right (307 μV), indicating that there is some conductive loss on the left (as a result of the 3 operations) as well as 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; [*large white arrowheads*]). CT scan from a normal subject is shown for comparison. The TORP device is clearly seen abutting the oval window on the *left*. The small white arrows show the head of the malleus. (Courtesy of Dr. John Harding-Smith, Central Sydney Imaging).

loss,[73] VEMPs can be abnormal,[74] either too small[75] or too large,[76] or can have altered tuning properties.[77] In some cases, glycerol dehydration can reduce theVEMPs that are too large and enlarge the VEMPs that are too small.[78–80] VEMPs can be used to monitor intratympanic gentamicin therapy in patients with Ménière's disease.[81]

## Vestibular Neurolabyrinthitis and Benign Paroxysmal Positional Vertigo

After an attack of vestibular neuritis, about one patient in three experiences posterior SCC benign paroxysmal positional vertigo (BPPV), usually within 3 months.[82]

The patients who experience BPPV after vestibular neuritis have intact VEMPs, whereas those who do not show absence of VEMP. In other words, an intact VEMP seems to be a prerequisite for the development of post–vestibular neuritis BPPV. The reason could be that in patients who experience post–vestibular neuritis BPPV, only the superior vestibular nerve—which innervates the anterior SCC, lateral SCC, and utricle—is involved. Because the inferior vestibular nerve innervates the posterior SCC and the saccule, the presence of posterior canal BPPV and the preservation of the VEMP imply that the inferior vestibular nerve must have been spared. Support for such an explanation comes from data showing preservation of posterior SCC impulsive vestibulo-ocular reflex in some patients with vestibular neuritis, patients who presumably have involvement of only the superior vestibular nerve.[83,84] The galvanic current–evoked VEMPs are generally abolished in those patients with vestibular neuritis in whom the click-evoked VEMPs are abolished, indicating that the site of lesion is truly in the vestibular nerve rather than, or as well as, in the labyrinth.[85] The VEMP can recover in patients with vestibular neuritis.[86]

## Vestibular Schwannoma

Although most patients with vestibular schwannoma (*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.[87] The VEMP, which is transmitted via the inferior vestibular nerve, is abnormal—of low amplitude or absent—in perhaps four of every five patients with acoustic neuromas.[88–90] Because the VEMP does not depend on cochlear or lateral SCC function, it can be abnormal in the presence of vestibular schwannomas 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 due to neurofibromatosis type 2, loss of VEMP is unrelated to tumor size.[91] The VEMP can be preserved after vestibular schwannoma surgery,[92] suggesting preservation of the saccular nerve.

## Multiple Sclerosis

VEMPs can also be abnormal in diseases affecting central vestibular pathways,[93] especially white-matter diseases such as multiple sclerosis,[94–97] which affect the medial vestibulospinal tract, the continuation of the medial longitudinal fasciculus, a site commonly involved by demyelination.

## Other Conditions

Initial experience with this technique suggests that the VEMP test can provide valuable information additional 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.[47] 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. The following case study describes a typical example.

### CASE STUDY 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 as well as 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. 9.7*A*).

*Electronystagmogram:* Minimal left-beating gaze-evoked nystagmus in the dark; normal caloric test results (Fig. 9.7*B*).
*Subjective visual horizontal:* More than 6 degrees to the left (Fig. 9.7*C*).
*Vestibular-evoked myogenic potentials:* Absent from left ear to clicks and to taps (Fig. 9.7*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 due to 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 VEMP values both returned to normal.

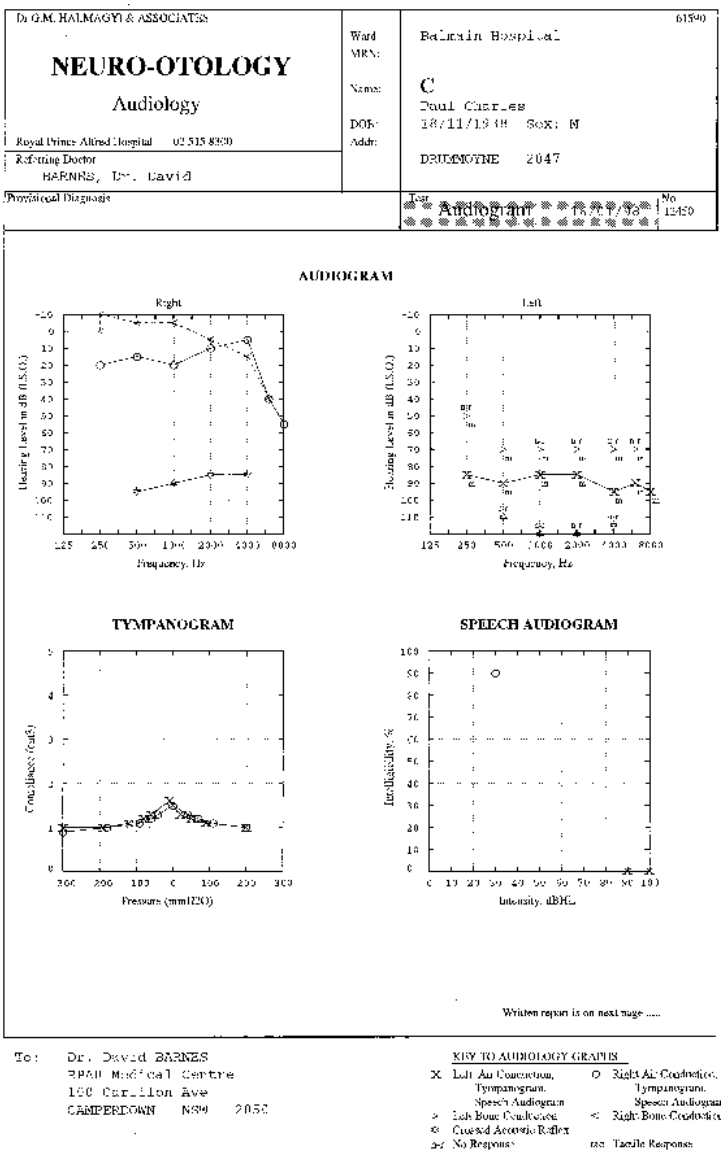



**Figure 9.7** (*A*) Audiogram from a 61-year-old man with left neurolabyrinthitis. There is a severe sensorineural hearing loss on the left with absence of acoustic reflexes and zero speech discrimination, all findings suggestive of a retrocochlear loss. On the right there is a slight conductive loss, which is asymptomatic and unrelated to the present problem.

*(continued on following page)*

## CASE STUDY 1 *(continued)*

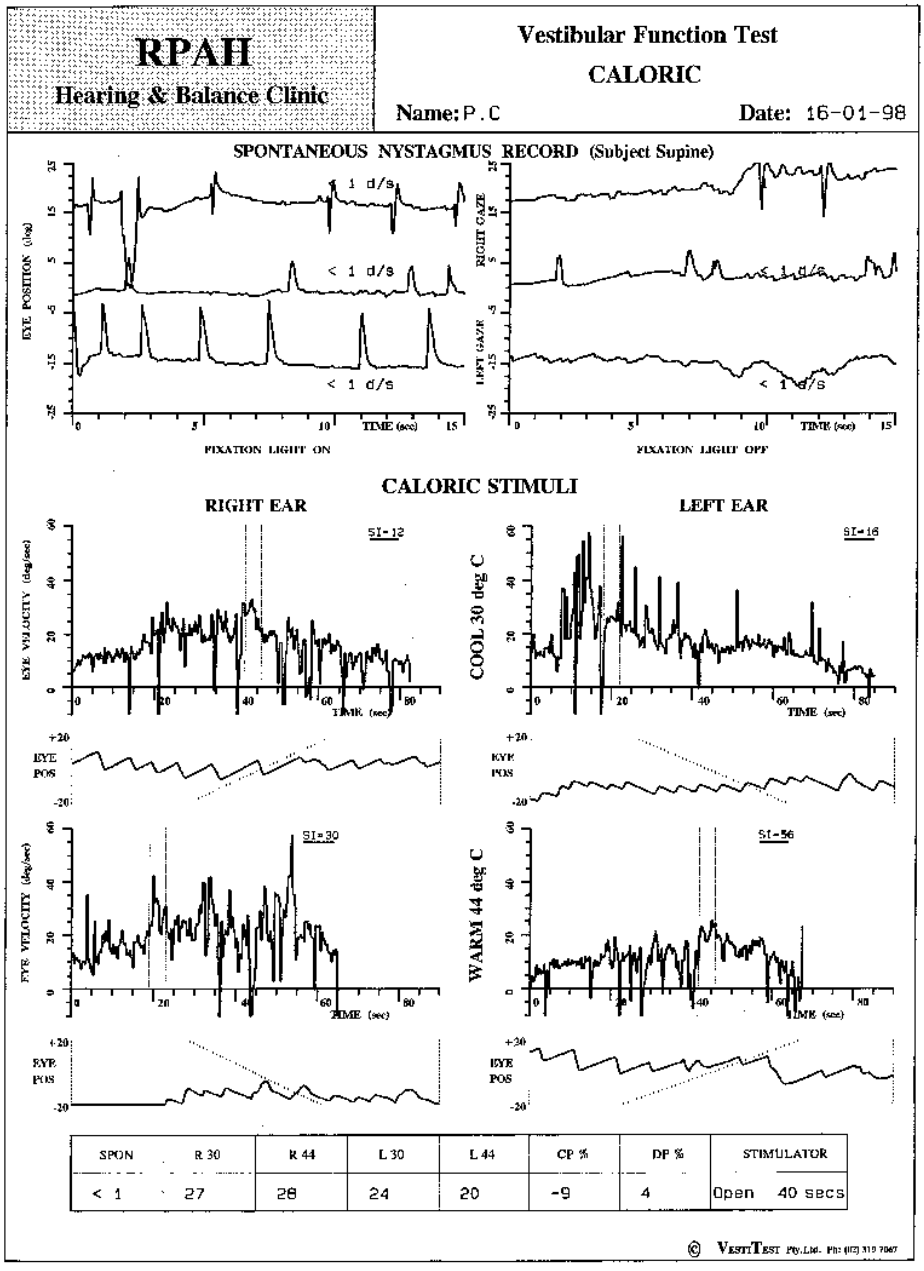
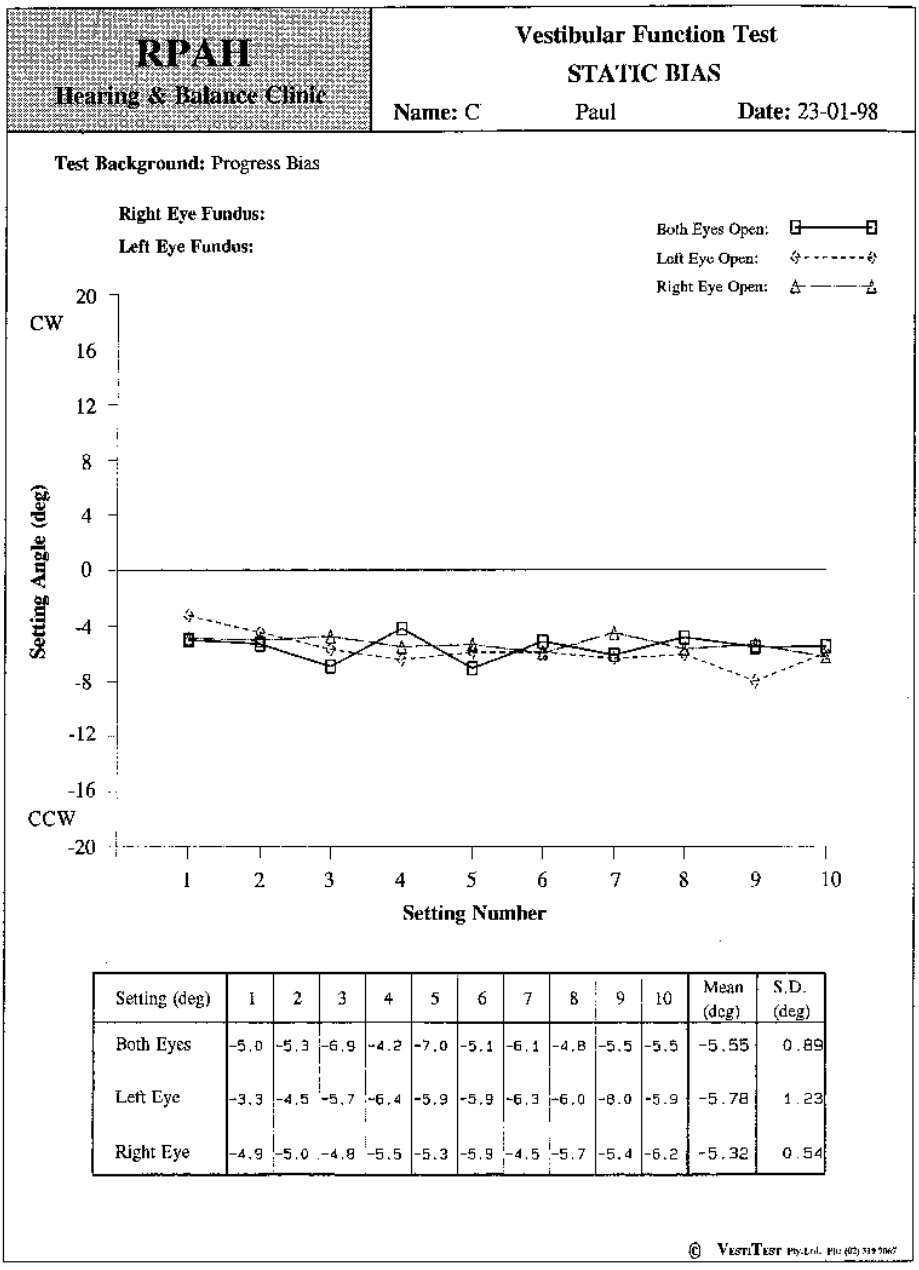



**Figure 9.7** ***(continued)*** (*B*) Electronystagmogram and caloric test from the same patient. There is minimal left-beating gaze-evoked nystagmus in darkness (slow-phase velocity $< 1$ deg/sec). Bithermal caloric tests show symmetrical slow-phase velocities from each ear. These findings indicate normal lateral semicircular canal function.




| Setting (deg) | 1 | 2 | 3 | 4 | 5 | 6 | 7 | 8 | 9 | 10 | Mean (deg) | S.D. (deg) |
|---|---|---|---|---|---|---|---|---|---|---|---|---|
| Both Eyes | -5.0 | -5.3 | -6.9 | -4.2 | -7.0 | -5.1 | -6.1 | -4.8 | -5.5 | -5.5 | -5.55 | 0.89 |
| Left Eye | -3.3 | -4.5 | -5.7 | -6.4 | -5.9 | -5.9 | -6.3 | -6.0 | -8.0 | -5.9 | -5.78 | 1.23 |
| Right Eye | -4.9 | -5.0 | -4.8 | -5.5 | -5.3 | -5.9 | -4.5 | -5.7 | -5.4 | -6.2 | -5.32 | 0.54 |

**Figure 9.7** ***(continued)*** (*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 on following page)*

## CASE STUDY 1 *(continued)*

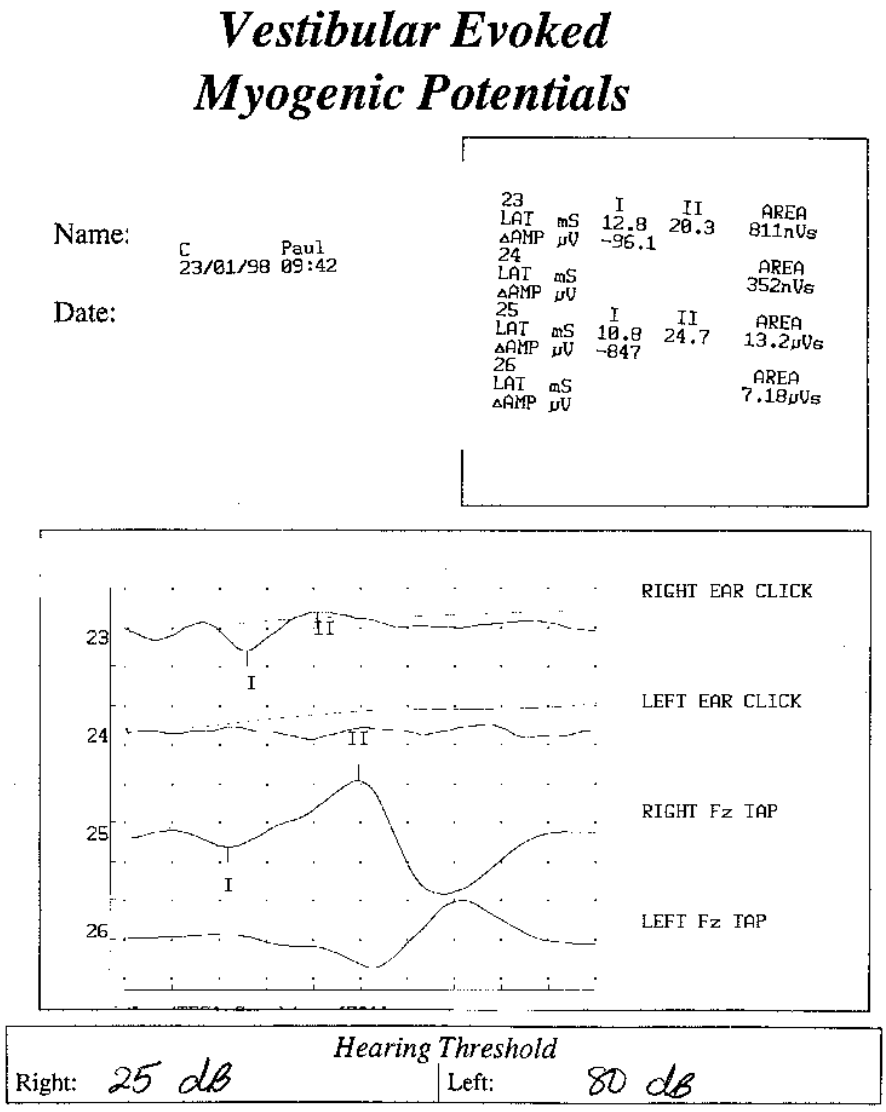



**Figure 9.7** ***(continued)*** (*D*) Vestibular evoked myogenic potentials (VEMPs) from the same patient. Channels 23 and 24 show the responses to 95-dB clicks; channels 25 and 26 show the response to forehead taps. The responses are absent from the left. The absence of tap responses shows that the absence of click response is due not to some conductive hearing loss in the left ear (that is being masked by the severe sensorineural loss), but due to loss of saccular function. The VEMP response to clicks has nothing to do with the audibility of the stimulus—it is present even if the click is not heard.

## 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 5).

## 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.

## Acknowledgments

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CHAPTER 10

# Auditory Examination

*Sharon Polensek, MD, PhD*

Many patients presenting to a clinician with a possible vestibular disorder have underlying auditory impairment. Because the ear houses the sensory organs for auditory as well as vestibular input, many otologic disorders (e.g., Ménière's disease, perilymphatic fistula, labyrinthitis, acoustic neuroma), not surprisingly, give rise to both auditory and vestibular symptoms. An understanding of the clinical methods of assessment and management of hearing loss is essential 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 the hearing acuity of any patient presenting with possible vestibular impairment, especially if audiological testing has not yet been performed. Obtaining a formal audiological assessment in 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 he or she frequently needs 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 of 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. 10.1*A*). If sound perception lateralizes to one side, then hearing loss is present (Fig. 10.1*B*).

To determine which ear has the loss, the examiner can perform a Rinne test with the fork. 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. If the patient can still hear the tone when the fork is placed by the ear canal, the result is normal (Fig.10.2). If the Rinne test result is abnormal on the side where the tone perception lateralized with the Weber test, the patient likely has a conductive hearing loss in that ear. On the other hand, if

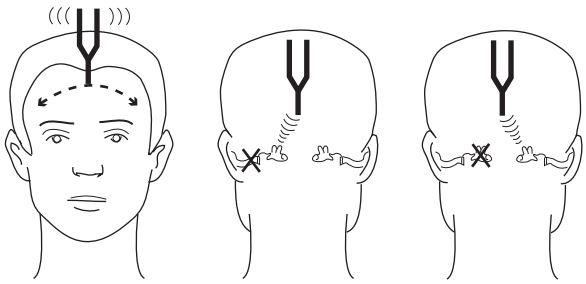

**Figure 10.1** The Weber tuning fork test. The sound of the vibrating tuning fork is perceived (*A*) in the ear with a conductive hearing loss or (*B*) in the ear contralateral to the ear with a sensorineural loss.

the Weber test result is abnormal and the Rinne test yields a normal result, the patient probably has a sensorineural hearing loss (SNHL) 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 in the evaluation of 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 of 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 a caloric test is performed 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 situation.

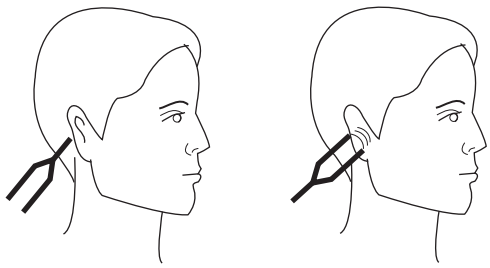

**Figure 10.2** The Rinne test. See text for explanation.

# 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 in 50% of presentations.[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. 10.3). 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 by means of 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 skull bony prominence, 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.

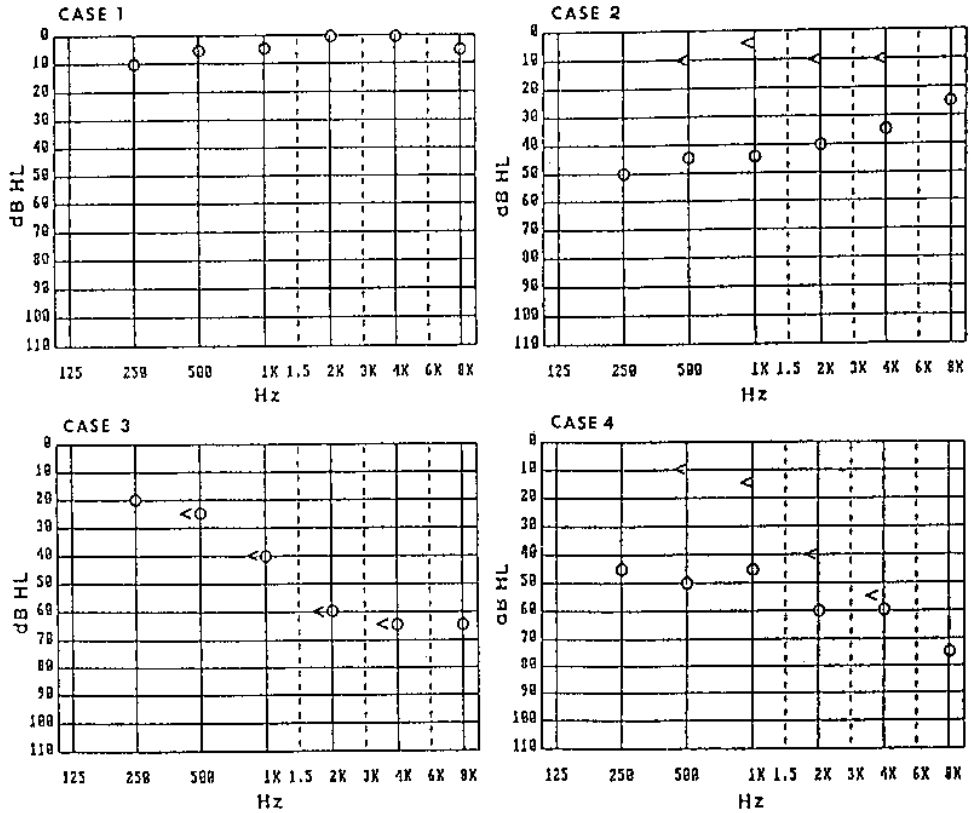


**Figure 10.3** Types of audiograms. *Case 1*, normal hearing; *case 2*, conductive hearing loss; *case 3*, sensorineural hearing loss; *case 4*, mixed type hearing loss. o = air conduction; < = bone conduction.

## 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 case 2 of Fig. 10.3. The presence of an air-bone gap indicates that the hearing loss may be due to a problem in either the outer or middle ear. Such a hearing loss is called a *conductive loss*. Causes of conductive losses include fluid in the middle ear, disruption or diminished mobility of the ossicular chain, and abnormal tympanic membrane compliance due to 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! Disorders that may cause 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 (SCC) 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, no air-bone gap is present, and the loss is of sensorineural origin (Fig. 10.3, case 3). A *sensorineural loss* (SNHL) is therefore due to a problem either in the cochlea, or further proximal in the auditory nerve or central nervous system. Common causes of SNHLs are 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, including meningitis, congenital syphilis, toxoplasmosis, cytomegalovirus, rubella, and herpes. Birth complications such as hypoxia, prolonged mechanical ventilation, and low birth rate also increase the risk for development of sensorineural loss.

When BC thresholds show hearing loss and AC thresholds indicate an air-bone gap, a *mixed-type loss* is present, meaning the loss has both conductive and sensorineural components (Fig. 10.3, case 4). Mixed-typed losses may be seen in advanced otosclerosis, severe chronic otitis media, and some genetic forms of auditory impairment.

In addition to classification as conductive, sensorineural, or mixed, hearing losses can also be further classified according to severity, the frequencies predominantly affected, or the threshold shape on the audiogram.

The severity of the hearing loss may be described as mild, moderate, moderately severe, severe, or profound according to the amount of threshold elevation above normal range (Table 10-1). In general, hearing thresholds are considered normal in adults if less than 20 dB HL. For children younger than 18 years, thresholds less than 15 dB HL are considered within normal limits. Hearing loss is considered mild if thresholds are less than 40 dB, moderate if 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 SNHLs 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, noise trauma leads to hearing loss in which sensitivity to 4000 Hz is most affected. This kind of hearing loss forms a V-shape on the audiogram, or what is sometimes referred to as a "4 KHz notch." Individuals with high-frequency hearing loss have 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 SNHLs are typically seen in patients with Ménière's disease and some forms of inherited deafness. An audiogram may also be described as being "U-shaped" or "cookie-bite" shaped. Thresholds in this type of audiogram form a U-shape when a hearing loss with either the least or most severely affected frequencies is in the mid-portion of the hearing range.

■ Table 10-1 **CLASSIFICATION OF HEARING LOSS**

| Pure-Tone Audiogram Result (dB) | Classification |
|---|---|
| <15 | 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 |

## 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* (SDT). The SDT should closely coincide with the average threshold of hearing sensitivity to pure tones at 500, 1000, and 2000 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 higher 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* (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 varies according to the number and type of words in the list, and normative ranges are available for the particular 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 of the eardrum 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 commonly 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, and hence, the compliance will remain constant at all pressure levels, thereby giving rise to a flat tympanogram (without a peak).

For routine immittance testing, there are several types of tympanograms, as depicted in Figure 10.4. 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 greater than normal. The $A_d$ pattern is typical for ossicular chain disruption or for a flaccid eardrum. The $A_{shallow}$ 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 from otosclerosis, tympanic membrane thickening, or 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 for approximately 10 seconds in response 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 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 in either the ipsilateral or contralateral ear.

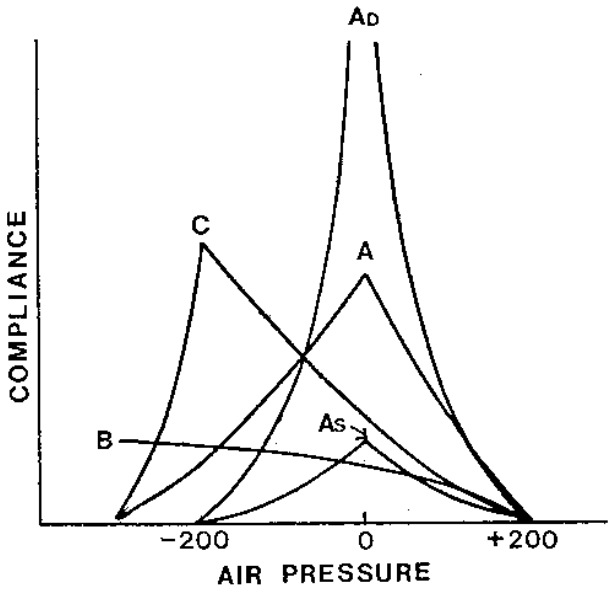


**Figure 10.4** Types of tympanograms. See text for explanation.

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.[3] 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 for the fitting of appropriate hearing aids.

Middle ear disease will inhibit the acoustic reflex as well, 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 differentiate impairment of the VIIth or VIIIth 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 presence of normal hearing threshold measurements should raise suspicion of VIIth cranial nerve disease. 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 in a patient whose ipsilateral reflexes are intact may in indicate brainstem disease.

The acoustic reflex threshold should never be less than the pure-tone threshold. Such findings, when they occur, are most likely due to either a nonorganic hearing loss or a technical error.

## Auditory Brainstem Response

Measurement of the auditory brainstem response (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 in patients unable to participate in routine audiometric testing, such as infants or the cogni-

tively impaired, and documentation of an audiological threshold in those feigning hearing loss. Information about the site of a lesion along the auditory pathway may be inferred from the ABR, because 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.

To elicit the ABR, both clicks and brief frequency-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 1000 to 3000 sweeps are averaged to obtain the ABR.

The morphology of the ABR is shown in Figure 10.5*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 VIIIth nerve in the cerebellopontine angle (CPA); 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, waves III through V have bilateral crossed inputs and no longer represent solely an ipsilateral response. Response latencies normally increase as stimulus amplitude decreases, but interwave intervals remain relatively constant regardless of stimulus intensity; hence, wave I–V and III–V intervals are routinely measured.

Norms are available for waveform response latencies, and abnormally long latencies or widened interwave intervals may be seen with lesions of the auditory nerve or brainstem. An example of abnormal ABR waveform from a patient with a left-sided acoustic neuroma is shown in Figure 10.5*B*.

## Otoacoustic Emissions

*Otoacoustic emissions* (OAEs) are sounds that are generated from healthy cochlear outer hair cells. Outer hair

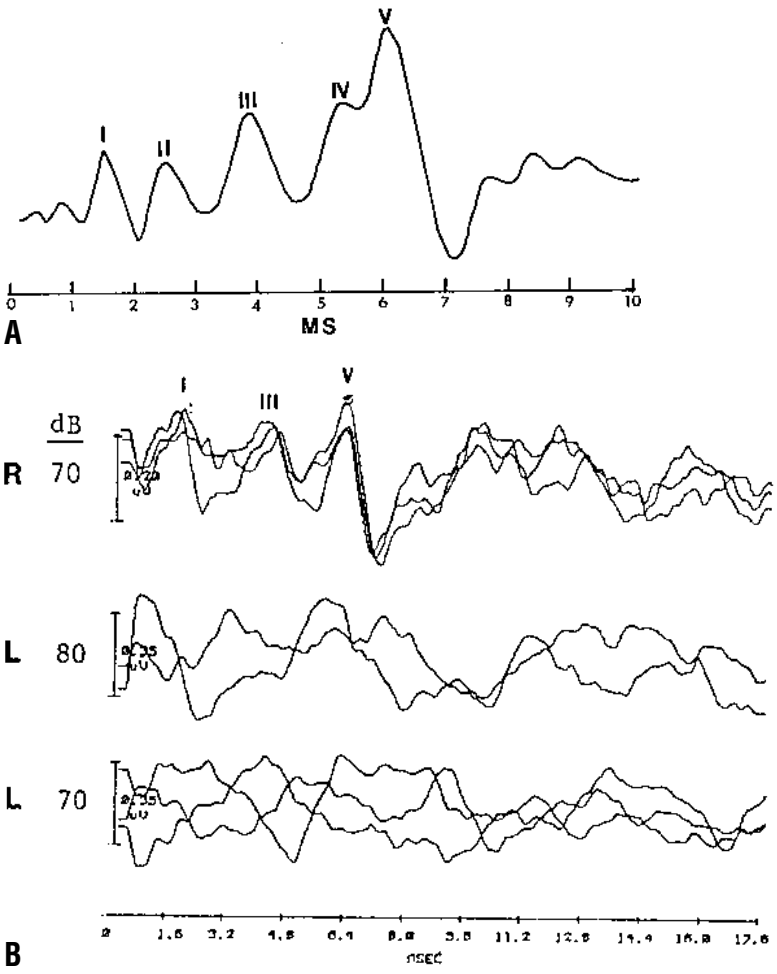


**Figure 10.5** (*A*) Basic morphology of auditory brainstem response (ABR). Wave I corresponds to the cochlea, acoustic nerve; wave II to the acoustic nerve, cochlear nucleus; wave III to the superior olivary complex; wave IV to the lateral lemniscus; and wave V to the inferior colliculus. (*B*) ABRs obtained from a 46-year old patient with a surgically confirmed acoustic neuroma on the left side. She had a moderate sensorineural hearing loss in the left ear. Repeated ABR testing on the left side at 70 dB and 80 dB normalized hearing level showed no discernible responses, but clear responses were obtained when the right ear was stimulated.

cells are motile and are thought to function as an amplifier of cochlear partition displacement during acoustic stimulation and, in so doing, generate acoustic byproducts, 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 remain stable over several years for any one ear. OAE measurements 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 emissions (SOAEs), which occur without acoustic stimulation of the ear, and evoked otoacoustic emissions (EOAEs), 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 EOAEs 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 OAEs 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.

## Electrocochleography

*Electrocochleography* refers to the measurement of neuroelectric events generated by the cochlea and auditory nerve in response to acoustic stimulation. 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. 10.6). The cochlear microphonic mimics the waveform of the sound stimulus, and the shift in its baseline (or direct current [DC]) is called the *summating potential*.

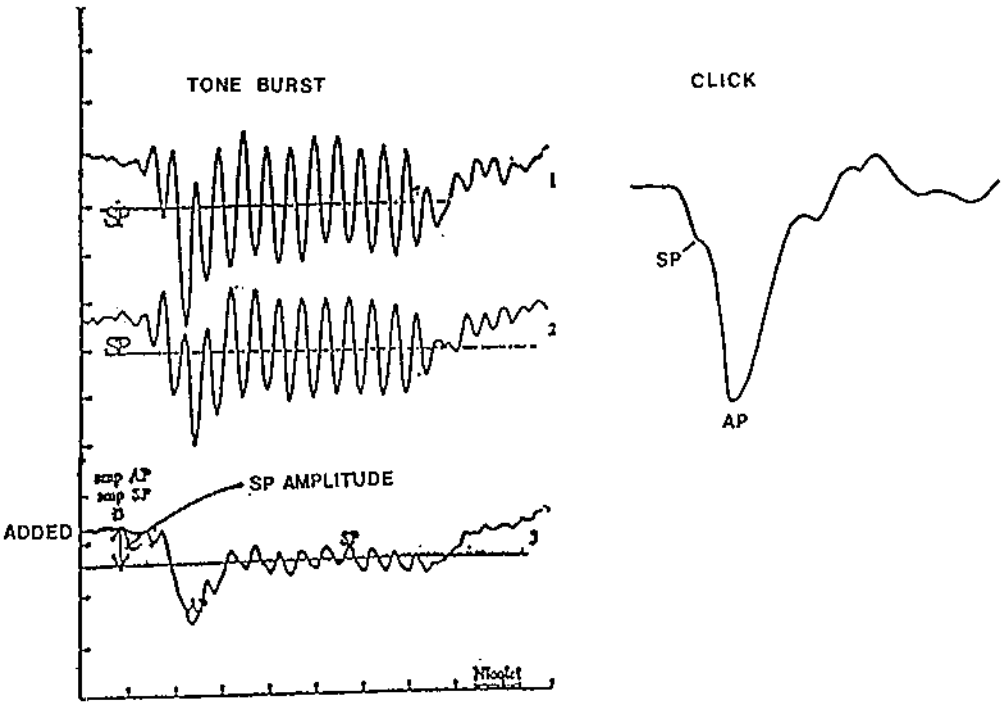


**Figure 10.6** Electrocochleograms from the normal ear.

The AP corresponds to wave I of the ABR. Ideally, electrocochleography is recorded with an electrode placed as close as possible to the round window of the cochlea. A transtympanic needle electrode referenced to another electrode placed on the forehead or tragus may be used, 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 evaluate for 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-to-AP ratio normally ranges from 10% to 50%, with higher ratios in many patients with Ménière's disease.

## *Audiological Management*

### Hearing Aids

The majority of individuals with hearing loss are treated with hearing aids (Fig. 10.7). Selection of proper amplification systems for hearing loss depends on many factors, including type and extent of the loss and whether it is bilateral and symmetric. In 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). Developments in hearing aids have allowed high-powered aids for more severe hearing losses to be smaller and less conspicuous. The smallest hearing aids are worn entirely in the ear canal (ITC), so that they are minimally visible at all, 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 their power supply may be larger, body aids 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 patients with vision or dexterity limitations that 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 utilize an oscillator that contacts skin overlying the mastoid bone, leading to transmission of sound via bone conduction and bypassing the middle and outer ear. They may be worn either attached to a headband or eyeglasses or like 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 that is surgically inserted into mastoid bone. It has a separate directional microphone and detachable external sound processor.

Lastly, some hearing aids are specifically manufactured to benefit those with unilateral hearing losses. The CROS (contralateral routing of signal) 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.

Traditional hearing aids receive sounds and convert them either into electrical signals, which are then amplified (analog conversion), or to digital signals. Digital hearing aids are more expensive but have the advantage of

**Figure 10.7** Types of hearing aids. *Left,* In-the-ear; *center,* behind the ear; *right,* in-the-canal.

being able to selectively amplify certain frequencies to suit an individual's needs and hearing loss characteristics.

### Aural Rehabilitation

*Aural rehabilitation* involves the administration of services to help people adjust to their hearing loss and make the best use of hearing aids. Exploring possible assistive devices and providing education about 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.

## Medical Testing in the Evaluation of Hearing Loss

The majority of hearing losses may be attributed to genetics, may be age-related, or both, and may be optimally treated with 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 must be able to recognize common medical causes underlying hearing impairment and, in addition, may need to maintain a low threshold of suspicion for uncommon, yet potentially treatable, causes of hearing loss.

### *Laboratory Testing*

Some systemic disorders manifest as hearing loss, such as autoimmune or infectious disease. For patients presenting with SNHL, either the fluorescent treponemal antibody absorption test (FTA-ABS) or the microhemagglutination test for *Treponema pallidum* (MHA-TP) should be performed, particularly because hearing loss related to syphilis is potentially treatable. Routine screening for autoimmune disorders is not routinely warranted, however, unless disease is suggested by information obtained from the history and physical examination. In particular, if other cranial neuropathies are present, lumbar puncture is warranted to perform the Venereal Disease Research Laboratory (VDRL) test for neurosyphilis, and also to evaluate cerebrospinal fluid (CSF) for cytology, cell count, and levels of glucose and protein, as well as to perform cultures. In endemic geographic areas, serum Lyme antibody titers should be checked. If the history or physical findings are otherwise suggestive of systemic disorders, serological tests for possible autoimmune causes, such as erythrocyte sedimentation rate measurement and tests for angiotensin-converting enzyme (ACE) level and presence of rheumatoid factor, cytoplasmic antineutrophil cytoplasmic antibody (c-ANCA), perinuclear ANCA (p-ANCA), and antinuclear antibody (ANA), may be helpful.

### *Radiological Imaging*

Radiography is warranted in selected patients with hearing loss. Magnetic resonance imaging (MRI) with gadolinium enhancement is currently the "gold standard" for evaluation of hearing losses of possible retrocochlear origin and particularly in making the diagnosis of a tumor of the VIIIth cranial nerve or in the cerebello-pontine angle (CPA) region. Computed tomography (CT) is useful in patients with suspected labyrinthine congenital anomalies, such as large vestibular aqueduct syndrome and Mondini dysplasia. CT also is useful in cases with suspected labyrinthine fistula or temporal bone fractures.

## Clinical Presentations of Auditory Impairment

### *Sudden Sensorineural Hearing Loss*

The cause of sudden sensorineural hearing loss (SSNHL) is not completely understood and has been hypothesized to be due to a variety of causes: hypoxia, intralabyrinthine membrane rupture, and viral, vascular, inflammatory, or metabolic disorders, among others. SSNHL is considered an otologic emergency because accepted guidelines for treatment are to institute pharmacological therapy within 2 weeks to achieve maximum benefit. Many physicians use as criteria for SSNHL 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 audiogram, age extreme, 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 (reviewed in reference 4).

Oral corticosteroids have been shown to be effective in some cases,[5] and a 2005 study recommends giving prednisone 60 mg/day for 14 days.[6] A review of the literature concluded that intratympanic corticosteroid treatment for patients in whom oral steroid therapy fails can be only weakly recommended.[7] Addition of an antiviral

agent to steroid treatment did not improve hearing outcome in one study,[8] but 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 caused by the degradation of the tissue-fluid environment within the inner ear by either 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 diagnosed clinically when patients present with a relatively sudden onset of SNHL 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 cause of deafness associated with meningitis.

Measles and mumps rarely cause hearing and vestibular loss in developed countries because of widespread vaccination against these diseases. 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 this condition is typically seen in an immunocompromised patient, such as one with 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 such vesicles are accompanied by an ipsilateral facial weakness, the condition is referred to as *Ramsay Hunt syndrome,* which is caused by a herpes virus. Facial paralysis is most commonly seen, but 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 rise with advancing age in association with an age-related decrease in cellular immune response to herpes zoster virus.[9]

### Syphilis

The incidence of hearing loss is as high as 80% in cases of symptomatic neurosyphilis, and 29% patients with asymptomatic neurosyphilis.[10] 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 due to syphilis is frequently indistinguishable from that in Ménière's disease, because it may fluctuate and may 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

Rapidly progressive SNHL 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 is confirmed by serologic titers. Treatment is with broad-spectrum antibiotics.

### Lyme Disease

Lyme disease is caused by the tickborne 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 cause in endemic areas. Recommended antimicrobial therapy for Lyme disease with neurological manifestations is with third-generation cephalosporins, as long as the patient is not allergic to these agents.

## *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 due to ototoxic drug therapy.

### Aminoglycoside

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 damage.[11] Hearing loss may be asymmetric and may 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 treatment, advanced age, and combined administration of other ototoxic drugs, particularly loop diuretics.[12]

### Aspirin

Aspirin toxicity may manifest as tinnitus and reversible SNHL, 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.[13] Caloric responses can also be reduced by salicylates.[14] Nonsteroidal antiinflammatory drugs (NSAIDs), such as naproxen, ketorolac, and piroxicam, have been known to cause reversible SNHL, but this side effect occurs far less commonly 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, virtually all patients who have received cisplatin probably will be found to have at least some degree of ototoxicity.[15] Occasionally the hearing loss is accompanied by vertigo or tinnitus. Other chemotherapeutic agents known to cause ototoxicity are 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.[16] 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 are polymyxin B, propylene glycol, acetic acid, and antifungal agents.[17,18]

### Other Drugs

Other drugs that have been reported to cause hearing loss are 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 SNHL, tinnitus, and visual disturbances.[12] 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 problem can frequently be surgically corrected by clearing the middle ear cavity of the structural disease, by replacement of the ossicles with a prosthetic device, or both. Disruptions in the tympanic membrane can also be treated with a tympanoplasty procedure. Rarely do causes of conductive hearing loss without a sensorineural component give rise to vestibular symptoms.

In contrast to management for conductive hearing loss, surgical treatment for SNHL is considerably less common, but notable approaches include cochlear implants and interventions for CPA tumors, SCC dehiscence, and other perilymphatic fistulas (PLFs). CPA tumors and PLFs commonly give rise to vestibular symptoms, and patients with cochlear implants may have vestibular as well as hearing loss; thus, clinicians managing vestibular disorders may not infrequently encounter patients with these lesions.

### *Cochlear Implants*

Cochlear implants are an option for a growing 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 SNHL took place only in the past four decades. A deafened auditory nerve

was first electrically stimulated by Djourno and colleagues[19] in 1957, but it was not until 1972 that a commercial device was developed for this purpose. Traditional criteria for implantation include bilateral profound-to-total SNHL, inability to benefit from conventional hearing aids, good physical and mental health, and the motivation and patience to complete a rehabilitation program.[20] More recently, cochlear implants have been advocated as treatment for hearing loss associated with enlarged vestibular aqueduct syndrome.[21,22] Implantation criteria continue to broaden as studies are now under way exploring simultaneous usage of acoustic hearing aids with short-electrode cochlear implants in the same ear.[23]

All cochlear implants have several elements in common (Fig. 10.8*A*). A microphone, usually at ear level, detects acoustic energy, which is encoded into an electrical signal by the external sound processor. The electrical stimulus is transmitted to the implanted electrode array either in the middle ear or inner ear through some form of signal coupler (Fig. 10.8*B*). The most commonly used


**Figure 10.8** (*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.)

commercially available implants in the United States are those with 22 active electrodes.

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 growing numbers of patients are found to benefit from the procedure. The rehabilitation process is still lengthy, however, because it involves adjustments of the speech processor and extensive aural training.

## Cerebellopontine Angle Tumors

Although the acoustic neuroma (acoustic neurinoma, vestibular schwannoma, acoustic neurilemoma) is the most common lesion seen in the CPA, a variety of other benign and malignant lesions may be found in this region. Patients with such lesions may also present with vertigo, unsteadiness, headache, twitching, and weakness or numbness of the face. Unilateral progressive SNHL is the most common first symptom, and predominantly high-frequency SNHL 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.[24]

Brainstem evoked response audiometry is the single most reliable audiometric diagnostic procedure in the diagnosis of CPA lesions. Specificity and sensitivity of the ABR range from 92% to 96% in the diagnosis of acoustic neuromas approximately 1 cm in diameter.[25] Vestibular hypofunction is demonstrated in 82% to 96% of patients with CPA lesions,[26] and positional nystagmus is a very common finding.[27] MRI of the brain with gadolinium contrast enhancement provides a reliable method of diagnosing acoustic neurinomas, which may be purely within the internal auditory canal, and is particularly useful for monitoring tumor growth and postoperative recurrence.

The acoustic neuroma arises most commonly from the superior division of the vestibular nerve, and next most commonly from the inferior division. In rare cases it arises from the cochlear nerve. 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, the most appropriate management in patients who are otherwise poor operative candidates may consist of merely monitoring the lesion. Patients with serviceable hearing who have unilateral tumors less than 2.0 cm in diameter may be offered either surgical removal with an attempt to save hearing or radiographic monitoring, surgery being reserved for an enlarging tumor. For small tumors in patients 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 younger than 65 years is likely. For large tumors in older patients without evidence of brainstem compression, radiographic monitoring is recommended, with surgery reserved for enlarging tumors and progressive symptoms. In patients with NF2 and bilateral tumors, the goal is to avoid bilateral deafness and vestibular hypofunction, so if hearing is good in both ears, one should proceed with surgical excision of one tumor in the attempt to preserve hearing.[28]

## Superior Semicircular Canal Dehiscence

SCC dehiscence is caused by a disruption in the bony capsule of the temporal bone overlying the superior SCC that renders the vestibular labyrinth sensitive to loud sounds or pressure changes. The condition is probably congenital because it is frequently bilateral, but it is sometimes asymptomatic. Diagnosis can be made through demonstration of characteristic torsional nystagmus induced by application or positive or negative ear canal pressure or by presentation of loud sound.

Frequently, patients with SCC dehiscence first present with a conductive hearing loss. Moreover, such patients 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 SCC.[29,30]

## Perilymphatic Fistula

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.[31] The onset of PLF commonly manifests as sudden onset of 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 with PLFs complain of tinnitus, regardless of hearing status. Hearing loss in present in 53% of patients and usually does not fluctuate. Moreover, patients do not typically present with episodic vertigo, tinnitus, or aural fullness, features that 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. When present, physical findings include positional nystagmus occurring in several head positions, but especially with the suspect ear down. Hennebert's sign—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—may be present with PLF. This sign may be secondary to stimulation of the utricular macula as a result of deformation of the utricular wall by the pressure change.

No specialized audiometric or electronystagmography result, including caloric testing, has been of any value in identifying PLF or the side of the lesion. Black and coworkers[32] reported posturography as being a highly specific and sensitive test for PLF, but this finding has not been corroborated well by others.[33] Other than rarely documenting intralabyrinthine air in association of perilymphatic leakage, neither CT nor MRI can positively identify a PLF.

In animals, it has been shown that most PLFs heal spontaneously, and some physicians choose nonoperative management for at least several days, allowing time for complete evaluation as well as possible spontaneous recovery. Conservative management comprises bedrest with the head of bed elevated to 30 to 40 degrees and use of a stool softener. Lifting, stooping, nose blowing, and 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 unlike PLF secondary to superior SCC dehiscence, which may be more readily diagnosed, PLF of the oval or round window may not always be identified, even intraoperatively, unless an obvious anatomical malformation is present.

### *Other Causes of Hearing Loss*

Many other disorders may exhibit hearing loss as a symptom (Table 10-1), 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 almost any intracranial process that disrupts the central auditory pathways. Paget's disease (osteitis deformans), although a rare disorder of bone, commonly manifests as hearing loss and can be treated with calcitonin or etidronate disodium.[34] Rarely, hearing loss may accompany blood dyscrasias, AIDS, 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 which are pertinent in the evaluation of this patient population are discussed, and standard audiological testing procedures and their indications are reviewed. The clinical presentations of the more common etiologies of hearing loss are outlined because of their relevance to the clinician who assesses and treats patients with vestibular disorders—the pathophysiology underlying vestibular disease also frequently gives rise to auditory symptoms. Familiarization with the clinical presentation and audiological features of 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 appropriate management and therapy.

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# T·H·R·E·E

# Medical and Surgical Management

CHAPTER 11

# Pharmacological and Optical Methods of Treatment for Vestibular Disorders and Nystagmus

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Patients with vestibular disease may complain of vertigo, oscillopsia, or the visual consequences of nystagmus.[1] *Vertigo* consists of the illusion of turning and implies vestibular imbalance. *Oscillopsia* consists of illusory, to-and-fro movements of the seen environment; when it occurs with head movements, it usually implies bilateral loss of vestibular function. Patients with spontaneous *nystagmus*, due to vestibular or other processes, may also complain of oscillopsia when their heads are still. In this chapter, treatments for vertigo, oscillopsia, and the visual consequences of nystagmus are reviewed, with an attempt to base therapies on known pathophysiology.

In trying to understand and treat the symptoms resulting from labyrinthine disorders, one must bear in mind 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 the head perturbations occurring during locomotion. This fact was first pointed out by the anonymous physician J.C.,[2] who had lost vestibular function due to aminoglycosides; 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."

Figure 11.1 summarizes the peak velocities and predominant frequencies of head rotations measured in 20 normal subjects as they walked or ran in place. Note that although peak head velocity is generally below 150 degrees per second (deg/sec), the predominant frequencies range from 0.5 to 5 Hz.[3] The latter value exceeds the frequencies that vestibular physiologists have conventionally used to test patients with vestibular disorders, except for the head-impulse test,[4] which has gained in popularity. Furthermore, in the design of exercises to rehabilitate patients with vestibular disorders, strategies should be applied to use head movements that contain these sorts of frequencies, which mainly result from transmitted heel-strike. Thus, in thinking about methods to improve vestibular symptoms, one must identify the functional goals for which the patient is aiming and so determine the physiological demands that will be made of the vestibular system to achieve them.

## Vertigo

### *Pathophysiology of Vertigo*

As distinct from one's perception of self-motion during natural locomotion, vertigo is a distressing, illusory sensation of turning that is linked to impaired perception of a

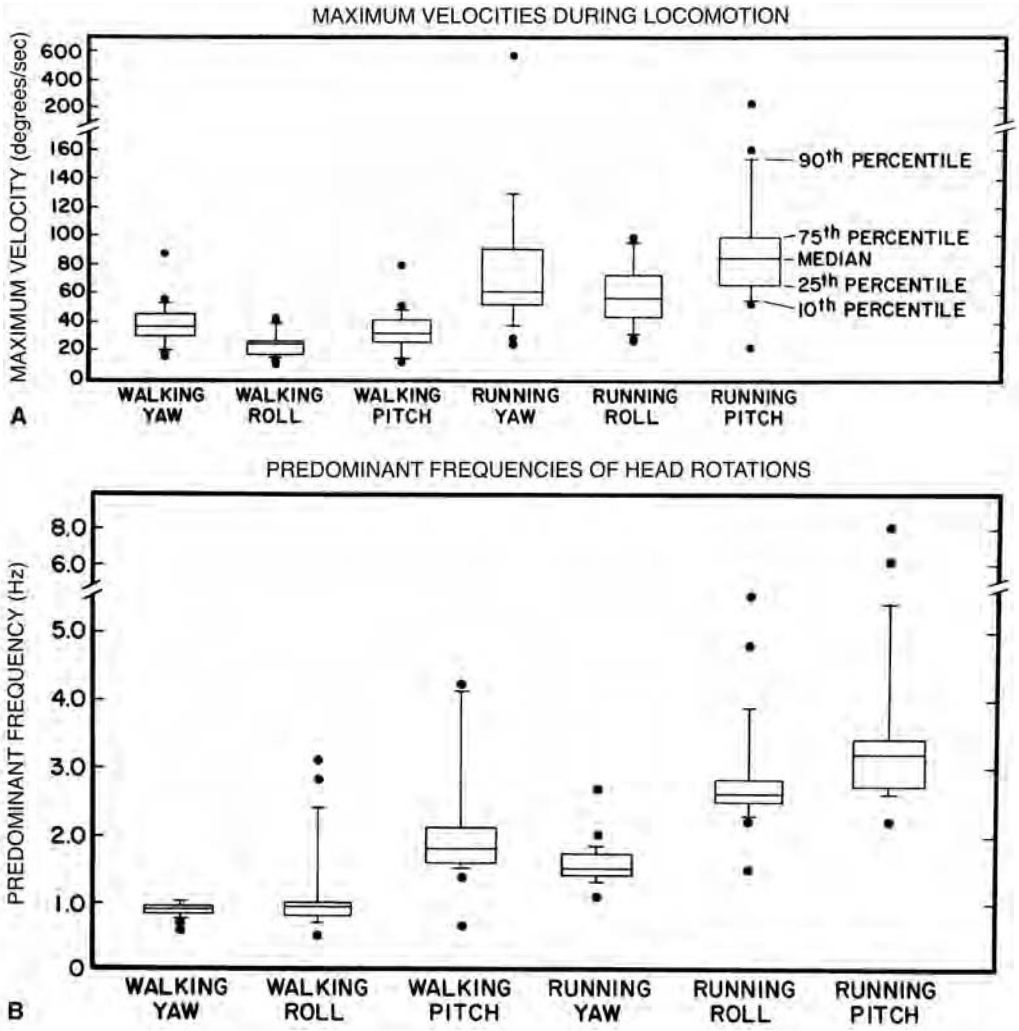


**Figure 11.1** Summary of the ranges of (*A*) maximum velocity and (*B*) frequency of rotational head perturbations that occur during walking or running in place. Distributions of data from 20 normal subjects are displayed as Tukey box graphs, which show selected percentiles of the data. All values beyond the 10th and 90th percentiles are graphed individually as points. (From Leigh and Zee, 2006.[1])

stationary environment. It is the mismatch between the actual multisensory inputs and the expected pattern of sensory stimulation with the head stationary that causes vertigo.[5] Although rotational vertigo connotes disturbance of the semicircular canals or their central projections, sensations of body tilt or impulsion (e.g., lateropulsion, levitation) imply otolithic disturbance.

Vertigo should be differentiated from other causes of "dizziness," such as presyncopal faintness, loss of stable balance, lightheadedness, and psychological disorders (such as agoraphobia, acrophobia, and phobic vertigo syndrome).[5] Thus, accurate identification of symptoms is essential before therapies are begun, although in practice, diagnosis may be difficult.[5–7] Furthermore, even when patients do experience true vertigo, it may not be due to organic disease. So, for example, certain individuals are prone to experience vertigo, unsteadiness, or malaise with motion, at height, and when assuming certain postures.[5]

Besides causing vertigo, sudden loss of tonic neural input from one labyrinth or vestibular nerve causes nystagmus and unsteadiness.[1,5–7] The nystagmus is typically mixed horizontal-torsional with slow phases directed toward the side of the lesion. The nystagmus is more marked 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. Patients with rotational vertigo due to acute, peripheral vestibular lesions are often

uncertain as to the direction of their vertiginous illusions. This is because their vestibular sense indicates self-rotation in one direction, but their eye movements (slow phases of vestibular nystagmus) cause visual image movements that, when self-referred, connote self-rotation to the opposite side. It is therefore worthwhile to evaluate a patient's vestibular sense alone by asking specifically about the perceived direction of self-rotation with the eyes closed, thus eliminating any possibly confounding visual stimuli. Although most patients with acute peripheral lesions recover within a month or two, some are left with recurrent vestibular symptoms, and others experience benign paroxysmal positional vertigo.

## Neuropharmacology of Vertigo and Nystagmus

Basic research has identified a number of neurotransmitters that appear to contribute to peripheral and central vestibular mechanisms.[8–12] The excitatory amino acid glutamate is the most likely neurotransmitter at the vestibular hair cell–vestibular nerve afferent synapse, and also at the synapse between the vestibular nerve and medial vestibular nucleus (MVN). In both cases, it seems that both kainate/AMPA (alpha-amino-3-hydroxy-5-methyl-4-isoxazole-propionic acid) and NMDA (*N*-methyl-D-aspartate) receptors are involved. There is also some evidence that the inhibitory neurotransmitter gamma-aminobutyric acid (GABA) has a role in the vestibular labyrinth. The neurotransmitter of the vestibular efferents, which project from brainstem to hair cells, is acetylcholine.

Both MVN and lateral vestibular nucleus (LVN) neurons possess muscarinic and nicotinic acetylcholine receptors. $GABA_A$ receptors on MVN type I neurons may be the mechanisms of inhibition for projections from MVN type II neurons that receive commissural projections, and cerebellar Purkinje cells of the cerebellar flocculus. A general principle concerning the *vestibular projections* is that inhibitory pathways to motoneurons mediating the vertical VOR use GABA, whereas the inhibitory pathways to motoneurons mediating the horizontal VOR utilize glycine.[12] Histamine, norepinephrine, dopamine, serotonin, and nitric oxide also appear to exert effects on vestibular mechanisms. Thus, a large number of neurotransmitters and neuromodulators have been identified within vestibular and ocular motor structures, but the functional role of most of these molecules remains unclear.

One approach that has clarified the functional significance of these findings has been to study the behavioral effects of *pharmacological inactivation* by microinjection of agents that inactivate certain neurotransmitters. Thus, ocular motor deficits have been induced by microinjection of the weak GABA antagonist bicuculline and the strong GABA agonist muscimol into the vestibular and adjacent prepositus nuclei of monkeys.[13,14] Depending on the site of injection, either agent could induced gaze-evoked nystagmus ("leaky neural integrator") with centripetal slow-phase drifts or centrifugal, increasing velocity waveforms ("unstable neural integrator"); sometimes, nystagmus implying a vestibular imbalance was also produced. Thus, the effects of these injections may represent varying combinations of vestibular imbalance and gaze-holding failure. These findings indicated that GABA is not just an important neurotransmitter for the vertical and horizontal VOR but is also involved in the gaze-holding mechanism, which depends heavily on the medial vestibular and prepositus nuclei. This finding led to clinical trials of GABAergic agents as treatment for forms of acquired nystagmus (described later).

There is also evidence that control of the dynamic property of the VOR, referred to as *velocity storage* (enhancement of vestibular time constant from that of the cupula to that of the VOR) by the nodulus and uvula, is achieved by inhibitory pathways that use GABA.[15] Experimental lesions of the nodulus and uvula cause prolongation of velocity storage and periodic alternating nystagmus (PAN).[16] The $GABA_B$ agonist baclofen is able to abolish PAN due to experimental or clinical lesions.[17]

Clinical evidence has also supported a role for nicotinic *acetylcholinergic mechanisms in vertical eye movements* that are probably mediated by the vestibular system. First, it has been shown that nicotine can produce upbeat nystagmus in normal subjects in darkness.[18] Second, intravenous physostigmine may increase the intensity of downbeat nystagmus.[19] Third, intravenous scopolamine suppresses downbeat nystagmus in some patients.[20]

## Treatment of Vertigo

The general goals in treatment are to eliminate vertigo (illusion of motion) 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 vertigo due to migraine, it may be possible to treat the underlying cause. Usually, however, the treatment is symptomatic.

In *acute vertigo* due to a peripheral vestibular lesion, functional recovery is the rule in the ensuing weeks. There is a consensus that drugs that have a "sedative effect" on the vestibular system should be used for only the first 24 hours.[5,7,21] Some drugs commonly used for treatment of vertigo, nausea, and vomiting are summarized in Table 11-1 and are discussed more fully in

## ■ Table 11-1 SOME COMMONLY USED VESTIBULAR SEDATIVES

| Drug | Class | Dosage | Comments | Precautions |
|---|---|---|---|---|
| Dimenhydrinate (Dramamine) | Antihistamine<br>Increases cAMP | Oral: 50 mg every 4–6 hr<br>IM: 50 mg; maximum of 200 mg in 24 hr | Mild sedative<br>Causes dryness<br>Moderate antiemetic | Asthma, glaucoma, prostrate enlargement |
| Promethazine (Phenergan) | Antihistamine<br>Anticholinergic<br>Phenothiazine | Oral: 25 mg, every 6 hr<br>Supp: 50 mg, every 12 hr<br>IM: 25 mg; maximum of 75 mg in 24 hr | More sedative<br>Moderate antiemetic | Asthma<br>Glaucoma<br>Prostate enlargement<br>Epilepsy |
| Meclizine (Antivert, Bonine) | Antihistamine<br>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<br>Less sedative | Asthma<br>Glaucoma<br>Prostate enlargement |
| Prochlorperazine (Compazine) | Antihistamine<br>Anticholinergic<br>Phenothiazine | Oral: 5–10 mg every 6 hr<br>Supp: 25 mg every12 hr<br>IM: 5–10 mg every 6 hr; maximum of 60 mg in 24 hr | Sedative and antiemetic<br>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<br>More antiemetic<br>Suitable for motion sickness<br>Can cause confusion, mydriasis, "dependency" | Asthma<br>Glaucoma<br>Prostate enlargement |
| Ondansetron (Zofran) | Serotonin 5-hydroxytryptamine$_3$ (5-HT$_3$) receptor antagonist | Oral: 4–8 mg every 8 hr<br>IV: 4 mg | Antiemetic; developed for patients receiving cancer chemotherapy<br>May be effective for nausea due to CNS disease | Headache<br>Constipation |
| Lorazepam (Ativan) | Benzodiazepine | 0.5 mg every 12 hr<br>IM: 1 mg; maximum of 6 mg in 24 hr | GABA modulator<br>May be habit-forming | Glaucoma<br>Additive with sedative drugs, scopolamine |

CNS = central nervous system; GABA, gamma-aminobutyric acid; IM, intramuscular; IV, intravenous; supp = suppository.

reviews.[5,9,21] 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. Evidence now suggests that methylprednisolone, but not the antiviral agent valacyclovir, may hasten recovery[22]; further studies are probably needed to determine the long-term outcome.[23]

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.[21] During this period, a course of specific vestibular exercises may be indicated (see Chapter 14). Patients in whom enduring vestibular symptoms develop may have an underlying central nervous system disorder, typically of the cerebellum,[24] and imaging studies are indicated. Other patients who complain of persistent symptoms may have either a phobic disorder[5] 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 due to migraine (including migraine without a headache) can usually be successfully managed medically (see Chapter 12). Recurrent vertigo due to a perilymphatic fistula or superior canal dehiscence syndrome may recover spontaneously, but some patients require surgical repair (see Chapter 14).

Vertigo due to Ménière's disease is often difficult to manage, although a low-salt diet and diuretics help some patients with the disorder.[25] 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 can be beneficial when vertigo persists, especially when it arises from a deaf ear.[25,26]

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. Sometimes a combination of agents, such an anticholinergic (e.g., scopolamine) and an antidopaminergic agent (e.g., prochlorperazine) will bring more relief than a single agent. Ondansetron, a serotonin 5-hydroxytrytamine$_3$ (5-HT$_3$) antagonist, helps some patients with vertigo due to brainstem stroke.[27]

*Benign paroxysmal positional vertigo* is effectively treated in most cases by specific vestibular exercises or maneuvers (see Chapter 17) [21]; drugs are not indicated in this condition.

# Oscillopsia

## *Pathogenesis*

Oscillopsia brought on or accentuated by head movement is usually of vestibular origin and reflects an inappropriate VOR gain or phase.[1,2,7,28] Vision becomes blurred 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 upon the retina (Box 11-1). Excessive retinal slip not only causes oscillopsia but also impairs vision. Oscillopsia with head movements may also occur as result of weakness of an extraocular muscle (e.g., abducens nerve palsy). Oscillopsia due to nystag-

Box 11-1

### ETIOLOGY OF OSCILLOPSIA[1]

**Oscillopsia with Head Movements: Abnormal Vestibulo-Ocular Reflex**

Peripheral vestibular hypofunction:
- Aminoglycoside toxicity
- Surgical section of VIIIth cranial nerve
- Congenital ear anomalies
- Hereditary vestibular areflexia
- Cisplatin therapy
- Idiopathic

Central vestibular dysfunction:
- Decreased vestibulo-ocular reflex (VOR) gain
- Increased VOR gain
- Abnormal VOR phase

**Oscillopsia due to Nystagmus**

Acquired nystagmus (especially pendular, upbeat, downbeat, see-saw, dissociated nystagmus)
- Saccadic oscillations (psychogenic flutter/voluntary nystagmus, ocular flutter, microsaccadic flutter and opsoclonus)
- Superior oblique myokymia (monocular oscillopsia)
- Congenital nystagmus (uncommon under natural illumination)

**Central Oscillopsia**

With cerebral disorders: seizures, occipital lobe infarction

From Leigh and Zee, 2006.[1]

mus and other ocular oscillations occurs when the head is stationary.

An abnormal VOR may lead to oscillopsia during head movements in three possible ways: abnormal gain, abnormal phase shift between eye and head rotations, and a directional mismatch between the vectors of the head rotation and eye rotation. Disease of either the vestibular periphery or its central connections may be the cause (see Box 11-1).

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, and the clinician can easily demonstrate this behavior at the bedside by testing visual acuity first with the patient's head stationary and then while rotating it from side to side at 1 to 2 cycles/second.[1]

Oscillopsia may also occur with disorders of the central nervous system that change the gain or phase of the VOR. Thus, disease of the vestibulocerebellum may cause "vestibular hyper-responsiveness," particularly in the vertical plane. This occurs in patients with the Arnold-Chiari malformation[1] and, occasionally, patients with cerebellar disorders are reported with increased gain of both the horizontal and vertical VORs.[29] 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.[30] 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.[31]

### *Treatment of Oscillopsia*

With time, compensation takes place in patients with oscillopsia due to vestibular loss. Compensation occurs by means of a variety of factors, including potentiation of the cervico-ocular reflex, pre-programming of compensatory eye movements, and perceptual changes.[2,32,33] Thus, drugs have little to offer. Exercises (see Chapter 13) and encouragement of the patient to resume activities such as walking are key. Rarely, oscillopsia due to a hyperactive VOR can be treated pharmacologically.[29] Paradoxically, patients who lack a VOR can read head-fixed visual display during locomotion better than normal subjects can.[34] 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, then it is easy to suppress vestibular eye movements. The practical application of this finding—in the future—might be development of head-fixed video displays of images obtained with cameras that can compensate for head perturbations.

Newer alternative biomedical approaches to loss over vestibular function include tactile sensory substitution methods, in which an angular accelerometer provides tactile stimulation to the patient's tongue.[35] With time, the patient learns to interpret mechanically sensory information in terms of tactile stimuli and can negotiate the environment better. However, it remains to be determined how well elderly patients can learn to perform this sensory substitution.

## 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 due to drift of images of stationary objects upon the retina, interfere with reading and watching television, and the oscillopsia is 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 Snellen optotypes), image motion in excess of about 5 deg/sec impairs vision.[36,37] On the other hand, the relationship between retinal image velocity and the development of oscillopsia is less consistent; the magnitude of oscillopsia is usually less than the magnitude of nystagmus. For example, in patients with downbeat nystagmus, oscillopsia is, on average, about one third the amplitude of the nystagmus.[38] 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. Nevertheless, if retinal image drift, in patients with nystagmus, can be reduced to less than about 5 deg/sec, oscillopsia is usually abolished and vision is improved.[39]

### *Treatments*

#### Drugs

Some of the various drugs reported to suppress nystagmus are summarized in Box 11-2. *Gabapentin* has been shown, in some patients, to suppress or abolish acquired pendular nystagmus that is associated with multiple sclerosis or follows brainstem stroke (oculopalatal tremor syndrome).[40,41] Acquired pendular nystagmus produces perhaps the most troubling visual disturbances of all

Box 11-2

## TREATMENTS FOR NYSTAGMUS AND ITS VISUAL CONSEQUENCES[1,17,41]

**Drugs**

- Gabapentin
- Memantine
- Baclofen
- 4-aminopyridine and 3,4-diaminopyridine
- Acetazolamide
- Clonazepam
- Valproate
- Trihexyphenidyl and benztropin
- 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

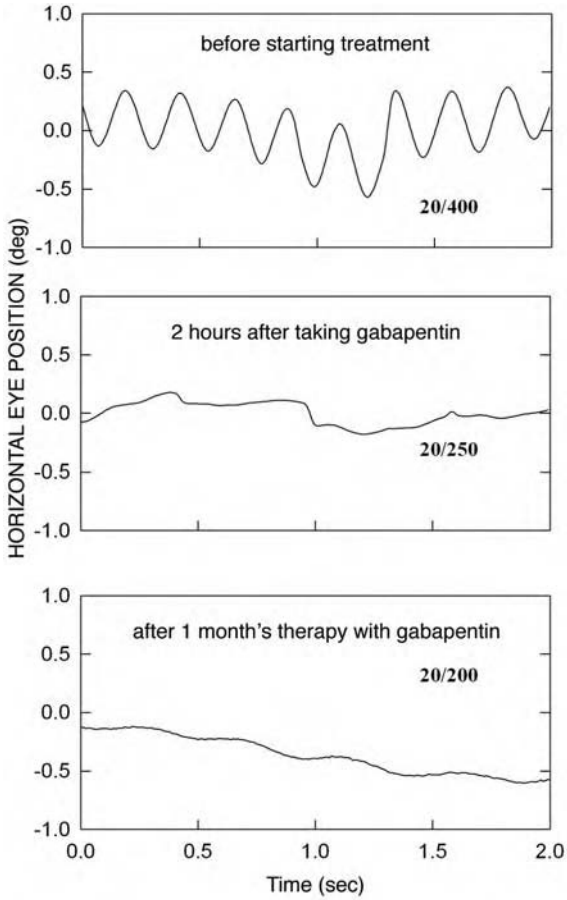


**Figure 11.2** Example of the effects of gabapentin on the horizontal component of pendular nystagmus in a 41-year-old woman with multiple sclerosis. Visual acuity measurements of the recorded eye are shown in each panel. After the initial session (*top panel*), the patient ingested 300 mg of gabapentin, which reduced her oscillations and improved her vision (*middle panel*). The effect was sustained 2 months later (*bottom panel*), when she was taking a dosage of 300 mg three times/day.

forms of nystagmus, and most patients who take gabapentin experience some relief (Fig. 11.2). However, not all such patients respond, and worsening of ataxia may be a troublesome side effect.

*Memantine*, an uncompetitive *N*-methyl-D-aspartate (NMDA) antagonist, has also been reported to suppress acquired pendular nystagmus in patients with multiple sclerosis and is generally well tolerated.[42] In 2004, memantine received FDA approval for treatment of memory failure in Alzheimer's disease. Physicians considering using memantine as off-label treatment for acquired pendular nystagmus should be aware that the required dose is up to 40 mg per day, which is twice the recommended dose for treatment of Alzheimer's disease.

*Baclofen* is less effective in treating acquired pendular nystagmus than gabapentin but, as previously discussed, is usually effective in treating PAN.[17] Neither baclofen nor gabapentin is reliable for treatment of downbeat nystagmus.[17,40] Other GABAergic agents, especially *clonazepam*, have been reported to be useful in the treatment of downbeat nystagmus.[17,43]

*Anticholinergic agents* may be effective in some patients with pendular or downbeat nystagmus. In a double-blind study, Barton and associates[20] administered scopolamine, benztropine, and glycopyrrolate (a quaternary agent devoid of central nervous system activity) intravenously. A single dose of scopolamine effectively reduced nystagmus and improved vision in all 5 patients with pendular nystagmus, benztropine was less effective, and glycopyrrolate had no significant effect. However, in a randomized, double-blind crossover trial of oral trihexyphenidyl and tridihexethyl chloride (a quaternary anticholinergic that does not cross the blood-brain barrier), only 1 of 10 patients showed a decrease in nys-

tagmus and an improvement of visual acuity with trihexyphenidyl.[44] Anticholinergic side effects included dry mouth, constipation, disturbed sleep, and tiredness. Furthermore, transdermal scopolamine is not reliable as treatment for acquired nystagmus and may actually make it worse in some patients.[45]

Two potassium channel–blocking agents, 4-aminopyridine and 3,4-diaminopyridine, have both been reported to suppress downbeat nystagmus in some patients.[46,47] Both agents have been used in the past for symptomatic treatment in multiple sclerosis. Of the two agents, 3,4-diaminopyridine is better tolerated and has a longer duration of action, although epileptic seizures and paresthesias may occur with its use. These agents may also be effective treatment of the syndrome of episodic ataxia type 2[48] although *acetazolamide* remains the mainstay of therapy.[17]

## Optical Devices

A number of optical devices have been suggested for treatment of nystagmus. One approach that often benefits patients whose nystagmus dampens while they are viewing a near target is convergence prisms. An arrangement that is often effective is 7.00 diopter base-out prisms with –1.00 diopter spheres added to compensate for accommodation.[17,49] 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 due to wearing of base-out prisms may be sufficient to qualify them for a driving license. Some patients with acquired nystagmus also benefit.[50] Occasionally, in patients in whose nystagmus is worse during near viewing, base-in prisms help.

Theoretically, it should be possible to use prisms to help patients whose nystagmus is quieter when the eyes are moved into a particular position in the orbit—the "null region." Thus, in patients with congenital nystagmus, there is usually some horizontal eye position in which nystagmus is minimized, and in patients with downbeat nystagmus, the eyes may be quieter in upgaze. 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 an optical system to stabilize images upon the retina during eye movements.[39,51,52] This system consists of a high-plus spectacle lens worn in combination with a high-minus contact lens. 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 upon 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. Another is that some patients with ataxia or tremor (such as those with multiple sclerosis) have difficulty inserting the contact lens.

A newer "hi-tech" approach is the use of an electro-optical device that measures ocular oscillations and moves prism devices to negate the effects of the nystagmus.[53] 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. Figure 11.3 summarizes the

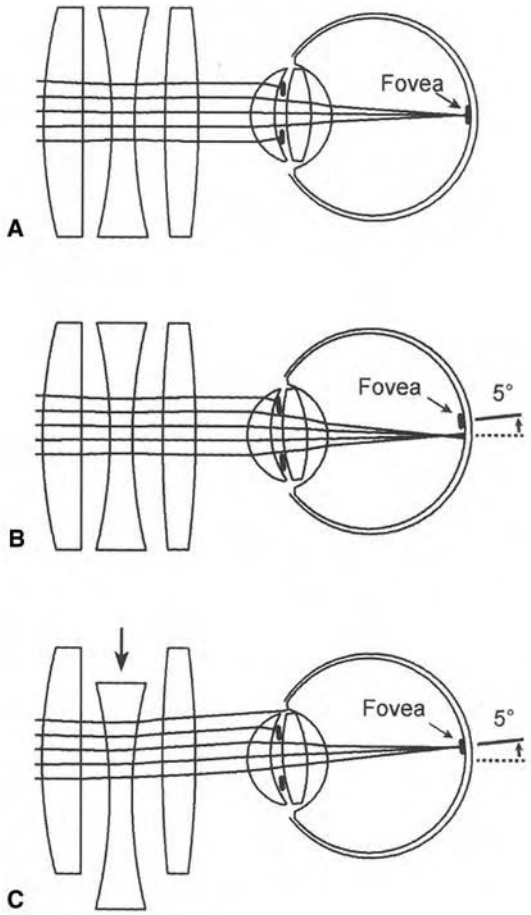


**Figure 11.3** Demonstration of how a three-lens image-shifting device can null the visual effects of ocular oscillations. Starting with the eyes and optics in a neutral position (a), light from a distant target is brought to the fovea of the retina. If the eye is rotated down (b), the image is displaced 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. (From Smith et al, 2004.[54])

image-shifting optics that are being used to develop a portable, battery-driven device.[54]

### Surgery

A range of operative procedures have been developed for treatment of congenital nystagmus[55] and have been proposed as treatment for acquired forms of nystagmus; formal evaluation is needed. Neurosurgery does have a clear role in the therapy of the Arnold-Chiari syndrome; suboccipital decompression is reported to improve downbeat nystagmus and to prevent progression of other neurological deficits.[56] Injection of botulinum toxin either into selected extraocular muscles or into the retrobulbar space temporarily abolishes or suppresses nystagmus, but the side effects ptosis and diplopia limit the therapeutic value, except in selected patients.[17,57,58]

## 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 due to loss of the vestibular sense often improves spontaneously, and medications do not aid the recovery; new sensory substitution methods may provide a new therapeutic strategy. Oscillopsia due to nystagmus may be helped if gabapentin, memantine, baclofen, clonazepam, or acetazolamide can reduce the ocular oscillations. 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 of the problem must be made before any of these treatments is attempted.

## Acknowledgments

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CHAPTER 12

# Migraine, Ménière's Disease, and Motion Sensitivity

*Ronald J. Tusa, MD, PhD*

Migraine is a common cause of episodic vertigo and dysequilibrium in children and adults. In practices treating patients with headaches, 27 to 33 percent 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) (Kuritzky et al, 1981).[3] In this chapter, 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 will be described.

## Incidence of Migraine

Migraine is an extremely prevalent disorder. An epidemiologic study involving over 20,000 individuals between 12 and 80 years of age found that 17.6 percent of all adult females, 5.7 percent of all adult males, and 4 percent 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 will be described later. Of those individuals with migraine, approximately 18 percent 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.[6]

## Symptoms of Migraine

### *Case Example*

Patient is a 50 year old with dizziness. Several months ago while standing looking out the window developed severe vertigo, N/V and fell backwards. This was followed by a mild headache. Vertigo lasted 10 minutes, H/A lasted for 1 day, felt washed out for days. Since then he has had 5 more spells of vertigo, photophobia, phonophobia followed by headache. Feels better if takes a short nap when spell occurs. He also notes increased motion sensitivity while wife drives car, while he played games on his computer or while he is driving a motorboat. In addition, he now has daily dizziness consisting of light-headedness and sense that 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 neurologic and otologic exam. He already had an audiogram, ENG, and MRI of head, all off which were normal. He

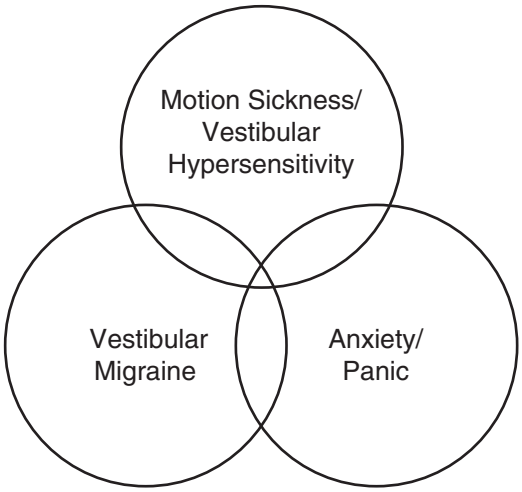


**Figure 12.1** Causes of symptoms in patients with migraine.

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. 12.1). This case example illustrates the overlapping symptoms often found in patients with vestibular migraine. He started with vertigo with a migraine headache–consistent 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 12.1 can occur separately, but frequently they occur together in individuals with vestibular migraine.

### *Symptoms during Vestibular Migraine Aura*

A questionnaire was given to 38 consecutive patients with vestibular migraine to determine the type of symptoms and incidence (Table 12-1). The symptoms were divided into different types of dizziness, 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).

■ Table 12-1 **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 |
| Bilateral paresthesia | 3 |
| Diplopia | 2 |
| *Chronic Symptoms* | |
| Motion sickness/vestibular hypersensitivity | 78% |
| Generalized anxiety disorder (GAD) | 57% |
| GAD + panic attacks | 47% |

## Classification and Criteria for Diagnosis

### *Migraine*

Migraine disorders are usually subdivided into several types. To help standardize terminology and diagnostic criteria, a classification system for headaches was developed by the IHS.[5] This classification was based on 2 years of discussion among 100 individuals with representatives from seven countries. The classification and criteria for headaches pertinent to neuro-otological disorders is summarized in Box 12–1. The general features of

Box 12-1

## INTERNATIONAL HEADACHE SOCIETY CLASSIFICATION OF HEADACHE

**1.1 Migraine without aura** (replaces common migraine):

A. At least 5 attacks fulfilling criteria B–D.

B. Headache attacks lasting 4–72 hours untreated. In children <15 years, attack may last 2–48 hours.

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. At least one of the following:

1. History and physical examinations do not suggest another disorder.
2. History and physical examinations do suggest such disorder, but it is ruled out by appropriate investigations (e.g., magnetic resonance imaging or computed tomography of the head).

**1.2 Migraine with aura** (replaces classic migraine):

A. At least 2 attacks fulfilling B:

B. At least 3 of the following:

1. One or more reversible aura symptoms indicating focal central nervous system dysfunction.
2. At least 1 aura symptom develops gradually over more than 4 minutes, or 2 or more symptoms occur in succession.
3. No aura symptom lasts more than 60 minutes unless more than 1 aura symptom is present.
4. Headache occurs either before, during, or up to 60 minutes after aura is completed.

C. Same as E above in criteria 1.1.

**1.2.2 Migraine with prolonged aura** (replaces complicated migraine):

Fulfills criteria for 1.2, but at least one symptom lasts more than 60 minutes and less than 7 days.

**1.2.4 Basilar migraine** (replaces basilar artery migraine):

Fulfills criteria for 1.2, but 2 or more aura symptoms of the following types:

Vertigo, tinnitus, decreased hearing, ataxia, visual symptoms in both hemifields of both eyes, dysarthria, double vision, bilateral paresthesia, bilateral paresis, decreased level of consciousness.

**1.2.5 Migraine aura without headache** (replaces migraine equivalent or acephalgic migraine):

Fulfills criteria for 1.2 but no headache.

**1.5 Childhood periodic syndromes** that may be precursors to or associated with migraine.

**1.5.1 Benign paroxysmal vertigo of childhood:**

A. Brief, sporadic episodes of disequilibrium, anxiety, and often nystagmus or vomiting.

B. Normal neurological findings.

C. Normal electroencephalographic findings.

**1.6.2 Migrainous infarction** (replaces complicated migraine):

A. Patient has previously fulfilled criteria for 1.2.

B. The present attack is typical of previous attacks, but neurological deficits are not completely reversible within 7 days and/or neuroimaging demonstrates ischemic infarction in relevant area.

C. Other causes of infarction are ruled out by appropriate investigations.

Adapted from the Headache Classification Committee of the International Headache Society, 1988.[5]

Box 12-2

**NEURO-OTOLOGICAL DISORDERS**

Neuro-otological disorders due to migraine:

- Paroxysmal torticollis of infancy
- Benign paroxysmal vertigo of childhood
- Basilar migraine
- Benign recurrent vertigo of adults
- Migrainous infarct resulting in vertigo

Neuro-otological disorders associated with migraine:

- Motion sickness
- Ménière's disease
- Benign paroxysmal positional vertigo

the relevant types of migraine are discussed briefly; the features of specific neuro-otological disorders are discussed in greater detail (Box 12-2).

## Migraine without Aura

Migraine without aura, which replaces "common migraine," consists of periodic headaches that are usually throbbing and unilateral, exacerbated by 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, 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 Migraine

Basilar migraine (IHS classification 1.2.4) was first described by Bickerstaff,[7] and has been subsequently reported by a number of individuals.[8,9] 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.[10]

## Benign Recurrent Vertigo in Adults

This disorder was not formally classified by the IHS. Based on the symptoms, it may either be referred to as basilar migraine (1.2.4), or migraine aura without headache (1.2.5). This was first described by Slater,[11] and is the most common neurotological syndrome caused by migraine. This disorder consists of spells of vertigo, occasionally with tinnitus but without hearing loss.[12,13] In some individuals, jerk nystagmus may occur during the spell (see Case Study 1 at end of chapter). Vertigo typically lasts between minutes and hours; the majority last less than 1 hour, and may occur with or without headache. The spells usually occur between the ages of 20 and 60. Peripheral and central vestibular deficits diagnosed by caloric and spontaneous eye movements during electronystagmography have been reported to occur in 5 to 80 percent of individuals with migraine in their headache-free period.[14–16] This variation in abnormality may be due to a difference in criteria used for diagnosing vestibular deficits.[15] In the majority of cases, audiograms and caloric studies are normal. In this author's opinion, concomitant Ménière's disease was not always excluded in all cases. Therefore, it is not clear that permanent vestibular deficits can occur in migraine disease. Some individuals develop exercise-induced spells from a variety of physical activities including sit-ups, heavy lifting,

intercourse, and strenuous aerobic exercises[17] (see Case Study 2). One needs to rule out Ménière's disease, benign paroxysmal positional vertigo, TIAs, vestibular epilepsy, and perilymphatic fistula before making a diagnosis of migraine-induced vertigo.

### Childhood Periodic Syndromes

There are two neuro-otological disorders in children due to migraine. (1) Benign paroxysmal vertigo of childhood (IHS classification 1.5.1), which was first described by Basser.[18] This disorder consists of spells of vertigo and dysequilibrium without hearing loss or tinnitus.[19–23] The majority occurs between 1 and 4 years of age, but can occur anytime during the first decade. Vertigo and dysequilibrium 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 normal physical examinations and normal EEGs. 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 Ménière's disease, vestibular epilepsy, perilymphatic fistula, posterior fossa tumors, and psychogenic disorders. (2) Paroxysmal torticollis of infancy was not specifically classified by the IHS, but may fit into the same classification as benign paroxysmal vertigo of childhood; these disorders frequently occur in the same patient.[22] This disorder was first described by Snyder,[24] and has now been reported by a number of individuals.[25–27] Paroxysmal torticollis of infancy 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 due to migraine auras without headache. Some individuals complain of headache when they become older. The differential diagnosis includes posterior fossa tumors and torticollis.

## *Disorders Associated with Migraine*

### Motion Sickness

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) characterize motion sickness. Motion sickness is partially due to a visual-vestibular conflict or mismatch.[28] Twenty-six to sixty percent of patients with migraine have a history of severe motion sickness compared to 8 to 24 percent 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.

#### *Generalized Anxiety Disorder*

Generalized and persistent unrealistic worry with motor tension, autonomic hyperactivity, apprehensive expectation and vigilance for over 6 months.

#### *Panic Attack*

Discrete spells of intense fear or discomfort and >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

# 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] As dopamine increases, yawning begins to occur. With further dopamine increase, more symptoms occur that includes 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 Ncol C allele in the *DRD2* gene is higher than the Ncol T allele in patients with migraine, generalized anxiety and panic disorders.[31,32] Perhaps this common allele in individuals with migraine and anxiety is the reason why these two entities frequently co-exist 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.

### 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 percent 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.[33] This same chromosome locus may be involved also in other forms of migraine aura.[34] Defects involving this gene are involved with other autosomal dominant disorders that have neurotologic symptoms (Table 12-2). The symptoms of these disorders overlap extensively.[35] Only 50 percent 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.[36,37] This may be the cause for increased motion sensitivity in individuals with migraine. 26-60% of patients with migraine have a history of severe motion sickness compared to 8-24% 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.[38] 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.[39] These agonists are the most effective drugs in aborting migraine headache.

■ Table 12-2 **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<br>GEN and DBN may persist after spells |
| Point mutation | Episodic ataxia-2 | Episodic ataxia and vertigo<br>GEN<br>DBN<br>Decreases in VOR cancel and pursuit<br>Normal VOR |
| CAG repeats | SCA 6 | Progressive ataxia<br>GEN<br>DBN<br>Decreases in VOR cancel and pursuit<br>Normal VOR |

DBN = downbeat nystagmus; GEN = gaze-evoked nystagmus; VOR = vestibulo-ocular reflex.

## Management

Management begins with identiftying to what extent the patient is suffereing from spells due to vestiblar migraine, or more chronic symptoms due to chronic anxiety and increased motion sensitivity.

### Treatment of Vestibular Migraine

Spells of vertigo and dysequilibrium 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, hypo-

glycemia, fluctuating estrogen, certain foods, and smoking.[40-42] 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 12–3). 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.[43] 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.[43, 44] 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.

Box 12-3

**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 prior to the headache and/or spell.
   c. Bring diary in with you on your next visit!
6. Medications.

### *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 (see Table 12-2). 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]

### *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" (Table 12-3) that they had during 24 hours prior to 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. There are a variety of drugs used in this capacity including beta-blockers, amitriptyline, calcium-channel blockers, lithium carbonate, aspirin, and ibuprofen.[47] All of these drugs have been found to be effective in reducing the frequency and severity of headache with or without aura. These drugs have also been found to be

## ■ Table 12-3 DIET FOR PATIENTS WITH MIGRAINE

| Food Category | Foods Allowed | Foods to Avoid |
|---|---|---|
| Beverages | Decaffeinated coffee, fruit juice, club soda, noncola soda<br>Limit caffeine sources to 2 cups/day (coffee, tea, cola) | Chocolate, cocoa, certain alcoholic beverages (red wine, port, sherry, scotch, bourbon, gin) |
| | | Excessive aspartame (Nutrasweet): no more than 24 oz/day of diet drink |
| Meats, fish, and poultry | Fresh or frozen turkey, chicken, fish, beef, lamb, veal, pork | Aged, canned, cured, or processed meats, including ham or game, pickled herring, salad and dried fish<br>Chicken liver, bologna<br>Fermented sausage<br>Any food prepared with meat tenderizer, soy sauce, or brewer's yeast<br>Any food containing nitrates or tyramine (smoked meats, including bacon, sausage, ham, salami, pepperoni, hot dogs) |
| | Limit eggs to 3/week | |
| | Tuna or tuna salad | |
| Dairy products | Milk: Homogenized, 2%, or skim | Milk: Buttermilk, sour cream, chocolate milk |
| | Cheeses: American, cottage, farmer, ricotta, cream, Canadian, processed cheese slice | Cheeses: Stilton, bleu, cheddar, mozzarella, cheese spread, Roquefort, provolone, gruyere, muenster, feta, parmesan, emmenthal, brie, brick, camembert types, cheddar, gouda, romano |
| | Yogurt (limit 1/2 cup/day) | |
| Breads, cereals | Commercial bread, English muffins, melba toast crackers, bagels<br>All hot and dry cereals | Hot fresh homemade yeast bread, bread or crackers containing cheese<br>Fresh yeast coffee cake, doughnuts, sourdough bread<br>Any product containing chocolate or nuts |
| Potato or substitute | White potato, sweet potato, rice, macaroni, spaghetti, noodles | None |
| Vegetables | Any except those to avoid | Beans such as pole, broad, lima, Italian, fava, navy, pinto, garbanzo<br>Snow peas, pea pods, sauerkraut, onions (except for flavoring), olives, pickles |
| Fruit | Any except those to avoid<br>Limit citrus fruits to1/2 cup/day (1 orange)<br>Limit banana to1/2 per day | Avocados, figs, raisins, papaya, passion fruit, red plums |
| Soups | Cream soups made from foods allowed in diet, homemade broth | Canned soup, soup or bouillon cubes, soup base with yeast or monosodium glutamate (MSG; read labels) |

*(continued on following page)*

■ Table 12-3 **DIET FOR PATIENTS WITH MIGRAINE** *(continued)*

| Food Category | Foods Allowed | Foods to Avoid |
|---|---|---|
| Desserts | Any cake, cookies without chocolate, nuts, or yeast<br>Any pudding or ice cream without chocolate or nuts<br>Flavored gelatin | Any product containing chocolate, including ice cream, pudding, cookies, cake or pies<br>Mincemeat pie |
| Sweets | Sugar, jelly, jam, honey, hard candy | Chocolate candy or syrup, carob |
| Miscellaneous | Salt in moderation, lemon juice, butter or margarine, cooking oil, whipped cream, white vinegar, commercial salad dressings | Pizza, cheese sauce, MSG in excessive amounts (including Chinese food and Accent), meat tenderizer, seasoned salt, yeast, yeast extract<br>Mixed dishes (including macaroni and cheese, beef stroganoff, cheese blintzes, lasagna, frozen "TV dinners")<br>Chocolate, nuts (including peanut butter)<br>All nonwhite vinegars<br>Anything fermented, pickled, or marinated |

Modifed from Diamond, 1991,[40] and Shulman et al, 1989.[45]

useful in preventing migraine-associated dizziness.[48,49] Daily valproate recently has been found to be effective in preventing migraine headaches[50] and has been approved for prophylactic treatment of migraine. To what extent this drug stops vertigo due to migraine is not known. With the exception of aspirin, ibuprofen, and valproate, the mode of action of these prophylactic drugs may be via their antagonist effect on 5-$HT_2$ receptors.[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/d. 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 hemiplegic migraine[52] and those with familial migraine with vertigo.[53]

## *Abortive Medical Therapy*

The drugs used to abort migraine headache include aspirin, ibuprofen, isometheptene mucate (Midrin), ergotamine, dihydroergotamine, and 5-HT agonists (serotonin).[47] There are several types of serotonin receptors. It has been postulated that the abortive drugs are 5-$HT_1$ agonists, whereas the prophylactic drugs are 5-$HT_2$ antagonists.[51] Table 12-4 lists the serotonin receptor medications and their uses. Sumatriptan is a potent agonist of the 5-$HT_1$ receptor but does not cross the blood-brain barrier well. Consequently, a host of other drugs are now on the market with similar receptor agonists including eletriptan, naratriptan, rizatriptan, and zolmitriptan. Dihydroergotamine is also effective with 5-$HT_1$ agonism. These drugs are primarily designed to abort headache. They have not been found to be effective in aborting migraine auras to date. In fact, subcutaneous sumatriptan is totally ineffective when given during the aura.[54] There are also now a host of selective serotonin 5-$HT_3$ receptor antagonists that prevent nausea and vomiting (see Table 12-4). These drugs, plus dexamethasone, are the most effective regimen for prevention of acute vomiting caused by cancer chemotherapy. They have been found to be effective in other causes of severe nausea and vomiting including central vertigo,[55] but they have not yet been approved for these conditions. They are expensive and should not be used beyond a 24-hour period.

**■ Table 12-4 SEROTONIN (5-HT) RECEPTOR DRUGS**

| Receptor | Chemical (Trade Name) | Dose | Indications | Side Effects |
|---|---|---|---|---|
| 5-$HT_1$ agonists | Sumatriptan (Imitrex) | 6 mg SC; 25–100 mg PO; 20 mg nasal spray | Abortive therapy for migraine | Flushing, chest discomfort |
| | Zolmitriptan (Zomig) | 2.5 mg PO | Abortive therapy for migraine | Flushing, chest discomfort |
| | Naratriptan | 2.5 mg PO | Abortive therapy for migraine | Flushing, chest discomfort |
| | Eletriptan | | Abortive therapy for migraine | Flushing, chest discomfort |
| | Rizatriptan | | Abortive therapy for migraine | Flushing, chest discomfort |
| 5-$HT_3$ antagonists | Ondansetron (Zofran) | 8 mg PO × 2; 32 mg IV | Prevention of severe nausea and vomiting from chemotherapy | Mild headache and dizziness |
| | Dolasetron (Anzemet) | 100 mg PO; 1.8 mg/kg IV | Prevention of severe nausea and vomiting from chemotherapy | Mild headache and dizziness |
| | Granisetron (Kytril) | 2 mg PO 10 mg/kg IV | Prevention of severe nausea and vomiting from chemotherapy | Mild headache and dizziness |

IM = intramuscular; IV = intravenous; PO = oral; SC = subcutaneous.

## Migraine versus Ménière's Disease

There is a great deal of confusion between migraine aura without headache and vestibular hydrops (vestibular Ménière's disease).[56, 57] Both can present with transient vertigo, ear fullness, or occasional tinnitus, but without any decrease in hearing (Table 12-5). A history of headaches associated with the spells of vertigo may help to distinguish these two syndromes, but occasionally the diagnosis is made only following the patient's response to a therapeutic trial (see Case Study 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 due to migraine aura without headache (see Case Study 4). Kayan and Hood[1] and Hinchcliffe[58] noted a higher than expected incidence of both migraine and Ménière's disease in the same individual. Whether there is a causal link between these two disorders is unclear.

**■ Table 12-5 MIGRAINE VERSUS MÉNIÈRE'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 |

## CASE STUDY 1

A 46-year-old owner of a blacktop paving company is referred for spells of vertigo, nausea, vomiting, oscillopsia, and diaphoresis for 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 life-long 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 that 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 artery migraine (IHS classification 1.2.4) was made and his spells of vertigo stopped after he was placed on a diet and propranolol.

## CASE STUDY 2

A 28-year-old real estate developer is referred for thirty 5 to 10 minute spells of dysequilibrium, vertigo, 15° 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, stairclimber, and stationary bicycle).

*Comment*

Because of the exercise-induced nature of these spells, they were believed to be due to 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 migraine (IHS classification 1.2.4) was made. These spells stopped after he was placed on the antimigraine schedule listed in Box 12–3.

## CASE STUDY 3

A 47-year-old medical transcriptionist is referred for a 10-year history of spells of nausea, dysequilibrium, 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. This entity usually 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 (IHS classification 1.1) and migraine aura without headache (IHS classification 1.2.5). 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 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 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. 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 aura without headache (IHS classification 1.2.5). 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 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 dysequilibrium 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 2-1/2 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 prior to menstrual periods. During the migraine aura, head movements usually provoke dizziness. In about 30 percent 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 due to canalithiasis of the horizontal semicircular canal and sustained ageotropic nystagmus can be due to 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 dysequilibrium 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 dysequilibrium from migraine should be treated by reducing these risk factors and, if necessary, by medical therapy.

## Patient Information

Glaxo Wellcome (1-800-377-0302). 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 or http://www. headaches. org/.

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CHAPTER 13

# Therapy for Mal de Débarquement Syndrome

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

*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 similar conditions have been described after extended airplane travel, train travel, and space flight.[1] Symptoms include sensations of 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.

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. After returning from the cruise, she experienced land-sickness, consisting of 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."

The single largest study of MDDS was conducted by Hain and associates,[2] who reported 27 cases. Nearly all patients were middle-aged women (26/27; mean age 49.3 years). Duration of symptoms ranged from 6 months to 10 years (mean 3.5, SD 2.5). Subjects got little or no relief from conventional antivertiginous medication such as meclizine and scopolamine patches. The most effective medication was a benzodiazepine (Klonopin). Only a small number of additional cases have been reported, all showing a similar pattern.[3–5]

MDDS has similarities to land-sickness. The features differentiating land-sickness from MDDS are a shorter duration of land-sickness, the association of land-sickness with motion sickness, and gender differences. Between 41% and 73% of persons disembarking from seagoing voyages experience a brief unsteadiness syndrome, or "land-sickness." [6–8] Males and females do not seem to differ significantly in the incidence, intensity, or duration of land-sickness symptoms.[7] Persons with land-sickness are also likely to have seasickness,[6] but persons with MDDS generally are untroubled by seasickness.

MDDS also has some similarities to motion sickness (mal de mer). However, motion sickness is easily distinguished by the relatively short duration of motion sickness and gender differences. 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 (such as sitting in a chair). Many persons with motion sickness find driving very difficult, as do persons with vestibular disorders.

## Cause of the Syndrome: Persistent Adaptation to Swaying Environments?

MDDS does not have the features of a "pathologic" disease, in the sense that it does not appear to follow an injury. Rather, it is provoked by exposure to motion that

does not trouble most individuals in a persistent manner. Movements such as those experienced in a boat expose a person to rhythmic upward and downward movements along with tilts to the left and right. During this time the brain must adjust motions of the legs and body so that they counter the rhythmic pattern of shipboard motion. Adapting to such movement is sometimes referred to as "gaining sea legs."

Persons with MDDS may be very good at adapting to unusual motion situations–gaining sea legs during ocean travel—but slow to give up their adaptation when they return to the stable ground.[5] In support of this idea is that a brief period of rocking can be induced in normal subjects by "sway referencing," a procedure resembling the situation on a boat.[9]

As discussed in Chapter 1, good balance requires integration of sensory input from the inner ear, eyes, ankles (somatosensory input), and internal models of orientation. One possibility, then, is that MDDS is a disorder of somatosensory integration. Some studies have suggested that persons with MDDS become increasingly reliant on somatosensory input after motion exposure and reduce the weighting of vision and vestibular input.[10] We suspect that this observation is a "red herring," as it is difficult to understand the rationale for such an adaptation in the context of boat travel.

Another possibility is that individuals susceptible to MDDS may become more reliant on visual and graviceptive information (and thus have *decreased* somatosensory weighting). Unsurprisingly, this occurs in normal subjects who are exposed to situations in which somatosensory feedback is distorted.[11] This particular theory is better grounded in physiology but contradicts available data, suggesting the exact opposite adaptation in MDDS.[10] Either adaptation might result in inaccurate land sensorimotor integration. Neither of these adaptations explains the rocking sensation of MDDS or the characteristic improvement upon driving a car.

A third mechanistic possibility for MDDS arises from internal model theory. On a boat, during quiet standing on deck, there are two main reasons why somatosensation at the ankles and foot might be altered. When the boat rotates (pitch), the deck tilts in space. Sheer force exerted on the soles of the feet alters somatosensation. In this situation, it would be best for balance to allow bodily inertia to keep the body upright and not react with an ankle torque. On the other hand, when the boat translates forward (as in surging forward), sheer is created. Here it would be best to rapidly respond with an ankle torque to prevent a fall. 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 nonsalient 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.

## Treatment

Conventional vestibular suppressants such as meclizine and transdermal scopolamine are not helpful in MDDS.[2] Benzodiazepines, such as clonazepam, are of the most benefit. Zimbelman and Watson[14] reported a case in which physical therapy was used in an attempt to treat MDDS; of course, it is not known how this patient would have fared without intervention. In the largest study of MDDS, many individuals underwent vestibular rehabilitation, again because of a lack of controls, but it was not possible to determine whether they did any better than subjects who were not treated.[2] Thus, the efficacy of vestibular rehabilitation for MDDS is currently unknown.

Our view is that in therapy of MDDS, the goal should be to modify an inappropriate internal model. The individual should be encouraged to seek out situations in which there is abundant and accurate sensory information regarding orientation in space and should practice balancing in a variety of sensory contexts. We advise patients to avoid going back on boats or in other vehicles incorporating periodic motion because symptoms frequently flare up after such experiences.

Although data that any intervention works are lacking, a number of physical therapy treatment strategies might be tried in MDDS. As already mentioned, two theories applicable to MDDS propose an inappropriate weighting of somatosensory input (one too much, one too little). Accordingly, if, during the assessment of the patient, the clinician determines that somatosensory weighting is inappropriate, he or she might attempt a specific intervention.

For example, the patient who disregards proprioceptive information should practice detecting joint position sense and kinesthetic awareness of the toes and ankles while sitting with the eyes closed. Once there is heightened awareness of joint position sense, the patient should use the sensory information during a dynamic task. While standing on a firm surface with the eyes closed, the patient may dynamically shift weight from anterior to posterior for 1 to 2 minutes and then from side to side for 1 to 2 minutes. He or she may then be asked to walk with eyes closed on mats with objects placed randomly

beneath the mats to increase focus on the somatosensory information. For persons who overweight somatosensory information, efforts might be reasonably directed toward up-weighting visual and vestibular inputs.

If MDDS is instead caused by an internal oscillator, the treatment strategy should logically aim at manipulation of cognitive variables. Patients need to ignore their aberrant internal signal, in the same way that persons with tinnitus deal with abnormal internally generated sounds. From this perspective, physical therapy that focuses attention on the rocking sensation could even be counterproductive. Controlled studies are needed regarding the effect of physical therapy on MDDS.

## Summary

MDDS is an uncommon disorder in which an inappropriate sensation of rocking develops that is paradoxically relieved by actual motion, such as driving. Research is needed to establish the role of physical therapy in the treatment of MDDS. As of this writing, strategies based on provision of abundant and accurate sensory information about balance, possibly combined with attempts to improve somatosensory integration, appear most reasonable.

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CHAPTER 14

# Surgical Management of Vestibular Disorders

*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, the patient living with the symptoms must make the decision whether to proceed with surgery. The physician should discuss the likelihood of a successful outcome, as well as the nature of potential complications, and must leave the ultimate decision up to the patient. In the author's 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.

## Acoustic Neuromas (Vestibular Schwannomas)

*Acoustic neuromas* are nerve sheath tumors occurring in the internal auditory canal or cerebellopontine angle.[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 or sudden hearing loss.[2]

An acoustic neuroma should be suspected in any patient with an unexplained unilateral sensorineural hearing loss, particularly if the patient has abnormal brainstem auditory responses or hypoactive (or absence of) 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.

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 very 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 2 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 repeated 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 are 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. 14.1). 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 and temporal lobe are 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. 14.2). The tumor is approached as in a standard mastoidectomy. The cortical and pneumatized bone of the mastoid are drilled away

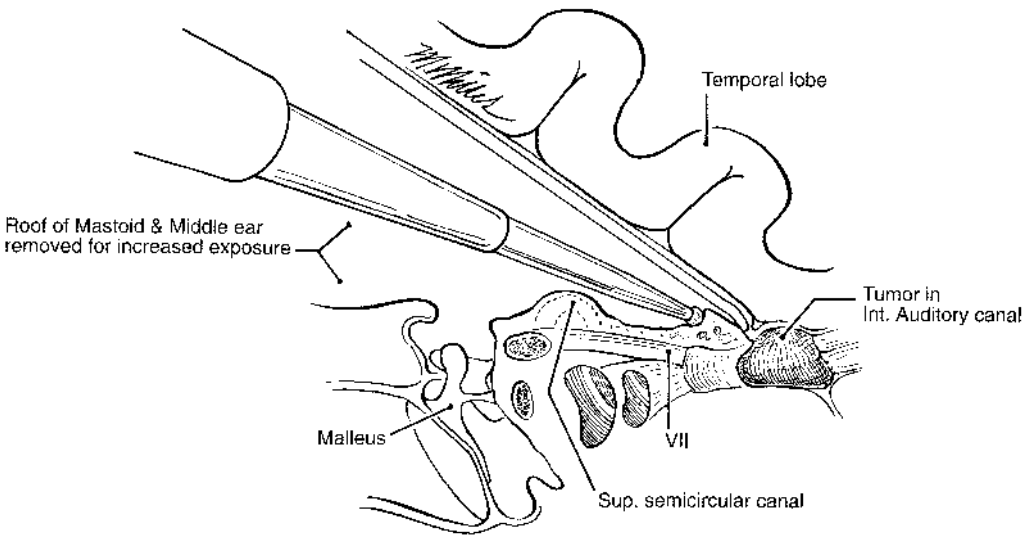


**Figure 14.1** Middle fossa. This coronal section through the internal auditory canal and middle ear shows the exposure of the internal auditory canal through the middle fossa. A drill is shown passing through a small temporal craniotomy. The temporal lobe is elevated extradurally. The superior semicircular 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.

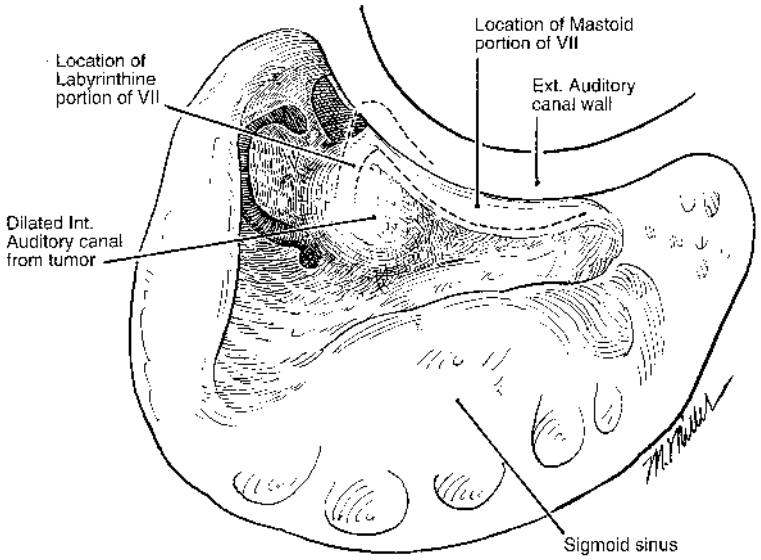


**Figure 14.2** Translabyrinthine approach for removal of acoustic neuroma. A translabyrinthine approach to the internal auditory canal is shown in surgical position with anterior—*top*; superior—*left*. The tumor is exposed after completion of a labyrinthectomy (see Fig. 14.5). The internal auditory canal has been dilated by the tumor. Removal of the last shell of bone over the tumor as well as the bone from the posterior fossa allows complete removal of the tumor.

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.

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 2.5 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 Craniectomy*

Suboccipital craniectomy gives the best exposure when the tumor is large or adherent to the brainstem (Fig. 14.3). 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 craniectomy 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 piece of the occipital skull is removed. This defect is reconstructed with prosthetic mesh at the end of the procedure.

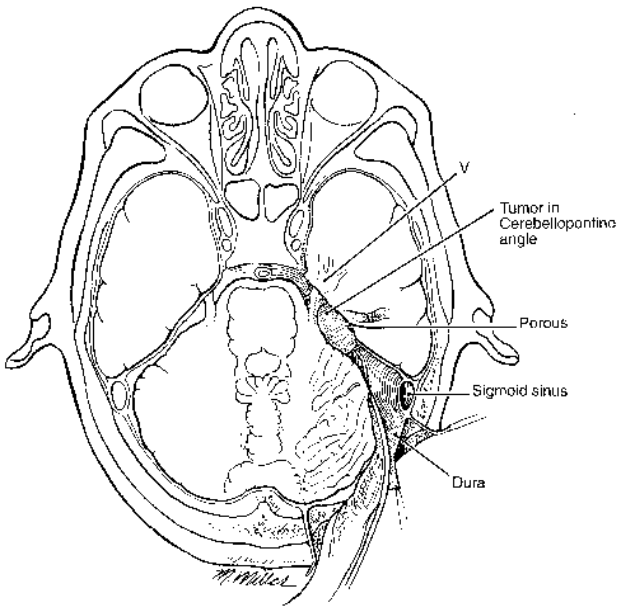


**Figure 14.3** Suboccipital craniectomy. This axial section demonstrates a posterior fossa craniotomy behind the sigmoid sinus. The cerebellum drops away from the posterior surface of the temporal bone. The tumor is exposed in the cerebellopontine angle. The posterior lip of the internal auditory canal must be drilled away to remove the tumor extending into the canal.

The cerebellum lies between the craniotomy and the cerebellopontine angle; 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 an ultrasonic aspirator or laser. After the tumor has been decompressed, the portion of the tumor within the internal auditory canal is removed by drilling away the posterior surface of the canal. The facial nerve is identified in the fundus of the internal auditory canal, where its anatomy is constant. The tumor is mobilized from the brainstem, and the VIIth 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*

Complications after acoustic neuroma surgery are relatively uncommon. Hearing loss always occurs in 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 while, with proper geometric planning, the surrounding neural and vascular structures are spared.

The initial 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 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 legitimate 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 them all. 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).[16]

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 surgi-

cal 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*

### 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.[18] 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 almost an understatement to say that endolymphatic sac surgery is controversial. The fluid spaces involved are minuscule, and the ability of mechanical means to improve function of the sac is doubtful. In a clinical trial, similar results were obtained with real and sham operations.[19] Nonetheless, the procedure controls the vertiginous attacks in one half to two thirds of patients and has the advantage of relative ease and safety.

### 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 60dB, 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 cranial 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. 14.4). In the retrolabyrinthine approach, a complete mastoidectomy is performed as described for the endolymphatic sac procedure.[20] In addition, all of the bone medial to the sigmoid sinus is removed to expose the posterior fossa dura. The dura is opened to expose the cerebellopontine angle. The vestibular and auditory branches of the VIIIth 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 VIIIth 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.[21]

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 (Fig. 14.5). 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, I 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.

## *Chemical Labyrinthectomy*

Chemical labyrinthectomy with aminoglycosides has gained popularity in the management of Ménière's dis-

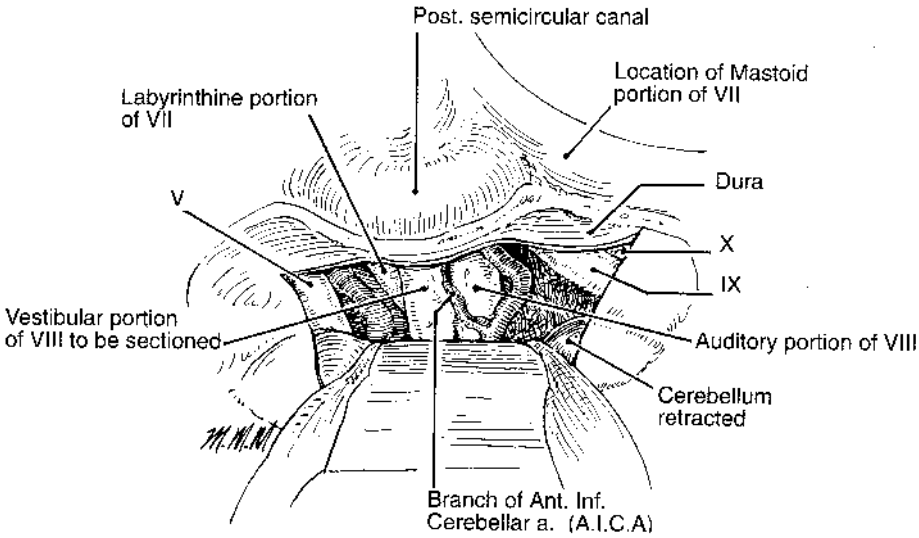


**Figure 14.4** Retrolabyrinthine vestibular nerve section. This surgeon's view of a right ear in surgical position (anterior—*top*; superior—*left*) shows the exposure for a retrolabyrinthine nerve section. The cerebellopontine angle is exposed by removal of the posterior surface of the temporal bone between the sigmoid sinus and the posterior semicircular canal. The dura is opened, and the VIIth and VIIIth 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 VIIIth nerve (superior half) is divided.

ease 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. [13,22] This procedure appears to produce good control of vertigo attacks but poses significant risk to hearing.[24] Cohen-Kerem and associates[25] 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.

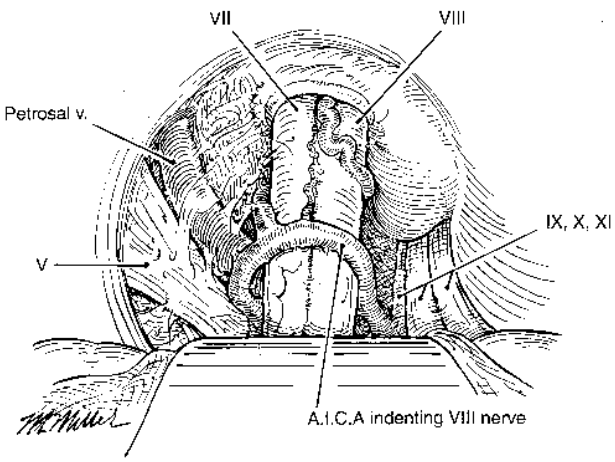


**Figure 14.5** Labyrinthectomy. A transmastoid labyrinthectomy of the right ear (anterior—*top*; superior—*left*). The three semicircular canals have been identified, and the lateral and posterior canals have been partially opened. The canals will be completely removed, and the vestibule will be opened to remove all neuroepithelium. AICA = anterior-inferior cerebellar artery; *Roman numerals* indicate cranial nerves.

Intratympanic administration of aminoglycosides has the great advantage of being nonoperative, thus avoiding the risks of anesthesia and surgery and the cost of hospitalization. Even though it is a simple procedure, however, 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 to a surgical intervention.

In all the procedures described, 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, however, are seldom, if ever,

indicated in patients who have active bilateral disease. Special care should be taken 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).[26]

## 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.[27] 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] The most common position is in a lateral or head-hanging position with the diseased ear down. The symptoms generally have a few seconds' latency 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 as well as 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 17).

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.[28] 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 not be violated. Bone is removed posterior and inferior to the round window membrane 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.[29] 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.[30,31]

## Perilymphatic Fistula

*Perilymphatic fistula* is a direct communication between the inner ear and the middle ear, usually through the round or oval window (Fig. 14.6).[29] Such leaks were initially described in association with barotrauma; however, spontaneous leaks are being described with increasing frequency. 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 some of the most 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 as well as protein analyses are being developed as potential markers for the presence or absence of the perilymph fistula.[33]

Middle ear exploration for perilymph fistula is straightforward. The middle ear is approached through the external auditory canal, and the tympanic membrane 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.[32]

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

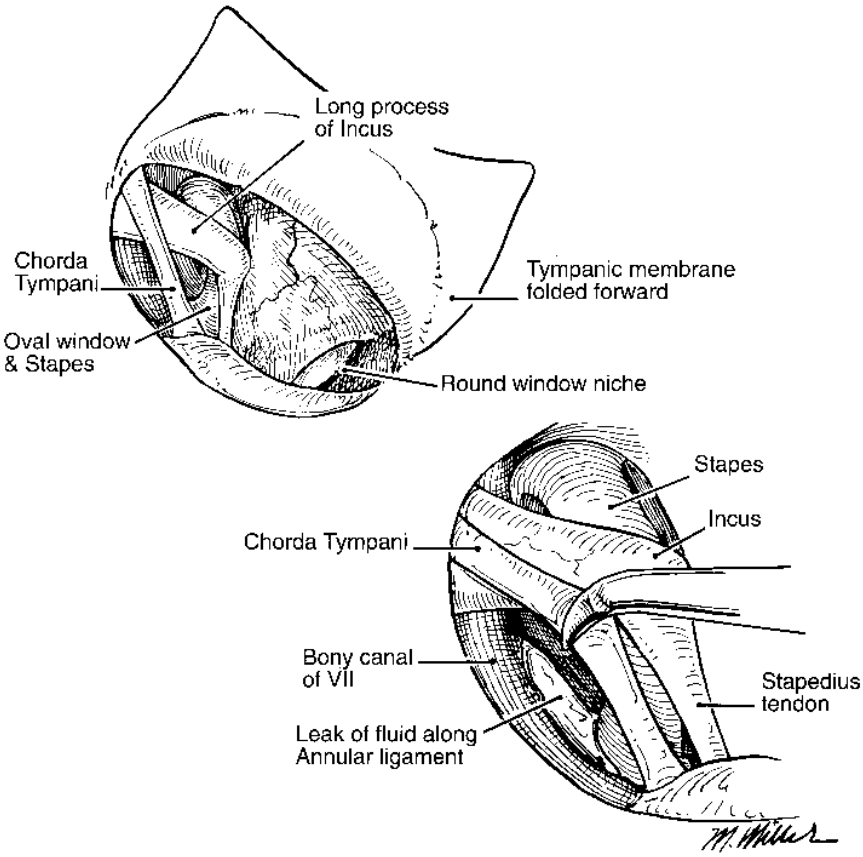


**Figure 14.6** Perilymphatic fistula. A right middle ear exploration for perilymph fistula is shown in the surgical position (anterior—*top*; superior—*right*). The tympanic membrane has been reflected forward to expose the oval and round windows. Close inspection of the annular ligament of the oval window demonstrates a leak of perilymph. This will be closed with an autologous tissue graft. VIIth = seventh cranial nerve.

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. 14.7). The first well-described vascular syndrome was hemifacial spasm, an uncontrollable twitching of one side of the face. This was found by Janetta and colleagues[34] 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.[35] The significance of these vascular loops is difficult to determine, because the symptoms overlap other diagnostic categories, including Ménière's disease and perilymphatic fistula. Furthermore, tortuous vessels are common in the cerebellopontine angle 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 radiologic confirmation can be obtained

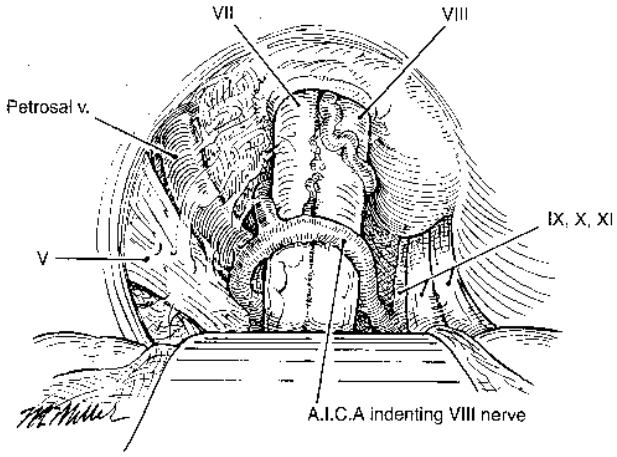


**Figure 14.7** Retrolabyrinthine exposure of vascular compression. The exposure is the same as shown in Figure 14–4. The anterior-inferior cerebellar artery (AICA) is seen compressing the VIIIth nerve. This compression is treated by careful elevation of the vessel and interposition of muscle or sponge material between the vessel and the nerve. *Roman numerals* indicate cranial nerves.

with the combination of high-resolution CT with intravenous and air contrast.

Microvascular loop decompression is performed through a standard posterior craniectomy. 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 toward the rehabilitation and functional restoration of the diseased inner ear.

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CHAPTER 15

# 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 them 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 non-vestibular 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 15-1 lists the prevalence in the United States.

■ Table 15-1 **PSYCHOLOGICAL DISORDERS IN THE U.S. POPULATION (NIMH)**

| Disorder | No. Affected in U.S. | % Affected in U.S. |
|---|---|---|
| Anxiety disorders: | 23.2 million | 12.6: |
| Phobia | | 10.9 |
| Obsessive-compulsive disorder | | 2.1 |
| Panic with and without agoraphobia | | 1.3 |
| Depressive disorders | 17.5 million | 9.5 |
| Somatization disorders | 400,000 | 0.2 |

### 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.[3–7] Few articles have sufficient detail to allow determination whether these deficits are truly due to 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 suggest 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.[5] Another article found discrepancies in the VOR but no caloric deficits in patients with panic disorder.[4]

## *Psychological Problems in Patients with Dizziness*

There is a high prevalence of unrecognized mood and psychological problems in dizzy patients, especially anxiety disorders.[8,9] Forty percent of all dizzy patients have psychological disorders.[1] Stein and coworkers[10] report that 15% of all dizzy patients meet the *Diagnostic and Statistical Manual of Mental Disorders*, 3rd edition (DSM-III), 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.[1,11] 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.[12] 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).[13] 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. "Space phobia,"[14] "motorist's disorientation syndrome,"[15] and phobic postural vertigo[16] have also been described as syndromes seen after vestibular defects. 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.[9] Patients without vestibular deficits more frequently have somatization disorders as well as more current and lifetime unexplained medical symptoms.

### Disability

Clark and associates[17] have 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 disorder 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.[18] 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 6th grade reading level (see Case Study 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.[19] The patient is given a paper form similar to the representation in Box 15-1. 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 (listed in Table 15-2).[20] A "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,[20] 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.[20,21] Whether there is a correlation between the sensory aspect

Box 15-1

**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.

## ■ Table 15-2 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 you 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.

of posturography and DHI is controversial.[21,22] 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[23] 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 15-3). 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 not used them in our clinic, but 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.[24]

Beck Anxiety Inventory[25] and the Beck Depression Inventory[26]: 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.

■ Table 15-3 **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 one 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.

## 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[27] have identified six characteristic clinical features that are useful in the diagnosis of psychogenic stance and gait disorders. Table 15-4 lists these features and their prevalence in the 37 patients in the Lempert study. This

■ Table 15-4 **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 |

review found psychogenic gait disorders in 9% of their neurologic inpatients. In 5 years, 47% had favorable outcomes.[16]

### 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.[28] 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.[29] 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.[30–32] Box 15-2 lists the key patterns in such patients. Figure 15.1 compares the sensory test results of a patient with a psychogenic balance disorder with those of a patient with an acute UVL.

Box 15-2

**KEY PATTERNS ON DYNAMIC POSTUROGRAPHY IN PATIENTS WITH PSYCHOGENIC BALANCE DISORDERS**

- Substandard performance on sensory tests 1 and 2[31,32]
- Better performance on harder tests (sensory 5 & 6) compared with easier tests (sensory 1 & 2)[31]
- Large intertrial variability[30]
- Repetitive anterior-posterior sway without falling (voluntary sway)
- Large amplitude (> 5 degrees) anterior-posterior sway without falling on sensory tests 4–6[30]
- Large amplitude lateral sway (> 1.25 degrees) on sensory tests 4 – 6[30]

# 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.[33,34] Panic attacks consists of discrete spells of intense fear or discomfort, in which at least four of the symptoms listed in Box 15-3 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 handwashing 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 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 2 and 3 at the end of this chapter).

## *Mood Disorders*

Minor depressive disorders have the following criteria: At least five of the symptoms listed in Box 15-4 must be present during the same day and must represent a change from previous functioning; in addition, the patient must have either "Depressed mood" or "Loss of interest or pleasure in usual activities, including social contact." The disturbance causes significant impairment in social or

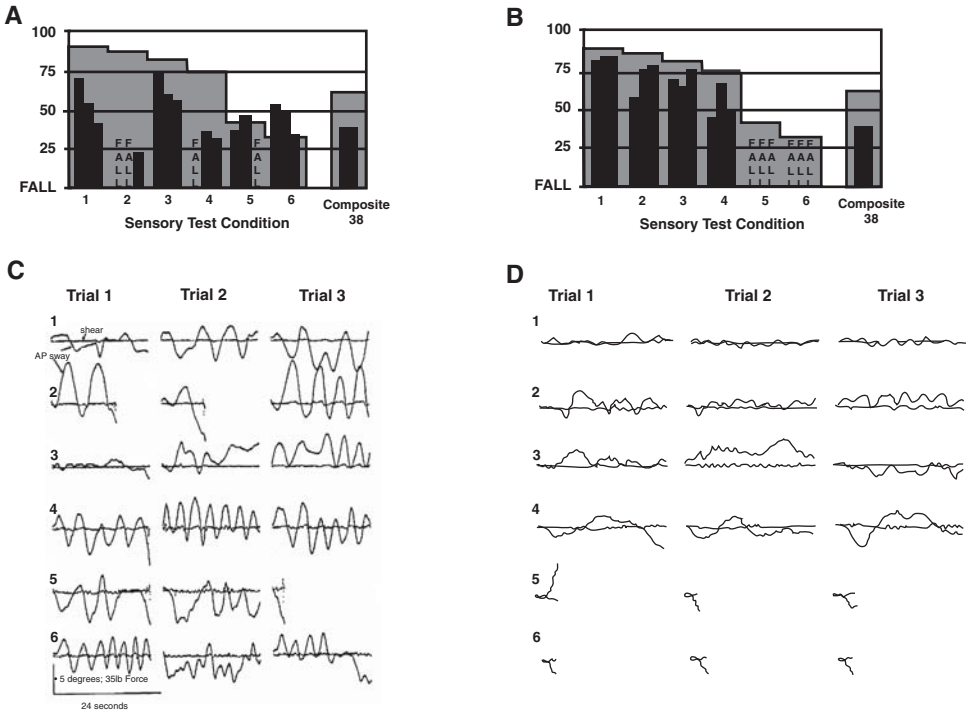


**Figure 15.1** Sensory tests during dynamic posturography in a patient with psychogenic balance disorder (*A* and *C*) and a patient with an acute unilateral vestibular loss (*B* and *D*). *A* and *B* show the mean sway on the 6 sensory test conditions repeated three times (100 indicates no sway; 0 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.

occupational functioning, or marked distress. There is no substance abuse and no manic episodes.

## *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.[35]

### 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 4).

Box 15-3

**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

Box 15-4

**SYMPTOMS CONSISTENT WITH DEPRESSION**

- Change in appetite or weight (down or up)
- Difficulty sleeping or sleeping too much
- Loss of energy, fatigability, or tiredness
- Psychomotor agitation or retardation
- Loss of interest or pleasure in usual activities, including social contact
- Feelings of self-reproach or excessive or inappropriate guilt
- Diminished ability to think or concentrate
- Recurrent thoughts of death or suicide
- Tearful or sad face
- Pessimistic attitude
- Brooding about past or current unpleasant events
- Preoccupation with feeling of inadequacy
- Resentful, irritable, angry, or complaining
- Demanding or clinging dependency
- Self-pity
- Excessive somatic concern

### *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 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.[36]

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 15-5).

Box 15-5

## 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.

### *Medications*

Physicians may want to start the patient on medication, as listed in Table 15-5. 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 15-5. After 3 weeks, I either taper the intermittent medication completely or limit its use to 1 or 2 days per week at most.

■ Table 15-5 **MEDICATION FOR PSYCHOLOGICAL PROBLEMS**

| 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 qam | |
| Zoloft (sertraline hydrochloride) | 50–100 mg qam | |
| Tofranil (imipramine) | 10–75 mg/day | |
| Norpramin (desipramine) | 50–100 mg/day | |

## CASE STUDY 1

A 51-year-old woman is referred for evaluation prior to 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 neurologic findings are otherwise normal except for gait, which is shown in video clip ‘Aphysiological gait 1’. 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; low gain and high phase for low-frequency sinusoidal chair rotation but normal values at high-frequency rotations. The patient’s Millon Inventory is shown in Figure 15.2.

*Comment*

This patient shows evidence of good central compensation of a left vestibular defect based on DVA and rotary chair testing (see Chapter 8 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 have 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 removal, 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.

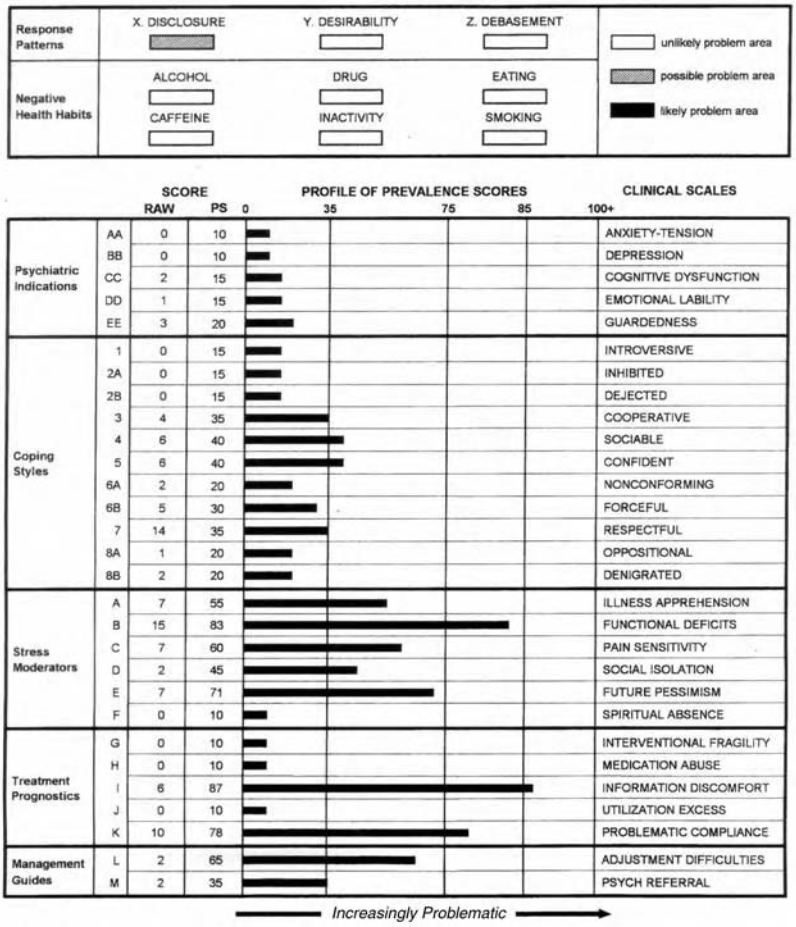


**Figure 15.2** Sample of the Millon Inventory in a dizzy patient.

## CASE STUDY 2

A 37-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, 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, 0.25mg qhs × 3 weeks and Paxil, 10 mg qam, and told to restart his exercise program.

## CASE STUDY 3

A 27-year-old repairman 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 VIIIth 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 are also discussed. He is encouraged to restart a regular exercise program to help reduce stress. He is started on Paxil, 10 mg qam, and Klonopin, 0.5 mg qpm; 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 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, results of both of which 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 neurologic disorder but no disorder was found. She has a psychogenic stance and gait disorder with several of the features described in Table 15-4. 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 5

A 45-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 neurologic 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 15-4). 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 fact 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 therapy as expected, the therapist should consider a significant psychological problem. Re-evaluation by the physician may be indicated.

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F·O·U·R

# Rehabilitation Assessment and Management

CHAPTER 16

# Physical Therapy Diagnosis for Vestibular Disorders

*Susan J. Herdman, PT, PhD, FAPTA*

As our profession assumes more independence in patient management, it becomes increasingly important that we prepare ourselves to screen patients for a multitude of problems and to make decisions about treatment and referral. "Dizziness" is one of the most prevalent complaints for which people seek medical help,[1] with an estimated 90 million Americans older than 16 years (roughly 42% of the population) having experienced it.[2] Dizziness can have significant consequences for an individual; 33% of patients with dizziness report that it affects their professional activities, and 14% that they have changed or abandoned their profession.[2] Although dizziness can be caused by many different medical conditions, it is estimated that as many as 45% of cases are due to vestibular disorders.[2]

This chapter provides a decision-making strategy that can be used when the clinician suspects that a patient may have a vestibular deficit. 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 therapy (PT) diagnosis differs from a medical diagnosis in that, rather than an attempt to identify a particular disease, a constellation of symptoms and signs is identified toward which physical and occupational therapy will be directed. Once the PT diagnosis is achieved, the vestibular exercise approach can be identified (Table 16-1). Certainly there are times when the medical diagnosis and the PT diagnosis are the same, for example, in benign paroxysmal positional vertigo (BPPV). However, there are also times when the diagnoses differ. For example, vestibular neuronitis is a medical diagnosis. As therapists, however, we do not treat the inflammatory process of vestibular neuronitis. A more appropriate diagnosis for PT purposes would be vestibular hypofunction.

## Physical Therapy Diagnosis and the International Classification of Functioning, Disability, and Health Model of Diasablement

The International Classification of Functioning, Disability, and Health Model of Disablement (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.[4] This scheme consists of three domains that can be used to describe the effect 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 16-1).

In the diagnostic schematic presented here, PT Diagnosis is determined from information gained at the level of the body (normal structure and function versus impairment). Modifiers for treatment are identified at the level of the individual (activities and limitations) and from contextual factors. Outcome of treatment would

**■ Table 16-1 DIAGNOSIS-DRIVEN TREATMENT**

| Diagnosis | Treatment Options |
|---|---|
| Benign paroxysmal positional vertigo | Canalith repositioning, Liberatory, Brandt-Daroff, Appiani maneuvers |
| Unilateral vestibular hypofunction | Adaptation, substitution (habituation) |
| Motion sensitivity | Habituation |
| Bilateral vestibular loss | Substitution, adaptation |
| Central vestibular | Habituation |

then be measured at the level of the individual and at the societal level.

The diagnostic schematic presented here is a "work in progress." It is offered as a framework for arriving at a physical diagnosis for patients with vestibular problems. Each of the PT diagnoses presented should demonstrate commonalities among all persons with that diagnosis. There are two phases to making the 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.

## History

Of special importance in the patient's history are the nature of the patient's symptoms and the temporal quality of those symptoms. The *nature of the symptoms* refers to whether the patient is complaining of vertigo (an illusion of movement, typically vertigo or spinning) or of disequilibrium (the sense of being off-balance). 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 episodes; for example, episodes of vertigo lasting seconds or less than 1 minute suggest BPPV.

## Clinical Examination

The first of the simple clinical tests of the vestibulo-ocular system 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 are used to diagnose positional vertigo that is peripheral (e.g., BPPV; see Chapter 17) 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 7 and 19). 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 after a head thrust to the right, the vestibular loss

**Box 16-1**

**HEALTH CONDITION**

| Normal Function and Structure vs. Impairment (body level) | Activities vs. Limitations (individual level) | Participation vs. Restriction (societal level) |
|---|---|---|

**Contextual Factors**

| Environmental Factors: | | Personal Factors: |
|---|---|---|
| Natural environment | | 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 |

is on the right. A person who makes corrective saccades with head thrusts to the right and to the left has 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 do not have a positive head-thrust test, but the patient who does have a positive test almost certainly has a vestibular deficit. The findings in the Dizziness and Balance Center at Emory University with patients with bilateral vestibular loss suggest that the sensitivity of the head-thrust test may be a higher for this patient group.

Another simple clinical test that can be used to assess vestibular function is the measure of visual acuity during horizontal head movement (dynamic visual acuity [DVA]; see Chapter 19). The sensitivity of the clinical DVA test for vestibular deficits is approximately 85%, and its specificity is 55%.[6]

## Diagnostic Flowchart

The first question one must ask is whether or not the patient has complaints of vertigo (Fig. 16.1). If the patient has a history of vertigo (spinning), the next step is

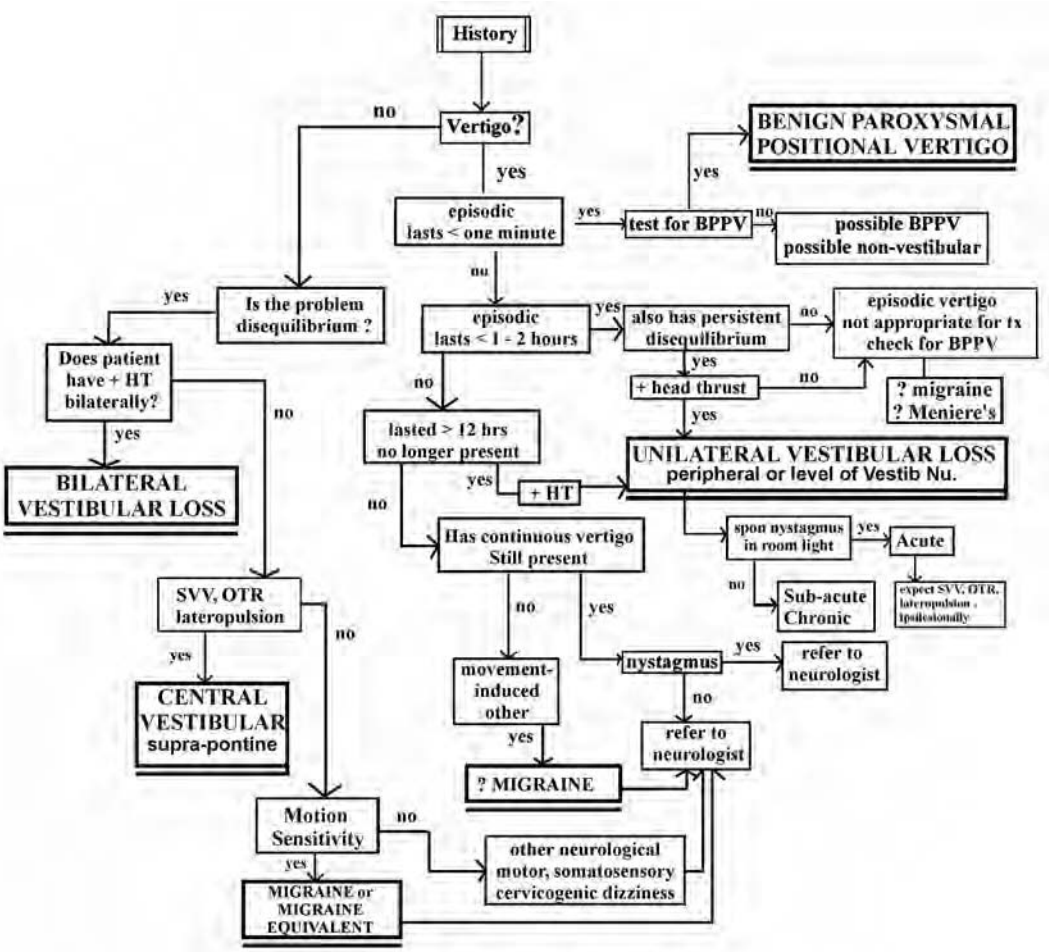


**Figure 16.1** This flow diagram begins with the history (*center*). Patients should be asked whether they have experienced vertigo (a sense of spinning). If the answer is "yes," the next series of questions deals with the temporal nature of that vertigo. (*Middle column*: <1 minute, 1–2 hours, >12 hours, continuous). The temporal quality of the vertigo then leads to diagnoses of benign paroxysmal positional vertigo (BPPV), episodic vertigo, unilateral vestibular loss (UVL), continuous vertigo, and motion sensitivity. Note that some of these diagnoses are appropriate for physical therapy treatment and others are not. If, during history-taking, 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 on basis of clinical findings (*left column*). Note how the presence or absence of corrective saccades to rapid head thrusts (HT) is used to identify unilateral and bilateral vestibular loss. OTR = ocular tilt reaction; SVV = subjective visual vertical; tx = treatment.

to determine the duration of the vertigo. Spells of vertigo lasting for brief periods (< 1 minute) suggest BPPV, the most common cause of vertigo due to a peripheral vestibular problem (see Chapter 17 for the diagnostic flowchart for BPPV). It is important to distinguish whether the nystagmus is typical for BPPV (combined torsional and vertical, or horizontal) or for a central lesion (pure vertical), or whether the history suggests another problem, such as a perilymphatic fistula.

One should note that positional testing may not provoke vertigo and nystagmus; however, this result 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, through history alone. Also, 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 lasts for extended periods 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 unilateral or bilateral vestibular hypofunction (see Chapters 20 and 21).

Vertigo that lasts for a few hours is most likely to be from Ménière's disease (see Chapters 6 and 12) or may be a migraine-related event (see Chapter 12). These types of episodic vertigo are not appropriate for treatment with vestibular rehabilitation and must be managed medically. The exception would be the patient who has persistent complaints of imbalance between these episodes of vertigo, in whom unilateral vestibular hypofunction might have developed with repeated episodes of Ménière's disease. The head-thrust test may help identify this problem, although formal vestibular function tests may be necessary.

Vertigo lasting 12 hours to days typically signifies the sudden onset of unilateral vestibular hypofunction (see Chapter 6). 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 whether there is central involvement.

Continuous vertigo has several possible explanations. First, the patient may be in the acute phase after 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. 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, and the patient should be referred to a neurologist. Unfortunately, looking for nystagmus with the patient looking at a Ganzfeld may not be sufficient for this examination unless the clinician 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 after 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 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 unilateral vestibular hypofunction (positive response to one side only) or bilateral vestibular loss (positive response to 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.

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 abnormal subjective visual vertical and an ocular tilt reaction), they probably have a central vestibular problem above the level of the vestibular nuclei (see Chapter 25). The complaint of disequilibrium may be used by a patient to describe motion sensitivity, leading to questions as to whether or not 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 on the basis of history and results of positional testing and the head thrust test, it is necessary to perform other assessments to identify other problems that will modify the exercise program for the individual patient. These modifiers

**■ Table 16-2 MODIFIERS OF TREATMENT**

| | |
|---|---|
| Activity level | Basic activities of daily living (ADLs)—bath, dress, meals, cleaning<br>Balance—gait speed, fall risk, endurance<br>Fall history—circumstance, frequency, injury<br>Driving—day, night<br>Work—harder to perform, changed jobs, not working |
| Personal factors | Visual—cataracts, macular degeneration, field<br>Somatosensory—peripheral neuropathy<br>Musculoskeletal—cervical, back, arthritis, strength, range of motion<br>Central nervous system—stroke (e.g., brainstem), cerebellar disease<br>Gender<br>Psychological:<br>Coping strategy<br>Anxiety, depression<br>Perception of disability<br>Somatoform, conversion<br>Severity of subjective complaints<br>Secondary gain issues |
| Environmental factors | Family support<br>Availability of transportaion<br>Location of stairs, bedroom, bathrooms |

**■ Table 16-3 FUNCTIONAL ASSESSMENT**

| Problem | Tests |
|---|---|
| Subjective complaints | Visual analog scales<br>Symptom scale<br>Dizziness Handicap Inventory (DHI) |
| Visual acuity during head movement | Clinical Dynamic V Acuity (DVA) test<br>Computerized DVA test |
| Balance in stance | Romberg test<br>Sharpened Romberg test<br>Single-leg stance |
| Balance while ambulating | Qualitative gait description<br>Dynamic Gait Index<br>Gait deviations<br>Gait with head turn |
| Quality of life | Vestibular disorders—Activities of Daily Living (ADL) Scale<br>DHI<br>Disability Scale<br>Medical Outcome Study 36-Item Short-Form Health Survey |

include disorders affecting other systems, subjective complaints, psychological factors, and the patient's premorbid activity level (Table 16-2). Finally, specific assessments 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 16-3).

## Summary

The use of the paradigm presented in this chapter should enable the therapist to arrive at a diagnosis in which all patients have a common group of symptoms and signs and that will indicate the appropriate treatments for an individual patient. The complete examination, including assessment of 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 17

# Physical Therapy Management of Benign Positional Vertigo

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Benign paroxysmal positional vertigo (BPPV) is perhaps the most common cause of vertigo in adults and, fortunately, is a very simple disorder to manage.[1,2] Symptoms remit with one treatment in more than 85% of cases. This chapter describes the etiology of BPPV, treatment for the various forms of the disorder, and the research that addresses treatment, and then provides suggestions for how to approach more complicated cases of positional vertigo.

## Characteristics and History

BPPV is characterized by brief episodes of vertigo when the head is moved into certain positions. It has been reported in adults of all ages (Table 17-1), although it is uncommon in children.[1,3] BPPV occurs spontaneously in many patients but may follow head trauma, labyrinthitis, or ischemia in the distribution of the anterior vestibular artery.[3] Spontaneous remission is common. For patients in whom the episodic vertigo persists, this disorder can be annoying, disruptive, and often leads to 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 are balance problems that may last for hours or days after the episodic vertigo has stopped as well as more vague sensations, such as lightheadedness and a feeling of floating (Table 17-2).

### *Mechanism*

Schuknecht,[2] in 1969, proposed that degenerative debris from the utricle (possibly fragments of otoconia) adhere to the cupula of the posterior canal, making the ampulla gravity sensitive. This theory, cupulolithiasis, was supported by the presence of basophilic deposits on the cupula of the posterior canal in patients with a history of BPPV. 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 (Fig. 17.1). The result is vertigo, nystagmus, and nausea. Because the cupula remains deflected as long as the patient is in the provoking position, the nystagmus and vertigo persist, although the intensity may decrease slightly because of central adaptation or because the patient may also have canalithiasis.[5] Thus, cupulolithiasis is characterized by (1) immediate onset of vertigo when the patient moves into the provoking position, (2) the presence of a nystagmus, which appears with the same latency as the complaints of

**■ Table 17-1 AGE DISTRIBUTION OF BENIGN PAROXYSMAL POSITIONAL VERTIGO (BPPV)***

| Age (yr) | Patients with Complaints of Dizziness (No.) | *Patients with BPPV* | |
|---|---|---|---|
| | | Percentage | No. |
| 0–9 | 9 | 0.0 | 0 |
| 10–19 | 32 | 3.1 | 1 |
| 20–29 | 64 | 3.1 | 2 |
| 30–39 | 191 | 17.8 | 34 |
| 40–49 | 261 | 16.5 | 43 |
| 50–59 | 207 | 22.2 | 46 |
| 60–69 | 298 | 26.2 | 78 |
| 70–79 | 376 | 23.7 | 89 |
| 80–89 | 176 | 33.1 | 58 |
| 90–99 | 14 | 50.0 | 7 |

*Cases seen at the Dizziness and Balance Center, University of Miami Dept of Physical Therapy and Bascom Palmer Eye Institute, 1994–98.

**■ Table 17-2 FREQUENCY OF COMPLAINTS IN 100 CONSECUTIVE PATIENTS WITH BENIGN PAROXYSMAL POSITIONAL VERTIGO (BPPV)**

| Complaint | 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 |

Adapted from Tusa and Herdman, 1998.[4]

vertigo, and (3) persistence of the vertigo and nystagmus as long as the patient's head is kept in the provoking position. This form of BPPV is relatively uncommon.

A second theory, canalithiasis, was proposed by Hall and colleagues,[7] who suggested that the degenerative debris is not adherent to the cupula of the posterior canal but, instead, is floating freely in the endolymph of the canal. Freely moving debris has been visualized through a microscope in patients in whom the membranous labyrinth has been exposed during surgery.[8] When the head is moved into the provoking position, the otoconia move to the most dependent position in the canal (see Fig. 17.1). The movement of the otoconia results in movement of the endolymph, which in turn pulls on the cupula and changes the firing rate of the neurons of that canal. The 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 extent of deflection of the cupula. Vertigo and nystagmus stop as the position is maintained because the endolymph ceases to move. Thus, BPPV from canalithiasis is characterized by (1) delay in the onset of the vertigo of 1 to 40 seconds after the patient has moved into 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, which increase and then decrease, disappearing within 60 seconds. BPPV from canalithiasis is the most common form of this disorder.

## *Semicircular Canal Involvement*

Initially, BPPV was believed to involve only the posterior semicircular canal (SCC), because the most commonly observed nystagmus, upbeat and torsional, would be produced by excitation of the posterior canal. It has now been recognized that involvement of anterior and horizontal canals also occurs.[9–14] The frequency of BPPV in each of the canals is shown in Table 17-3. Which canal or

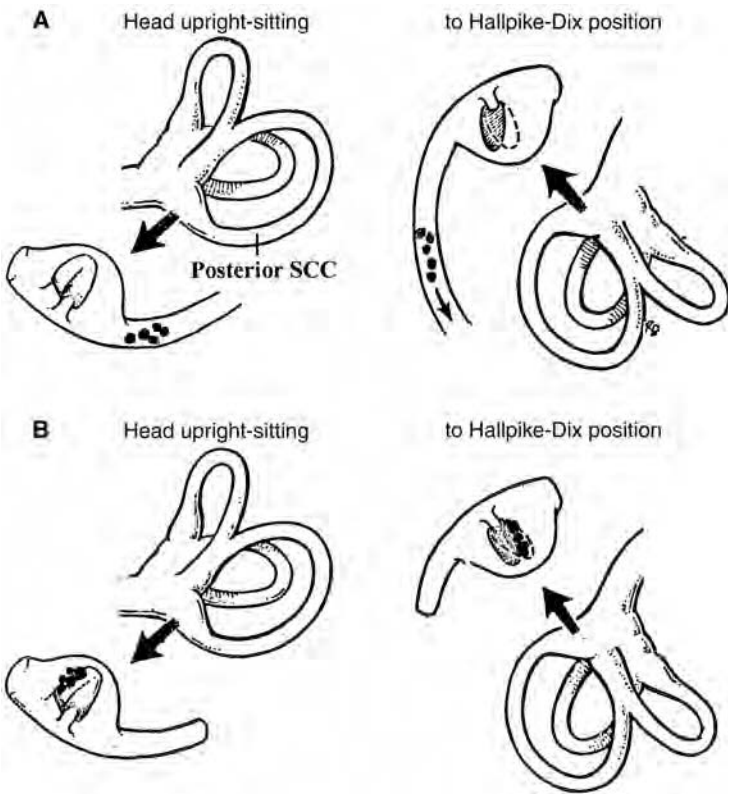



**Figure 17.1** (*A*) In canalithiasis, the calcium carbonate crystals are floating freely in the long arm of the canal (shown here for posterior canal). When the head is moved into the head-hanging position (Hallpike-Dix test), the debris moves to the most dependent portion of the canal. The movement of the debris causes the endolymph to move, in turn overcoming the inertia of the cupula, and an abnormal signal is sent to the central nervous system. (*B*) In cupulolithiasis, the debris is adhering to the cupula of the semicircular canal (shown here for posterior canal). With movement into the head-hanging position, gravity displaces the weighted cupula, again resulting in an abnormal signal from that canal. (Modified from Herdman SJ et al, 1993.[5])

canals are involved can be determined by the direction of the nystagmus when the patient moves into the provoking position (Table 17-4). Observation of the direction of the reversal phase of the nystagmus and the direction of the nystagmus when the patient returns to a sitting position can also be used to identify which canal is involved (see Table 17-4). Proper identification of the involved canal and determination whether cupulolithiasis or canalithiasis is the underlying problem dictate which treatment is appropriate.

## Diagnosis

Three different maneuvers can be used to test for the presence of BPPV. Because observation of the direction and duration of the nystagmus is critical to development of a treatment plan, the patient must understand what to expect. The patient should keep the eyes open and should try to stay in the provoking position. The examiner should explain to the patient that the vertigo will stop or decrease if he or she stays 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 slower performance decreases the likelihood of provoking the nystagmus.

■ Table 17-3 **CANAL INVOLVEMENT IN BENIGN PAROXYSMAL POSITIONAL VERTIGO (BPPV) IN 200 CONSECUTIVE PATIENTS SEEN IN OUR CLINIC**

| Semicircular Canal | % of Patients |
|---|---|
| Posterior | 76 |
| Anterior | 13 |
| Horizontal | 5 |
| Posterior or anterior SCC? | 6 |

Dizziness and Balance Center, Johns Hopkins University and University of Miami.

All tests for BPPV can be performed in room light, but the use of Frenzel lenses or an infrared camera system

## ■ Table 17-4 IDENTIFICATION OF CANAL INVOLVEMENT BASED ON DIRECTION OF NYSTAGMUS DURING RIGHT HALLPIKE-DIX TEST

| Canal | Right Hallpike-Dix Test | Reversal Phase | Return to Sitting |
|---|---|---|---|
| Right posterior | Upbeating and rightward torsional[a] | Downbeating and leftward torsional | Downbeating and leftward torsional |
| Right anterior | Downbeating and rightward torsional | Upbeating and leftward torsional | Upbeating and leftward torsional |
| Left anterior | Downbeating and leftward torsional | Upbeating and rightward torsional | Upbeating and rightward torsional |

[a]*Torsional* is defined as the direction movement of the superior pole of the eyes (e.g. in rightward torsional, the superior poles of both eyes move to the patient's right [quick phase]).

to prevent fixation suppression of horizontal and vertical nystagmus will raise the likelihood of observing nystagmus. Torsional nystagmus is not suppressed by fixation.

### *Dix-Hallpike Test*

The Dix-Hallpike test, sometimes called the Barany maneuver or the Nylen-Barany maneuver, is the test most commonly used to confirm the diagnosis of BPPV.[15] In this test, the patient's head is turned 45 degree horizontally while the patient is in the sitting position (Fig. 17.2 *position 1*). The patient then quickly lies down with the head hanging over the edge of the treatment table approximately 30 degrees below horizontal (Fig. 17.2 *position 2*). This maneuver places the posterior canal on 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 down, resulting in vertigo and nystagmus. In most cases of BPPV, nystagmus and vertigo occur within a few seconds of position change, but occasionally, they appear only after 30 seconds because of debris plugging within the canal. Therefore, the patient should be kept in this position for at least 30 seconds.

If the patient has BPPV, vertigo and nystagmus will be provoked when the affected ear is inferior. Then the patient can be slowly returned to a sitting position. If vertigo was provoked when the patient was moved into the head-hanging position, he or she may experience vertigo again when returned to a sitting position. The test can then be repeated with the patient's head turned to the other side. Note that the anterior canal of the downside ear is also in a more dependent position during this test, so the maneuver may also trigger vertigo due to anterior canal involvement of the downside ear (see Fig. 17.2

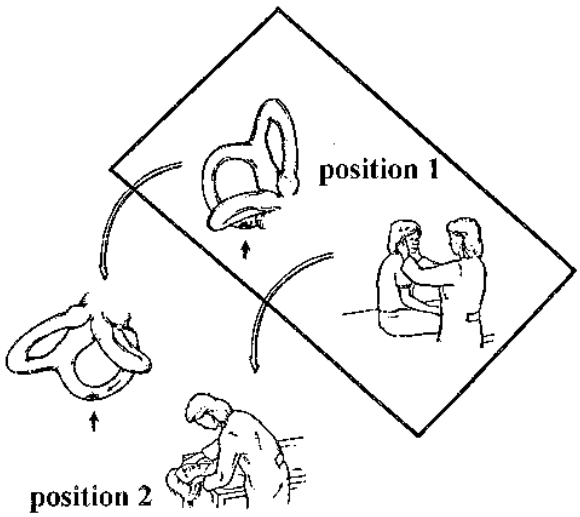



**Figure 17.2** Dix-Hallpike test for anterior or posterior canal benign paroxysmal positional vertigo. The patient sits on the bed or examination table, and the head is turned 45 degrees horizontally toward the labyrinth to be tested (*position 1*). The examiner quickly brings the head and trunk straight back "en bloc," so that the head is hanging over the edge of the examination table by 20 to 30 degrees (*position 2*). The examiner asks whether the patient has vertigo and observes for nystagmus. The patient's upper body is then brought up slowly to a sitting position with the head still turned 45 degrees, and nystagmus is sought again (*not shown*). This test then is repeated with the head turned 45 degrees in the other direction. This figure also shows the right labyrinth with free-floating otoconia in the right posterior semicircular canals (*large black arrows*). During the Hallpike-Dix test, this debris would move, resulting in nystagmus and vertigo when the test is performed to the right side but not when the test is performed to the left side. (Modified from Tusa and Herdman, 1998.[4])

*position 2*). The nystagmus in anterior canal BPPV is downbeat and torsional.

## Side-Lying Test

In the side-lying test, the patient sits on the side of the treatment table (Fig. 17.3 *position 1*). The head is turned 45 degrees to one side, and the patient then quickly lies down on the opposite side (Fig. 17.3 *position 2*). This again puts the posterior canal on the downside ear in the plane of the pull of gravity and may provoke a response in either canalithiasis or cupulolithiasis. Similarly, debris in the anterior canal of the downside or inferior ear also moves, provoking vertigo and downbeat and torsional nystagmus. The patient then returns to a sitting position. After waiting to be sure that the patient does not experience any vertigo in sitting, the test is repeated to the opposite side. The side-lying test can be substituted for the Dix-Hallpike test if the patient cannot extend the head sufficiently (head-hanging position in the Dix-Hallpike test) or cannot lie on the back.

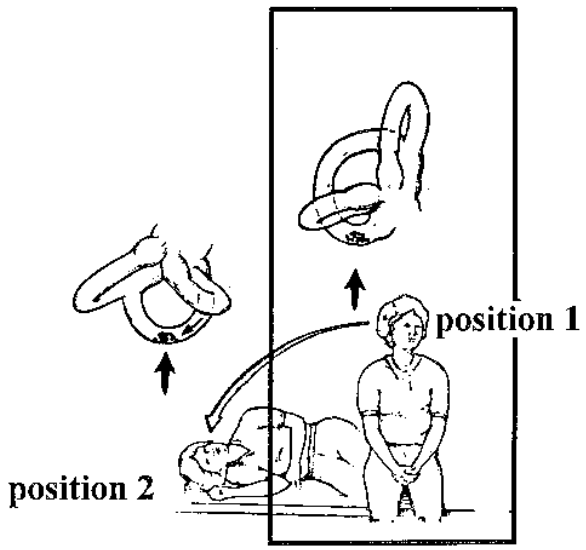



**Figure 17.3** Side-lying test for anterior or posterior canal benign paroxysmal positional vertigo. The patient sits on the bed or examination table with the legs over the side, and the head is rotated 45 degrees horizontally away from the labyrinth to be tested (*position 1*). The examiner then quickly brings the patient's head and trunk down on the side opposite to the direction the head is turned (*position 2*). The patient is asked to report any vertigo and is observed for nystagmus. The patient is then brought to a sitting position with the head still turned 45 degrees, and the examiner rechecks for nystagmus and vertigo (*not shown*). The test is repeated with head turned 45 degrees horizontally to the other side. This figure also shows the right labyrinth with free-floating otoconia in the right posterior SCC (*large black arrows*). During the Side-lying test, the debris would move, resulting in nystagmus and vertigo when the test is performed to the affected side but not when it is is performed to the unaffected side. This test is also useful for anterior canal BPPV, because debris in this canal would move when the test is done on the affected side. (Modified from Tusa and Herdman, 1998.[4])

## Roll Test

In patients with horizontal canal BPPV, the Hallpike-Dix test may not provoke vertigo and nystagmus.[11] The best maneuver would be one that moves the patient's head in the plane of the horizontal canal, which the roll test does. The patient lies supine with the head flexed 20 degrees (Fig. 17.4*A*). Then the head is quickly rolled to one side and kept in that position for up to 1 minute to test whether the patient experiences any vertigo (Fig. 17.4*B*). The head is then slowly rolled back to midline (still in slight flexion) (Fig. 17.4*C*) and then quickly rolled to the other side (Fig. 17.4*D*). In horizontal canal BPPV, vertigo and nystagmus occur when the head is turned to both the right and the left, because the debris moves back and forth within the canal. The slow-phase eye velocity, duration of the nystagmus, and the patient's subjective complaints are believed to be worse when the head is turned toward the affected ear.[11] The direction of nystagmus in horizontal canal BPPV depends on whether the debris is free-floating (canalithiasis) or adhering to the cupula (cupulolithiasis) (Fig. 17.5).[13] In a patient with canalithiasis of the horizontal canal, the nystagmus is geotropic (quick phases of nystagmus beat to earth) and will fatigue, whereas in a patient with cupulolithiasis, the nystagmus is apogeotropic (beats away from earth) and is persistent.

## Test Series

One can easily assess all three canals for BPPV quickly with the following test procedure. For sake of discussion, assume that the patient complains of vertigo when lying on the right side. The series is as follows:

1. Perform the Dix-Hallpike test on the left side.
2. Perform the Dix-Hallpike test on the right side.
3. If the patient has no vertigo: Before sitting the patient up from the right side, perform a roll test by having the patient turn the head quickly to the left.
4. After 30 seconds, have the patient quickly turn the head back to the right.
5. After 30 seconds, have the patient sit up.

If, at any time during this series of tests, nystagmus and vertigo appropriate for BPPV occur, the testing should be stopped, and the patient treated.

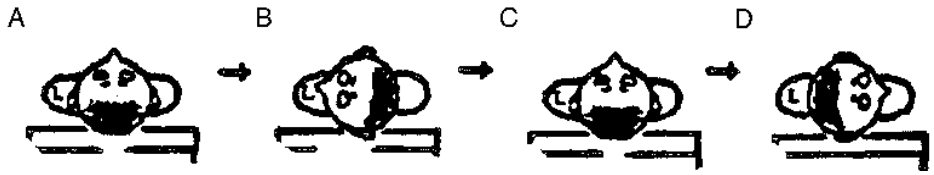



**Figure 17.4** Roll test for horizontal canal benign paroxysmal positional vertigo. (*A*) The patient is laid supine with the head flexed 20 degrees. (*B*) The head is quickly rolled to one side, nystagmus is looked for and the patient is asked to report any vertigo. (*C*) The head is then slowly rolled back to a supine position. (*D*) The head is then quickly rolled to the other side, nystagmus is looked for, and the patient is asked to report any vertigo. (Modified from Tusa and Herdman, 1998.[4])

## *Treatment*

Successful treatment depends 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, the Liberatory maneuver, and the Brandt-Daroff habituation exercises. Variations of these treatments have been developed depending upon which canal is involved. Studies on the efficacy of these treatments indicate that all three treatments facilitate recovery.[5,16–24] 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 authorities have reported spontaneous recovery within 3 to 4 weeks,[22,25] although Brandt and Daroff[26] suggest that the vertigo may disappear spontaneously after several months even if left untreated.

The choice of which exercise is most appropriate for a particular patient depends on which canal is involved as

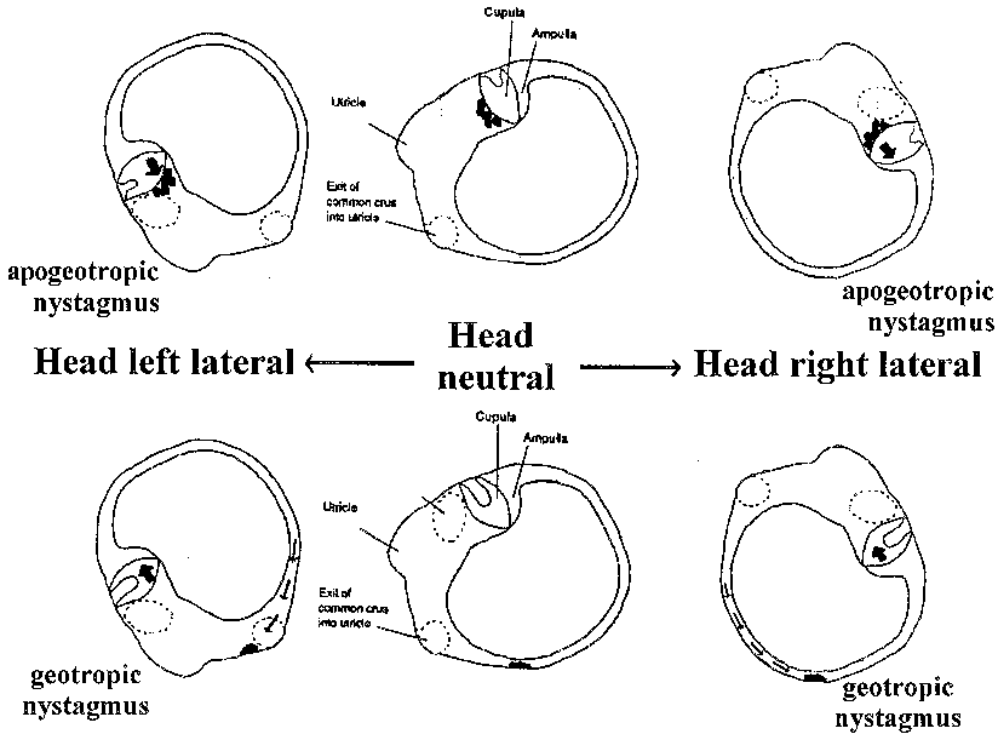



**Figure 17.5** Direction of nystagmus in horizontal canal (HC) benign paroxysmal positional vertigo. For both HC cupulolithiasis (*A*) and HC canalithiasis (*B*), the patient would have nystagmus and vertigo when the head is rolled to either side, but the duration and direction of the nystagmus would differ in these two types of BPPV. For cupulolithiasis, the nystagmus is persistent, and the direction of the quick phases is away from earth (apogeotropic). For canalithiasis, the nystagmus is transient, and the direction of the quick phases is toward the earth (geotropic). (Modified from Baloh et al, 1993.[11])

## ■ Table 17-5 TREATMENT FOR BENIGN PAROXYSMAL POSITIONAL VERTIGO: OPTIONS AND INDICATIONS*

| Semicircular Canal Involved | Severe Canalithiasis | Mild Canalithiasis | Cupulolithiasis |
|---|---|---|---|
| Posterior | CRT*<br>Liberatory maneuver<br>Brandt-Daroff exercises | Brandt-Daroff exercises*<br>CRT<br>Liberatory maneuver | Liberatory maneuver *<br>Brandt-Daroff exercises |
| Anterior | CRT*<br>Liberatory maneuver—AC<br>Brandt-Daroff exercises | Brandt-Daroff exercises*<br>CRT<br>Liberatory maneuver—AC | Liberatory maneuver—AC *<br>Brandt-Daroff exercises |
| Horizontal | Bar-B-Que roll treatment*<br>Forced prolonged position | Bar-B-Que roll treatment*<br>Forced prolonged position | Bar-B-Que (quick movements) |

*Preferred method is listed first.
AC = anterior canal; CRT, canalith repositioning treatment.

well as whether the patient has the canalithiasis or cupulolithiasis form of BPPV (Table 17-5). We typically use the canalith repositioning treatment (for canalithiasis) or the Liberatory maneuver (for cupulolithiasis) first. The Brandt-Daroff habituation exercises are used for milder residual complaints.

## *Treatment of the Most Common Form of BPPV: Posterior Canal Canalithiasis*

Consider the following case:

> The patient is a 66-year-old woman who experienced severe vertigo when she rolled over in bed in the morning about 12 days ago. The vertigo lasted only seconds, although she continued to feel off-balance for the next 24 hours. She avoided lying down after that incidence and has been sleeping on three pillows at night. Three days ago, she was seen by a neurologist, who diagnosed BPPV on the basis of the upbeat and rightward torsional nystagmus with concurrent vertigo that was elicited when she was moved into the right Dix-Hallpike position. He treated her with CRT and referred her to you for follow-up care. On examination today, she still has an upbeat and rightward torsional nystagmus with vertigo that lasts for 8 seconds when she is moved into the right Dix-Hallpike position. She states, however, that it is 50% better than it was when the neurologist treated her 3 days ago. You are concerned that the treatment was not done correctly.

In this section, we first describe the original components of the canalith repositioning procedure and then detail the modifications that have been proposed over the years. We then give instructions on how to perform CRT correctly.

### The Original Canalith Repositioning Procedure

The canalith repositioning procedure (CRP) was developed for treatment of posterior SCC BPPV and consisted of the following five key elements[17,26]:

**Premedication of the Patient.** Approximately 1 hour before treatment, the patient is given transdermal scopolamine or diazepam in order to reduce nausea and prevent vomiting during testing and treatment. The patient is then seated in a treatment chair for the actual maneuver.

**Specific Positions Used in the Maneuver Itself.** The specific sequence of head positions recommended involves moving the head in 90-degree increments so the "debris" will move through the long arm of the posterior canal and into the utricle (Fig. 17.6). Note that with each change in head position, the debris should move away from the cupula of the posterior SCC.

**The Timing of Shifts from One Position to Another.** The timing used for each change in position is based on the duration of the nystagmus observed during the maneuver. 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 no nystagmus is observed, the amount of time the head is maintained in that position is based on the duration of the

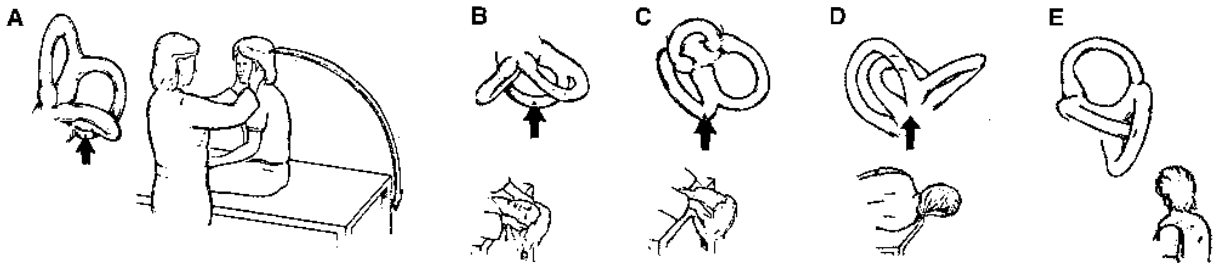



**Figure 17.6** Canalith repositioning treatment for treatment of posterior or anterior semicircular canal (SCC) benign paroxysmal positional vertigo. The patient is first moved into the Hallpike-Dix position toward the side of the affected ear (shown here for left) and kept in that position for up to 1 minute (*A* to *B*). Then, the head is slowly rotated through moderate extension toward the unaffected side and kept in the new position briefly (*C*) before the patient is rolled to a side-lying position with the head turned 45 degrees down (toward the floor) (*D*). In each of these positions, the patient may experience a short spell of vertigo and nystagmus with the same characteristics as the original nystagmus, indicating that the debris is moving through the posterior canal. With the head kept deviated toward the unaffected side and pitched down, the patient then slowly sits up (*E*). Some clinicans recommend having the patient wear a soft collar for the remainder of the day to ensure that the patient does not bend over, lie back, move the head up or down, or tilt the head to either side. If a soft collar is used, the examiner should tell the patient to turn the head from side to side every hour to avoid muscle spasm. *Black arrows* indicate location of free-floating debris in the posterior SCC. Note that the movement of the patient's head will gradually shift the debris away from the cupula and into the common crus. (Modified from Tusa and Herdman, 1998.[4])

last occurrence of nystagmus. In general, each position is maintained for between 6 and 13 seconds.[17] The maneuver is repeated until either nystagmus is no longer observed or there have been no changes in nystagmus for two repetitions.

**Use of Vibration during the Maneuver.** Vibration is applied to the mastoid region on the affected side during the repositioning maneuvers. In the original procedure, Epley[17] suggested that two different applications be used: 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.[17] 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.

**Post-procedure Instructions.** The last component of the CRP was the post-treatment instruction. The patient was advised to keep the head upright for 48 hours after the treatment. The follow-up visit for reassessment and another treatment, if needed, would be scheduled for 1 week later.

## Modification of the Canalith Repositioning Procedure

As more and more clinicians have used the CRP, numerous modifications of the procedure have been proposed (Table 17-6). These modifications encompass elimination of premedication regimens, as well as changes in positions used, timing, use of vibration, and post-treatment instruction. Some of the modifications resulted in less optimal outcome, and others simplified the CRP as proposed by Epley[17] without affecting outcome. We use the term *canalith repositioning treatment* (CRT) to distinguish the modified treatment approaches from the originally proposed procedure.

**Premedication.** 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 proposed by Epley,[17] most reports do not mention medicating patients prior to treatment.[5,21,24,27–29] We recommend that an exception would be the patient with a history of emesis associated with BPPV. In that situation, the use of an antiemetic medication such as promethazine (Phenergan) or prochlorperazine (Compazine) 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 nystagmus and vertigo occur during positional testing (Dix-Hallpike or Sidelying test), the patient should immediately be taken through the CRT rather than being subjected to repeated provoking tests.

## ■ Table 17-6 MODIFICATIONS OF THE CANALITH REPOSITIONING TREATMENT AND THEIR OUTCOME

| Study and No. of Subjects | Premedication | Positions | Timing of Position Changes | Mastoid Oscillation | Repetitions | Post-Treatment Instructions | Remission Rate |
|---|---|---|---|---|---|---|---|
| Epley, 1992[17]<br>n = 30 | Transdermal scopolamine or diazepam | 5-position cycle—includes nose down | Sum of latency and duration of nystagmus (typically 6–13 seconds) | 700 Hz for one cycle*; 80 Hz for at least one cycle | Until no nystagmus or no change over two repetitions | Upright for 48 hours; treatment repeated at weekly intervals | 100% |
| Epley, 1994[64] | Transdermal scopolamine or diazepam | 5 position cycle—includes nose down | Typically 6–13 seconds | 80 Hz applied indirectly; optional | Until no nystagmus or no change over two repetitions | Upright for 48 hours; treatment repeated weekly or in 1–2 days | 90% |
| Epley, 1996[26]<br>n = 400 | Transdermal scopolamine or diazepam | 5 position cycle—includes nose down | Typically 6–13 seconds but may be > 30 seconds | 80 Hz applied indirectly | Until no nystagmus or no change over two repetitions | Upright for 48 hours; treatment repeated weekly or in 1-2 days | 80% in one treatment; 95% overall |
| Herdman et al, 1993[5]: | | | | | | | |
| n = 30 | None | 4-position cycle to opposite H-D | 4 minutes in each position | None | One maneuver only | Upright for 48 hours | 57% |
| n = 30 | None | 4-position cycle to opposite H-D | 4 minutes in each position | None | Repeated maneuvers in same session | Upright for 48 hours | 92% |
| n = 30 | None | 5-position cycle to nose down | 3–4 minutes in each position | None | One maneuver only | Upright for 48 hours | 83% |
| Wolf et al, 1999[29]<br>n = 107 | None | 4-position cycle to opposite H-D | 4 minutes in each position | None | One maneuver only | Upright for 48 hours | 80% |
| Li, 1995[18]: | | | | | | | |
| (n = 10) | | | | No vibration | | | 0% |
| (n = 27) | | | | Vibration | | | 70% |
| Hain et al, 2000[33]: | | | | | | | |
| n = 50 | | | | No vibration | | | No significant difference |
| n = 44 | | | | Vibration | | | No significant difference |
| Sargent et al 2001[34]: | None mentioned | 5 position cycle to nose down | Pausing in each position | No vibration | Until no nystagmus or no change over two repetitions | Upright for 48 hours | 73% |
| n=104 | | | | Vibration (60 Hz) | | | 84% (p = 0.151) |
| n = 64 | | | | | | | |

*Later dropped from protocol.
H-D = Hallpike-Dix position

**Positions.** The original series of position changes suggested by Epley have remained unchanged. Herdman and colleagues[5] 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.[16] 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 or when the four-position maneuver was repeated several times, the outcome was significantly better. Herdman and colleagues[5] also showed that when the patient was moved once through all the positions suggested by Epley, remission of BPPV was 83%. When patients were moved through four positions multiple times, a 93% remission rate was achieved. This latter finding has been confirmed by Wolf and associates,[29] who found a 93.4% remission rate in patients treated with only the first four positions. Therefore, it seems that either approach—using five positions or using four positions but repeating the maneuver multiple times—is effective.

An interesting, and quite different, series of position changes has been suggested by Furman and coworkers.[30] Called the "heels-over-head" rotation, the patient sits in a treatment chair that can be rotated in pitch 360 degrees. The head is positioned at each stage of the maneuver so the movement is always in the plane of the posterior canal. All of the 11 patients in this study had complete remission of symptoms with the first treatment. Although effective, this approach requires relatively sophisticated equipment and seems to be unnecessarily complicated and awkward for the patient.

**Timing.** The timing of the position changes in the CRP was based on the combination of the latency until nystagmus begins plus the time until the nystagmus stops. If nystagmus is not observed in any given position, which happens frequently, then the timing is based on the duration established during the initial Dix-Hallpike maneuver. Several studies have used a longer period between each change in position,[5,29,31,32] with essentially the same results as by Epley[17]. 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 prevent actual emesis.

**Mastoid Oscillation.** Later studies have failed to identify any difference in outcome when a vibrator is used or not.[33, 34] In our practice, we do not routinely use mastoid oscillation. The exception is the patient who has shown no response to CRP. We think that in some patients (<5%), the debris does not move easily through the SCC. When that happens, the treatment is unsuccessful. We have found that keeping the orientation of the labyrinth well below the horizon during the position change from the initial Dix-Hallpike position (see Fig. 17.6*B*) to the second Dix-Hallpike position (see Fig. 17.6*C*) facilitates movement of the debris through the canal. We do this by either hyperextending the patient's neck 30 degrees during position *B* and *C* in Figure 17.6, or, in patients with limited neck range of motion or with cervical pain, by lowering the head end of the treatment table. If that fails, we use mastoid oscillation during the treatment. Contrary to other investigators,[33,34] we believe it is important to recognize that in a case of "canal jam," there would be no nystagmus or vertigo with change in head position, because the debris would block movement of the endolymph, and therefore, the cupula would not be displaced.

**Post-treatment Instructions.** Keeping the head upright for 48 hours, presumably to prevent the loose otoconia in the utricle from moving back into the posterior SCC, 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.[31,37,38] Massoud and Ireland[31] examined the need for the post-treatment instruction to stay upright for 48 hours. Two groups of patients were given CRT or another treatment (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 treatments but were not given any post-treatment instructions other than to avoid brisk head movements for 1 week. All patients had follow-up visits 1 week after treatment. These researchers found no statistical difference among the four groups (analysis of variance, $P > .2$). The success rate of the maneuvers was 88% to 96%. Similar results were reported by Nuti and colleagues,[38] who examined 52 patients at 20 minutes, 24 hours, and 7 days after the Liberatory maneuver for posterior canal BPPV. After the maneuver, the patients were asked to remain in a seated position for 20 minutes; 47 of the patients had no vertigo when retested at 20 minutes after the maneuver. After this first test, they were given *no* post-maneuver instructions. At 24 hours, 2 of the 47 patients had a recurrence and were re-treated. At 7 days, all 47 patients were still in remission. Nuti and colleagues[38] concluded that post-maneuver instructions are not necessary, including the use of a soft collar. We found similar results when we asked patients to sleep upright for 24, 48, or 72 hours (Fig. 17.7).[37]

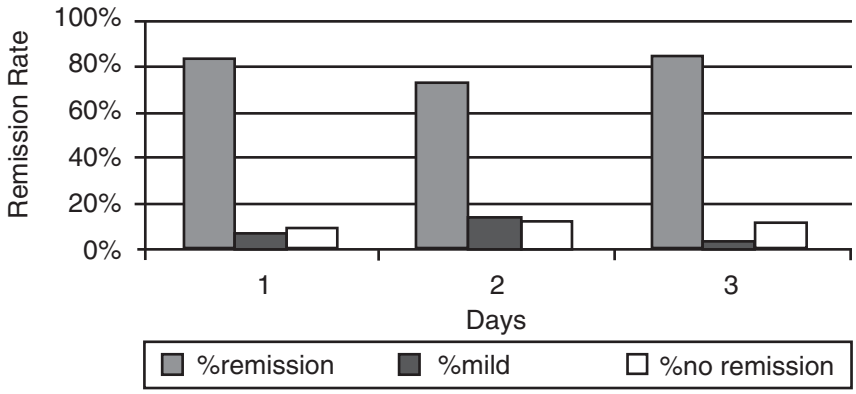



**Figure 17.7** Bar graph showing the outcome of canalith repositioning treatment for posterior semicircular canal benign paroxysmal positional vertigo based on the number of days the patients stayed upright after treatment. All patients were seen 7 days later. Outcome was determined from the presence of posterior canal nystagmus during the Dix-Hallpike test using video-infrared goggles and the occurrence of vertigo during the test. Cure was defined as no nystagmus and no vertigo, mild as nystagmus but no vertigo, and no cure as nystagmus and vertigo.

In summary, there appears to be no need to keep patients upright overnight after either CRT or Liberatory maneuver. Having patients stay upright for as little as 20 minutes after the maneuver is sufficient for remission of BPPV. Keeping patients upright for 3 days does not improve the efficacy of the treatment. These results raise the question what happens to the otoconia once they are moved out of the posterior SCC into the utricle. Is the free otoconium reabsorbed into the macule once it is placed back into the utricle, or does it simply dissolve within the endolymph of the utricle? Further studies are needed to settle this issue.

## Specific Instructions for the Canalith Repositioning Treatment for Posterior SCC BPPV

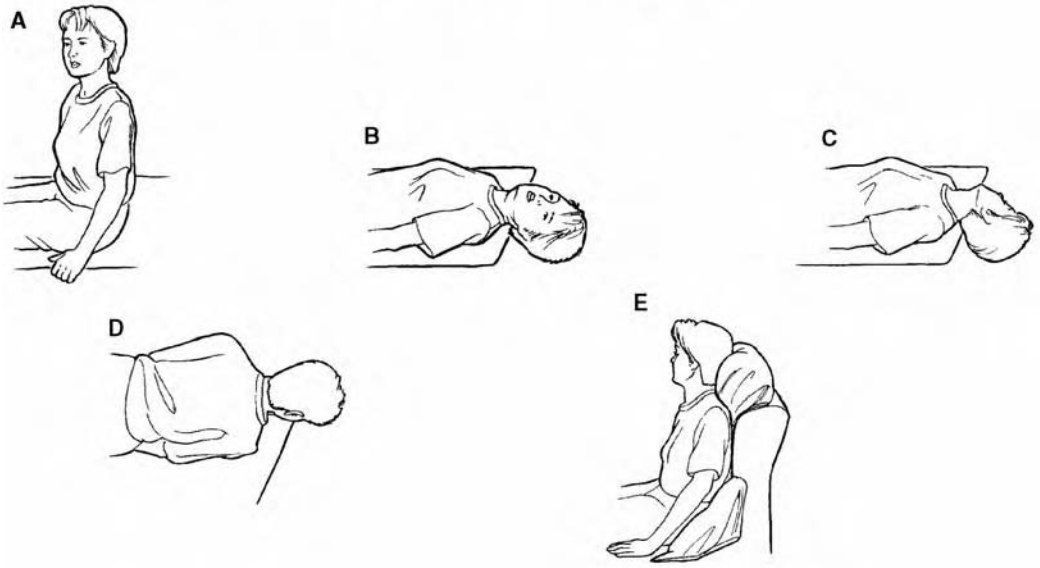
**The CRT Procedure.** During the CRT, the patient first is moved from sitting into the Dix-Hallpike position toward the side of the affected ear and then remains in that position until the nystagmus ceases (see Fig. 17.6). In some cases, the patient may remain in that position until any residual nausea decreases before the head (labyrinth) is shifted to the next position. The second phase of the treatment is either to rotate the patient's head slowly through moderate extension of the neck toward the unaffected side or to roll the patient's entire body onto the side if the treatment table has been prepositioned so the head is below horizontal. Again, the patient stays in the new position until the nystagmus stops. If there is no nystagmus, the position is maintained for approximately 20 seconds. The patient is then rolled to a side-lying position with the head turned 45 degrees down (toward the floor) and kept in that position for 20 seconds. Finally, keeping the head deviated toward the unaffected side and pitched down, the patient slowly sits up. Some patients experience vertigo shortly after returning to the seated position.

**Use of a Soft Collar.** To make certain the otoconia stay in the utricle after the treatment, the therapist can fit the patient with a soft collar and tell the patient not to bend over, lie back, move the head up and down, or tilt the head to either side for the rest of the day. The soft collar serves as a reminder to the patient to avoid those head positions and does not have to be fitted with the same rigor as one would fit a cervical collar on someone with a neck injury. The soft collar should be removed before bedtime. We no longer use a soft collar unless the patient requests one.

**Home Treatment.** The patient is told to perform the treatment at home. We review the treatment with the patient and any friends or family members who came to the clinic by demonstrating the treatment again and by giving the patient a handout with illustrations (Fig. 17.8). We ask the patient to repeat the treatment on his or her own the next morning. The patient is instructed that if he or she still experiences vertigo during the treatment, the treatment should be repeated once each morning until the vertigo is not elicited during the treatment. We have patients perform this home therapy for two reasons. First, patients learn how to do the treatment, which is useful if they have a recurrence of BPPV. Second, it raises the success rate of the treatment. We schedule a follow-up clinic visit a few days later to make certain the patient is in true remission.

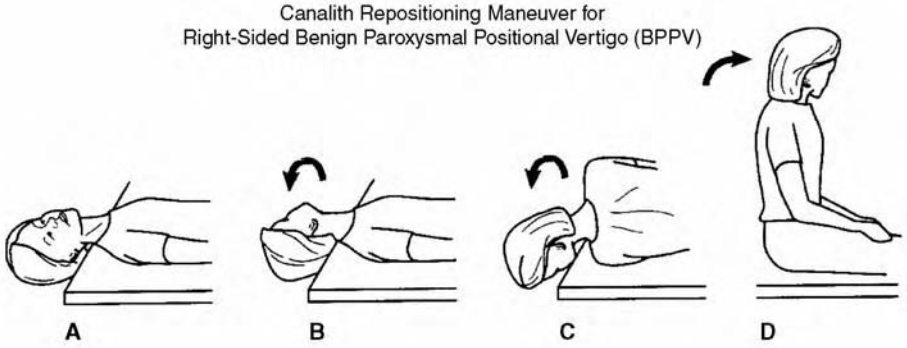
Some patients cannot perform the CRT on their own. In that situation, we teach them how to perform the

Canalith Repositioning Maneuver for
Left-Sided Benign Paroxysmal Positional Vertigo (BPPV)




A) Turn head 45deg to the left.
B) Lie down with the head hanging 20deg down over the edge of the bed. Keep head rotated 45deg to left.
Stay in this position for 20 secs or until dizziness stops, whichever is longer.
C) While keeping the head tilted back 20deg, rotate head such that it is 45deg to right.
Stay in this position for 20 secs or until dizziness stops, whichever is longer.
D) Roll over onto right shoulder and rotate head such that it is 45deg down.
Stay in this position for 20 secs or until dizziness stops, whichever is longer.
E) Slowly sit straight up with head still rotated to the right.
Straighten head and stay in an upright position for the rest of the day.

Canalith Repositioning Maneuver for
Right-Sided Benign Paroxysmal Positional Vertigo (BPPV)




A) Turn head 45deg to the right and lie down with the head hanging 20deg down over the edge of the bed. Stay in this position for 20 secs or until dizziness stops, whichever is longer.
B) While keeping the head tilted back 20deg, rotate head such that it is 45deg to left.
Stay in this position for 20 secs or until dizziness stops, whichever is longer.
C) Roll over onto left shoulder such that head is 45deg down.
Stay in this position for 20 secs or until dizziness stops, whichever is longer.
D) Slowly sit straight up with head still rotated to the left.
Straighten head and stay in an upright position for the rest of the day.

**Figure 17.8** Handouts given to patients for home treatment (canalith repositioning) for posterior SCC BPPV. *A*, Diagram shows treatment for left-sided problem, and *B* shows treatment for right-sided problems. (Adapted from Tusa and Herdman, Emory University, Atlanta, 1999.)

Brandt-Daroff habituation exercises. Radtke and associates[28] compared the success of using two different home self-treatments, Brandt-Daroff habituation exercises, and CRT. They found CRT to be much more successful.

### Common Sense Recommendations for Treatment

**Preventing Visual Fixation.** The diagnosis of BPPV is made by eliciting nystagmus and vertigo after the head is moved in the plane of one of the SCCs. Using a device to prevent visual fixation (such as Frenzel glasses, 20-diopter lenses, or infrared goggles) is important. Torsional nystagmus may not be suppressed by visual fixation, although both horizontal and vertical nystagmus can be suppressed. Without accurate identification of the presence, direction, and duration of the nystagmus, an incorrect diagnosis may be made. Additionally, observation of nystagmus during the treatment will identify whether the debris is moving in an appropriate direction at all stages of the treatment. With each position change, the debris should move away from the cupula. Therefore, the direction of the nystagmus should be the same with each change in head (labyrinth) position. A reversal or change in the type of nystagmus would suggest that the debris has moved back toward the cupula or into another canal.[39]

**Educating the Patient.** It is important to educate the patient before performing the testing maneuver or treatment. The therapist needs to know whether or not the patient has experienced extreme vertigo or nausea with changes in head position. If the patient describes having severe vertigo, it may be best to reduce the speed used for the Dix-Hallpike or side-lying maneuvers. If there is no vertigo when the test is performed slowly, the test can be repeated at a faster speed. The patient can also be premedicated.

Patients should be instructed as follows:

1. They should keep their eyes open and should not look around during the test.
2. They may experience vertigo during the test or treatment.
3. They must remain in the test position until the vertigo has stopped.
4. If they absolutely cannot remain in the position, the therapist will help them slowly return to a sitting position.
5. The therapist will not let them fall.

If not educated about the procedure, the patient may become frightened, close the eyes, or attempt to sit up when the vertigo is provoked.

**Efficient Testing Reduces Patient Discomfort.** We typically use the Dix-Hallpike maneuver as the first test for BPPV. If the patient has no vertigo or nystagmus after testing on one side, he or she is slowly returned to the sitting position. It is important to bring the patient slowly to sitting to avoid or minimize orthostatic symptoms. The Dix-Hallpike maneuver is then performed to the other side. If the patient has no vertigo or nystagmus on the second side, we lift the patient's head slightly and perform the roll test for horizontal canal BPPV. If that also is negative, we slowly bring the patient up to a sitting position. We then test the patient using the side-lying test. Patients are often willing to lie down faster on their sides than on their backs, an observation that may explain why the side-lying test result may be positive in some patients with a negative Dix-Hallpike test result. If the patient knows which side down produces vertigo, it is best to check the opposite side first during the test to keep nausea to a minimum.

**Focus on Moving the Labyrinth.** In patients with neck pain, limited neck range of motion, or cervical dysfunction that prohibits neck extension and rotation, it is helpful to remember that you are trying to move the labyrinth into a particular position, not the head and neck. The required position of the labyrinth can be obtained by tilting the entire support surface first. Then, when the patient lies down, the labyrinth is in the appropriate alignment to provoke the signs and symptoms of BPPV.

**Dealing with Balance Problems.** Complaints of imbalance are common in people with BPPV, although testing for balance problems and the possibility of treatment for imbalance can be deferred until after resolution of vertigo has been achieved.[40,41] In our experience, vague complaints of imbalance often resolve without intervention in about 2 weeks. Persistent balance problems or high risk for falling are indications that intervention is needed.

## *Treatment of Posterior Canal BPPV: Cupulolithiasis*

### The Liberatory Maneuver

The single treatment approach known as the Liberatory maneuver was developed by Semont and associates.[22] For the sake of discussion, let us assume that BPPV in the patient being treated has been identified to be in the right posterior canal. The Liberatory maneuver is performed as follows:

1. The patient is told to sit sideways on the examination table, and the therapist turns the head 45 degrees to the left.

2. The patient is quickly moved onto the right side while keeping the head turned left; the patient is kept in that position for 2 to 3 minutes (Fig. 17.9).
3. The patient is then rapidly moved up through the sitting position and down into the opposite side-lying position, while the therapist maintains the alignment of the neck and head on the body (in this case with the head always turned to the left). Thus, the final position will have the patieng lying with the face at a 45 degree angle toward the table.
4. Typically, nystagmus and vertigo reappear in this second position. If the patient does not experience vertigo in this second position, the head is abruptly shaken once or twice, through a small amplitude, presumably to free the debris. The patient stays in this position for 5 minutes.
5. The patient then slowly moves into a seated position.

Patients must remain in a vertical position for 48 hours (including while sleeping) and must avoid the provoking position for 1 week following the treatment. Like CRT, the liberatory maneuver usually requires only a single treatment. Reportedly, this approach works by floating the debris through the canal system to the common crus, but it may also dislodge debris adhering to the cupula. It is not clear why the procedure calls for the patient to be kept in each position for such an extended period. We typically use only 1 minute in each position, but even that duration may be excessive. The issue has not been examined in a controlled study.

## Brandt-Daroff Habituation Exercises

Proposed by Brandt and Daroff,[16] this treatment requires the patient to move into the provoking position repeatedly several times a day. It is performed as follows:

1. The patient first sits over the edge of the table and turns his or head 45 degrees toward the side

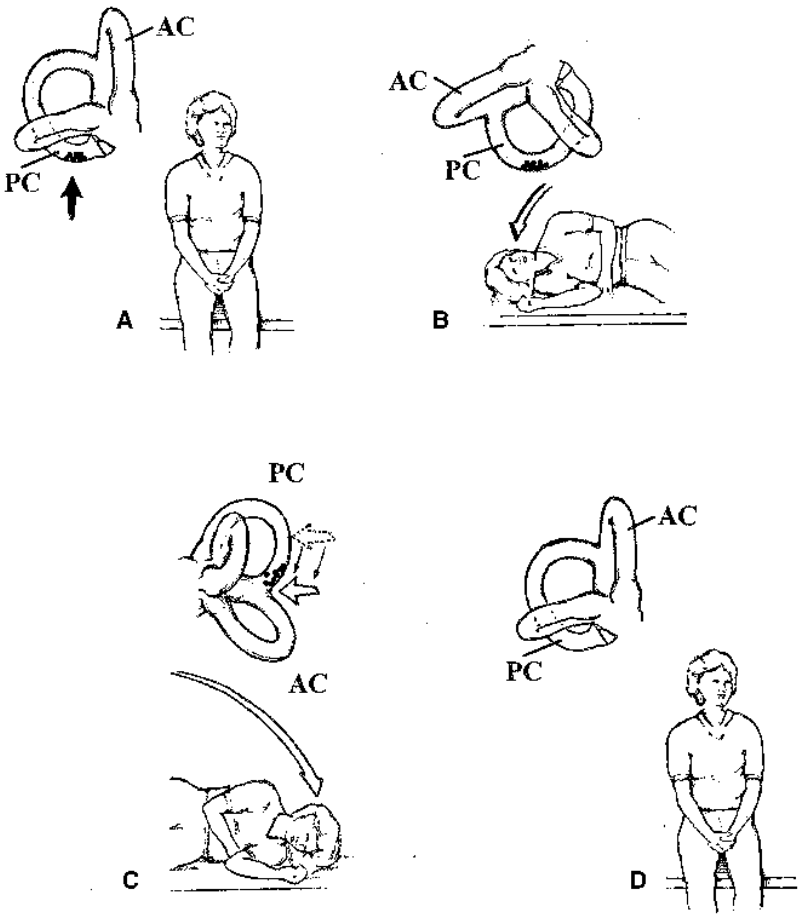



**Figure 17.9** Liberatory maneuver for treatment of posterior semicircular canal (SCC) benign paroxysmal positional vertigo (BPPV), shown for here for right posterior SCC BPPV. (*A*) The patient sits on the examination table sideways, and the head is rotated 45 degrees toward the unaffected side. (*B*) The patient is then moved quickly onto the affected side (parallel to the plane of the affected posterior canal) until the head is hanging 20 degrees down. (*C*) After 1 minute, the patient is rapidly moved through the initial sitting position to the opposite side with the head still positioned 45 degrees toward the unaffected side (the nose will now be angled 45 degrees down toward the floor). (*D*) The patient holds this position for 1 minute and then moves slowly to a sitting position. A soft collar is then placed, and the patient is given the same instructions as that described for canalith repositioning treatment (see Fig. 17.6). AC = anterior canal; PC = posterior canal. *Arrows* indicate position and movement of debris. (Modified from Tusa and Herdman, 1998.[4])

that is symptom free and then is moved rapidly into the side-lying position that causes the vertigo (Fig. 17.10). A torsional and upbeating nystagmus occurs with the onset of the vertigo. The severity of the vertigo will be directly related to how rapidly the patient moves into the provoking position.
2. The patient stays in that position until the vertigo stops and then sits up again. Usually, moving to the sitting position also results in vertigo, although less severe and of a shorter duration. Nystagmus, if reoccurring, will be in the opposite direction.
3. The patient remains in the upright position for 30 seconds, turns his or her head 45 degrees in the opposite direction and then moves rapidly into the mirror-image position on the other side, stays there for 30 seconds, and then sits up.
4. The patient then repeats the entire maneuver until the vertigo diminishes.
5. The entire sequence is repeated every 3 hours until the patient has 2 consecutive days without vertigo.

It is not clear why these exercises result in a decrease in the vertigo and nystagmus. One explanation is that the debris becomes dislodged from the cupula of the posterior SCC and moves to a location that no longer affects the cupula during head movement. A second possibility is that the debris dissolves in the endolymph, much like a lump of sugar dissolves with stirring. A third possibility is that central adaptation occurs, reducing the

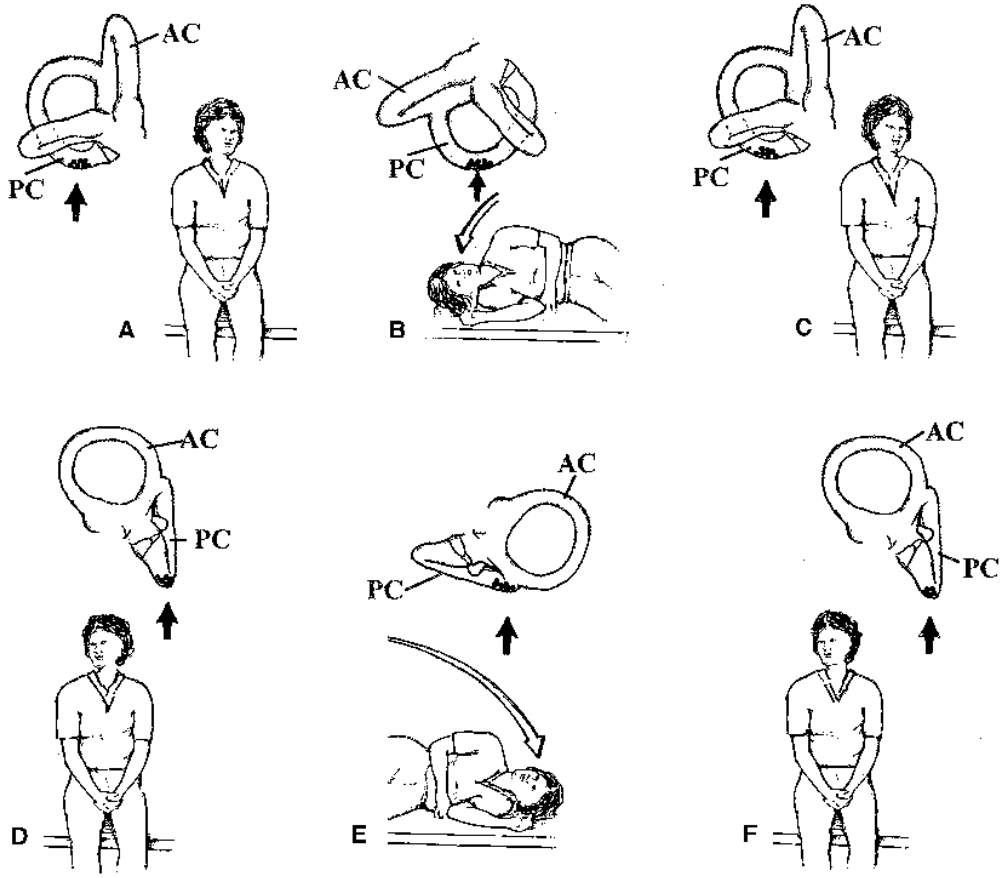



**Figure 17.10** Brandt-Daroff treatment for treatment of posterior semicircular canal benign paroxysmal positional vertigo. (*A* to *B*) The patient is moved quickly into the side-lying position on the affected side (shown here as right side) and stays in that position until 30 seconds after the vertigo 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 seconds 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.[4])

nervous system response to the signal from the posterior canal. Brandt and Daroff[16] argue against central adaptation as a mechanism for recovery because many patients recover abruptly with these exercises.

The Brandt-Daroff exercises for posterior canal BPPV can be modified for horizontal canal cupulolithiasis. The patient performs rapid, repetitive movements in the plane of the horizontal canal (the head stays facing forward all the time). Although this approach appears to be an effective treatment, only anecdotal evidence for it exists at this time because of the relatively low occurrence of horizontal SCC BPPV. Presumably this treatment works by dislodging the debris from the cupula.

## Variations in SCC Involvement

Up to now, we have discussed BPPV involving the posterior SCC, but BPPV also can occur in the anterior and horizontal SCCs (see Table 17-3). In 6% of our patients, we could not determine whether BPPV involved the anterior or posterior SCC because only the torsional component of nystagmus was present. The high prevalence of posterior SCC BPPV is likely determined by the ease in which otoconia can enter into this SCC when the patient lies down. Which SCC the debris is in can be determined from the direction of the nystagmus elicited by the Dix-Hallpike or Side-lying tests for BPPV (see Table 17-4).

### Horizontal SCC BPPV

In some cases of horizontal SCC BPPV, the Dix-Hallpike test does not elicit nystagmus or vertigo.[9] The optimal test for eliciting horizontal SCC BPPV is a roll in the plane of the horizontal SCC. For screening purposes, the patient is laid supine, and the head is then quickly rolled to one side. If no vertigo occurs, the head is quickly rolled to the other side. If vertigo does occur, the roll test needs to be more precise to determine the side of the defect.

One can improve the precision by always starting the roll from a nose-up position (see Fig. 17.4*A* and *B*). The head is rolled quickly to one direction, and nystagmus and vertigo are checked for. The head is then slowly rolled back to center (see Fig.17.4*C*). Nystagmus and vertigo are sought again after the patient is rolled to the other side (see Fig. 17.4*D*). In a patient with horizontal SCC BPPV, nystagmus and vertigo occur for rolls in both directions, but slow-phase eye velocity and duration of nystagmus are greater when the patient is rolled toward the affected ear.[11] The direction of the nystagmus depends on whether the otoconia is free-floating (canalithiasis) or fixed to the cupula (cupulolithiasis) (see Fig. 17.5).[43] In canalithiasis of the horizontal SCC, nystagmus is geotropic and fatigues; in cupulolithiasis, nystagmus is apogeotropic and persists.

#### *Bar-B-Que Roll Treatment for Horizontal Canalithiasis*

The CRT has been modified for horizontal SCC BPPV into a treatment referred to as the bar-b-que treatment (see Table 17-5).[37,43,44] We have found this treatment optimal for severe forms of horizontal SCC BPPV due to canalithiasis.[37] It can also be used as an alternative treatment for horizontal SCC BPPV due to mild canalithiasis. To begin the bar-b-que treatment, the patient lies on their back on the examination table or bed with the affected ear down (Fig. 17.11)—one should recall that the affected ear is identified as the side that causes more nystagmus and vertigo during the roll test. The patient moves from one position to the next with the clinician's assistance, as follows:

1. The patient's head slowly rolls away from the affected ear until the face is pointed up; this position is held for about 15 seconds, or until the dizziness stops.
2. The patient continues to roll the head in the same direction until the affected ear is up; this position is held for about 15 seconds, or until the dizziness stops.
3. The patient rolls the head and body in the same direction until the face is down.
4. After 15 seconds, the patient slowly sits up, keeping the head level or pitched down 30 degrees.

As with regular CRT, the patient may be fitted with a soft collar. The follow-up treatment is the same as for regular CRT, in that the patient may lie down at night and then perform the bar-b-que roll treatment each morning.

#### *Liberatory Maneuver for Horizontal Canal Canalithiasis*

Although called "Liberatory maneuver" by Appiani and colleagues,[45] it does not parallel Semont's Liberatory maneuver for posterior canal BPPV cupulolithiasis. Instead, this treatment was developed for patients with horizontal SCC canalithiasis. Like the CRT treatment, the theory underlying the treatment is that the head is moved in such a way as to cause debris floating freely in the horizontal SCC to move into the utricle. It is performed as follows:

1. The patient starts in a sitting position, looking straight ahead, and then quickly lies on the unaffected side (Fig. 17.12).
2. After holding that side-lying position for 2 minutes, the patient quickly turns the head 45 degrees down and remains in that position for 2 minutes before sitting up.

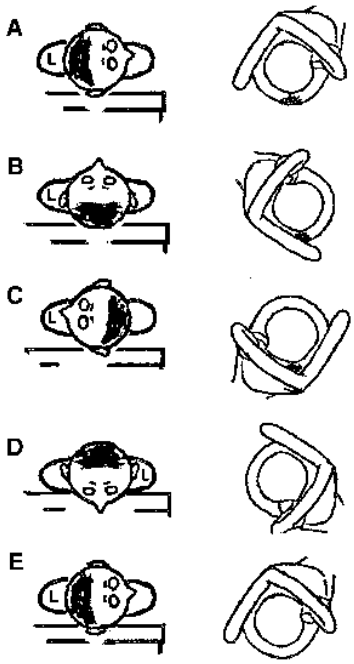



**Figure 17.11** Bar-B-Que roll or Canalith repositioning treatment (CRT) for horizontal SCC BPPV. (*A*) The patient lies supine on the examination table or bed with the affected ear down (shown here for right horizontal SCC BPPV). (*B*) The patient's head is then slowly rolled away from the affected ear until the face is pointed up; this position is maintained for about 15 seconds 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 seconds 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 seconds. 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. (*E*) 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 degrees. 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 treatment. (Modified from Tusa and Herdman, 1998.[4])

### *Forced Prolonged Position (FFP) for Horizontal Canal BPPV*

Another treatment for horizontal SCC canalithiasis BPPV is forced prolonged position.[14] In this treatment, the patient lies on the ear that contains the debris (side of BPPV) for 20 seconds, and then slowly rolls towards the healthy ear until the healthy ear is down. The patient remains in this position all night. The forced prolonged position can be combined with the bar-b-que treatment by having the patient perform the bar-b-que treatment at night prior to performing the FFP.

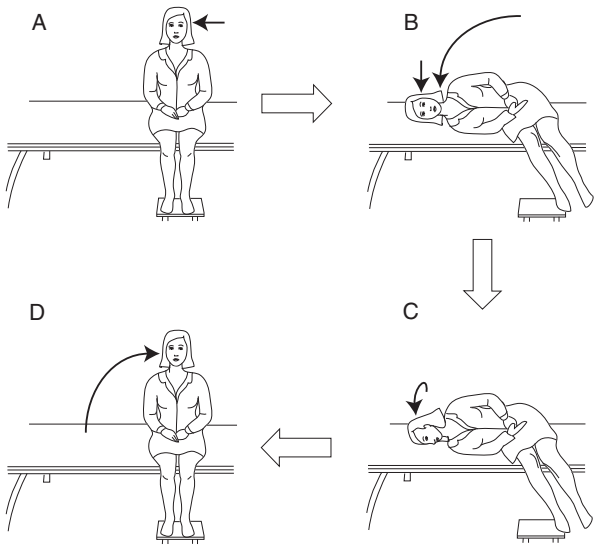



**Figure 17.12** Liberatory maneuver for horizontal canal canalithiasis. The patient starts in a sitting position looking straight ahead and then quickly lies on the *unaffected* side. After holding that side-lying position for 2 minutes, the patient quickly turns the head 45 degrees down, and remains in that position for 2 minutes before sitting up. *Arrows* point toward the affected side. (Modified from Appiani et al, 2001.[45])

### *Quick Bar-B-Que Roll Treatment for Cupulolithiasis BPPV*

In order to dislodge otoconia attached to the cupula of the horizontal SCC (cupulolithiasis), we follow the same procedure as for horizontal SCC canalithiasis (described previously), but we ask the patient to perform each head turn as quickly as possible. After this quick bar-b-que roll treatment, we ask patients to keep the head upright for the rest of the day, but we do not ask them to sleep upright. We have patients repeat this treatment once each morning until they experience no symptoms during the treatment.

### *Modified Semont Maneuver for Horizontal Canal Cupulolithiasis*

Another method for dislodging otoconia from the cupula of the horizontal SCC has been proposed by Casani and associates.[46] The patient moves quickly from sitting to side-lying position on the affected side. The patient then quickly turns the head so the nose is down 45 degrees and remains in that position for 2 to 3 minutes before sitting up again (Fig. 17.13). The actual movement is similar to

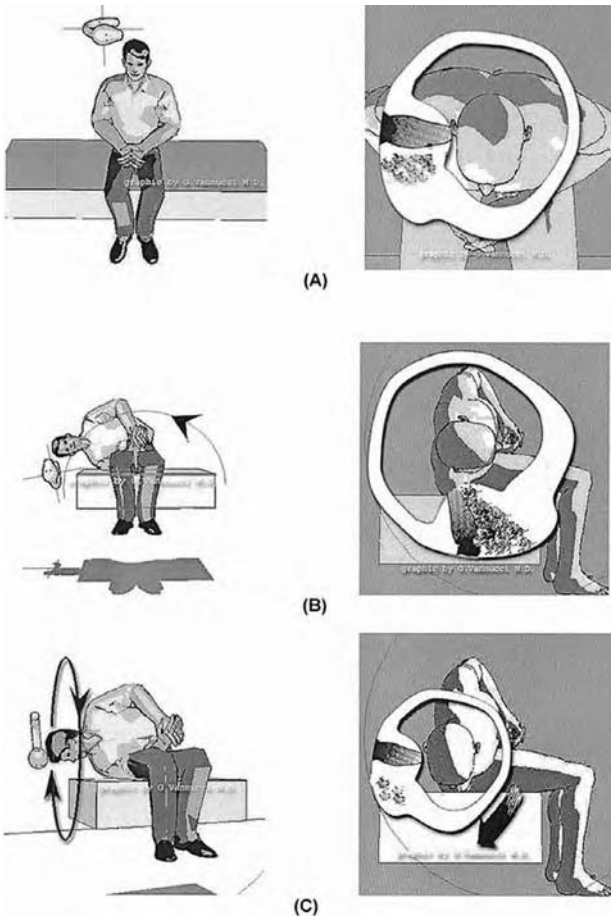



**Figure 17.13** Modified Semont maneuver for horizontal canal cupulolithiasis. The patient moves quickly from sitting to lying on the *affected* side. The patient then quickly turns the head so the nose is down 45 degrees, and stays in that position for 2 to 3 minutes before sitting up again. (Adapted from Casani et al, 2002.[46])

the Appiani Liberatory" maneuver for horizontal SCC canalithiasis, except that for cupulolithiasis, the patient lies on the *affected* side. Casani and associates[46] report remission of symptoms in 75% of patients with one maneuver.

## Anterior SCC BPPV

For anterior SCC BPPV, the nystagmus is primarily downbeat and torsional (during quick phases of nystagmus, the superior pole of each eye torts toward the dependent or inferior ear while lying).

### *Canalith Repositioning Treatment for Anterior SCC BPPV*

Anterior SCC BPPV is treated the same way as posterior SCC BPPV. The difficulty lies in deciding which side to treat. The best way to decide is based on the direction of the nystagmus rather than on the side of the dependent labyrinth (ear) during the Dix-Hallpike test. During the Dix-Hallpike test, any debris in the anterior canal of the dependent labyrinth moves away from the cupula, resulting in downbeating and torsional nystagmus (see Fig 17.14). One can imagine, however, that if the head were positioned into more extension than usual, debris in the opposite labyrinth might move, also away from the cupula.

As shown in Figure 17.14, for example, debris in the anterior canal of the right labyrinth would move away from the cupula when the patient is positioned in the right Dix-Hallpike position. This would result in a downbeat and rightward torsional nystagmus. If there were debris in the left anterior canal, either it would not move or, with more neck extension, the debris might move away from the left cupula of the anterior canal, resulting in a downbeat and leftward torsional nystagmus. That pattern would suggest left anterior canal BPPV and could be confirmed by the response when the patient is moved into the left Dix-Hallpike position.

### *Liberatory Maneuver (Semont or Brisk Treatment) for Anterior SCC BPPV*

For anterior SCC BPPV, the procedure must be modified in order to move the head in the plane of the anterior SCC. For sake of discussion, assume that the patient has debris in the right anterior SCC. The procedure is as follows:

1. The patient sits on the examination table sideways, but the head is rotated 45 degrees toward the right side.
2. The patient is then moved rapidly onto the right side (parallel to the plane of the affected anterior SCC).
3. After 1 minute, the patient is rapidly moved through the initial sitting position to the left side with the head still positioned 45 degrees toward the right side (nose is now angled 45 degrees up toward the ceiling).
4. The patient holds this position for 1 minute and then moves slowly to a sitting position.

### *Brandt-Daroff Habituation Exercises for Anterior SCC BPPV*

Brandt-Daroff exercises can be used to treat anterior SCC BPPV, because the head is moved in the plane of the posterior SCC on one side and of the anterior SCC on the other side.

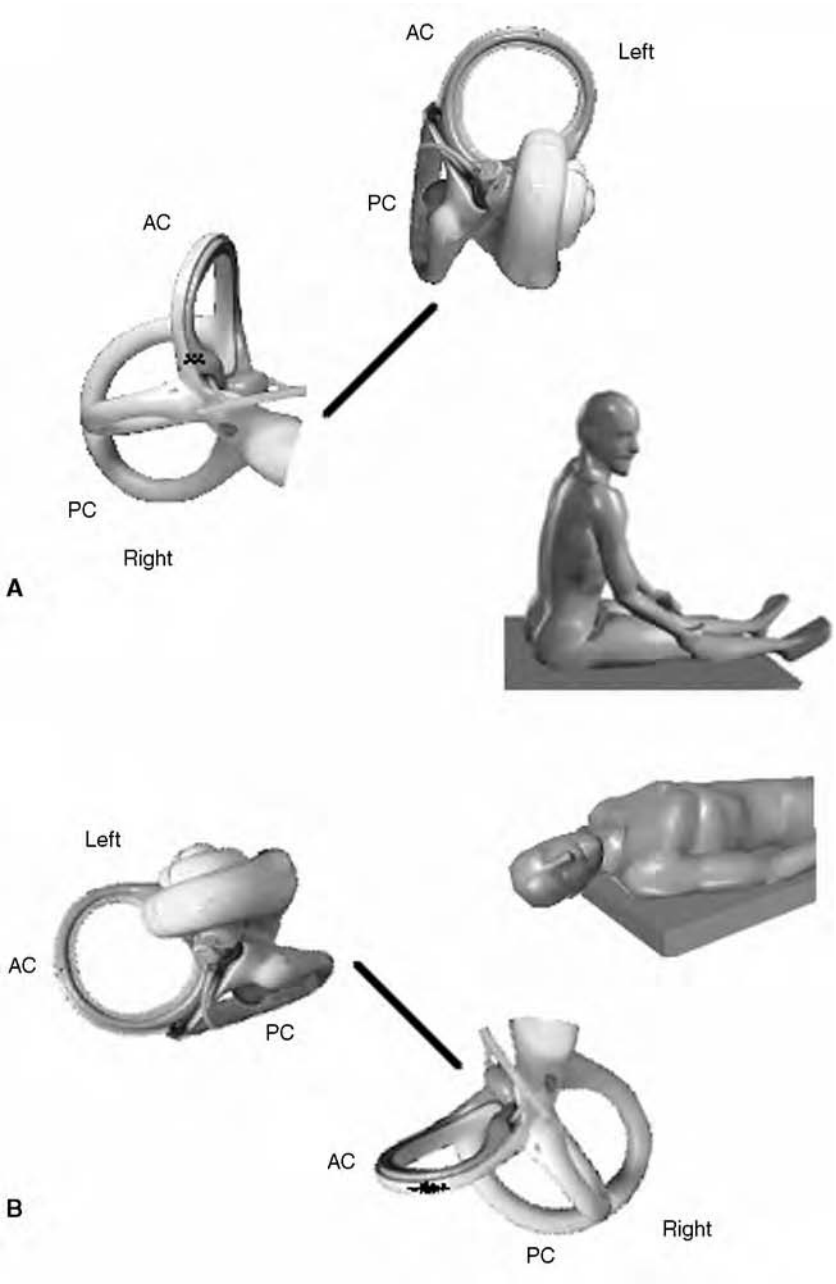



**Figure 17.14** Position testing for anterior semicircular canal (SCC) benign paroxysmal positional vertigo. (*A*) In the Dix-Hallpike test, the patient sits on the examination table, and the head is turned 45 degrees horizontally. (*B*) The head and trunk are quickly brought straight back "en bloc," so that the head is hanging over the edge of the examination table by 20 degrees. Nystagmus is observed, and the patient is asked whether he or she has vertigo. Although not shown in the figure, the patient is then brought up slowly to a sitting position with the head still turned 45 degrees, and nystagmus is observed again. This test then is repeated with the head turned 45 degrees in the other direction. This figure also shows movement of free-floating otoconia (canalithiasis) in the right anterior SCC during the Dix-Hallpike test. In this example, the patient would have a downbeat and rightward torsional nystagmus and vertigo when the test is done on the right side, but not when the test is done on the left side. AC = anterior canal; PC, = posterior canal.

## Algorithm for Treatment of BPPV

Figure 17.15 is an algorithm for arriving at the appropriate treatment for BPPV. Identification of the affected side, which canal is involved (based on direction of nystagmus), and whether the problem is due to canalithiasis or cupulolithiasis leads to specific treatments or to consideration that the nystagmus may be of central origin.

## Evidence-Based Practice

### *Quality of Available Evidence to Use Repositioning Maneuvers to Treat BPPV*

The quality of evidence that CRT successfully treats posterior SCC BPPV includes randomized controlled studies, randomized noncontrolled studies, prospective studies, and retrospective studies.[47] A brief summary of

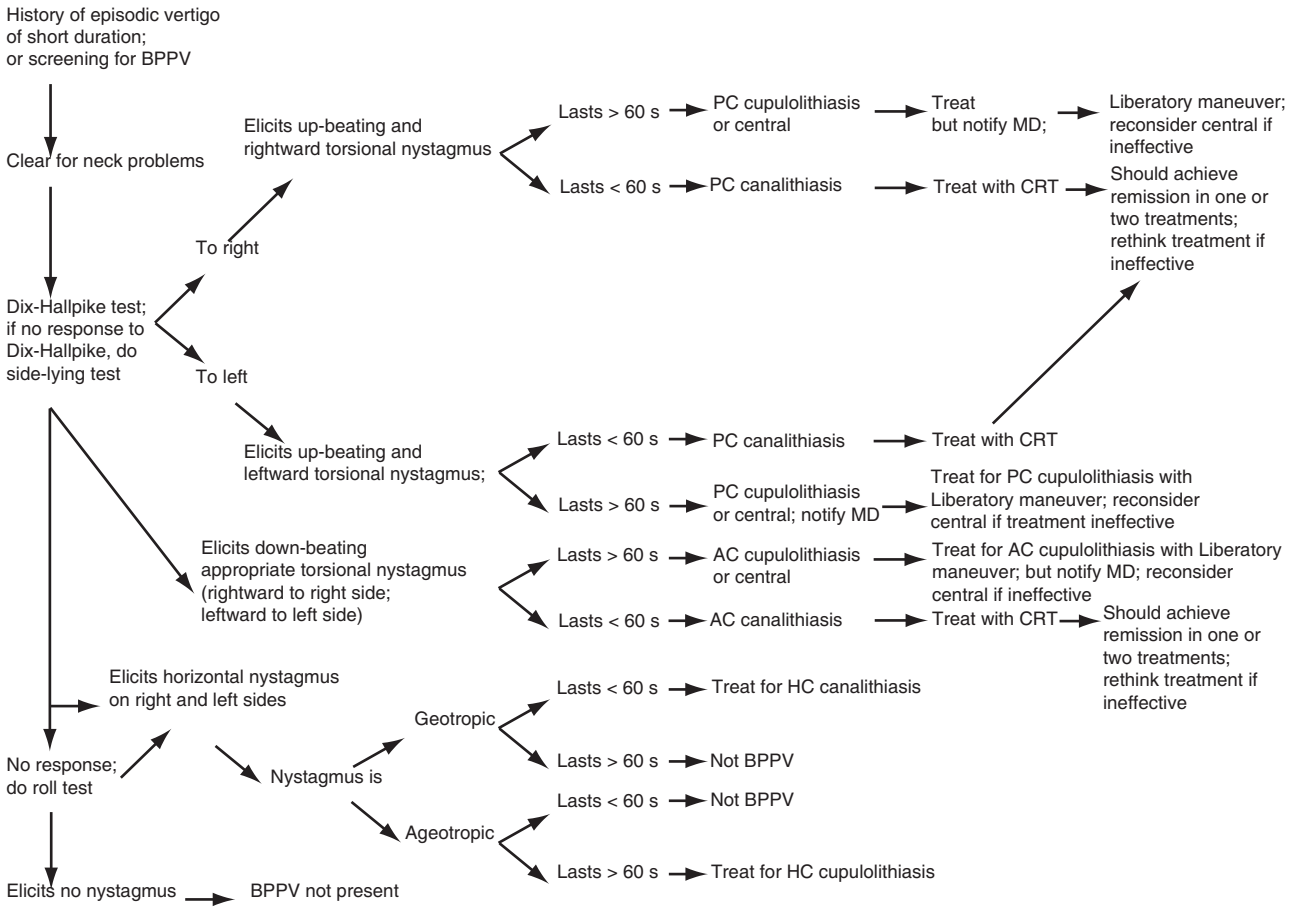



**Figure 17.15** Algorithm for assessment leading to treatment of benign paroxysmal positional vertigo (BPPV). Identification of the direction and duration of the nystagmus leads to the determination of the canal involved and whether the BPPV is from canalithiasis or cupulolithiasis. This information directs the appropriate choice of treatment. AC = anterior canal; BPP = benign paroxysmal positional vertigo; CRT = canalith repositioning treatment; MD = medical doctor; PC = posterior canal.

the randomized, controlled studies is given in Table 17-7. Overall, these studies indicate that successful remission of the signs and symptoms of posterior canal BPPV is obtained in 67% to 94% of all patients treated with CRT, compared with none to 48% for the control treatment. Blakely[51] published the only study that showed no difference in outcome between patients treated with CRT and those not treated at all; this study was criticized because the maneuver used was not clearly stated, follow-up was delayed (1 month after treatment), and the follow-up step did not examine the patients (telephone follow-up only). Li[18] stated that vibration was necessary during CRT for a good outcome, but the majority of studies used no vibration. Despite the number of excellent studies that show the effectiveness of CRT in treating BPPV, patients are still told that there is no treatment or are given medication for their dizziness. Hopefully this misconception will diminish with more publications of randomized controlled trials. For a review of the randomized controlled trials on CRT of BPPV, see Tusa and coworkers.[47]

## *Quality of Available Evidence to Use the Liberatory Maneuver to Treat BPPV*

Semont and colleagues[22] reported on a series of 711 patients with BPPV treated over an 8-year period. They found a "cure" rate of 84% after a single treatment and of 93% after two treatments. Recurrence of the symptoms was infrequent (4%). Other studies have reported somewhat lower remission rates. Hausler and Pampurik,[52] for example, examined the efficacy of the Liberatory manuever in a randomized controlled trial of 77 subjects. At 18 days after treatment, 51% of the patients treated with the Liberatory maneuver were in remission, com-

## ■ Table 17-7 RANDOMIZED CONTROLLED STUDIES USING REPOSITIONING MANEUVERS FOR BPPV

| Type of Study | Procedure and Remission Rate (%) | No. Subjects | Study |
|---|---|---|---|
| Single-blind, randomized, controlled | CRT (89%) vs. no treatment (27%) at 1 month (follow-up evaluator was blinded to the treatment type) | 36 | Lynn et al, 1995[21] |
| Single-blind, randomized, controlled | CRT (71%) vs. sham treatment (0%) at 1 week (patient did not know which treatment was to be given, crossover study was done) | 29 | Lempert et al, 1997[44] |
| Single-blind, randomized, controlled | CRT (67%) vs. untreated (38%) at 10 days (follow-up evaluator was blinded as to treatment type) | 50 | Froehling et al, 2000[1] |
| Single-blind, randomized, controlled | CRT (76%) vs. no treatment (48%) at 1 week (follow-up evaluator was blinded as to treatment type) | 58 | Yimtae et al, 2003[48] |
| Randomized, controlled | CRT (50%) vs. no treatment (43%) at 1 month | 38 | Blakley, 1994[51] |
| Randomized, controlled | CRT with mastoid vibration (92%) vs. CRT without mastoid vibration (60%) vs. no treatment (0%) at 1 week | 60 | Li, 1995[18] |
| Randomized, controlled | CRT (93.4%) vs. no treatment (50%) by 21 days (1 treatment per week) | 41 | Wolf et al, 1999[49] |
| Randomized, controlled | CRT (94%) vs. no treatment (?) at 1 month | ? | Asawavichianginda et al, 2000[50] |

BPPV = benign paroxysmal positional vertigo; CRT = canalith repositioning treatment.

pared with 18% of the untreated control group.[52] Levrat and coworkers[53] reported remission of symptoms in 62.6% of their subjects treated with only one maneuver. In a prospective, randomized study that included 30 subjects treated with the Semont maneuver, we reported remission of symptoms and signs or significant improvement in 90% of the cases after a single treatment.[5]

In a later study, Ireland[23] examined the effectiveness of the Liberatory maneuver in a series of patients using the patients as their own controls. The patients were first treated with the Liberatory maneuver but on the unaffected side. None of the patients had any relief from vertigo. The patients were then treated with only the routine post-maneuver instructions to keep the head upright for 48 hours, including sleeping in a sitting position. Again, at the end of 1 week, all patients were symptomatic. Then 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 number of subjects in this study is small (n = 10), the results do support the effectiveness of the Liberatory maneuver as treatment for BPPV.

### *Quality of Available Evidence to Use Brandt-Daroff Habituation Exercises to Treat BPPV*

Brandt and Daroff[16] studied a series of 67 patients with BPPV of 2 days to 8 months in duration. None of these patients had evidence of other neurological or neurotological disease. These investigators reported that 98% of the subjects had no symptoms after 3 to 14 days of exercises. The only subject who did not show response to treatment had a perilymphatic fistula requiring surgical repair. Recurrence was low, affecting only 3% of the patients.

In our experience with a series of 20 patients with BPPV who were treated with exercises similar to those

advocated by Brandt and Daroff,[16] the time until the patients were symptom-free (n = 12) or had at least a moderate reduction in symptoms (n = 7) was more protracted, extending from 1 week to 6 months (this last case involving a patient who was afraid to lie down; the treatment course was therefore several extra months while he worked at home just to achieve a supine position). Patients in whom there was only partial recovery complained most frequently of an intermittent "swimming" sensation rather than of true vertigo. One patient experienced no change of vertigo. All of these patients had histories of BPPV extending from a few days to 35 years. The more protracted the course, the more resistant the BPPV may be to treatment.

## Managing Persistent Imbalance in Patients with BPPV

More than 50% of patients with posterior SCC BPPV subjectively report imbalance.[37,54] The presence of imbalance in such subjects, relative to age-matched controls, has been quantified with computerized static and dynamic sway platform testing.[40,41,55–59] The CRT and liberatory maneuver may completely resolve positional nystagmus and vertigo based on repeat Dix-Hallpike test, and improve balance, based on repeat computerized platform testing,[40,41,56,59] but 8% to 14% of patients may still complain of imbalance.[5,60,61]

Benyon[61] has postulated that this imbalance may be due either to persistent BPPV that is not sufficient to deflect the cupula during testing or to vestibular hypofunction that was not detected during the initial evaluation. We do not believe that the persistent imbalance is caused by either one of these problems. We quantified the severity of imbalance in patients with posterior canal BPPV using computerized dynamic posturography before and after CRT in 33 patients.[41] Patients who had vestibular hypofunction confirmed by bithermal water caloric testing, bilateral BPPV, or head trauma were excluded. Figure 17.16 shows the mean posturography scores for patients with BPPV before CRT, for the same patients 1 week after CRT, and for age-matched controls. Analysis of the balance tests before CRT shows that scores for the patients with BPPV, as a group, did not differ significantly from normative scores on tests 1, 2, and 3. However, the scores for these patients on tests 4, 5, and 6 were significantly worse than the normative values. Balance improved in the patients with BPPV as a group after CRT. Scores in about 33% of patients with imbalance improved to normal values after CRT; scores in the remaining patients either did not improve or improved to subnormal values. The patients with persistent imbalance had no evidence of BPPV based on symptoms or on signs using video infrared cameras during the Dix-Hallpike test. Di Girolamo and colleagues[40] reported similar results on posturography in patients with BPPV

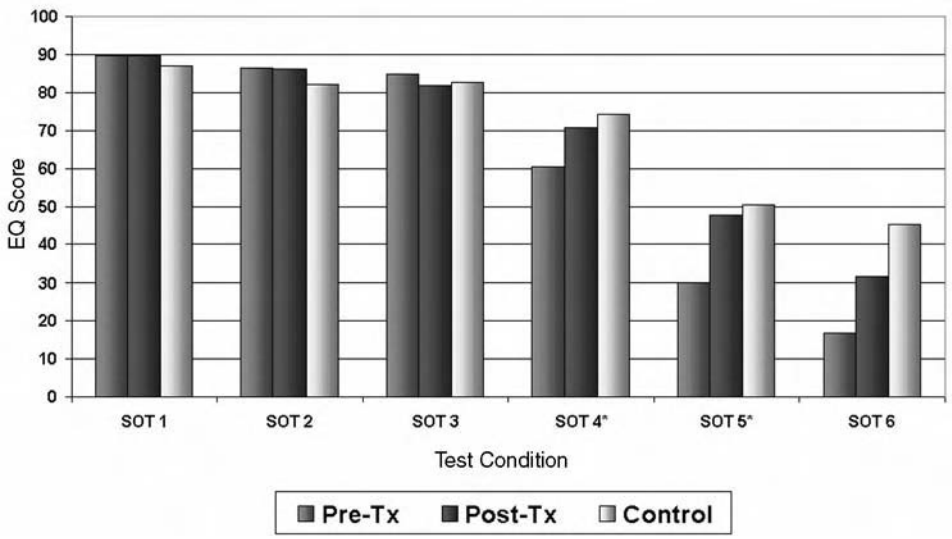



**Figure 17.16** Mean posturography scores for 33 patients with benign paroxysmal positional vertigo (BPPV) before canalith repositioning treatment (CRT) (pre-tx = pre-treatment) and 1 week after CRT (post-tx = post-treatment) compared with age-matched controls. * = significant difference in anterior-posterior sway amplitude. Blatt PJ, Georgakakis GA, Herdman SJ, et al: With permission from Am J Otol 2000;21:356.

and no evidence of vestibular hypofunction with use of the bithermal water caloric test.

So what is the cause of persistent imbalance in patients whose BPPV is in remission? One possibility is the age of the patient. Those patients who improved in the Di Giralamo study were younger ($P < .05$).[40] No difference was found between groups for time from onset of BPPV, history of falls, or patient ratings of the intensity of vertigo, although the difference between groups in patients' subjective rating of the intensity of disequilibrium approached significance. Therefore, the increased postural sway in some individuals on these tests may be due to a separate, pre-existing 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 recover fully after CRT. This latter explanation is supported by the study of Giacomini and coworkers,[59] who found that balance on a static platform did not fully recover in patients with BPPV until 12 weeks after CRT. This finding 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.

## Contraindications to the Assessment and Treatment of BPPV

Because the Dix-Hallpike test and CRT require that the head be rotated 45 degrees and extended 20 to 30 degrees, they should not be performed in patients with certain types of neck disorders. Humphriss and colleagues[62] consider the following disorders to be absolute contraindications to performing the Dix-Hallpike test, and presumably the treatment: history of neck surgery, recent neck trauma, severe rheumatoid arthritis, atlantoaxial and occipitoatlantal instability, cervical myelopathy or radiculopathy, carotid sinus syncope, Chiari malformation, and vascular dissection syndromes. They state that the test is otherwise safe and can be done without causing injury in individuals who do not have these disorders. They recommend that the following parameters be met in every patient before the Dix-Hallpike test is performed:

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?

Injury of patients during testing for BPPV is virtually nonexistent. However, clinicians must be cautious in performing the Dix-Hallpike test and should follow the parameters discussed by Humphriss and colleagues.[62]

In patients who cannot extend the head backward owing to neck disease, we perform the side-lying test (described previously) instead of the Dix-Hallpike test.[37,54] We also perform this alternative test in elderly patients who cannot extend the head backwards and in patients with low back pain or recent back surgery. In patients for whom rotating or extending the head is contraindicated, a modified side-lying test can also be done. In these patients we use a soft neck collar to help stabilize the neck.

1. To test the right posterior SCC, we sit the patient on the side of a table and then rotate the body and head together 45 degrees to the left.
2. We then lay the patient's body and head down together to the right to reach a side-lying position.

This modification avoids moving the head independent of the body. To test the left posterior SCC, we rotate the body and head 45 degrees to the right before laying the patient down to the left. This whole-body movement method has also been used to treat BPPV in patients with orthopedic problems in the neck and spine.[63]

We published a small study on the complications of the CRT.[39] Of 85 consecutive patients treated with CRT for posterior SCC BPPV, we found that two patients developed anterior SCC and 3 patients developed horizontal SCC BPPV at testing 7 days later (Table 17-8). Others have also noted the conversion of posterior SCC BPPV to horizontal SCC BPPV after CRT.[42,44,48] Another complication was a stiff neck from keeping the head upright. Finally, some patients experienced severe vertigo and nausea within 10–20 minutes after CRT or the Liberatory maneuver had been performed.

To reduce these complications, we now take the following measures: To avoid conversion of BPPV from posterior SCC to anterior and horizontal SCC, we always now use a tilt table to be certain that the patient's head is tilted at least 30 degrees down from horizontal during CRT. Since we began doing this, none of our patients experienced such conversions except for occasional patients who performed the treatment themselves at home. We believe that if the head is not tilted down enough during CRT, otoconia may enter into one of these other SCCs instead of the utricle. To avoid a stiff neck

■ Table 17-8 **COMPLICATIONS OF CRT FOR POSTERIOR SCC BPPV IN 85 CONSECUTIVE PATIENTS**

| Complication | No. of Patients Affected | Percentage of Patients Affected |
|---|---|---|
| Neck pain | 5 | 5.8 |
| Conversion to anterior SCC BPPV | 2 | 2.3 |
| Conversion to horizontal SCC BPPV | 3 | 3.5 |
| Emesis | 1 | 1.2 |

BPPV, benign paroxysmal positional vertigo; CRT, canalith repositioning treatment; SCC, semicircular canal.
Adapted from Herdman and Tusa, 1996.[39]

after CRT, we instruct all patients to move the head horizontally a few times every hour while they awake for at least 24 hours. To avoid severe vertigo, nausea, and vomiting, we instruct all patients to sit still for a couple of minutes after the treatment and for 10 minutes longer in the waiting room before leaving the clinic. Also, in patients with a history of severe nausea from BPPV, we perform the Dix-Hallpike test slowly; if BPPV is found, we immediately stop all further testing and perform the CRT slowly.

## Unraveling Complicated Cases

Unfortunately, not all patients with BPPV have obvious signs and symptoms. The following case studies illustrate some of the more confusing presentations. All patients in these case studies were tested with Frenzel lenses or infrared goggles. Figure 17.17 presents an algorithm that may provide some assistance is making decisions when confronted by difficult cases of BPPV.

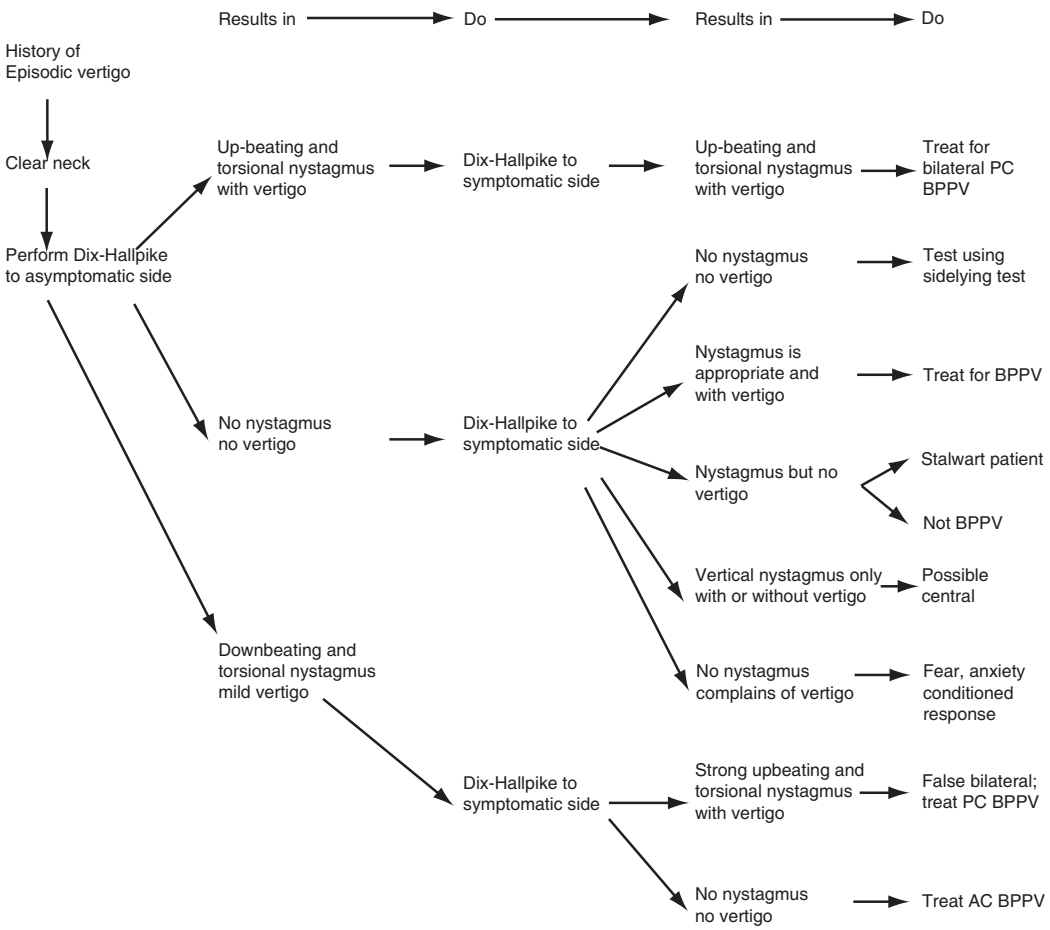



**Figure 17.17** Algorithm for assessing, interpreting, and treating complicated cases of benign paroxysmal positional vertigo.

## CASE STUDY 1

The patient has a persistent right-beating nystagmus without vertigo when in the right Dix-Hallpike position, and an upbeating and leftward torsional nystagmus with vertigo when in the left Dix-Hallpike position. This latter nystagmus resolves in 25 seconds, and then a persistent right-beating nystagmus (without vertigo) is again observed.

*Explanation*

The patient may have a left vestibular neuritis affecting the superior branch of the vestibular nerve. This would result in horizontal canal hypofunction and would explain the presence of the 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 the posterior canal canalithiasis form of BPPV.

*Resolution*

Treat the left-sided BPPV first. When the patient comes back and there is remission of BPPV, ask the patient whether the treatment has resolved all of the symptoms. If it has not and the patient reports imbalance or vertigo with quick head movements, ask whether any episode of vertigo has lasted for a day or longer (consistent with vestibular neuritis). Then perform the 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.

## CASE STUDY 2

The patient has no vertigo when moved into the left Dix-Hallpike position but does have a persistent downbeat nystagmus. When moved into the right Dix-Hallpike position, the patient has an upbeating and rightward torsional nystagmus (with vertigo) that decreases over 15 seconds but then becomes a persistent downbeat nystagmus. The vertigo lasts only 15 seconds.

*Explanation*

The upbeat and torsional nystagmus with vertigo lasting only 15 seconds is consistent with the posterior SCC canalithiasis form of BPPV. The downbeat nystagmus observed after the upbeat and torsional nystagmus resolves is not simply a reversal of the primary nystagmus of BPPV. First of all, it is persistent and 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. If the patient has a spontaneous downbeating nystagmus when sitting as well as when in the Dix-Hallpike positions, the physician must be informed.

## CASE STUDY 3

In the left Dix-Hallpike position, the patient has a vigorous upbeating and leftward torsional nystagmus associated with intense vertigo lasting 25 seconds. In the right Dix-Hallpike position, there is a downbeating and rightward torsional nystagmus with mild vertigo lasting 15 seconds.

*Explanation*

There are two possibilities—either the patient has posterior canal (PC) BPPV on the left side and anterior canal BPPV on the right side, or this is an example of false bilateral BPPV. In the latter situation, the patient actually has PC BPPV on the left side; the mild downbeating and torsional nystagmus with mild vertigo that occurs when the patient is in the right Dix-Hallpike position is actually produced by the PC BPPV on the left side. The orientation of the labyrinth when the patient is in the right Dix-Hallpike position is such that it causes the otoconia in the left posterior canal to drift toward the cupula. False bilateral BPPV is more common than true bilateral BPPV.

*Resolution*

The patient should be treated for left PC BPPV. If the nystagmus and vertigo resolve on both sides, the patient had false bilateral BPPV. If, after resolution of the PC BPPV on the left, the patient still has downbeating and rightward torsional nystagmus when in the right Dix-Hallpike position, the patient has true bilateral BPPV, and so should be treated for the anterior canal BPPV.

## CASE STUDY 4

In the right and left Dix-Hallpike positions, the patient has no nystagmus but complains of dizziness and becomes diaphoretic.

*Explanation*

It is quite possible that the patient had BPPV but it has already resolved. The patient, however, has become fearful and has a conditioned autonomic nervous system response when moved into a position that previously provoked the vertigo. It is also possible that the patient is being histrionic; he or she should be given the benefit of the doubt, however.

*Resolution*

The patient still needs to be able to tolerate lying down. This is a situation for which the habituation approach would be appropriate. The patient can begin by moving into a semi-supine position—one that elicits very mild symptoms—and gradually works toward becoming more comfortable when lying down.

## CASE STUDY 5

You have been treating a patient for BPPV for 3 weeks but her signs and symptoms have not yet been resolved. CRT was performed on two visits during the first week, and she also performed the treatment at home. When you asked the patient to demonstrate how she is doing the treatment, you noticed that she lifted her head when rolling from the initial Dix-Hallpike position to the opposite side. You corrected this problem, but after a week of performing the CRT at home, she was still not better. You then performed the Liberatory maneuver and taught the patient's husband how to do the treatment at home. This approach was tried daily for another week without success. The husband was able to demonstrate the treatment correctly, so faulty performance was not the problem. You then had the patient perform the Brandt-Daroff habituation exercise at home for another week, still without resolution.

*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 here, the patient should be re-evaluated by the referring physician for another disorder, such as central positional vertigo.

## Summary

Based on several randomized, controlled trials, both CRT and the Liberatory maneuver are effective treatments for posterior canal BPPV. CRT and the Dix-Hallpike test can be modified in patients with neck disease that prevents neck rotation and/or extension. There is no need to have patients sleep upright after the treatment is completed. Instead, they may need to sit upright for only 20 minutes, which is best done in the clinic. Because BPPV is a chronic disease (recurrences do occur), teaching the patient how to perform the treatments at home is an effective way to reduce repeat visits. The few complications of the treatments can be avoided through careful attention to the technique used for CRT and post-treatment instructions. Some patients have persistent imbalance after treatment; therefore, all patients should be questioned about persistence of imbalance, and appropriate testing and physical therapy given to those in whom imbalance persists.

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# APPENDIX 17A
# Differential Diagnosis: Mimicking BPPV

*Ronald J. Tusa, MD, PhD*

A number of causes of positional dizziness and positional nystagmus may be confused with benign paroxysmal positional vertigo (BPPV) (Appendix Table 17-1).

## Central Positional Vertigo with Nystagmus

### *Episodic Signs and Symptoms (Benign)*

Central positional vertigo (CPV) that occurs as spells is usually due to migraine or a migraine equivalent spell. Nystagmus may be vertical or horizontal. The nystagmus is usually sustained. See the Appendix Case Study.

### *Persistent Signs and Symptoms (Pathological)*

#### Injury near Cerebellar Nodulus

Persistent CPV is a rare entity that has been attributed to cerebellar tumor[1] or to hemorrhage dorsolateral to the fourth ventricle.[2,3] It produces a position-induced nystagmus (downbeat) and vertigo that (1) usually persists as long as the head position in maintained, (2) does not fatigue, and (3) does not habituate with repeated testing. The nystagmus in CPV does not have a torsional component. In addition, CPV is induced in all position changes (supine, left ear down, right ear down).

#### Vertebral Artery Compression

Vertigo and nystagmus can occur during head rotation from vertebral artery insufficiency[4] either from extreme head rotation[5] or in individuals with cervical spondylosis.[6] Transcranial Doppler ultrasonography can be used to demonstrate decreased posterior circulation flow in symptomatic individuals.[7] In our experience, vertebral artery compression during Dix-Hallpike testing and canalith repositioning treatment is extremely rare, most likely because the extent of rotation is relatively small (maximum 45 degrees horizontal rotation and 30 degrees head extension).

## Central Positional Nystagmus without Vertigo

There are various types of central positional nystagmus (CPN) that are usually not associated with vertigo. In these cases, nystagmus is usually sustained and is not suppressed by visual fixation. Unidirectional nystagmus (vertical, horizontal, or torsional) is more commonly associated with a central lesion than with a peripheral cause. These patterns of nystagmus are usually due to lesions in central vestibular pathways.

### *CPN Present Only When Patient Is Supine (Benign)*

Usually in elderly patients, downbeating or upbeating nystagmus may be elicited only when the patient is supine and there are no other abnormalities on examination. The etiology of this condition is unknown. Perhaps it represents mild trauma to the craniocervical junction from arthritis.

## ■ Appendix Table 17-1 DIFFERENTIAL DIAGNOSIS FOR BENIGN PAROXYSMAL POSITIONAL VERTIGO

| Category | Circumstance | Most Common Cause(s) |
|---|---|---|
| Central positional vertigo with nystagmus | Episodic signs and symptoms during position testing | Usually benign: Migraine |
| | Persistent signs and symptoms during position change | Usually pathologic: Tumor or stroke near cerebellar nodulus, or vertebral artery compression |
| Central positional nystagmus without vertigo | Present only when patient supine | Usually benign: Nothing seen on magnetic resonance imaging of head |
| | Present when patient supine or seated (pathologic) | Usually pathologic: Abnormality within central vestibular pathways (cerebellar or brainstem disorders) |
| Peripheral positional vertigo with nystagmus other than BPPV | Dizziness induced during Dix-Hallpike testing, but also usually induced by pressure changes between inner and middle ear (Valsalva maneuver, Hennebert's sign, Tullio phenomenon) | Perilymphatic fistula, superior canal dehiscence, hypermobile stapes |
| | Persistent, mild vertigo in certain head positions while patient is supine | Labyrinthine hypofunction |
| Positional dizziness without nystagmus | Dizziness induced when patient bends over, sits up, or stands; never when patient is lying down | Orthostatic intolerance |
| | Dizziness usually when patient sits or stands with head pitched or turned to an extreme position | Head extension dizziness or extreme rotation dizziness |

### *CPN Present When Patient Is Supine and When Sitting (Pathological)*

In some cases, downbeating or upbeating nystagmus occurs while the patient is seated and is usually enhanced while the patient is supine. The patient may also have mild cerebellar signs. The etiology depends on whether the nystagmus is downbeat or upbeat, as follows.

#### Downbeating Nystagmus

Downbeating nystagmus in both supine and sitting positions can occur from increased activity of central anterior semicircular canal (SCC) pathways relative to the posterior SCC.[8] This occurs with cerebellar degeneration, Chiari malformation, and selective lesions involving the cerebellar flocculus or the medial longitudinal fasciculus in the floor of the fourth ventricle, such as multiple sclerosis, tumor, and stroke.

#### Upbeating Nystagmus

Upbeating nystagmus in both supine and sitting positions is due to increased activity of central posterior SCC pathways relative to the anterior SCC. This occurs with lesions involving the brachium conjunctivum in the midbrain or ventral tegmental tract in the dorsal pontomedullary junction, including multiple sclerosis, tumor, and stroke.[9]

## Peripheral Positional Vertigo with Nystagmus Other Than BPPV

### *Pressure-Induced Disorders*

As in BPPV, perilymphatic fistula (PLF), SCC dehiscence, and hypermobile stapes can cause positional vertigo and nystagmus. Unlike BPPV, these disorders are usually associated with hearing loss. Valsalva maneuver,

change in pressure to the external ear (Hennebert's sign), mechanical movement of the tragus of the external ear, or loud noises (Tullio's phenomenon) may induce vertigo and nystagmus in these disorders.

### Perilymphatic Fistula

A perilymphatic fistula is a hole between the inner and middle ear caused by trauma, a cholesteatoma, otic syphilis, or displaced middle ear prosthesis.[10,11] As in BPPV, it can cause positional vertigo and nystagmus; unlike BPPV, it is usually associated with hearing loss. Valsalva maneuver or a change in pressure to the external ear frequently induces vertigo and nystagmus.

### Superior Semicircular Canal Dehiscence

SCC dehiscence is congenital thinning of the roof of the superior SCC.[12] During mild head trauma or Valsalva maneuver, the bony canal wall may erode and the superior SCC then comes in contact with the dura above the petrous bone. This results in a "third" membranous window into the inner ear that is susceptible to any pressure changes within the labyrinth.

### Hypermobile Stapes

The stapes may become hypermobile following head trauma or a stapes prosthesis may become dislodged and move into the oval window.[13]

### Labyrinthine Hypofunction

Direction-changing, static, positional nystagmus may occur in individuals with chronic unilateral vestibular hypofunction.[14] The nystagmus may either be geotropic (left-beating with the left ear down and right-beating with the right ear down) or apogeotropic (beats away from the earth). Individuals with this type of nystagmus may have mild vertigo during the position change, but the vertigo is much less than that found in BPPV.

## Positional Dizziness without Nystagmus

### *Orthostatic Hypotension*

The history of orthostatic hypotension consists of transient dizziness when the patient gets out of bed, stands up quickly, or bends over quickly. The patient may report episodic lightheadedness, weakness, impaired cognition, visual blurring, tremulousness, and vertigo that are aggravated by prolonged standing, or exercise, or that that occurs 15 to 45 minutes after eating (postprandial hypotension). Chronic fatigue may be present.

The physical examination in a patient with such a history should include a blood pressure check as follows. Supine blood pressure should be measured after 10 minutes. In a patient with orthostatic hypotension, the systolic blood pressure drops by at least 20 mm Hg within 3 minutes of standing up, and the patient is symptomatic. A tilt table test should be performed in individuals in whom results of the bedside blood pressure check is not clear (may be helpful in young individuals with syncope). Treatment consists of elimination of diuretics, nitrates, calcium-channel blockers, and beta-blockers if possible. If this step is not successful, the patient should drink at least 20 oz water and should salt food excessively during each meal. If necessary, fludrocortisone (Florinef) up to 0.6 mg/day can be prescribed. If this treatment is not effective, midodrine 10 mg up to three times a day can be tried. The patient should be instructed to sleep on a 30-degree wedge pillow to avoid supine hypertension.

### *Head Extension Dizziness and Extreme Rotation Dizziness*

The otoliths are outside their optimal functioning range when the head is pitched back.[15] This head position can cause dizziness in individuals when they are seated or standing. Normal individuals who rotate the head to an extreme position laterally also may experience vague dizziness. This unusual posture may be due to a sensory mismatch[16] or to a disturbance in spatial localization. When the Dix-Hallpike test result is negative in a patient complaining of consistent positional dizziness, it is useful to ask the patient to demonstrate the head posture that most commonly causes the dizziness. This can help confirm the diagnosis of head extension or extreme rotation dizziness.

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## CASE STUDY

The patient is a 45-year-old woman with a 3-month history of positional vertigo (getting out of bed), blurred vision, oscillopsia, imbalance, and throbbing headache. She has had migraine headaches since age 27. Clinical findings are normal. Magnetic resonance imaging and caloric test results are both normal. She visits the clinic on the day of her next spell. Physical findings are normal except during positional testing using a video infrared camera to block fixation.

The video shows sustained left-beating nystagmus with the left ear down and sustained right-beating nystagmus with the right ear down. She also has transient upbeat and right torsional nystagmus with the right ear down. She had mild vertigo during the sustained portions of nystagmus, and moderate vertigo during the transient upbeat nystagmus.

*Comment*

This patient has sustained geotropic nystagmus and right posterior canal BPPV. The sustained geotropic nystagmus does not fit the pattern for horizontal canal BPPV; therefore, it is most likely due to migraine aura and represents central positional vertigo (benign). She undergoes treatment for the BPPV and is started on a migraine program (see Chapter 12). On the follow-up visit in 2 days, she has no positional nystagmus or vertigo.

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CHAPTER 18

# 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 6000 degrees/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 velocity, acceleration, and frequency. 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 (preprogrammed saccades [PPSs]) 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. 18.1),[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 PPSs also function to improve gaze stability associated with ipsilesional head rotations. The influence of the PPS 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 VIIIth cranial neurectomy for removal of a vestibular schwannoma. The investigators 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

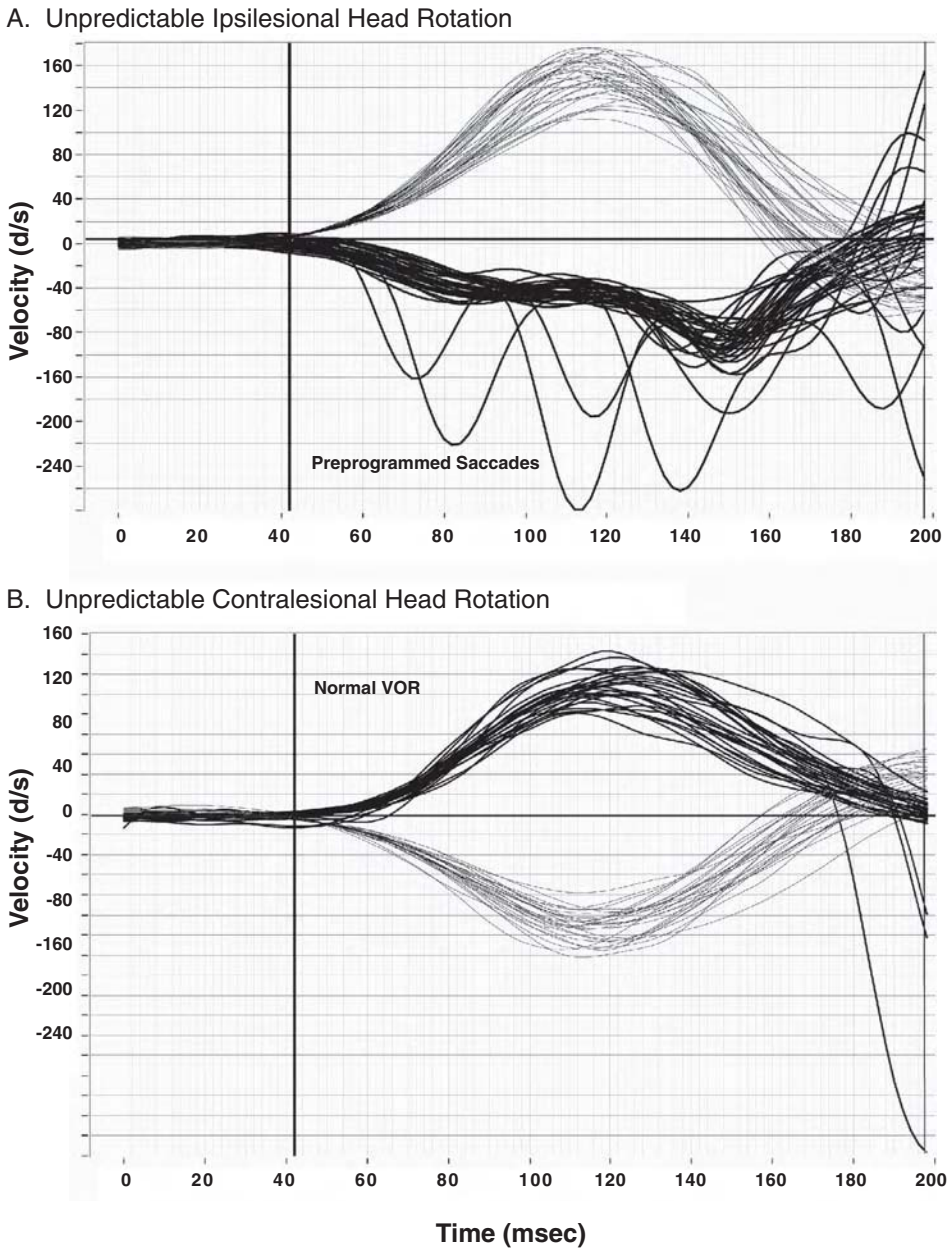


**Figure 18.1** Preprogrammed saccades vs. normal vestibular-ocular reflex (VOR) in a person with unilateral vestibular hypofunction (UVH). *Black traces* reflect head velocity, and *gray traces* eye velocity. The *vertical line* marks the onset of the head rotation. (*A*) The slow eye velocity is deficient in relation to the head velocity (low VOR gain), and preprogrammed saccades (PPSs) are recruited. Note that the direction of the PPSs 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.

similar synergistic relationship between slow-component vestibular and saccadic eye movements for the purpose of improving gaze stability.

The translational VOR (tVOR; otolith-mediated) has also been shown to benefit from the symbiotic relationship between VOR and saccades. Shelhamer and associates[20] adaptively increased the gain of the tVOR in human subjects 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 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 were asked to follow 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 head rotation (yaw) away from a centrally positioned visual target, a saccade is generated in the opposite direction to the head rotation, back toward the target.[14,18–19,21–23] Most of the investigators agree that these types of saccades (preprogrammed saccades):

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

The studies also demonstrate a great deal of variability in use of preprogrammed saccades. Some patients appear to use PPSs only for high-acceleration or large-amplitude head motions.[14,16,19,23–24]

It has also been shown that the latency to generate a preprogrammed saccade during an ipsilesional unpredictable head rotation (yaw, 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 movement in the place of the deficient vestibular system. The difference, however, is that the eye motion from the COR is generated from receptors in the joints and ligaments of the upper cervical vertebrae.[25] 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 a direction opposite to the head position. 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 rightward static head position would 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 rabbits when the trunk was rotated beneath a still head.[26] 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.[27,28] Recording from cat abducens motor neurons, Hikosaka and Maeda[25] 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,[29] 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 ($0.4 \pm 0.04$ for 0.5 Hz and $0.33 \pm 0.05$ for 2.3 Hz). This study showed that neurons in the vestibular nuclei are sensitive to neck rotation in primates. More importantly, these results appear to support the earlier study in cats concluding that the COR serves as a compensatory mechanism.

The identification of a COR as a means of gaze stabilization in humans is controversial. Part of the controversy is due to the methods used to study the COR, and part to the great variability in reported function of the COR. See Chapter 29 for a review of cervical vertigo.

### 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, 30–33] 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, 30–33] Of particular interest was a case study involving a subject in whom complete loss of vestibular function was diagnosed from caloric irrigation.[32] 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.[34] 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. 18.2). 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.

## 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

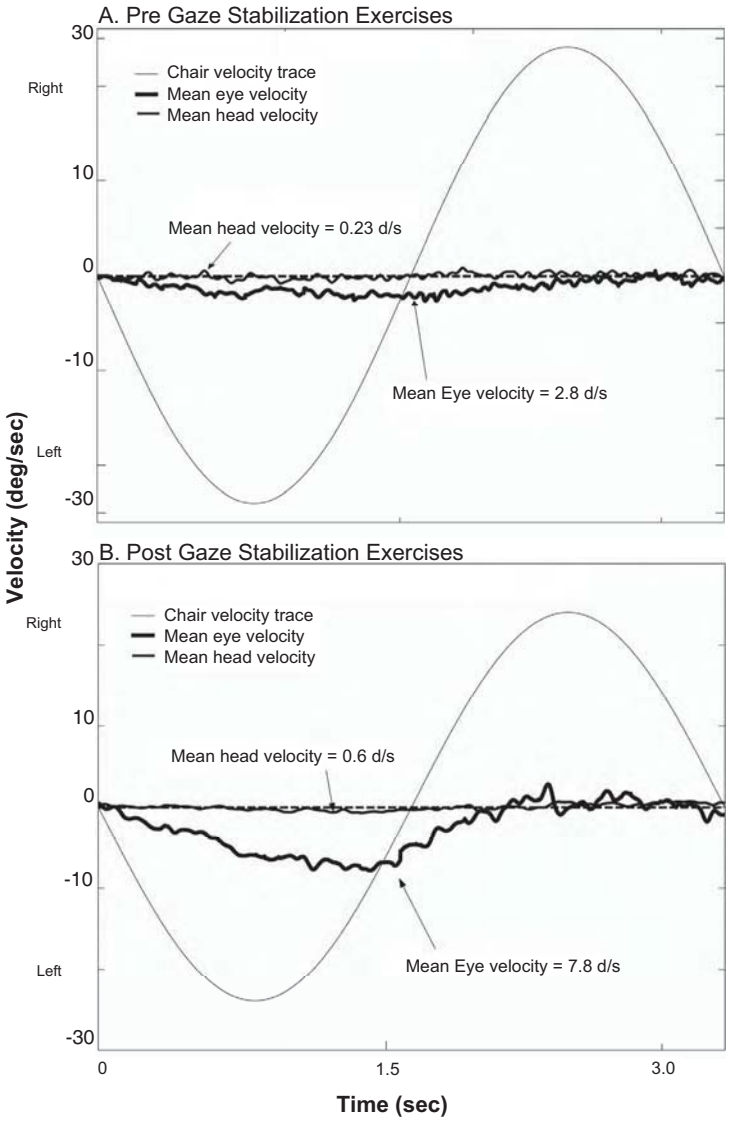


**Figure 18.2** Cervico-ocular reflex (COR) in a patient with unilateral vestibular hypofunction (UVH). The chair, eye, and head velocities are plotted. The subject's head was fixed to a post to prevent head motion. (*A*) The COR gain (eye velocity/chair velocity) was at 0.1 before the patient started a gaze stabilization exercise program. 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.

head movements toward the defect than during unpredictable head movements toward the defect in individuals with vestibular hypofunction.[15,17,35–39] Second, visual acuity during head motion (dynamic visual acuity [DVA]) is better during predictable head rotations than during unpredictable head rotations.[12,40] Enhancement of gaze stability and DVA with predictable head movement is believed to be due to 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. 18.3*A*).[15] These unique eye rotations occur with velocities greater than the velocity limit imposed by inhibitory cutoff (@ 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. 18.3*B*).

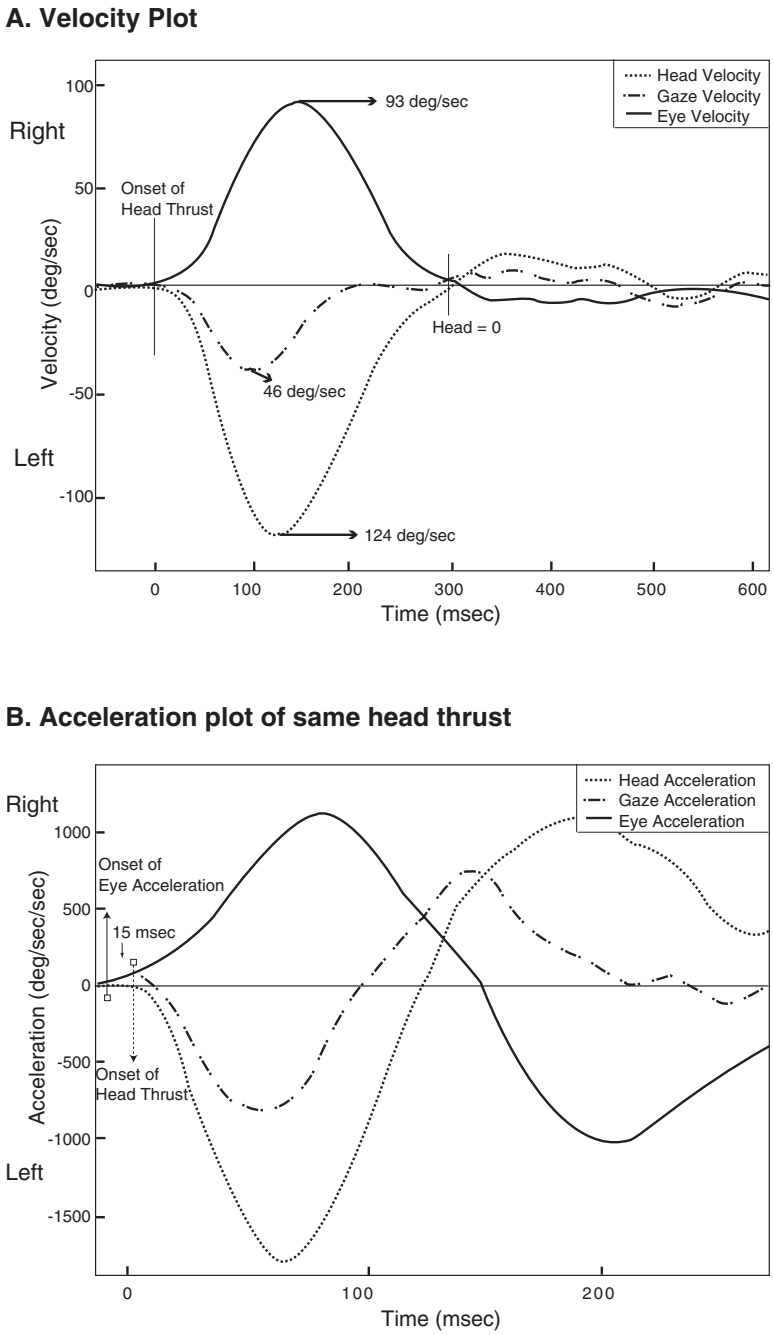


**Figure 18.3** Velocity and acceleration plots of a high-velocity slow phase during a predictable head thrust in a subject with BVH. (*A*) The head is thrust to the left at 124 deg/sec, above the level of inhibitory cutoff, yet the subject generates a slow eye velocity of 93 deg/sec, for a gain of 0.74. (*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.[41,42] 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.[43]

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 associates,[44] 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.

## Summary

Evidence strongly supports the notion that people with vestibular hypofunction use different strategies for gaze stabilization. These data demand that rehabilitation approaches be customized to meet the unique adaptive strategies and capacities of each patient.

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CHAPTER 19

# Physical Therapy Assessment of Vestibular Hypofunction

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Patients with peripheral vestibular hypofunction differ with respect to the onset and clinical course of their disability as well as 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 effect 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 19-1).

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."

## 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 19-2) and from the results of sensory mismatch and physical deconditioning.[13]

### *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 move-

**Box 19-1**

## HEALTH CONDITION[12]

| 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<br>Support and relationships<br>Attitude of family<br>Attitude of society<br>Services, systems, policies<br>Products and technology | e.g. | Gender, age<br>Co-morbidities<br>Social background<br>Education and profession<br>Past experience<br>Coping and character style |

ment. 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.

**Box 19-2**

## 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.

Other eye movements, such as saccades and pursuit, are not affected by vestibular loss. Saccadic eye movements are rapid voluntary 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]

### 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, and the patient experiences a spinning sensation even when he or she is not moving at all. In patients with a chronic vestibular dysfunction, the asymmetry becomes a problem only 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.[18] In addition, information about the orientation of the body with respect to the support surface and gravity is essential to postural control. 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 producing a unilateral vestibular neurotomy in baboons induces asymmetrical excitability in ipsilateral and contralateral spinal reflexes. Similarly, Allum and Pfaltz,[21] using support surface rotations, 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. 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.[22,23]

### 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.[24–27] Walking with head movement appears to be particularly complex for persons with vestibular dysfunction,[28] and limited activity is often the result. In addition, clinical observation of their gait commonly reveals deviations such as veering left or right, a widened base of support, decreased gait speed, shortened step lengths, decreased arm swing, a diminished ability to perform multiple tasks while walking, occasional head or trunk tilting, an impaired perception of vertical, and decreased head and trunk motion.[29–31] For some patients, the use of an assistive device such as a cane reduces gait instability by acting as an additional proprioceptive cue. Work by Jeka and Lackner[32–34] seems to support this concept.

## Participation versus Restriction

As a result of many factors, including fear of falling, embarrassment about staggering while walking, personality, and the uncomfortableness of head movement–induced symptoms, patients may adopt a more sedentary lifestyle, frequently abandoning premorbid exercise routines or recreational activities.[10,35] 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 19-1 lists a variety of measurement tools according to ICF domain.

■ Table 19-1 **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 |
| Dizziness Handicap Inventory | | | X |
| Vestibular Disorders Activities of Daily Living Scale | | | X |
| Disability Score | | | X |

These tools are discussed in the context of the full examination.

## *History*

### Medical History

Physical therapy is usually initiated after vestibular laboratory testing and the physician's determination of the patient's diagnosis. The diagnosis, vestibular laboratory test results, other diagnostic test results, and 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 evaluation. 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, could affect and prolong the patient's functional recovery. Diabetes, heart disease, old neck and back injuries, a history of migraines, and preexisting or long-term visual dysfunction 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 is vital because many medications can produce or enhance dizziness (see Chapters 7 and 11). Certain medications 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. Some people need medication in order 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.[36,37] They may not be able to tolerate rehabilitation efforts without such medication to control their dizziness.

### 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 19-2). The use of a questionnaire that the patient can fill out prior to the initial visit is often helpful and saves time (see Appendix 19-A). A complete description of the patient's symptoms should be documented, so that functional progress can be later assessed. Knowing what positions, movements, or situations aggravate the patient's symptoms may be of importance in treatment planning. In addition, the patient should be asked questions about the type, frequency, duration, and intensity of symptoms as well as 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 dysequilibrium can be measured by means of a visual or verbal analog scale similar to that used in the assessment of pain.[38,39] Some therapists use a 0 to 10 scale, and others use a 0 to 100 scale.[39,40] 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 rate their sensation of dizziness, 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.

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 Medium 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.[41–46] Other therapists may use the Sickness Impact Profile or other health status measures.[47] 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 learn whether the patient has actually improved.[48]

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 has been misdiagnosed by the medical community.[49] 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 15). Showing the patient brochures from the Vestibular Disorders Association (VEDA) often validates the condition, and the patient begins to understand that many other people have a similar impairment.

Several tools that have been developed to define the patient's subjective symptoms of dizziness in an objective manner.[50] 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 (see Appendix 19-A).[9,50–54] Items relate to functional, emotional, and physical problems that the patient may have, and these items reflect all three domains of the IFC—function, activity, and participation. 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. The DHI is consistent with high test-retest reliability ($r$ = 0.97).[49] Responses to various items on the DHI can lead the clinician to suspect benign paroxysmal positional vertigo (BPPV).[55] 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.[56] 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]

Shepard and colleagues[57] have suggested the use of a disability scale to objectively document a patient's perceived level of disability (Table 19-2). Test-retest reliability is high (ICC 1.0[57a]). This 6-point Disability Scale has descriptors that range from having no disability to having long-term disability. *Long-term disability* is defined as the inability to work for more than 1 year.[57] 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,[57] can be used at discharge to determine whether the patient perceives a change in disability (Table 19-3). The Disability Scale is also useful in predicting treatment outcome; Shepard and colleagues[57] found that patients who rated their disability as a 4 or 5 were less likely to show significant improvement with rehabilitation.

■ Table 19-2 **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 8:459, 1990.

■ Table 19-3 **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.

## Fall History

Some patients with vestibular dysfunction fall.[1,25,58–60] Taking a 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 floor. 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.[61] The patient can also be asked about having "near falls." *Near falls* are be 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.[61,62] 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 exercises programs can reduce fall risk, even in the frail older adult.[63] Hall and associates[64] 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 19-3). 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. Cohen and coworkers'[10,65] Vestibular Activities of Daily Living Scale is an excellent example of a tool that will assist the therapist in identifying functional limitations.

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.[66,67] 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[68] have reported that 50% of the people referred to the investigators' tertiary physical therapy clinic stated that they were always wary of heights.

Box 19-3

**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?
- Using a computer?

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.[69] After the assessment is complete, the therapist and the patient should discuss whether these goals are realistic and attainable. Many times, 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 so 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 19-4) 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 with indications, where possible, for which conditions different portions of the examination would be unnecessary. Many of the elements of the therapist's assessment are also discussed in Chapter 7. Key elements of the clinical examination are listed in Box 19-5.

## *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 prior to referral for physical therapy. It is therefore not always included in the physical therapy assessment.

First, the patient is observed for the presence of spontaneous nystagmus in room light. In patients with unilateral peripheral vestibular hypofunction, spontaneous nystagmus will be observable in room light during the acute stage after onset of the lesion. Spontaneous nystagmus occurs because of an imbalance in the tonic or

Box 19-4

## 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, 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, diadochokinesia, 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**

Hallpike-Dix 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

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.

Smooth pursuit is tested by asking the patient to track a moving object with the eyes while the head is stationary. This tracking test should assess the patient's entire visual field. Typically this test also assesses the motor function of cranial nerves III, IV, and VI. Inability to perform downgaze is not a sign of vestibular deficits but can occur with other neurological problems (e.g., progressive supranuclear palsy). Patients with this problem may have difficulty seeing objects on the ground as they walk and with descending steps. During the test of smooth-pursuit eye movements, the presence of gaze-evoked nystagmus and the quality of the eye movement should be noted. Saccadic pursuit, especially in younger individuals, or asymmetric pursuit should be noted.

For the patient with nystagmus, however, determining the quality of pursuit eye movements may be difficult. Care must be taken to distinguish gaze-evoked nystagmus from end-point nystagmus. Gaze-evoked nystagmus occurs when the eyes are 30 degrees eccentric. Direction-changing, gaze-evoked nystagmus is a sign of a central lesion. End-point nystagmus, which is normal, occurs when the eyes are at the extreme end of their range

Box 19-5

## 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

**Range of Motion**

Neck (rotation, extension, flexion, lateral flexion)

**Positional Testing**

Hallpike-Dix 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, singleton to right and left, 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

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 difficulty with smooth pursuit, which can confound the results of the testing.

Patients can also be tested for ocular alignment, particularly for skew deviations that can occur during the acute stage of a unilateral vestibular loss. 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. By convention, the skew is named according to the side of the elevated eye (e.g., *right hypertropia* means the right eye is elevated, although in reality the left eye has dropped). As with spontaneous nystagmus from UVL, skew deviations from UVL should resolve within 3–7 days after onset.

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.

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 vestibulo-ocular cancellation (VORc) and is a function of the parietal lobe. Results should agree with the observations made during the smooth-pursuit test.

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 involves an unpredictable, high-acceleration, small-amplitude head thrust in the horizontal plane.[70,71] 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 (5-10 d) but high-acceleration (3000–4000 deg/sec$^2$) thrust is applied horizontally. When the head impulse stops, the eyes are observed for a corrective saccade, a rapid eye movement that returns the eyes to the target. 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 target helps reduce the false-positive results seen in some older individuals because of

poor ability to accommodate to a near target. Testing is always performed with appropriate visual correction (glasses) for the distance tested. The sensitivity of the test has been reported to be 54%, and specificity to be 100%.[72] People without vestibular disease will be able to maintain fixation during both slow and rapid head movements. 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. During the acute stage or with severe deficits, corrective saccades occur even with slow head rotations.

Another evaluation of VOR function is to measure the degradation of visual acuity that occurs with head movement.[73,74] We use a modified ETDRS chart with SLOAN letters (Lighthouse Distance Visual Acuity Tests, Long Island City, New York) for this test. A hand-held Snellen card is not as appropriate because it measures the patient's vision at a distance of only 18 inches, at which distance older patients in particular have difficulty accommodating. 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 gently oscillated at 2 Hz. Using a metronome helps standardize the test. In normal individuals, visual acuity changes at most by one line. In patients with uncompensated, unilateral vestibular loss, visual acuity degrades by three or four lines. A computerized system for measuring visual acuity during head movement is now available (see Chapter 8). Brandt[75] suggests that distance acuity poorer than 20/50 has a significant effect on postural stability. Additionally, visual field loss can also affect balance,[76] and patients with monocular vision may have particular difficulty with depth perception, which would affect their ability to walk up and down stairs.

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, via infrared cameras that 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,[77] tragal pressure–induced nystagmus, hyperventilation-induced nystagmus, and positional nystagmus (see Chapter 7).

During this assessment, the patient is asked to report any symptoms of blurred vision or dizziness. Tests that involve repeated head movements (VOR, head shaking–induced nystagmus) 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.

## 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.[78,79] These tests may not be performed in all subjects but should be considered especially in older individuals.

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.[80] 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 the gain of the VOR occurs with aging, at least at higher frequencies, as well as a more limited adaptive capability.[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 as well as 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.

## Range of Motion and Strength

The patient's range of motion and strength must be assessed in the initial evaluation. 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 (VOR during rapid head thrusts, head shaking–induced nystagmus, 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.[23] 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 not uncommon in the elderly patient to see distal weakness, 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. Postural abnormalities are more commonly seen in people with central disorders of the nervous system.

## 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 19-4). 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 (Fig. 19.1).[82] This test is most commonly utilized in patients who complain of vertigo only when they move into certain positions, but it should be included in almost all assessments. Vertigo and nystagmus occurring when the patient is moved into the Dix-Hallpike position is used to diagnose BPPV (see Chapter 17). In the Dix-Hallpike maneuver, the patient typically sits with the head turned to one side. The patient then is moved backward so that the head is extended over the end of the table approximately 30 degrees below horizontal. The maneuver is performed to both the right and left sides. The patient should be cautioned in advance that the maneuver can cause dizziness or vertigo but nonetheless should be performed. Two variations of the Dix-

■ Table 19-4 **TYPICAL POSITIONS AND MOVEMENTS THAT PROVOKE SYMPTOMS***

| 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 position | | | |
| 6. Return to sit from left Hallpike-Dix position | | | |
| 7. Right Hallpike-Dix position | | | |
| 8. Return to sit from right Hallpike-Dix 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 20 for further information.

Hallpike test, the side-lying test and the roll test, should also be performed (see Chapter 17). It is also important to perform the Dix-Hallpike test because some patients will not have a specific diagnosis but rather a "diagnosis" of dizziness or vertigo. After the Dix-Hallpike test is performed, it may become obvious that these patients have benign paroxysmal positional nystagmus. There are also people who have "subjective" BPPV, as reported by Haynes and associates,[83] who describe symptoms typical of BPPV yet do not exhibit the typical 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.[83] Recognizing benign paroxysmal positional nystagmus and providing the proper intervention can significantly enhance the patient's quality of life.[84]

Benign paroxysmal positional vertigo is a common cause of vertigo.[85] This peripheral vestibular deficit is easily and effectively treated with physical therapy.[86, 87] In contrast, vertebral artery compression is relatively rare. It often causes similar, yet somewhat different symptoms to 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.

Examples of other movements that can be tested are rolling, supine to sit, reaching in sitting toward the floor, and sit to stand. All of these movements should 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. A more formalized assessment of what positions and movements induce symptoms in patient is the Motion Sensitivity Quotient (see Chapter 20). The speed of the activity often affects

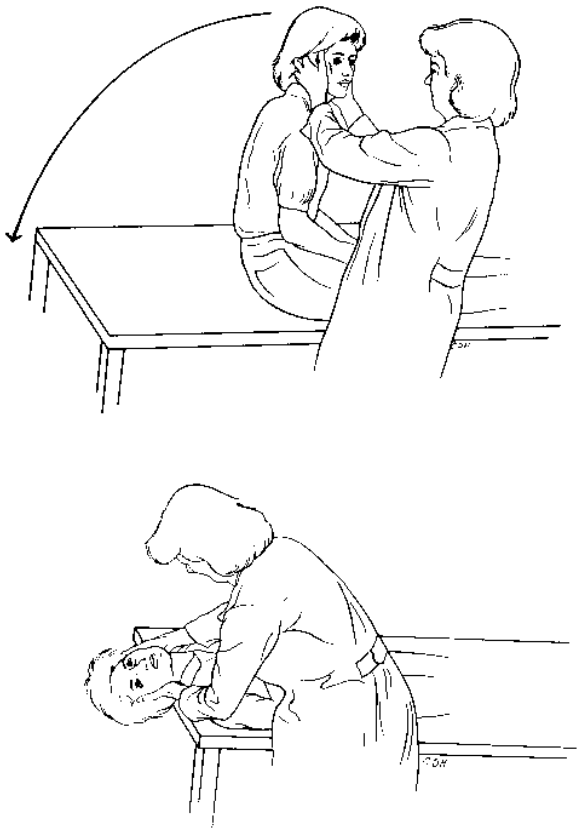

**Figure 19.1** Hallpike-Dix test used primarily to evaluate for benign paroxysmal positional vertigo. The head is turned to one side and the patient is moved from sitting into a supine position with the head hanging over the end of the table. The patient is then observed for nystagmus, and complaints of vertigo are noted. The patient is then returned to the upright position. (From Herdman, 1990.[81])

the patient's symptoms. For example, a quickly performed movement could increase the patient's symptoms, whereas the same movement done 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. 19.2)!

In addition to testing positions and movements that incorporate multiple body segments, the patient is asked to perform 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 also been used in an attempt to better quantify the patient's symptom intensity.[38,39] The same movements are tested in the standing position or during gait, if the patient can tolerate further testing.

## *Balance Assessment*

### Sitting Balance

Many patients with chronic vestibular disorders do not have difficulty with balance in sitting, but for some patients, including an assessment of sitting balance may be appropriate. Patients should be tested while they are leaning anteriorly and posteriorly as well as right and left; tests should be performed both actively and passive-

**Figure 19.2** 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.

ly. 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.

## Static Balance

Static balance tasks have been used clinically to objectively document balance function.[88–92] 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.[92–94] 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.[92] The Romberg test has been shown to have low intrasubject variability when measures are repeated over a 5-day period.[93]

Patients with vestibular deficits may have normal performance on these tests.[94] 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. 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 19-5 lists the expected results for static and dynamic balance tests in patients with acute and compensated unilateral vestibular loss.

## 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 or of simple tools such as a sway grid.[95,96] 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[75] 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

■ Table 19-5 **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 |
| Clinical Test for Sensory Interaction in Balance (CSTIB)—foam, eyes closed | Most cannot perform | Normal |
| Fukuda's stepping tets | 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 |

patient stands from the target should be standardized and should be within 1.5 me. Kirby and associates[97] 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.[51,53] 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[98] 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.[26,98–100] 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. 19.3), 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 CTSIB, 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 CTSIB 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 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.[102–104].

## 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


**Figure 19.3** 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.

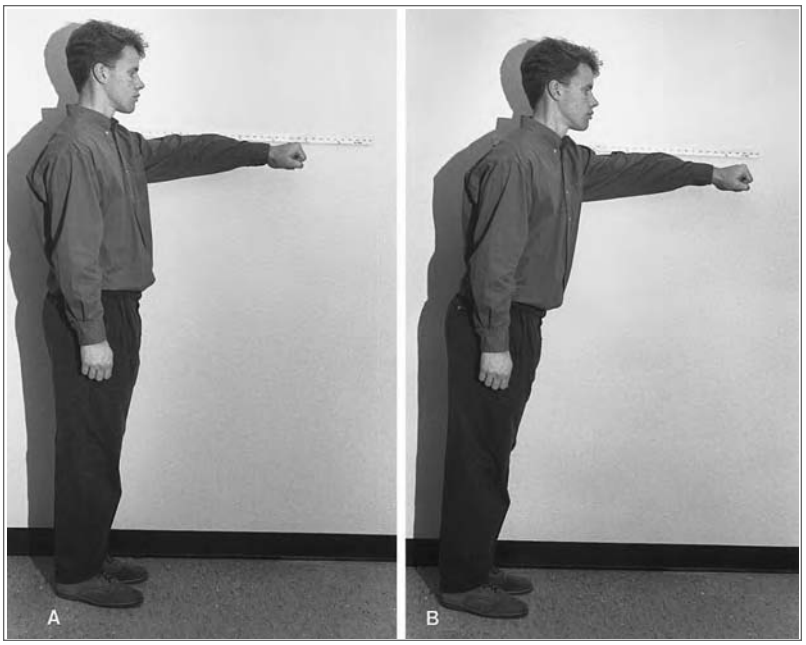


**Figure 19.4** The distance a patient can reach is one measure of functional balance.[106] (*A*) The patient's acromion is lined up with a yardstick while the patient's arm is held parallel to the yardstick. (*B*) The patient reaches forward as far as possible while keeping both feet flat on the ground.

the patient moves freely and symmetrically.[105] 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 standing reach test has been developed as a way to assess the subject's balance and willingness to reach outside the base of support.[51,106–111] 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. 19.4). Duncan and coworkers[108] 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 might be modified so that reaches in different directions are documented (Fig. 19.5).[105] Functional reach has also been shown to be related to falls in older male veterans.[107]

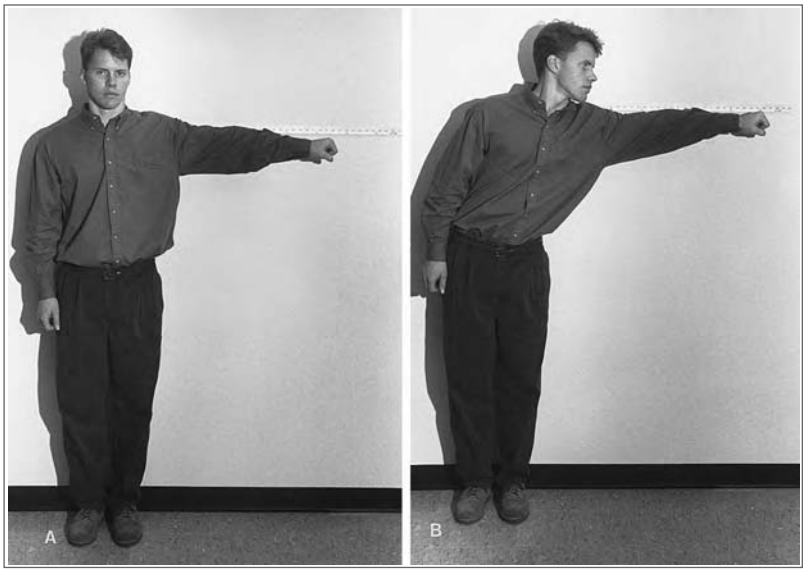


**Figure 19.5** The distance a patient can reach to the side is another possible functional measure of balance. (*A*) The patient's acromion is lined up with the edge of a yardstick. (*B*) The patient reaches to the side as far as possible while keeping both feet flat on the floor.

## ■ Table 19-6 BALANCE CODING

| Criterion | Code |
|---|---|
| *Static Stress* | |
| Unsafe while seated | 0 |
| Safe while seated, unsafe standing | 1 |
| Steady while standing for 20 sec with aid | 2 |
| Steady while standing, 20 sec, no aid, wide base | 3 |
| Steady while standing, 20 sec, no aid, narrow base | 4 |
| Steady while standing, 20 sec, no aid, long base | 5 |
| Steady while standing, 20 sec, no aid, long base, eyes closed | 6 |
| *Rotational Stress* | |
| For those who can stand for 20 sec with or without aid<br>Subject should stand with feet in most stable position | |
| Steady while turning head from right to left<br>(Tested three times, 5 seconds of rest after each trial) | a |
| Can turn 360° without staggering or grabbing onto furniture | b |
| *Sagittal Stress* | |
| Subject can arise from chair (with help if necessary), is immediately steady, and can stand for 20 sec without help except for aid if uses one | x<br>(No code given if cannot perform) |
| *Directional Preponderance* | |
| Coded if subject tends to fall or overbalance in one direction consistently during the above tests | Anterior: (A)<br>Posterior: (P)<br>Right: (R)<br>Left: (L) |

Modified from Gabell and Simons, 1982.[112]

Generally, scores of 6 inches or less indicate that the person is at high risk for falling.[107] 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,[109] although Wernick-Robinson and colleagues[111] suggest that functional reach measures have less value in patients with vestibular dysfunction.

Gabell and Simons[112] also developed a functional balance test to assess elderly clients at risk for falling. Their assessment examines static positions as well as rotational and sagittal movements (Table 19-6) and includes criteria for successful performance of each task. For example, to succeed in one of the rotational stress tasks, the patient must rotate 360 degrees once without staggering or grabbing onto furniture.

Fukuda's stepping test assesses stability during the self-initiated movement of marching, which the patient performs first with eyes open and then with eyes closed.[113] The test is easily administered in the clinic and can be quantified by the use of a polar coordinate grid placed on

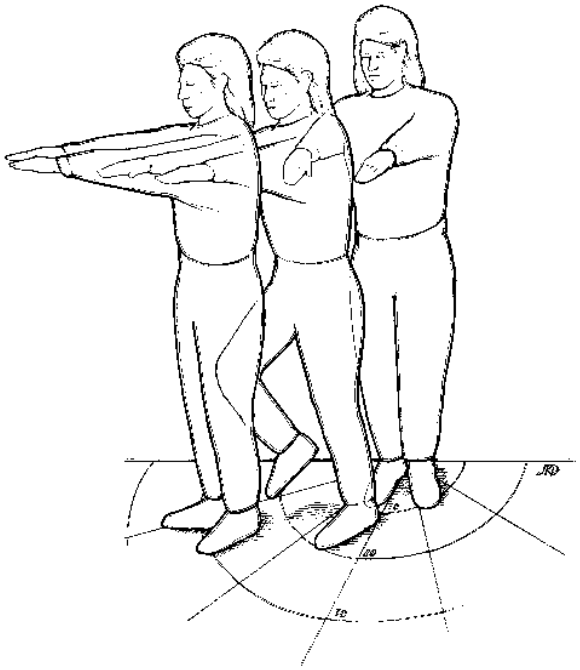

**Figure 19.6** Fukuda's stepping test assesses balance while the patient marches in place, first with eyes open and then with eyes closed. The forward progression of the patient as well as the amount and direction of turning are recorded. Normal subjects will have moved forward less than 50 cm and turned less than 30 degrees at the end of 50 steps. Patients with unilateral vestibular deficits often turn excessively. A polar coordinate grid, placed on the floor, enables easy scoring of the subject's response.

the floor (Fig. 19.6). The test is not specific for vestibular dysfunction, but patients with unilateral vestibular deficits often turn excessively when stepping with the eyes closed.[113] Bonnani and Newton[114] reported that the 50-step test is more reliable than the 100-step test in people without vestibular disease. There appear to be many false-positives and false-negative results for the Fukuda stepping test, so it is important to consider the results as only one aspect of the clinical picture of the patient.[113]

The standing reach test,[101] Gabell and Simon's functional assessment,[112] and Fukuda's stepping test[113] are administered easily and require very little equipment. Such functional measures of balance can easily be used in the clinic or home care setting to document whether the patient has made gains in physical therapy.

## Movement Strategy

During the balance assessment, the therapist should observe and document the patient's movement strategy. Three types of movement strategies have been described for controlling anterior-posterior displacements of the center of mass.[113] The ankle strategy produces shifts in the center of mass via rotation of the body about the ankle joints. According to Horak and Nashner,[115] 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 difficult, 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 lifespan, 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 difficult. 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.[116]

## *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, or computerized motion analysis. 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 noncrowded hallway may yield very different 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 nonpreferred 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"[117] 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),[118] 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.

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 enhance task difficulty, the patient may be asked to perform the same task with the eyes closed. Another such task, called the Singleton test, requires the patient to walk quickly and pivot to the right or left 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. 19.7) as well as 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 therapist. For example, the therapist may decide at which point in the course to throw a ball toward the patient (Fig. 19.8). 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 of 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

**Figure 19.7** 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.

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[25,28,118–121] is very helpful in quantifying gait dysfunction in people with vestibular disease (Box 19-6). 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,[119,121] a tool to assess balance. Scores of 19 or less on the DGI have been related to falls in community-living elderly adults.[119] 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.[121–124] Not all patients with peripheral vestibular disorders have balance

**Figure 19.8** 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 19-6

## 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; or 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 >3 seconds and stops with no loss of balance.

*(continued on following page)*

Box 19-6 *(continued)*

- Moderate impairment: Turns slowly, requires verbal cuing, requires several small steps to catch balance after turn and stop.
- 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.[118]

disorders.[87] Scores of 36 and less on this test indicate a 100% risk of falling in older adults.[119,125] 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.[121]

Another tool that might be helpful to assess gait over time is the Timed "Up and Go" test.[126] 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 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.[28] 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.[28] The TUG has been modified to include components that assess an individual's ability to allocate attention. 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.

## 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 19-7). It is possible for individuals to be referred to a balance and vestibular clinic with undiagnosed central nervous system disease. Acoustic neuromas, multiple sclerosis, brainstem transient ischemic attacks (TIAs), cerebellar disorders, and migraines are but a few of the disorders that have been diagnosed in patients 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.

## Transition from Assessment to Treatment

The following points are offered as guidelines for the development of a treatment program based on the assessment.

Box 19-7

**"RED FLAGS" DURING ASSESSMENT THAT INDICATE A NEED TO ASK FURTHER QUESTIONS**

Numbness
Tingling
Weakness
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 two weeks
Vertical nystagmus without torsional component (not benign paroxysmal positional vertigo)

### *Is There a Documented Vestibular Deficit?*

The results of the 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 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. 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 8, 9).

### *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 benign paroxysmal positional vertigo (see Chapter 17) will not help the patient with a unilateral vestibular loss (see Chapter 20).

### *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.

### 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 upon 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 in order 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." [31,40,127,128] 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. For example, Brown and colleagues[102] defined clinically significant change for the DHI as a change greater than 18 points, whereas Weiner and associates[129] defined it as a change greater than 2 points. In neither case did the investigators 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.[73,130] 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.[118–121] 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. For example, Whitney and colleagues[59] used a composite score based on the ABC, DHI, and DGI scores. Finally, criterion for improvement can be based on change in score that is beyond the within-subject variability of the measures.[64] 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 as well as 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.

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# APPENDIX 19A

## 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)[2]*

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<br>very slightly<br>or not at all | 2<br>a little | 3<br>moderately | 4<br>quite a bit | 5<br>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

Please pick the one statement from the following list that best describes how you feel[1]:
_____ 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[1]:
_____ 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[2] 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[1]:* _____

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 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° 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 Hallpike-Dix maneuver: nystagmus _____ vertigo _____

F. L Hallpike-Dix 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 (video-taped? Y N)*

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)

E. Fukuda's stepping test: eyes closed _____ (FP, turn) Normal? _____

### *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. Singleton test: To right _____ To left _____

I. Fall Risk (Dynamic Gait Index): _____

## *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 _____

## Appendix References

1. Shepard NT, et al: Habituation and balance retraining therapy: a retrospective review. Neurol Clin 1990;8:459.
2. Watson D, et al: Positive and negative affectivity and their relation to anxiety and depressive disorders. J Abnorm Psychol 1988;97:346.

# APPENDIX 19B
# Dizziness Handicap Inventory*

Name: _____ Date: _____

The purpose of this scale is to identify difficulties 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? | _____ | _____ | _____ |
| 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? | _____ | _____ | _____ |

*From Jacobson GP, Newman CW: The development of the dizziness handicap inventory. Arch Otolaryngol Head Neck Surg 1990;116:424. 

| | Yes | No | Sometimes |
|---|---|---|---|
| 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* | _____ | _____ | _____ |
| | (24) | (20) | (22) |

*Total:* _____ F _____ E _____ P _____
(38) (36) (28)

CHAPTER 20

# Interventions for the Patient with Vestibular Hypofunction

*Susan J. Herdman, PT, PhD, FAPTA*
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Vestibular rehabilitation is considered an appropriate and valuable treatment approach for patients with vestibular hypofunction. There are now many randomized controlled studies that have demonstrated the value of vestibular exercises compared with placebo exercises,[1–4] general exercise,[5,6] no exercise,[7] and other vestibular exercises.[8] In addition, numerous observational studies report patient outcomes before and after vestibular rehabilitation, and although these studies lack a control group, they provide information about what factors may affect recovery and the effect of vestibulopathy at the participation/restriction level of function.[9–15] New information about how the nervous system works and about the mechanisms of recovery after vestibular loss has led to the development of more specific exercises than those proposed by Cawthorne and Cooksey in the 1940s.[16,17] This chapter provides the reader with the background necessary to treat patients with vestibular hypofunction. The similarities and differences among the various treatment approaches are examined. Case studies are provided to illustrate the decision-making process in the development of exercise programs.

## Mechanisms of Recovery

Several different mechanisms are involved in the recovery of function after unilateral vestibular loss (UVL). These mecahnisms include cellular recovery, spontaneous reestablishment of the tonic firing rate centrally, vestibular adaptation, the substitution of other strategies, and habituation.

### *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 after aminoglycoside-induced loss.[18,19] There appears to be some functional recovery related to the anatomic recovery, although there is also a persistent deficit.[20] It is unclear at this time whether recovery of hair cells is a significant factor in recovery of vestibular function in human beings.

### *Reestablishment of Tonic Firing Rate*

Asymmetrical disturbances of static vestibular function (nystagmus, skew deviation, and postural asymmetries in stance) recover spontaneously.[21,22] These symptoms and signs are caused by the disruption of tonic vestibulo-ocular and vestibulospinal responses. In the normal individual, when the head is stationary, the tonic firing of the neurons in the vestibular nuclei on each side of the brainstem is balanced. Unilateral loss of the input from the semicircular canals (SCCs) results in an asymmetry in that activity. That asymmetry is interpreted as head movement. For example, loss of the signal from the SCCs

on one side results in a slow-phase eye movement away from the intact side as if the intact side were excited by movement. The slow-phase eye movement is interrupted by a quick-phase eye movement in the opposite direction. This quick-phase eye movement resets the eye position, creating a 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.[23]

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,[24] and in a postural asymmetry, which can be detected clinically.[25] These signs and symptoms resolve within 3 to 14 days after onset of the unilateral vestibular deficit. The timing of the disappearance of the symptoms parallels the recovery of the resting firing rate of the vestibular nucleus neurons.[26] Although visual cues can also be used to suppress spontaneous nystagmus and the postural asymmetry, several studies have demonstrated that recovery of spontaneous nystagmus does not depend on visual inputs per se.[27] Nystagmus decreases at the same rate in animals kept in the dark immediately after unilateral labyrinthectomy as in animals kept in a lighted environment. Spontaneous recovery is probably due to the development of denervation supersensitivity and to axonal sprouting.

## Recovery of the Dynamic Component

UVL also results in disequilibrium and visual blurring, especially during head movements. These symptoms are due to the disruption of the vestibular response to head movement. This results in a dramatic decrease in vestibulo-ocular reflex (VOR) gain during head movements. During the acute stage, the gain of the VOR is decreased by as much as 75% for head movements toward the side of the lesion and by 50% for head movements away from the side of the lesion in patients with unilateral vestibular deficits.[28] Disturbances of the dynamic vestibulo-spinal response are distinguished by a gait ataxia. Typically, patients ambulate with a widened base of support, frequently side-step, and may drift from one side to another while walking. They decrease trunk and head rotation while walking because these rotations would make them less stable. Head movement would result in an asymmetrical vestibular signal that increases their sense of disequilibrium and the 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.[22,29–32] VOR gain does not recover when cats or monkeys are kept in the dark after unilateral labyrinthectomy.[22,31] Recovery of VOR 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, and the recovery period is prolonged (Fig. 20.1).[32]

## Vestibular Adaptation

It was initially believed that the recovery of the dynamic vestibulo-ocular responses were due 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. VOR adaptation can be induced

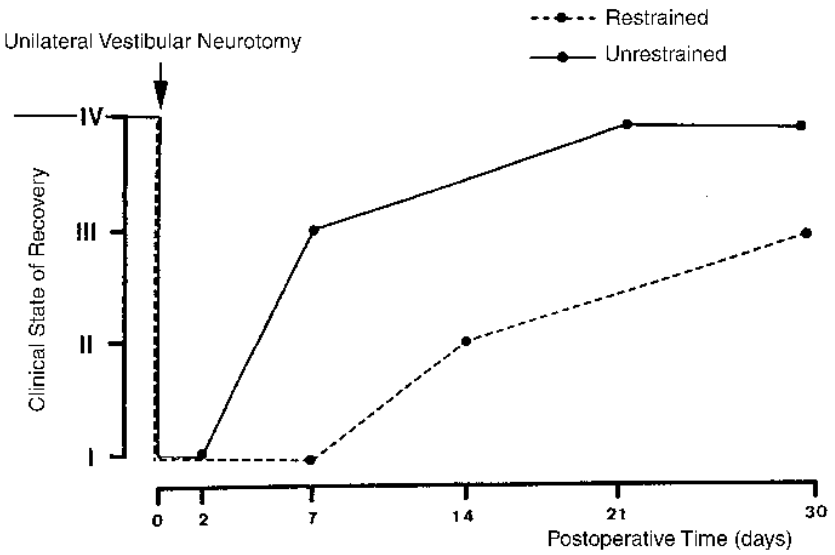


**Figure 20.1** Effect of restricting mobility on rate of recovery after unilateral transection of vestibular nerve. Baboons that were restrained after unilateral vestibular nerve section had a delayed onset of recovery and a more prolonged recovery than those animals that were allowed free movement. (From Lacour et al, 1976.[32])

after unilateral labyrinthectomy in cats as early as the third day after surgery.[33] The vestibular system in human beings can also be adapted during the acute stage after UVL. Pfaltz[34] found an increase in the VOR gain in patients with UVL stimulated optokinetically compared with untreated patients. More recently, Szturm and colleagues[8] reported greater VOR gain in patients after a course of vestibular adaptation exercises but not in a control group.The signal for inducing vestibular adaptation is *retinal slip,* or the movement of a visual image across the retina.[35] This slip results in an error signal that the brain attempts to minimize by increasing the gain of the vestibular responses.

Although retinal slip induces adaptation of the VOR, it may not be the mechanism behind recovery in patients with vestibular hypofunction. Research now suggests that the substitution of alternative strategies may be behind functional recovery or at least contributes significantly to recovery.

## Substitution

The final mechanism involved in recovery after vestibular lesions is the substitution of other strategies to replace the lost function. Enhancement of gaze stability and dynamic visual acuity during predictable head movement is believed to be from either central preprogramming or efference copy of the motor command. For example, patients with vestibular hypofunction make a saccade during the head thrust itself. These preprogrammed saccades occur at a latency that is too short to be voluntary or reflexive[36,37] and are not present in normal subjects.[38] Another mechanism that has been described is anticipatory slow-phase eye movements. These slow-phase eye movements are initiated more than 5 msec 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 are probably initiated by the intact vestibular system, on the basis of the latency to onset of these eye movements (less than 10 msec after the head thrust), which is within the range of the VOR.[39]

Recovery of postural stability may be due to 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 is 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—functions across frequencies of up to 20 Hz[40,41] for activities such as walking and running—and must stabilize gaze during both unpredictable and predictable head movements (Fig. 20.2). Preprogrammed saccades and anticipatory eye movements occur primarily with predictable head movements and not during unpredictable head movements. Other type of eye movements do not function across the necessary frequency range. For example, pursuit eye movements are limited to less than 1 Hz and a maximum eye velocity of 60 degrees per second (deg/sec).[42] 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.[43] The cervical ocular reflex also does not operate in the appropriate frequency range, and well-controlled studies have failed to identifiy a cervico-ocular reflex, in which the eye movements are in the compensatory direction.[44,45] Similar limitations exist for the substitution of somatosensory and visual cues for lost vestibular function (Fig. 20.3). Again, the vestibular system operates through a wider frequency and velocity range than do vision and somatosensation.[40–41,46–48]

Patients may also restrict head movements as a means of seeing clearly or maintaining their balance. This strategy is not particularly desirable, because it

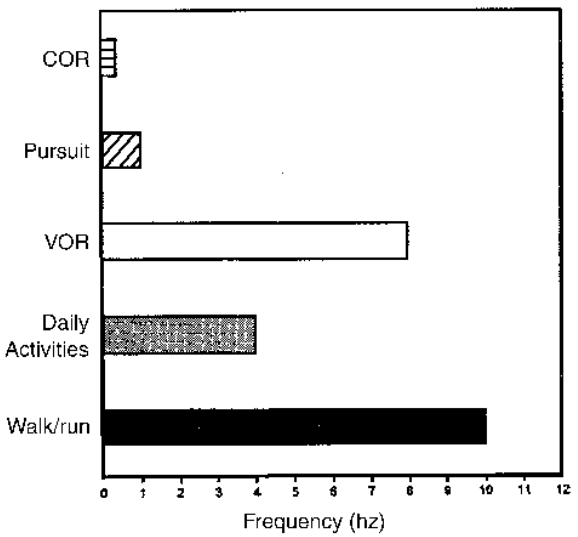


**Figure 20.2** Frequency range over which the cervico-ocular reflex (COR), smooth-pursuit eye movements, and the vestibulo-ocular reflex (VOR) can contribute to gaze stability compared with the frequency and velocity ranges for daily activities, walking, and running. Only the normal VOR operates over the frequency and velocity ranges of normal activities. (Modified from Herdman, 1998.[49])

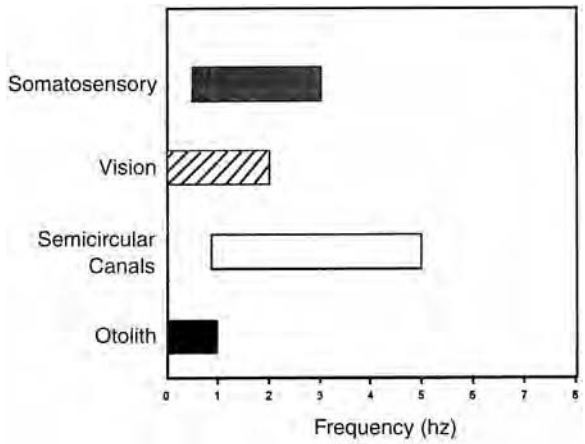


**Figure 20.3** Frequency ranges over which somatosensory, visual, SCC, and otolith inputs contribute to postural stability. This information has been determined only for stability during quiet stance and after sudden perturbations. (Modified from Herdman, 1998.[49])

would result in limited activity but would not provide a mechanism for seeing clearly or for maintaining balance during head movements.

### *Habituation*

*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.

## Evidence that Exercise Facilitates Recovery

The use of exercises, especially if supervised, adds to the health care costs of the patient. It is necessary, therefore, to be able to demonstrate the effectiveness of these exercise approaches in the rehabilitation of patients with vestibular hypofunction. Animal studies support the concept that visuomotor experience facilitates the rate of recovery and improves the final level of recovery after vestibular dysfunction.[30–32] Several animal studies have suggested that exercise may facilitate the process of vestibular compensation. Igarashi and associates[30] found that, following unilateral labyrinthectomy, squirrel monkeys exercising in a rotating cage had less spontaneous nystagmus than a nonexercise control group. In another similar study, Igarashi and associates[50] found that locomotor equilibrium compensation occurred faster in a group of squirrel monkeys exercising in a rotating cage (7.3 days) than in a nonexercise group (13.7 days). Similar findings have been observed in cats after unilateral labyrinthectomy.[29]

Several randomized, controlled studies support the benefits of vestibular exercises in the rehabilitation of patients with vestibular disorders.[1–8] Horak and colleagues[5] compared the effectiveness of vestibular rehabilitation (customized programs consisting of gaze stability, habituation, and balance exercises), general conditioning exercises, and vestibular suppressant medications on dizziness and imbalance in patients with chronic vestibular dysfunction. In this prospective, blinded study, patients were randomly assigned to one of the groups and were followed up for 6 weeks. Dizziness was assessed through a calculation of movement sensitivity based on intensity and duration of symptoms provoked by specific movements and positions, and by indicating whether the symptoms had improved, worsened, or were unchanged. Postural stability was assessed by measuring peak-to-peak anterior-posterior (AP) sway under different sensory conditions. A functional balance measure, single-leg stance (SLS), was also used. These 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 AP sway and increased SLS time. It is understandable that the group receiving medications did not show improvement; both meclizine and diazepam are vestibular suppressants. A later study has shown that long-term use of these medications actually prolongs the recovery period.[51] One criticism of the Horak study is that it did not use an untreated group of patients for comparison with the group performing vestibular exercises. An untreated group, however, would not be an appropriate control, because the subjects would clearly know they were not being treated and they would not have parallel contact with the physical therapists.

In another study of patients with chronic vestibular deficits, Shepard and Telian[52] compared the efficacy of customized vestibular exercise programs with that of a more generic exercise program. They did not use a control group but instead used a delayed treatment paradigm. All subjects were assessed at baseline and again at 1 month before initiating any exercises, to serve as a control for spontaneous recovery. Subjects who did not show spontaneous recovery were then stratified according to age and pretreatment disability levels to ensure that the two groups were similar. After 3 months of therapy, both groups showed a decrease in self-reported levels of dizziness, with 85% of those in the vestibular rehabilitation group and 64% in the generic exercise group reporting complete or dramatic improvement. Only the vestibular rehabilitation group, however, 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 scores, 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.

Herdman and coworkers[1] examined whether patients with acute UVL after resection of acoustic neuroma would benefit from vestibular rehabilitation. This prospective, double-blinded study compared the effect of vestibular adaptation exercises with those of exercises designed to be "vestibular neutral" (smooth-pursuit eye movements performed with the head still). Both groups were instructed in safe ambulation every day. Exercises were initiated on postoperative day 3 in both groups. These researchers found no difference in subjective complaints of vertigo between the groups over the course of the study. This finding 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 patients in the control group. Differences between the groups were also noted for gait pattern, especially with horizontal head movement. All of the control subjects had increased ataxia or developed some ataxia when asked to turn the head while walking. In contrast, only 50% of the vestibular exercise group showed this gait disturbance.

One criticism of this study is that the extent of vestibular dysfunction and vestibular compensation prior to surgery was not known. There was no difference, however, between the two groups on the basis of clinical examination preoperatively. A second criticism is that the study involved only a small number of subjects who might not be representative of patients with recent removal of acoustic neuromas. More recently, however, another randomized controlled trial with a larger cohort of subjects (n = 65) confirmed the findings.[4] 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 after removal of an acoustic neuroma.[3] There were differences, however, in the outcome measures and the exercises used, which may explain the different conclusions; clearly, however, more research is needed on the effect of time from onset on recovery.

The original Cawthorne-Cooksey exercises, developed to treat patients with vestibular deficits, were not customized for the individual patient.[16,17] These exercises are often given as a handout to patients who are simply instructed to "go home and do them." Szturm and colleagues[8] compared the effectiveness of an unsupervised program of Cawthorne-Cooksey exercises and of a customized, supervised program of vestibular adaptation exercises. They found that a greater percentage of patients improved in the vestibular adaptation exercise group than in the Cawthorne-Cooksey exercise group.

All of the studies described here are important because they provide the first evidence that vestibular exercises result in better function than a placebo treatment. As with many investigations, the studies are limited in certain ways as follows:

1. In none of these studies was the exercise approach described in sufficient detail to allow replication of the exercises used.
2. Although the study designs included analysis of group data, typically no information was provided on the individual level—for instance, the number of participants within a group who showed no improvement.
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, and environment, on recovery.

## Goals of Treatment

The goals of physical therapy intervention in vestibular hypofunction 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 participation in society, and (6) reduce the patient's social isolation. Patients are usually seen by the physical therapist on an outpatient basis, although, in some cases, the initial treatments occur while the patient is still in the hospital. An important part of the rehabilitation process is the establishment of a home exercise 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 for the patient the treatment goals as well as the potential effects of exercise.

## Treatment Approaches

Several different approaches have been advocated in the management of patients with vestibular hypofunction. Four different approaches are presented here, although there are elements common to all. Table 20-1 provides a

■ Table 20-1 **COMPARISON OF DIFFERENT EXERCISE APPROACHES FOR THE PATIENT WITH A PERIPHERAL VESTIBULAR DISORDER**

| Feature | Adaptation Exercises | Substitution Exercises | Cawthorne-Cooksey Exercises | Habituation Exercises |
|---|---|---|---|---|
| Incorporate head and neck exercises into the treatment approach | × | × | × | × |
| Use a functional evaluation to assess the symptoms of the patient | × | × | × | × |
| Incorporate principles of motor control and learning into designing a treatment program | × | | | × |
| Practice mental exercises to increase concentration | × | | × | × |
| Have the patient work in a variety of environments and task contexts | × | × | × | × |

summary and comparison of the different approaches. Two case studies are used to demonstrate the basis for specific exercises used in treatment and the progression of the patient's exercise program.

## *Adaptation Exercises*

*Vestibular adaptation* refers to the long-term changes that occur in the response of the vestibular system to input. Adaptation is important during development and maturation as well as in response to disease and injury. Although adaptation of residual vestibular function may not underlie recovery, the use of exercises based on generating a retinal slip error signal does appear to induce recovery.

### Guidelines for Development of Adaptation Exercises

The following points should be considered in the development of exercises based on adaptation for the patient with a unilateral peripheral lesion.

**Consider the Stimulus.** 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 vestibular system. The best stimuli 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 as effectively as head movement combined with a visual stimulus.[35,53–55] Figure 20.4 shows two simple exercises that can be used as the basis for an exercise program for patients with unilateral vestibular lesions. In each, the patient is required to maintain visual fixation on an object while the head is moving.

**Response to Exercise Takes Time.** The early studies on vestibular adaptation used paradigms in which the stimulus was present for several hours or more.[57,58] 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.[34,59] 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 20.4, for instance, should be performed for 1 minute without stopping. The time for each exercise can then be gradually increased to 2 minutes.

**Adaptation of the Vestibulo-Ocular System is Context-Specific.** Therefore, for optimal recovery, exercises must stress the system in different ways.[46] For example, adaptation of the vestibular system is frequency dependent.[60,61] 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.

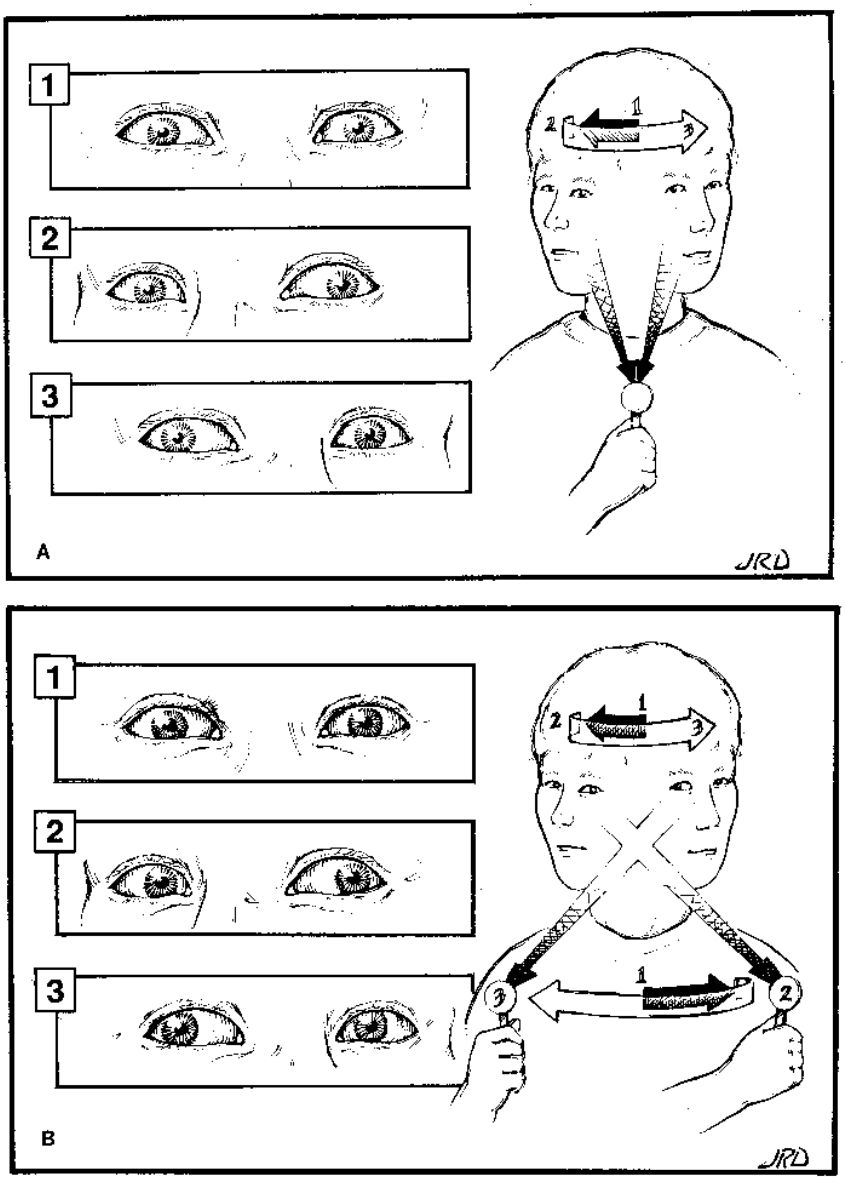


**Figure 20.4** Exercises to increase the gain of the vestibular system can include a ×1 viewing paradigm (*A*) and a ×2 viewing paradigm (*B*). In the ×1 viewing paradigm, the visual target is stationary and the subject moves the head back and forth while trying to maintain visual fixation on the target. In the ×2 viewing paradigm, the target and the head move in opposite directions while the subject again keeps the target in focus. These exercises are performed with 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 et al, 1993.[56])

**Adaptation is Affected by Voluntary Motor Control.** VOR gain can be increased even in the dark if the subject simply imagines looking at a stationary target on the wall while the head is moving. Although not increasing the gain as much as head movement plus vision combined, these results suggest that mental effort will help improve the gain of the system. Patients should be encouraged to concentrate on the task and should not be distracted by conversation and other activities.[62,63]

**Patients Should Always Work at the Limit of Their Ability.** Although patients' morale can be lifted through activities that they can perform relatively easily, most exercises should stress their 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.

## Substitution Exercises

Mechanisms other than adaptation are involved in recovery and should be included in a well-rounded exercise program. 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 gaze and postural stability. For example, balance exercises should "stress" the system by having the patient work with and without visual cues or should alter

somatosensory cues by having the patient stand on foam. Removing or altering cues forces the patient to use the remaining cues. Thus, if the patient is asked to stand on foam with eyes closed, the use of vestibular cues will be fostered.

## Cawthorne-Cooksey Exercises

The Cawthorne-Cooksey exercises were developed in the 1940s.[16,17] At the time, Cawthorne was treating patients with unilateral vestibular deficits and postconcussive disorders. In conjunction with Cooksey, a physiotherapist, Cawthorne developed a series of exercises that addressed their patients' complaints of vertigo and impaired balance. The Cawthorne-Cooksey exercises include movements of the head, tasks requiring coordination of eyes with the head, total body movements, and balance tasks (Box 20-1).

## Guidelines for Development of Cawthorne-Cooksey Substition Exercises

The following points should be considered when one is using the Cawthorne-Cooksey exercises in the treatment of the patient with a unilateral peripheral lesion.

- Cawthorne and Cooksey recommended that the exercises be performed in various positions and at various speeds of movement.
- Patients were required to perform the exercises with the eyes open and closed. According to Cawthorne and Cooksey, 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.[16,17]
- Cawthorne and Cooksey also recommended that patients be trained to function in noisy and crowded environments. These situations may be very difficult for patients with vestibular disorders to manage.
- To encourage active participation, Cawthorne and Cooksey had patients exercise together in daily group sessions. They believed that a group exercise format would be more economical and more fun for patients and also that it would facilitate identifcation of any malingerers. Hecker and colleagues,[65] utilizing Cawthorne-Cooksey exercises to treat a group of patients with vestibular disorders, reported that 84% of the patients responded favorably. They also emphasized the importance of performing the exercises

Box 20-1

### CAWTHORNE-COOKSEY EXERCISES FOR PATIENTS WITH VESTIBULAR HYPOFUNCTION

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 3 ft to 1 ft 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, 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 around 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, stretching, and aiming, such as skittles, bowling, 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.

From Dix, 1979.[64]

regularly.[65] In addition, they noted that emotional stress seemed to affect a patient's progress.

Cooksey[17] stressed that patients should be encouraged to move into positions that provoke symptoms. She believed that with repeated exposure to a stimulus, the patient would eventually tolerate the position without experiencing symptoms. This treatment philosophy is remarkably similar to the philosophy supported by many physical therapists today who use habituation exercises. Most physical therapy clinics that treat patients with vestibular deficits use some component of the Cawthorne-Cooksey exercises.

### Habituation Exercises

The habituation exercise approach is based on the concept that repeated exposure to a provocative stimulus will result in a reduction in the pathological response to that treatment. In the 1980's, Norré[66,67] proposed the use of vestibular habituation training for 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, produces the symptoms experienced by patients with unilateral peripheral vestibular loss.

The Motion Sensitivity Test, 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 (UVH) (Table 20-2).[51,52,68] The patient rates the intensity of the symptoms and indicates the duration of symptoms.

### Guidelines for Development of Habituation Exercises

The following points should be considered when one is developing exercises based on habituation for the patient with a unilateral peripheral lesion.

- Up to four movements are chosen from the test results to form the basis for these exercises. The patient performs these movements two or three 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.
- As habituation occurs, the movements can be performed more rigorously.
- The patient should rest after each movement until the symptoms 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 are usually performed for at least 2 months and then can be gradually decreased to once per day.
- This treatment 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.
- If treatment fails and the patient still experiences debilitating symptoms during leisure or work activities, counseling is advisable regarding changing those activities or reorganizing the work area.

## Expectations for Recovery

Recovery from unilateral vestibular lesions is usually quite good, and patients should expect to return to normal activities. Several factors can affect the final level of recovery, and the therapist should keep them in mind when talking to patients about their progress and anticipated recovery.

Do all subjects improve? Most studies on treatment of patients with vestibular hypofunction examine outcomes for the entire group and focus on those patients who show improvement with intervention. However, several studies on treatment of patients with vestibular hypofunction clearly state the reality that some patients do *not* experience improvement.[2,6,15,69,70] Between 10% and 30% of reported subjects with UVH do not experience improvement, depending on what outcome measure is used.[2,70] Of patients with BVH, outcome is worse, with between 25% and 66% of patients failing to have improvement (SJ Herdman, personal communication, 2006). Identifying and assessing these "nonresponding" patients is extremely important, because we clinicians are obliged to identify these patients, seek the reasons for poor outcome, and develop methods to reverse it.

### *Factors Affecting Outcome*

An important part of treatment is the education of the patient about the possible final level of recovery. This process enables the patient (and the therapist) to set realistic goals. As one might expect, patients with less initial disability and those seen earlier after onset of vestibular

## ■ Table 20-2 MOTION SENSITIVITY QUOTIENT TEST FOR ASSESSING PATIENTS WITH DIZZINESS*

| Baseline Symptoms | Intensity (0–5) | Duration+ | Score |
|---|---|---|---|
| 1. Sitting to supine | | | |
| 2. Supine to left side | | | |
| 3. Supine to right side | | | |
| 4. Supine to sitting | | | |
| 5. Left Hallpike-Dix position | | | |
| 6. Return to sit from left Hallpike-Dix position | | | |
| 7. Right Hallpike-Dix position | | | |
| 8. Return to sit from right Hallpike-Dix 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 | | | |

*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.
From Shepard and Telian, 1995.[52]

hypofunction have better recovery.[12,52] Patients with stable unilateral vestibular deficits, or patients with partially compensated deficits whose symptoms are provoked only by movement, also have a better prognosis.[68]

Only one randomized controlled study has examined the influence of multiple factors on the outcome measured, and only a few factors could be examined because of the small number of subjects in the study.[2] As a group, patients who performed vestibular exercises showed a significant improvement in visual acuity during head movements ($P = .00001$), whereas those performing placebo exercises did not ($P = .07$). Stepwise regression analysis showed the leading factor contributing to improvement to be vestibular exercises, which accounted for 42% of the variance in visual acuity during head movement. Age, time from onset, initial visual acuity during head movement, oscillopsia, and duration of treatment did not contribute to recovery of visual acuity during head movement. However, observational studies provide additional insight about factors that may influence outcome in patients with vestibular disorders. Although these results are limited because of the small numbers of participants studied, or because they included patients with both vestibular and non-vestibular causes of "dizziness," they provide some idea of individual relationships that affect outcome.

**Subject Age and Gender.** Results from several 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 who undergo a course of vestibular exercises.[2,10,12,71,72] Less is known about the role of gender in recovery. Within the group of patients who underwent surgical removal of acoustic neuroma, gender was a significant factor in the patients' postoperative perception of handicap, with male patients reporting a greater perception of handicap at 3 months, but there was no gender effect at 1 year after surgery.[71] In patients with a variety of diagnoses, male patients reported worse disability scores.[51]

**Physical Comorbidities.** Relatively little is known about the effect of comorbid problems on outcome in patients with vestibular hypofunction. Those studies examining specific comorbidities have shown that for some comorbidities (e.g., migraine), patients have an equivalent improvement in physical activities but have greater perceived handicap than patients without the comorbidity.[73–75] Similar findings have been reported by Betchen and associates,[76] who found that persistent headache lowered quality of life and affected return to work in patients who had had resection of acoustic neuroma. Patients with vestibular symptoms after head injury were unlikely to show significant improvements in subjective complaints or balance with vestibular rehabilitation.[12] Patients with cerebellar dysfunction and dizziness also seem to demonstrate less improvement after vestibular rehabilitation than those with other central nervous system conditions (Whitney et al, personal communication, 2006). Perhaps the best guideline now is that patients with multiple comorbidities do not show as great an improvement as those with only one other etiological factor.[77]

**Psychiatric and Psychological Factors.** With any chronic illness, quality of life is an important outcome variable. One would hope that improvements in functional status would lead to meaningful changes in quality of life for a given patient. However, quality of life is complicated by several factors, including anxiety, depression, and locus of control/coping strategies (i.e., internal vs. external control or active vs. passive coping). In some populations, the variable most likely to influence quality of life is depression (e.g., see Jia and associates[78]). However, Grunfeld and colleagues[79] noted that the presence of a vestibular lesion per se is not associated with depression. Somewhat in contrast, Yardley and coworkers[80] found that negative mood, rather than true depression, was a factor contributing to self-perceived handicap in patients with dizziness.

Although there may be no direct association between vestibular deficits and depression, the presence of depression can have a serious effect on the potential for recovery. Krebs and associates[6] observed that some patients with vestibular hypofunction experienced depression that prevented their participation in vestibular rehabilitation. This observation is supported by others, who noted that patient adherence is low for treatments that appear to decrease quality of life[81] whereas better outcomes and improved quality of life are associated with greater treatment adherence.[82] 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.[83,84] 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.[84]

**Coping Strategies.** Several studies have documented that maladaptive coping strategies are associated with decreased quality of life and with greater depression in patients with chronic medical illness.[85,86] One study of patients with episodic vertigo identified the following four primary strategies for coping with vertigo: problem-focused information-seeking, distraction, denial, and relinquishing responsibility.[84] The investigators found that "relinquishing responsibility" predicted handicap even when the analysis controlled for other factors such as emotional distress. Because of the nonepisodic nature of the symptoms in patients with vestibular hypofunction, the relationship of coping strategies and psychological factors with outcome may be different from what has been already reported.

**The Vestibular Deficit Itself.** There appears to be a difference in final level of recovery (at the activity and participation level) between patients with UVH and patients with bilateral vestibular hypofunction (BVH).[12,69,87] The extent to which the severity of deficit (e.g., incomplete or complete) and the rate of development of the problem (e.g., sudden unilateral, simultaneous bilateral, or sequential bilateral) affect the final level of recovery is not clear. Some evidence points to the importance of the underlying etiology to the outcome. For instance, within the group of subjects who had had surgical removal of acoustic neuroma, tumor size and degree of preoperative canal paresis were significant factors in the patients' postoperative perception of handicap.[71] In contrast, patients with surgically induced vestibular loss have a similar level of recovery to that of patients with unilateral vestibular neuritis or labyrinthitis.[12]

**Time from Onset.** Two studies of patients with acute UVH have suggested that delaying the initiation of vestibular exercises results in a delay in the initiation

of recovery or slows compensation.[1,4] In two studies of patients with acute loss of vestibular function after resection of acoustic neuroma, those who started a vestibular rehabilitation program 3 days postoperatively and had better postural stability and less disequilibrium than a control group who performed placebo exercises.[1,4] However, a third study found no difference in outcome between patients performing vestibular exercises and a placebo group.[3] Differences in the outcome measures and the exercises used may explain the different conclusions.

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.[5,8,86,88] This has been shown directly through analysis of time from onset as a factor that might be involved in recovery.[6,12] Up to 30% of patients with chronic vestibular hypofunction, however, do not experience improvement during a course of vestibular exercises.[2,6] This fact raises the suspicion that time from onset combined with other factors may affect the potential to benefit from these exercises

**Medication.** Several investigators have suggested that vestibular suppressant medications such as diazepam may delay or slow recovery,[51,89] but few studies have actually 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.[5] Patients who are treated with steroids for vestibular neuritis, on the other hand, appear to have less perception of handicap after treatment than patients who are not.[90]

**Limit or Delay of Recovery.** If the patient restricts head movement or if visual inputs are minimized during vestibular rehabilitation, recovery may be delayed or limited.[27,31–32] 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 may limit the final level of recovery may be the use of medications that suppress vestibular function.[89]

**Is Recovery Sustained after Rehabilitation is Stopped?** Both anecdotal evidence and systematic studies suggest that recovery after 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.[52] Patients need to be aware of this possibility and should understand that it does not indicate a worsening of the underlying disease. Little is known as to whether patients who have improved function after vestibular rehabilitation need to continue with exercises in order to sustain that improvement. Krebs and associates[6] examined patients with UVH and BVH at 1 year after a course of vestibular rehabilitation. They found that frequency of performing a home exercise program over the course of that year correlated with sustaining improvements in gait speed.[6]

# Treatment

## *General Considerations*

First, treatment should begin early. As mentioned, when visuomotor experience is prevented during the early stages after UVL, there is a delay in recovery.[27,31] Furthermore, the initiation of the recovery of postural responses is delayed, and the course of recovery prolonged, when motor activity is restricted.[32]

Second, exercise can be done for brief periods initially. Pfaltz's study[34] is also 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.

Third, although patients with chronic vestibular deficits often do not have vertigo or vomiting (the exception being patients with episodic vestibular disorders, such as Ménière's disease), they frequently have limited their movements, or at least their head movements, in an attempt to avoid precipitating the symptoms of disequilibrium and nausea. Head movements must be encouraged in these patients both to induce compensation (and thereby improve the function of the remaining vestibular system) and to habituate the symptoms provoked by movement.

Fourth, many of the exercises provided to the patient may, at first, worsen the patient's symptoms. The reason is that the exercises involve head movement, which the patient has been avoiding because moving the head provokes the 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 help 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 whether the exercises can be modified or should be discontinued until the patient is formally reevaluated.

Excessive exacerbation of the patient's symptoms can also be avoided through careful exercise prescription. Initially, the patient may be provided with only a few key exercises. The patient is instructed to attempt the exercises three to five 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. Patients who become excessively "dizzy" while performing the exercises may think that the exercises are making them worse and may refuse to continue with rehabilitation. On subsequent visits, patients are reevaluated and the exercise program is expanded so that all of the physical therapy goals are addressed.

Fifth, it is reasonable to expect improved function within 6 weeks in patients who are compliant with the exercises, but anecdotally at least, the longer the problem has existed, the longer the time needed to see improved function. Once the patient is able to perform the initial vestibular exercises, it may be necessary to take the patient with chronic vestibular deficit through more complex movements in order to habituate the response to movement or at least to make the patient less fearful that movement will precipitate vertigo. Cawthorne's exercises, such as moving from sitting to standing and turning around in between, and the habituation exercises can be useful at this stage (see Box 20-1 and Table 20-2).

## 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 would differ from that for a patient with vestibular neuronitis. Specifically, the treatment program for a patient with Ménière's disease would not address the symptom of vertigo. Compensation of vertigo in Ménière's disease is difficult to achieve because of the fluctuating nature of the disease process itself. Instead, the physical therapy program would focus on improving any persistent imbalance in a variety of task and environmental situations, preventing physical deconditioning, and, if indicated, education in environmental modification and safety awareness.

A problem-oriented treatment approach can incorporate adaptation, substitution, habituation, and the Cawthorne-Cooksey exercises according to what is needed. In addition, it should incorporate functional activities and contemporary principles of motor learning and motor control. As with many of the other approaches, the general treatment progression includes the following:

- Increasing and alternating the speed of the exercises
- Performing exercises in various positions and activities (i.e., head movements performed while sitting, then standing, during walking, and during walking with upper extremity manipulation)
- Performing exercises in situations of decreasing visual and/or somatosensory input (i.e., balance exercises with eyes open and eyes closed)
- 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 Gaze Stabilization*

Visual blurring and dizziness when performing tasks that require visual tracking or gaze stabilization during head movements are most likely due to decreased VOR gain resulting in visual blurring during head movement and also visual-vestibular sensory mismatch. Adaptation exercises can be used to improve the gain of the VOR and therefore improve gaze stabilization.

Some caution should be taken with the use of adaptation exercises during the acute stage after UVL. 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. Good visual inputs (bright room lights, curtains open) also should be encouraged during the first days after the acute onset of a vestibular deficit.

After 1 to 3 days, the symptoms of nausea and vertigo should resolve, and the spontaneous nystagmus should be decreasing as the resting state of the vestibular system readjusts centrally. Patients can begin exercises to facilitate adaptation of the vestibular system (compensation) as early as 1 or 3 days after the onset of the vestibular loss, using gentle, active head movement. Horizontal head movement while fixating on a small (foveal), stationary target is performed for only 1 minute, followed by a period of rest ( X1viewing; see Fig. 20.4 and Box 20-3). The exercise is then repeated using vertical head movements. As patients improve, they should try to perform the exercises for up to 2 minutes each. Although patients may complain of increased vertigo or disequilibrium with head movements, neither is a reason to stop the exercises. Vomiting and significant nausea, however, are reasons for modifying the exercises.

VOR gain in the acute stage after UVL is poor (0.25 to 0.5), and relatively slow head velocities and low fre-

Box 20-2

## EXERCISES TO IMPROVE POSTURAL STABILITY

Many different balance exercises can be used. These exercises are devised to incorporate head movement (vestibular stimulation) or to foster the use of different sensory cues for balance.

1. The patient stands with the feet as close together as possible and with both or one hand helping maintain balance by touching a wall if needed. The patient then turns the head to the right and to the left horizontally while looking straight ahead at the wall for 1 minute without stopping. The patient takes the hand or hands off the wall for longer and longer periods while maintaining balance. The patient then tries moving the feet even closer together.
2. The patient walks, with someone to assist if needed, as often as possible (acute disorders).
3. The patient begins to practice turning the head while walking. This will make the patient less stable, so the patient should stay near a wall while walking.
4. The patient stands with the feet shoulder-width apart with eyes open, looking straight ahead at a target on the wall. The patient progressively narrows the base of support from feet-apart to feet-together to a semi–heel-to-toe 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 seconds before the patient does the next most difficult exercise. The patient practices for a total of 5 to 15 minutes.
5. The patient stands with the feet shoulder-width apart with eyes open, looking straight ahead at a target on the wall. The patient progressively narrows the base of support from feet-apart to feet-together to a semi–heel-to-toe position. The exercise is performed with eyes closed, at first intermittently and then for longer and longer periods. 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 seconds, and then the patient tries the next position. The patient practices for a total of 5 to 15 minutes.
6. A headlamp can be attached to the patient's waist, shoulders, or head and the patient can practice shifting the weight to place the light into targets marked on the wall. This home "biofeedback" exercise can be used with the feet in different positions and with the patient standing on surfaces of different densities.
7. The patient practices standing on a cushioned surface. Progressively more difficult tasks might involve hard floor (linoleum, wood), thin carpet, shag carpet, thin pillow, sofa cushion. Graded-density foam can also be purchased. Backward walking can be attempted cautiously.
8. Make walking more difficult by asking the patient to count backwards while walking. This can be made progressively more difficult by having the patient perform the exercise while walking on different surfaces.
9. 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, then gradually touching only intermittently, and then not at all.
10. The patient practices turning around while walking, at first making a large circle but gradually making smaller and smaller turns. The patient must be sure to turn in both directions.
11. The patient can practice standing and then walking on ramps, either with a firm surface or with more cushioned surface.
12. The patient can practice maintaining balance while sitting and bouncing on a exercise 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.
13. Out in the community, the patient can practice walking in a mall before it is open, and therefore quiet. The patient can practice walking in the mall while walking in the same direction as the flow of traffic or walking against the flow of traffic.

quencies 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 used previously. Within 1 or 2 weeks after onset, patients can begin the vestibular adaptation exercises that require them to maintain fixation on a visual target that is moving in the opposite direction as the head movement (*X*2 viewing; see Fig. 20.4 and Box 20-3). This exercise should be performed with somewhat 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 *X*1 and the *X*2 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 VOR adaptation exercises while standing, as a preparation for walking and turning the head as well.

Ultimately, the adaptation paradigms 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. 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. 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. The electronic "Simon Says" game requires patients to watch and remember a sequence of lights that flash in rapid succession in front of them; this task incorporates Cawthorne's suggestion to include mental exercises in the exercise regimen.

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 add tasks, such as grocery shopping or walking through a shopping mall.

## Problem: Exacerbation of Symptoms

If a patient experiences exacerbation of symptoms during the head movement assessment, habituation exercises are incorporated into the exercise program. During the physical therapy assessment, positions or movements that provoke the patient's symptoms are identified according to the results of the Motion Sensitivity Test (see Chapter 15). These positions and movements are then incorporated into the patient's exercise program.

Movements such as bending forward may exacerbate the patient's symptoms (see Table 20-2). If the patient has significant symptoms, habituation exercises may be performed initially in an easy poistion, such as sitting rather than standing. Later, the patient can perform the exercises while standing or during walking. If straight-plane movements of the head produce few symptoms, neck diagonal movements, with or without a combination of trunk movement, are prescribed. During the head movement/position exercises, the patient is instructed to hold each position for at least 10 seconds or until the symptoms dissipate. In addition, the patient is instructed to perform each exercise three to five times. It is particularly important that the initial prescription of these exercises should avoid those movements/positions that produce severe symptoms. Many therapists have found that too aggressive an initial approach may lead to patient noncompliance. Instead, therapists should select movements and positions that produce minimal to moderate symptoms.

Attempts are also made to include habituation exercises in 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 an object or place it on a high shelf (Fig. 20.5).

## Problem: Static and Dynamic Postural Instability

Balance and gait exercises prescribed for a patient are based on the problems identified during assessment and vary considerably according to patient history and medical diagnosis. Recovery from unilateral vestibular neuronitis, labyrinthitis, or a surgical procedure, such as vestibular nerve section, typically takes 4-6 weeks, although it may take up to 6 months in some patients. Patients who have undergone 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 greater 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 inten-


**Figure 20.5** Neck movements are incorporated into the patient's normal daily routine. In this example, the patient is instructed to incorporate a neck diagonal movement when loading and unloading the dishwasher.

sity (opening a door to the outside or walking through intermittent shadows, such as of 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. Rarely do these patients report an inability to stand on one leg. 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 should be task directed. Balance and gait training are inseparable and therefore considered together in this discussion.

On the basis of 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. 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 are walking backward, sidestepping, and braiding performed with the eyes closed, marching in place on foam performed with the eyes open and later with the eyes closed (Fig. 20.6 and Box 20-2), and walking across an exercise mat or mattress in the dark. It is important that the therapist recognize that there are preparatory exercises the patient should master before attemping these more difficult exercises.

Another functional deficit is the instability patients experience when faced with situations that require movements of the head during gait. 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). In the grocery store, the patient may try this for one or two aisles initially, without a cart.

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

**Figure 20.6** 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.

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. 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 have 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

Box 20-3

## EXERCISES TO IMPROVE GAZE STABILITY

**Acute and Subacute Stages (also used with chronic, uncompensated vestibular loss)**

1. A business card or other target with words on it (foveal target) is taped on the wall in front of the patient so the patient can read it. The patient moves the head gently back and forth horizontally for 1 minute while keeping the words in focus.
2. This is repeated with the head moving vertically for 1 minute.

The patient should repeat each exercise at least three times a day, gradually increasing to five times a day.

The duration of each of the exercises is extended gradually from 1 minute 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 minute to 2 minutes, resting between exercises.

**Chronic Stage**

1. Depending on whether this induces any nausea, the exercise is then repeated using a large pattern such as a checkerboard (full-field stimulus) with the head moving horizontally.
2. The exercise with the checkerboard is then repeated with the head moving vertically.
3. The patient holds a business card in front of face so able to read it. The patient moves the card and the head back and forth horizontally in opposite directions, keeping the words in focus, for 1 minute without stopping.
4. This is repeated with vertical head movements and with a large, full-field stimulus.

The duration is gradually extended from 1 minute to 2 minutes. The patient should repeat each exercise at least three, preferably five, times a day.

5. The patient fixates on a visual target placed on the wall in front of the patient while gently bouncing up and down on a trampoline (otolith stimulation).

with balance and gait activities that challenge the patient's postural control system in a variety of ways.

## Problem: Progression of Balance and Gait Exercises

Recovery of postural stability occurs more gradually than recovery of gaze stability. After 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 can usually stand with the feet together and the eyes closed within 4 to 5 days after onset, although they still have increased sway. Gait is 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 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, and for postoperative patients, bending over must be avoided.

Gait exercises can be more challenging for patients with chronic vestibular disorders, although, as with all cases, the starting point depends on the problems identified during the assessment (see Box 20-3). Patients can be taken through a series of exercises that stress their balance by gradually decreasing the 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 UVL, however, rarely perform the Sharpened Romberg's test 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 another. 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 walking against it), 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.

## Problem: Physical Deconditioning

Physical deconditioning because of inactivity may be a significant problem for many patients with UVL. Most patients are advised to begin a regular walking program, the purpose of which is two-fold: (1) to prevent deconditioning of the patient and (2) to provide realistic balance challenges to the patient's central nervous system. Tasks such as walking on uneven terrain, walking through a shopping mall, and crossing the street challenge the patient in ways that cannot be simulated by a therapeutic exercise program (Fig. 20.7). 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


**Figure 20.7** Patients may have difficulty walking when they must conform to temporal constraints, such as when crossing a street before the traffic light changes.

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 daily. Over subsequent weeks, the patient is instructed to increase to a 30-minute walk or longer. 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. Patients are advised to experience as many challenges as possible when walking in these situations. For example, riding an escalator in a shopping mall may provide an interesting challenge. 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 and golf, that 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 BVL must be more cautious, because without visual cues, they will have difficulty knowing "which way is up" when under water. Ocean swimming in particular may not be advisable. Swimming with goggles will help, but not in murky water.) For all patients, the precaution all people should follow needs to be observed: Never swim alone.

### *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 with 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 an empty parking lot. Driving at night and driving on high-speed roads should be delayed until the patient is comfortable with the quieter roads. Drivers' training is often beneficial, and its cost may be covered through insurance.

## CASE STUDY 1

A 56-year-old woman experienced onset of severe vertigo with nausea and vomiting 6 months ago. At that time, the vertigo appeared to be positional. She reports she had to crawl to the bathroom and that she "passed out" for an unknown period. She was hospitalized for 8 days, during which time she was treated for dehydration and worked up for cardiac problems and stroke. A caloric test performed at that time showed right canal paresis (63% decrease on right). She underwent a course of vestibular rehabilitation at a different site, which she states was not beneficial. Currently she complains of lightheadedness and dizziness with head movement. She is using a cane for ambulation, which she did not need prior to the onset of the vertigo. Review of records from 6 months ago show that at that time, the patient was ambulating with a walker and that her clinical visual acuity was 20/20 with her head stationary but deteriorated to 20/200 during 2-Hz horizontal head oscillations. Additionally, she had restricted participation in her normal social activities, no longer going to church function or out to dinner with friends.

#### *Past Medical and Social History*

Significant weight gain in last 6 months. No history of trauma, exposure to ototoxic medications, heart disease, diabetes, thyroid disorder, hypertension, arthritis, or treatment by a psychiatrist.

*(continued on following page)*

## CASE STUDY 1 *(continued)*

*Comment*

UVH developed in this patient 6 months ago (based on the caloric test); the description of the vertigo as being "positional" is misleading but may indicate a superior vestibular artery lesion resulting in horizontal canal hypofunction and utricular degeneration with sparing of the posterior canal. This would lead to horizontal SCC paresis and posterior SCC 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 and she may have not complied with it. Clearly, she has made some improvement since the onset of the problem—she is walking now with a cane instead of a walker.

*Subjective Complaints*

The patient rates her disequilibrium as 6.0 on a visual analogue scale from 0 (normal) to 10 (worst it could be); she rates her oscillopsia during ambulation as 8.4 on a similar scale. Her dizziness affects her activities 80% of the day.

*Neurological Examination*

Normal findings except as follows:

1. Patient has decreased response to vibration (125 Hz) and abnormal kinesthesia in her toes. 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 test with eyes open or closed for 30 seconds.
4. Walks slowly with widened base of support; turns "en bloc."

**Comment**

The peripheral neuropathy, although mild, may affect the patient's postural stability, especially in the dark, as well as the final level of functional recovery. It is unusual for the Romberg test result to be abnormal. It may be due to her peripheral neuropathy (if it is a profound deficit) or to 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 the following:

- 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.
- VOR is normal during slow head rotations, but patient makes large corrective saccades with head thrusts to the right.
- No spontaneous or gaze-holding nystagmus.
- Dynamic Visual Acuity testing using the computerized system could not be performed because the patient experienced 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 demonstrates the following:

- Horizontal head-shaking resulted in a left-beating nystagmus; there was no vertical head shaking–induced nystagmus.
- Movement into the Hallpike-Dix position to the left had negative result, but with same movement to the right, the patient had 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 deficit. The normal-appearing VOR with slow head movements may be due to the function of the intact left side, residual function of the right vestibular system, or 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 pres-

ence of a left-beating nystagmus after horizontal head shaking is consistent with a right-sided lesion. The left-beating nystagmus, without vertigo, when the patient was moved into the right Hallpike-Dix position is from the asymmetry in the utricular signal produced by movement into that position and is not uncommon in patients with UVL. It may explain why an earlier diagnosis of positional vertigo was made.

### *Balance and Gait*

- The patient walks with a slow cadence (0.63 ft/sec; normal for age is >3.59ft/sec) with and without the cane, a widened base of support, and decreased rotation through the trunk and neck. She is unable to turn her head while walking and keep her balance.
- Fall risk: Her dynamic gait index is 14/24, indicating a risk for falling.

### *Plan for Vestibular Rehabilitation*

**Problems**

- Disequilibrium and postural instability induced by head movement
- Decrement in visual acuity with head movement
- Limited tolerance for head movement
- Ambulation with a cane
- Slow gait speed
- Is at risk for falling
- Decreased participation level
- Social isolation

**Plan**

The patient was started on vestibular adaptation exercises, static and dynamic balance exercises, and some exercises based on functional activities. The exercises consisted of the *X*1 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 the symptoms of dizziness to decrease before starting the next exercise. She used both horizontal and vertical head movements. This exercise was to be performed 3 times daily initially with an increase to 5 times daily in one week if tolerated. 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 increase the gain of the remaining vestibular system, to decrease the patient's symptomatic response to head movement, and to encourage her 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. It was made clear to the patient that she needed to perform the balance exercises in a safe place in her home, such as in a corner facing out. Unfortunately, she lives alone, so a family member is not available to help her. She was instructed to practice walking without her cane and without touching the furniture or walls. This latter exercise she was to perform 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 while walking. The patient was also asked to begin a walking exercises program, increasing her walks to 20 minutes daily.

The patient performed all exercises at least once during this initial clinic visit to ensure that (1) she was able to perform them correctly and (2) they 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 the clinic every 2 weeks.

### *One-Month Reevaluation*

**Subjective Complaints**

The patient rated her disequilibrium as a 3.0 on an analogue scale from 0 (normal) to 10 (worst it could be); she rated her oscillopsia during ambulation as a 2.9 on a similar scale. These scores were a marked improvement over those on the initial visit.

**Balance and Gait**

She could perform the Romberg test with eyes open and with eyes closed for 30 seconds but could not perform the Sharpened Romberg test with eyes open. She was no longer 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 2.75 ft/sec. When asked to turn her head from side to side while walking, there

*(continued on following page)*

## CASE STUDY 1 *(continued)*

was a slight 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, on stairs, and on grass with and without head rotation. She had resumed local driving during 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 after her UVL. Her inability to perform the Sharpened Romberg test with eyes open is normal for age (although some people in her age range can perform this test). A home program of exercises should be continued until the patient's recovery has reached a plateau.

## CASE STUDY 2

A 46-year-old man has complaints of dizziness and imbalance; he is scheduled for resection of left vestibular schwannoma within 1 month. Magnetic resonance imaging (MRI) with gadolinium contrast showed a 1.5-cm tumor extending into the left cerebellopontine angle with no 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 falling. Patient appears quite concerned about the scheduled surgery. He expresses concerns about dying or being unable to return to work after surgery.

### *Past Medical History*

Headache (sinus or migraine), allergic to hay; no hearing in left ear, and a minimal high-frequency loss in right with a speech reception threshold of 35 and no speech discrimination loss. Medications: none. Allergies: No known drug allergies. Patient is independent in basic activities of daily living and drives day and night with no limitations.

### *Subjective Complaints*

He rated his disability as 0/5 (Shepard Disability Scale), indicating that he has negligible symptoms. The Positive and Negative Affective Scale (PANAS) score indicated both anxiety and depression. Head movement did not induce dizziness—visual analog scale (VAS) score 0/10—and only slightly increased his oscillopsia (VAS score 0.1/10) and disequilibrium (VAS score 2/10).

### *Oculomotor Examination*

Positive result of head-thrust test to the left. With Frenzel 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. Quantified visual acuity during active head oscillations between 120 and 180 deg/sec was normal for rightward and leftward head movements.

### *Balance*

Romberg test result was normal, and the patient could maintain the Sharpened Romberg position with eyes open for 30 seconds. He could not perform the Sharpened Romberg test 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 (3.25 ft/sec, compared to a normal value of 3.72 ft/sec). He stated that he normally walks slowly. He did not appear to use excessive visual fixation to maintain his balance while ambulating. Fall risk: Dynamic Gait Index (DGI) = 22/24.

### *Comments*

This patient is typical of many patients with acoustic neuromas, in that his initial complaint was decreased hearing. The absence of vertigo or even disequilibrium in such patients is due to 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, indicating an inadequate vestibular

response. The only abnormal finding in the assessment of his balance was an inability to perform the Sharpened Romberg test with eyes closed. We have found that the Sharpened Romberg test with eyes closed is sensitive to UVL but not specific for vestibular deficits.

The primary concern expressed by the patient is that he might die during surgery or might not be able to work again after surgery.

### *Intervention*

Prior to surgery, the typical postoperative rehabilitation course was discussed with the patient and his wife. Because of his concerns about the operation, an appointment was set up with the neurologist, who prescribed an antianxiety and antidepressant medication for short-term use only.

Surgical resection of the acoustic neuroma was performed 3 weeks later via a translabyrinthine approach because the patient had no usable hearing in the ear on the side of the tumor (see Chapter 14). 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 worsened when he looked to the right. He expressed considerable relief that the opertion was over and no longer appeared to be anxious or depressed.

On postoperative day 3, the patient 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. 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 sidestepped. He appeared to use excessive visual fixation while he walked for balance and slowed his gait when he turned his head while walking. Romberg test result was negative; Sharpened Romberg test was not performed.

Vestibular adaptation exercises were initiated on postoperative day 3. The initial exercise used was the *X*1 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 and vertical head movements. The patient was instructed to perform the exercises 3 to 5 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 after resection of a vestibular schwannoma. Many patients experience vertigo immediately after the surgery because of the sudden asymmetry in vestibuar function with removal of the tumor. The surgery also accounts for the spontaneous nystagmus in this patient. His facial paresis could be a problem when he performs the adaptation exercises, because he cannot blink to lubricate his eye.

### *Early Postoperative Course*

By postoperative day 6, the Sharpened Romberg test result was normal with eyes open but the patient could not perform the test with eyes closed. He was ambulating independently but still had a widened base of support. 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 *X*1 and the *X*2 viewing paradigms, using both a foveal stimulus and a full-field stimulus, which he was to perform standing 5 times a day. He also was instructed to begin practicing turning his head while walking, being careful because it would make him less stable. Each exercise period would take approximately 45 minutes. At this stage, patients are still not allowed to bend over or lift anything weighing more than 5 lb (risk of cerebrospinal fluid leak). The patient was discharged from the hospital on postoperative day 6.

### *Comment*

Rapid recovery for patients with UVL is typical. The effect of the vestibular loss is still obvious (e.g. still has difficulty walking while moving his head). Most patients with UVL are never able to perform the Sharpened Romberg test with eyes closed. We typically do not give these patients any kind of assistive device to use when walking. For some patients, walking with a cane might be helpful for 2 or 3 days while they are in the hospital. Purchasing a cane for these patients is difficult to justify, because it will not be needed at discharge. There are exceptions to this rule, of course.

*(continued on following page)*

## CASE STUDY 2 *(continued)*

### *Outpatient Follow-Up*

At 4 weeks after resection of left vestibular schwannoma, the patient has complaints of dizziness and imbalance, which he notes are worse in the dark. He denies falls. Past medical history is unchanged except for the recent surgery and facial paresis with left eye irritation secondary to it. Medications: Ophthalmic ointment (Puralube) for the eye. He is independent in basic activities of daily living and drives during the day but has not resumed driving at night, primarily because of eye irritation.

**Subjective Complaints**

He rates his disability as 1/5, indicating that his symptoms are bothersome. Head movement VAS = 0.1/10; oscillopsia VAS = 0.1/10; disequilibrium VAS = 0/10. Balance confidence (Activities-specific Balance Confidence scale) = 82.5%.

**Balance**

Fall Risk: DGI score = 22/24 (difficulty keeping his balance when walking with head movements.). Gait speed = 3.10 ft/sec (normal >3.72 ft/sec).

**Visual Acuity**

DVA = 0.036 for head movements to the left (normal < 0.162).

### *Comment*

The patient is 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 has an appointment 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 after this procedure is 1 year, 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 with 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 even then, patients are cautioned to practice gentle rather than forceful facial movements. Another approach would be to send the patient for electromyography biofeedback instruction. 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.

## CASE STUDY 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 falling. She states that she still occasionally staggers, stumbles, and sidesteps when walking. Past medical history is noncontributory. Current medications 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, however, returned to college classes.

### *Neurological Examination*

The general neurological findings were normal except for the vestibular system. MRI and audiogram findings 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 greater difficulty experienced by the patient on all six of the sensory organization tests is unusual, but not unheard of, during the chronic stage of unilateral vestibular deficit. In some patients, this finding may reflect a functional component of 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**

The patient rates her disability as a 1 on a scale from 0 to 5.[68] 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); with predictable head movements to the right: LogMAR = 0.125.

**Static Balance**

Negative Romberg test result; Sharpened Romberg eyes open = normal, eyes closed = 4 sec (normal for age = 60 sec).

**Gait**

Decreased head movement, occasional stagger.

**Fall Risk**

DGI score = 23/24 (increased fall risk indicated by scores $\leq$ 19/24).

**Impression**

Subjective complaints of head movement–induced dizziness, oscillopsia, and imbalance; abnormal visual acuity with 120 deg/sec head movements to right; abnormal gait pattern (decreased head movement); decreased activities (driving); all consistent with subacute UVL.

*(continued on following page)*

## CASE STUDY 3 *(continued)*

*Treatment Goals*

- 75% decrease in head movement–induced symptoms (dizziness, oscillopsia, and disequilibrium) in 6 weeks
- Normal visual acuity during head movement to right in 6 weeks
- Normal static balance with eyes closed except for SR
- Return to all normal activities in 6 weeks

*Comment*

The patient's complaints of disequilibrium with head movement, blurred vision, and a gait disturbance as well as 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 while standing with eyes open, adaptation exercises, and a walking program. She was told to perform the head and adaptation exercises 5 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" of the home exercise program. The following exercises were added to her home program: walking with head movements, walking with pivot turns, and circle walking. She was also given additional eye/head exercises (see previous discussion of 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 4 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. Animal studies and anecdotal evidence in human beings suggest that exercises facilitate recovery of vestibular system function. Early intervention also seems to be important in optimizing recovery. Restricting movement, preventing visual inputs, and the use of vestibular suppressant medications may delay the onset of recovery and limit the final level of recovery.

This chapter has presented treatment strategies for the physical therapy management of patients with unilateral peripheral vestibular hypofunction, who 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 vestibular compensation. Several case studies were presented to illustrate the rehabilitation management of patients with unilateral peripheral vestibular hypofunction.

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CHAPTER 21

# Assessment and Interventions for the Patient with Complete Vestibular Loss

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Patients with unilateral and bilateral vestibular hypofunction (BVH) typically have subjective complaints of imbalance, and they frequently complain of oscillopsia—a visual blurring or jumping of the environment during head movement. These are all serious problems, resulting in decreased activity level, avoidance or modification of driving with resultant diminished independence, limited social interactions, and poor quality of life. Bilateral loss of vestibular function potentially has a more profound effect on a person's ability to participate in the normal activities of daily living than would a unilateral loss of vestibular function, and patients with bilateral vestibular loss (BVL) often restrict their activities and can become isolated.

The main points this chapter addresses are as follows:

1. The assessment and physical therapy treatment appropriate for people with bilateral vestibular hypofunction (BVH) or loss (BVL).
2. The evidence that vestibular rehabilitation can improve postural stability and decrease the sense of disequilibrium in many patients, enabling them to resume a more normal life.[1–4]
3. Preliminary evidence that visual acuity during head movement also improves.
4. Expectations for the outcome of the rehabilitation process, including the observation that not all patients experience improvement.
5. The necessity for patients with BVH to continue with exercises or with activities that challenge balance in order to sustain their improvement.

In addition, several case studies are used to illustrate different points.

## Primary Complaints

### *Balance*

Patients with BVL are primarily concerned with their balance and gait problems. During the acute stage of their disease, they may feel off balance even when lying or sitting down. More typically, however, their balance problems are obvious only when they are standing or walking. Patients in whom BVL develops after the use of an ototoxic medication—the most common cause of BVL—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.

Even with full compensation, balance problems 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 Chapter 20, Figs. 20.2 and 20.3). Normal postural stability while walking requires the combined use of at least two of three sensory cues: visual, vestibu-

lar, and somatosensory. Patients who have no vestibular function, therefore, 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. Most patients are able to prevent falls even though they may sidestep or stagger occasionally.

### *Oscillopsia*

Another problem for patients with BVL is the visual blurring that occurs during head movements. Initially, loss of vestibular function results in a decrement in visual acuity even when the patient is stationary, if the head is not supported.[5] Even after the best compensation, 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 also affects postural stability because decreased visual acuity affects the person's ability to use visual cues for stability.[6]

### *Sense of Disequilibrium or Dizziness*

Patients often complain of "dizziness," "heaviness," or 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. This dizziness or disequilibrium may diminish as a result of compensation, but for many patients, it remains a serious and debilitating problem that can lead to decreased physical activity, social isolation, and depression.

### *Physical Deconditioning*

Poor physical condition can be a significant problem for patients with BVL. It can be caused directly by a decreased activity level because of either the patient's fear of falling or by the increased dizziness that occurs with movement. It is especially a problem for patients whose vestibular loss is secondary to ototoxic medications, who are already debilitated because of severe infection. Many patients undergoing 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 are described here. Physical therapy assessment of patients with BVL must address the intensity of their subjective complaints, postural instability, and oscillopsia, 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 BVL is presented in Box 21-1.

### *History*

#### Etiology

Bilateral loss of vestibular function can occur for several reasons (see Chapter 6). Most common is the effect of an ototoxic medication such as gentamicin. Bilateral vestibular loss was once considered to be an idiosyncratic response to gentamicin; initial studies indicated that less than 3% of people who received gentamicin have a vestibular deficit.[7] Subsequent studies, however, showed that the incidence of aminoglycoside ototoxicity ranges from 9% to 15%.[8–10] These are most likely conservative estimates based on relatively small studies (fewer than 150 participants in each study) and the fact that vestibular loss was assessed with electronystagmography. The prevalence is between 10% and 20%. In patients who have renal impairment, are older than 65 years, are taking loop diuretics, or have previous vestibular loss, it rises to 20% if they undergo renal dialysis and receive gentamicin.

The significance of knowing the underlying etiology of the BVL lies in the accompanying problems that the patient may have. The patient who has a spontaneous or sequential BVL 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. Furthermore, the patient who has a loss of vestibular function, with its resultant balance and visual problems, secondary to ototoxic medication may also have to deal with significant anger and depression.

#### Fall History

Patients with BVH are more likely to fall than normal subjects in their age range and than patients with unilateral vestibular hypofunction.[11] It is imperative that infor-

Box 21-1

## TEST RESULTS IN PATIENTS WITH BILATERAL VESTIBULAR LOSS (BVL)

**Subjective Complaints**

- Disability score: Continues to be abnormal with symptoms typically interfering with both work and leisure activities.
- Activities of daily living (ADLs): Most people with BVL will be independent in their basic ADLs, such as dressing, bathing, getting on and off a toilet, preparing a simple meal, and light housekeeping. Bathing often is modified, however, because of the difficulty getting into and out of a bathtub and/or maintaining balance in a shower when the person closes the eyes.
- Balance confidence: Patient may or may not achieve a criteria improvement to an average confidence in balance $>$ 80% across activities.

**Oculomotor Examination**

- Abnormal findings in room light, including poor vestibulo-ocular reflex (VOR) to slow and rapid head thrusts; visual acuity with head stationary is usually normal, but during gentle oscillation of the head, acuity could change to 20/100 or worse.
- With Frenzel lenses: No spontaneous, gaze-evoked, head shaking–induced, tragal pressure–induced, hyperventilation-induced, or positional nystagmus.
- Dynamic visual acuity, as measured with the computerized system, remains abnormal during active head movements in at least 25% of patients (see Chapter 8).

**Sensation**

Somatosensory and visual information is critical to functional recovery and must be carefully evaluated.

**Coordination**

Should be normal.

**Range of Motion**

Should be normal, but patients may voluntarily restrict head movement because it makes them less stable and also results in poor vision.

**Strength (Gross)**

Should be normal unless patient has become inactive.

**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 while sitting during the acute stage but should not during the compensated stage.

**Static Balance**

- Romberg test: Abnormal result during acute stage in many patients.
- Sharpened Romberg test: Patients with complete or severe bilateral loss will not be able to perform this with eyes closed.
- Single-leg stance: Difficult to perform even during compensated stage, with eyes open.
- Standing on rail: Usually not tested.
- Standing on foam surface: Difficult to perform with decreasing base of support. Should not be attempted in many patients.
- Force platform: During compensated stage, anterior-posterior sway should be normal or close to normal with eyes open and closed 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 result with eyes open during compensated stage; patient 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 walking cannot be performed with eyes closed.
- Gait speed: Preferred gait speed remains slower than normal for age in at least 30% of patients with BVL
- Walk while turning head: Gait slows and becomes ataxic.
- Singleton test: Loss of balance is expected. Uneven surfaces or poor light will result in increased ataxia and, possibly, loss of balance.
- Fall risk: There can be clinically important improvement in scores, but the majority of patients (73%) remain at risk for falling on the basis of Dynamic Gait Index scores.[3]

mation about frequency of falls, when the most recent fall occurred, conditions under which the fall occurred, and whether or not an injury was sustained be obtained from the patient. It is also important to identify whether or not the patient has had any "near falls," times when falling to the ground was prevented because they grabbed onto an object or a person or because someone caught them.

## Comorbidities

While taking the patient's history, the clinician should identify the presence of progressive disorders, especially those affecting vision such as macular degeneration and cataracts, and those affecting sensation in the feet, such as diabetes. These disorders lead to a gradual decrease in available sensory cues and will have an adverse effect on balance in the future.

## Subjective Complaints

The patient's complaints of disequilibrium and oscillopsia can be assessed using a visual analog scale (VAS). The Dizziness Handicap Inventory[12] and the Activities of Daily Living Assessment for Vestibular Patients[13] are useful tools for assessing the patient's perception of disability or handicap as well as the patient's functional abilities and problems.

## Vestibular Function

One important consideration in designing a treatment program is the presence or absence of remaining vestibular function. Vestibular function can be documented with tests such as the rotational chair and caloric tests. This information is then used to determine which exercises to give the patient. If no vestibular function remains, the exercises must be directed at the substitution of visual and somatosensory cues to improve gaze and postural stability.

The presence of remaining vestibular function can be used as a guide to predict the final level of recovery for patients.[2,3] Patients with incomplete BVL 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 cannot drive at all because of the gaze instability. Activities such as sports and dancing may be limited by the visual and the balance problems.

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.[14, 15] Certain aminoglycoside antibiotics are selectively taken up by vestibular hair cells, leading to a gradual loss of vestibular function. Typically, loss of vestibular function continues even after the medication is stopped. Some improvement in vestibular function may occur if some hair cells were affected by the ototoxic drug but not killed. 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.[16]

## Visual System

Assessment of visual function should include at least a gross test of visual field and a measure of visual acuity, because both can affect postural stability.[6] 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, a retinal slip of 3 degrees per second (deg/sec) would cause visual acuity to change from 20/20 to 20/200.[17] As such, in patients with no 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 [DVA]) can be performed either clinically or with a computerized system. The advantage of the computerized system is that the test and result are standardized and more reliable.[18] Clear differences in DVA scores occur among normal subjects, those with dizziness from non-vestibular causes, and patients with known vestibular loss (Table 21-1). The results of the computerized DVA test are both sensitive (90% for those older than 65 years and 97% for those younger than 65 years) 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 its efficacy (Fig. 21.1).[19]

## Somatosensory System

Particular attention should be paid to assessment of vibration, proprioception, and kinesthesia in the feet. Although mild deficits in sensation in the feet may have no effect on postural stability in otherwise normal individuals, but 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 progres-

**■ Table 21-1 DISTRIBUTION OF NORMAL AND ABNORMAL DVA SCORES BASED ON A COMPUTERIZED SYSTEM**

| Type of Subject | DVA (LogMAR) | Normal DVA Score (%) | Abnormal DVA Score (%) |
|---|---|---|---|
| Normal ($n$ = 51) | 0.040 + 0.045 | 96.1 | 3.9 |
| Dizzy, non-vestibular ($n$ = 16) | 0.097 + 0.099 | 87.5 | 12.5 |
| Unilateral vestibular loss ($n$ = 53) | 0.282 + 0.140 | 11.3 | 88.7 |
| Bilateral vestibular loss ($n$ = 34) | 0.405 + 0.134 | 0 | 100 |

DVA = Dynamic Visual Acuity; LogMAR = logarithm of the mminimum angle of resolution.

sive 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 test results. In the compensated stage, the Romberg result is usually normal. Although some patients are able to perform the Sharpened Romberg test with eyes open, they cannot do so with eyes closed. Patients with bilateral vestibular deficits also have difficulty performing tests in which both visual and somatosensory cues are altered. An example would be Fukuda's stepping test, in which

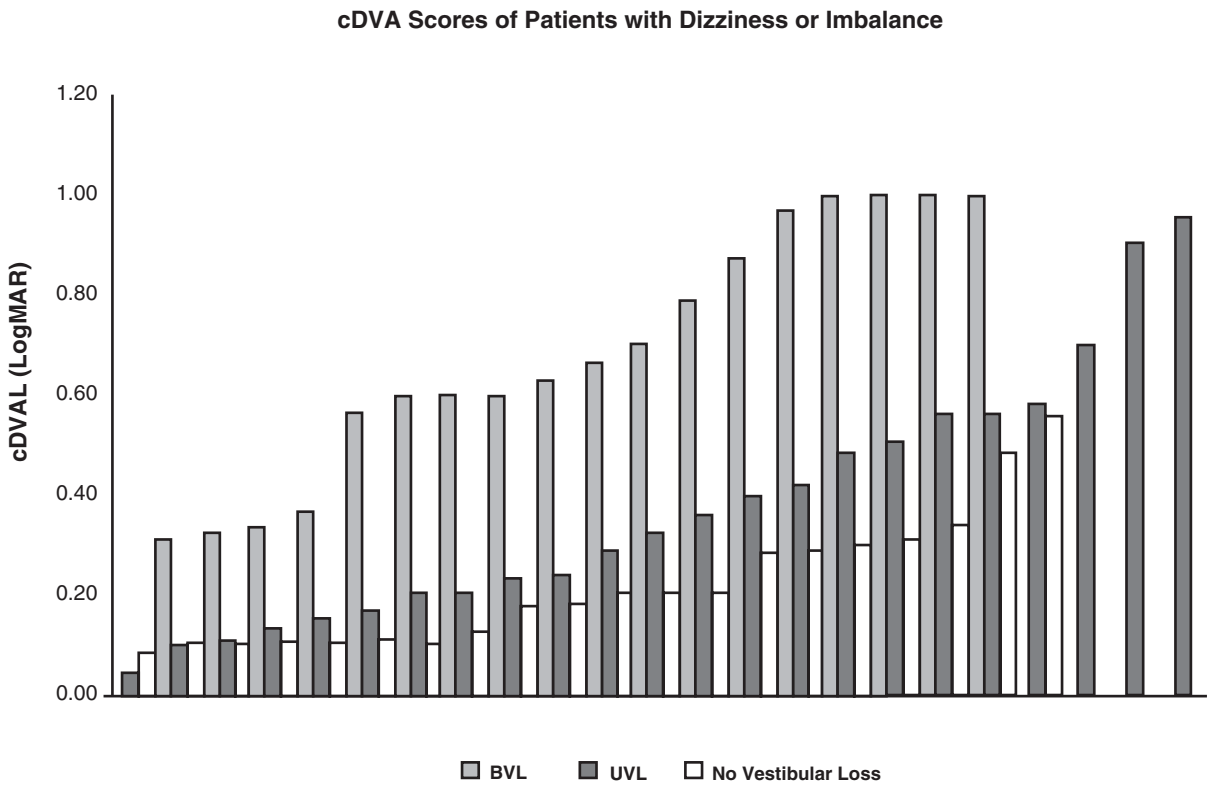


**Figure 21.1** Distribution of dynamic visual acuity scores (shown as the logarithm of the minimum angle of resolution [LogMAR]) obtained with the clinical test in patients with unilateral vestibular loss (UVL) and bilateral vestibular loss (BVL) and in normal subjects. (From Venuto, et al, 1998.[19])

the eyes are closed and the patient is marching in place. Patients may have normal responses with eyes open during this test but will fall with eyes closed.

Determining how well patients use different sensory cues to maintain balance and whether they depend on particular sensory cues is critical. It is important to recognize that these features may vary considerably from patient to patient and can change over the course of recovery. Bles and colleagues[20] have shown that patients with BVL are initially more dependent on visual cues than on somatosensory cues for balance. With time, their ability to use somatosensory cues improves. This improvement varies from patient to patient, however, and must be carefully assessed (Fig. 21.2).

The gait of patients with bilateral vestibular deficits is often wide-based, slow, and ataxic. Patients decrease their trunk and neck rotation in an effort to improve stability by avoiding head movements. Arm swing is similarly decreased. Many patients use excessive visual fixation and therefore have increased difficulty if asked to look up 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.

## Mechanisms of Recovery

The mechanisms used to stabilize gaze in the absence of vestibular inputs have been well studied (Box 21-2). The mechanisms involved in maintaining postural stability are still somewhat less well understood, although research is being done in this area.

### *Gaze Stability*

Subjects without vestibular function must develop different mechanisms to keep the image of the target on the

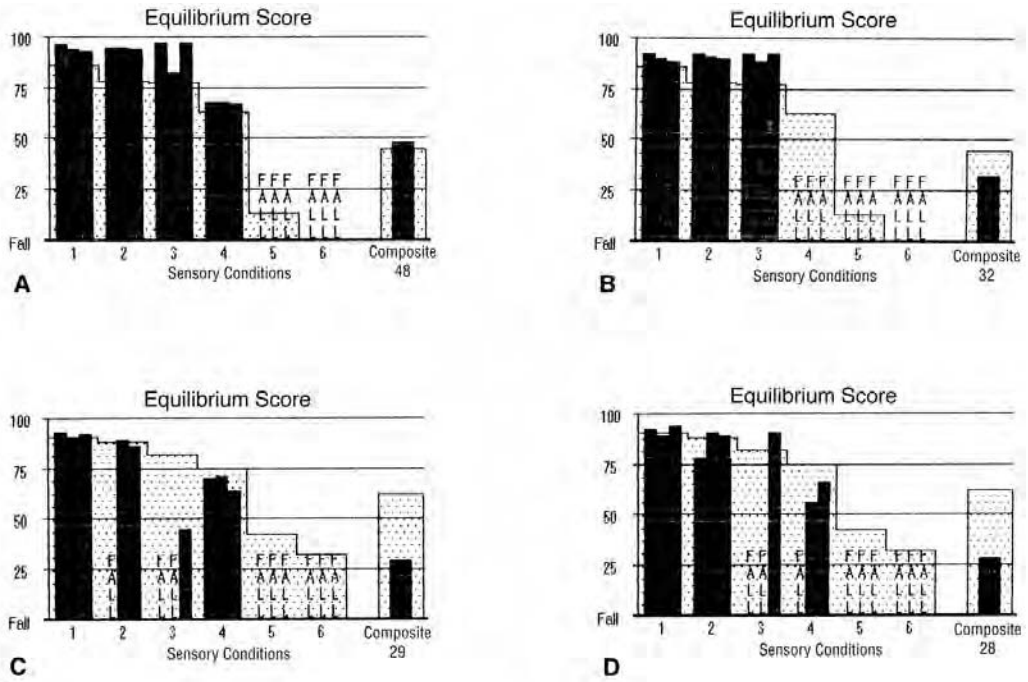


**Figure 21.2** Posturography test results from patients with bilateral vestibular loss demonstrating the differences in ability to maintain postural stability when different sensory cues are altered or removed. Patients are tested using the following six different conditions:

| | Available cues: | Unavailable or altered cues: |
|---|---|---|
| Test condition 1 | Vision, vestibular, somatosensory | — |
| Test condition 2 | Vestibular, somatosensory | Vision absent |
| Test condition 3 | Vestibular, somatosensory | Vision altered |
| Test condition 4 | Vision, vestibular | Somatosensory altered |
| Test condition 5 | Vestibular | Vision absent, somatosensory altered |
| Test condition 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*).

Box 21-2

**MECHANISMS USED TO STABILIZE GAZE**

- Change in amplitude of saccades
- Use of corrective saccades
- Modification of pursuit eye movements
- Central preprogramming of eye movements

fovea during head movements (see Box 21-2). Central preprogramming of movements is probably the primary mechanism by which gaze stability is improved in patients with BVL. The contribution of central preprogramming has been assessed by comparing the gain of compensatory eye movements[21,22] or by comparing visual acuity during predictable and unpredictable head movements.[23] 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 walking.

Other mechanisms used to improve gaze stability are modifications in saccadic and pursuit eye movements.[21,24] 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 after a head movement.[21] 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 depend on the nature of the stimulus (predictable vs. unpredictable; sinusoidal vs. constant-velocity). In general, for sinusoidal stimuli, smooth pursuit works well at frequencies of up to 1 Hz. For constant-velocity stimuli, smooth pursuit can work well up to target velocities of 100 deg/sec. Later evidence demonstrates that patients with bilateral vestibular deficits have higher smoothpursuit gains than normal controls, although the patients' performance is still within the normal range.[24]

At one time, potentiation of the cervico-ocular reflex (COR) was thought to contribute to the recovery of gaze stability in patients with BVL.[21,25,26] 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 head movement frequencies up to 0.3 Hz, well below the frequency range of head movements during normal activities. Therefore, although the COR is increased in patients with BVL, it does not actually operate at frequencies that would contribute significantly to gaze stability during the head movements that would occur during most activities.

Kasai and Zee[21] 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 21-2). No mechanism to improve gaze stability fully compensates for the loss of the VOR, however, and patients continue to have difficulty seeing during rapid head 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 upon which they rely.[20] 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-year study, the subjects recovered the ability to maintain balance to within normal limits in the testing paradigm except at high frequencies. The vestibular system functions at higher frequencies than the visual or somatosensory systems, a difference that would explain 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 and colleagues[27] 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, whether kinesthetic signals from the neck, which would occur during head movement, affect postural stability. Certainly patients with bilateral vestibular

dysfunction become less stable when asked to turn their heads while walking. This observation may indicate that kinesthetic cues do not contribute significantly to dynamic postural stability. Another interpretation is that such patients rely more 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 depend 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 and associates[28] reported a case in which the patient had a complete BVL 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 fixation on a visual target more than 1 m away), his postural stability deteriorated significantly. Again, visual and somatosensory cues do not substitute fully for the lost vestibular contribution to postural stability (see Chapter 20).[29–31]

### *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 patients become comfortable walking in the environment.

## Evidence that Exercise Facilitates Recovery

There is some evidence that experience facilitates recovery after bilateral ablation of the labyrinth. Igarashi and coworkers[32] trained monkeys to run along a straight platform. Performance was scored by counting the number of times the monkeys moved off the straight line. A two-stage ablation of the labyrinth was then performed. After the unilateral ablation, animals given specific exercises recovered faster than nonexercised animals, but all animals eventually achieved preoperative functional levels. After ablation of the second labyrinth, all monkeys had difficulty with the platform run task. The control group reached preoperative balance performance levels in 81 days, whereas the exercise group did so in 62 days. This result was not significantly different owing to the large variation in individual animals. These researchers also measured how long the animals took to reach 8 consecutive trial days in which they could keep their balance at preoperative levels. The exercise group achieved this criterion in 118 days. The control group took longer. One animal took 126 days, another took 168 days, and one animal had not achieved that criterion at 300 days. The conclusions from this study are that (1) recovery from bilateral deficits occurs more slowly than recovery from unilateral lesions, (2) exercise affects that rate of recovery in bilateral and unilateral lesions, and (3) the final level of function may be improved if exercises are given after bilateral lesions.

Several studies have studied the effectiveness of vestibular exercises on postural stability during functional activities for patients with chronic bilateral vestibular deficits. Krebs and colleagues,[1] in a double-blinded, placebo-controlled trial, found that the patients performing customized vestibular and balance exercises had better stability while walking and during stair climbing than 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 and colleagues[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 results for the patients with BVL ($n = 51$) were combined with those for patients with UVL ($n = 33$), 61% of the patients demonstrated significant improvements in gait.

In a retrospective chart review of 13 patients with BVL, Brown and associates[3] noted that as a group, the

patients 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). Again, not all patients benefited to the same extent. These investigators noted that 33% to 55% of the patients demonstrated what were considered clinically significant changes on the different measures. In another retrospective study, Gillespie and Minor[2] found that 63% of the patients with BVL who received vestibular rehabilitation demonstrated improvements (defined as reported increased activity levels, reduced symptoms, and demonstrated normal gait velocity, normal Romberg test result, or normal DVA score).

### Expectations of Level of Recovery

Several studies of patients with vestibular hypofunction clearly state that some patients do not improve.[3,4,33–36] In patients with UVH, depending on what outcome measure is used, between 10% and 30% of subjects do not improve.[34,35] In patients with BVH, outcome is worse, with between 25% and 66% failing to show improvement[3] (Herdman preliminary data). These findings are extremely important, because as clinicians, we deal with individuals, not groups.

### Treatment Approach

Not all exercise approaches are appropriate for patients with BVL, however. Telian and coworkers[37] 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 subjects after 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 to improve gaze or postural stability.

## Treatment

The treatment approach for patients with complete loss of vestibular function involves the use of exercises that foster (1) the substitution of visual and somatosensory information to improve gaze and postural stability and (2) the development of compensatory strategies that can be used in situations in which balance is maximally stressed (Boxes 21-3 and 21-4). Patients with some remaining vestibular function may benefit from vestibular adaptation exercises to enhance remaining vestibular function (Fig. 21.3, page 347). For both groups, postural stability can be improved by fostering the use of visual and somatosensory cues. This approach is also used in the treatment of patients unilateral with vestibular hypofunction (see Box 21-4).

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 extent 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 his or her eventual return to normal activities. Head movement, a component of all exercises, increases 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; the Head Movement VAS can be used to quantify this problem. Additionally, inability of the patient to sustain head movement for 1 minute helps define the initial exercise program.

It is important to explain to the patient that some increase in dizziness is expected at the beginning of the exercise program and with any increase in the intensity of the exercises. Only one exercise involving head movement should be prescribed initially. Other exercises can be added and the frequency and duration of the exercises can be increased as the patient improves. The patient should perform at least one set of all the exercises at the time of the clinic visit. Patients should also be taught how to modify the exercises if the dizziness becomes overwhelming (Box 21-5, page 347). They should be strongly encouraged to contact the therapist if they are having difficulty. In patients for whom dizziness continues to be a problem, we suggest meditation and relaxation techniques to try to reduce the effect of the dizziness on the patient's life.

### *Progression of Exercises*

The reported number and frequency of patient visits to the clinic varies tremendously from study to study. Improvement in patients undergoing vestibular rehabilitation has been reported when patients were seen once a day for 3 days[38], two or three times a week for months, once a week for 4 or 5 weeks,[34,35,38], once a month,[3,39-41] or even once in several months.[42,43] It is difficult to determine appropriate practice patterns by comparing these studies, however, because different exercise approaches,

Box 21-3

## PATIENT INSTRUCTIONS FOR 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, keep 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, the physical therapist should watch the patient's eyes closely. If the patient is making corrective saccades, he or she should slow the head movement down.
2. Active eye-head movements between two horizontal targets to foster the use of saccadic or pursuit strategies and central preprogramming:
   - 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 (saccade 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 that when you are looking directly at one, you can see the other with your peripheral vision. Practice for 2–3 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 whether 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 2–5 minutes, resting if necessary during the exercise.

different outcome measures, and different patient populations were used. However, two randomized clinical trials demonstrated significant benefits in function in patients with BVH who were supervised in the clinic only one visit per week and performed an extensive home exercise program for 8 weeks.[1,4]

Changing the duration of any given exercise, the frequency of performance, and how many different exercises are given (Table 21-2 page 348) can modify the intensity of the exercise program to improve gaze stability. Patients 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 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. As with other types of exercises, the initial program and the rate of progression should be customized for each patient.

### *Guidelines to Treatment and Prognosis*

The physical therapist should keep the following factors in mind when working with the patient with a bilateral vestibular deficit:

1. The patient's perception of dizziness can be the major deterrent to his or her return to normal activities.
2. Recovery after bilateral deficits is slower than after unilateral lesions and can continue for as long as 2 years. This is not to suggest that the patient should be actively engaged in rehabilitation for the entire 2 years but, rather, that imparting this knowledge should help the patient with BVH understand that improvement will most likely be a slow process.
3. Recovery is easily upset by other medical problems, such as having a cold or receiving chemotherapy.

Box 21-4

## PATIENT INSTRUCTIONS FOR EXERCISES TO IMPROVE POSTURAL STABILITY*

The purpose of these exercises is to force you to develop strategies for 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. For all of 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 1 inch at a time.

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.

_____ 3. Repeat #_____ above but while standing on a foam pillow.

(*Note:* Unusual for patients with bilateral vestibular loss to do this exercise.)

_____ 4. 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.

_____ 5. 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.

_____ 6. Walk and turn your head to the right and to the left as you walk while you count backwards out loud from 100 by 3s.

Try to focus on different objects as you walk.

Gradually turn your head faster.

Practice for _____ minutes.

_____ 7. 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.

*Therapist uses the blanks to mark which exercises the patient should perform as well as to specify durations and variations.

4. To maintain recovered function, patients may always need to be doing some exercises, at least intermittently.[4]
5. The patient's postural stability will never be completely normal. The patient may demonstrate a negative Romberg test result and may be able to maintain the Sharpened Romberg position with eyes open, but will not be able to do so 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

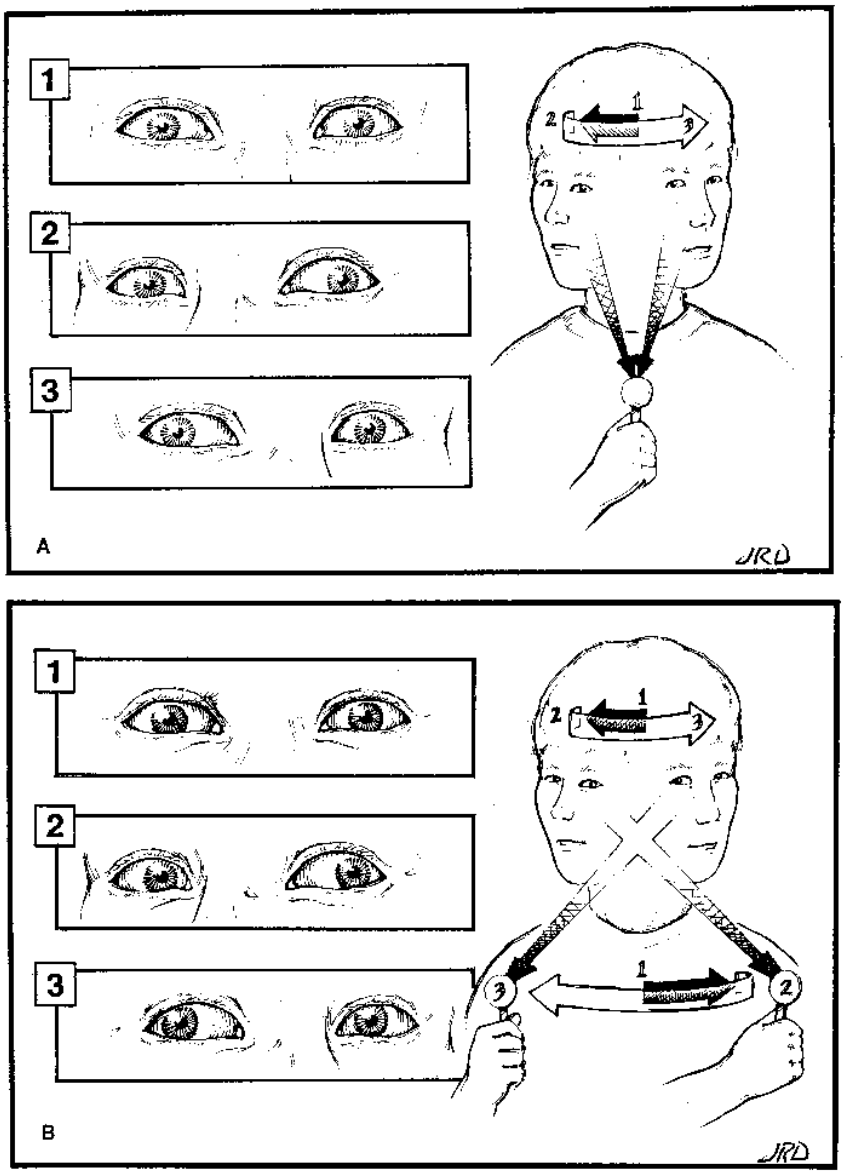


**Figure 21.3** Exercises that enhance the adaptation of the vestibular system should be used for any patient who has remaining vestibular function. These exercises should be performed many times a day and can be done while the patient is sitting and, eventually, standing. (Modified from Tusa and Herdman, 1993.[46])

Box 21-5

## ADJUSTMENTS MADE TO EXERCISES BECAUSE OF SEVERE DIZZINESS

- Perform the exercises fewer times each day.
- Move the head more slowly.
- Perform each exercise for a shorter period.
- Rest longer after each exercise.

some of the time. Most patients, however, are eventually able to walk without any assistive devices. Ambulation during the acute stage is wide-based and ataxic with shortened stride length and sidestepping to the right and left. The patient will turn "en bloc" and turning the head will increase instability. Ambulation will improve, but it, too, will not be normal.

7. Patients will be at increased risk for falls.[11] Patients will need to be careful when walking in low-vision situations, over uneven surfaces, or when they are fatigued.

## ■ Table 21-2 SUGGESTED PROGRESSION FOR GAZE STABILITY EXERCISES

| Exercise | Duration | Frequency | Position |
|---|---|---|---|
| *X*1 viewing paradigm against plain stationary background; horizontal or horizontal-and-vertical head movement | Maybe for <1 minute each time | 2 or 3 times daily | Sitting until can perform the head movements easily and then standing |
| *X*1 viewing paradigm against plain stationary background; horizontal or horizontal-and-vertical head movement | Increase to 1 minute each exercise | Increase to 5 times daily | Standing* |
| *X*1 viewing paradigm with target held in hand against a plain background; horizontal and then vertical head movements | 1 minute each exercise | Up to 5 times daily | Standing* |
| Add eye-head exercises, horizontal and vertical | No specific duration | 2 or 3 times daily | Sitting at first and then standing |
| *X*1 viewing paradigm with target held in hand and also with target at distance | 1 minute each exercise | 2 or 3 times daily | Standing* |
| Eye-head exercises, horizontal and vertical | No specific duration | 2 or 3 times daily | Standing* if possible |
| *X*1 viewing paradigm with target held in hand and also with target at distance | 1 minute each exercise | 4 times daily | Standing* |
| Eye-head exercises, horizontal and vertical | No specific duration | 4 times daily | Standing* |
| Add remembered target exercise | No specific duration | 4 times daily | Sitting at first and then standing |
| *X*1 viewing paradigm with target held in hand and also with target at distance | 1 minute each exercise | 4 times daily | Standing* |
| Eye-head exercises, horizontal and vertical | No specific duration | 4 times daily | Standing* |
| Remembered target exercise | No specific duration | 4 times daily | Standing* |
| Some patients may be able to progress to *X*2 viewing paradigm with target held in hand and also with target at distance | 1 minute each exercise | 4 times daily | Standing* |
| Eye-head exercises, horizontal and vertical | No specific duration | 4 times daily | Standing* |
| Remembered target exercise | No specific duration | 4 times daily | Standing* |
| *X*1 with target held in hand and also with target at distance | 1 minute each exercise | 4 times daily | Standing* |
| Eye-head exercises, horizontal and vertical | No specific duration | 4 times daily | Standing* |
| Remembered target exercise | No specific duration | 4 times daily | Standing* |
| Add finding numbers written randomly on large (6 ft × 5 ft) checkerboard pattern placed on wall | No specific duration | 2 times daily | Stand and step to touch number |

*The exercise can be made more difficult by changing the base of support (e.g., from feet apart to feet together).

## Future Directions

Currently research is being conducted to determine whether individuals with BVL may benefit from some form of sensory stimulation to replace the absent vestibular signals. Although this work is in its infancy, the preliminary results are promising. Kentala and colleagues[44] have used a rate gyroscope and linear accelerometer to encode an individual's anterior-posterior sway. This information is then fed back to the individual via a vibrotactile array. The amount of vibration and the location of the vibration is related to the degree and direction of sway. These investigators found that individuals with UVL and BVL demonstrated a marked reduction of anterior-posterior sway on the Sensory Organization Test component of the computerized dynamic posturography test when they were using the vibrotactile array.

Rather than a vibrotactile cue, Hegeman and colleagues[45] used auditory feedback to indicate either the amount of sway or the sway velocity of patients with BVL. Anterior-posterior and side-to-side sway was measured and encoded in four different stance conditions: eyes open–firm surface, eyes closed–firm surface, eyes open–compliant surface, and eyes closed–compliant surface. These researchers reported improvements, to normal levels, in single-limb stance with eyes open on a firm surface when patients used the auditory feedback. They noted no improvements, however, when tests were conducted with eyes closed on a compliant surface. Although these initial results are encouraging, it remains to be seen how useful these devices will be and how the devices will affect the approach to rehabilitation.

## CASE STUDY 1

A 64-year-old woman with a history of acute unsteadiness is referred for treatment. Six weeks prior to this clinic visit, she had a chronic bladder infection that had not responded to other antibiotics and was started on IV gentamicin at home q8hr for 10 days. She had no history of renal failure, nor was she taking other antibiotics or a loop diuretic at that time. She began to complain of imbalance 2 days after the last dose of gentamicin. She had no complaints of hearing loss, tinnitus, or vertigo. Her imbalance was severe, and she was using a walker. She was unable to walk independently. Standing, walking or moving her head exacerbated the balance problem.

### *Comment*

This patient's history suggests an ototoxic reaction to gentamicin. Fewer than 15% of all individuals treated with gentamicin experience BVL, and she had no known risk factors that would increase the likelihood of an ototoxic reaction, such as renal failure and taking a loop diuretic.

### *Pertinent History*

The patient had been treated for depression and anxiety. Current medications were Zoloft, Valium, and Premarin.

### *Neurological Examination*

Neurological findings were normal except for visual acuity of 20/20 – 3 OU with head stationary, which decreased to 20/80 – 4 during 2-Hz head oscillations (clinical DVA test). Patient made corrective saccades with slow and rapid head thrusts, worse to the right. She had a positive Romberg response. Gait was extremely slow and cautious without the walker. She stopped frequently and could not turn around without stopping. Patient also had a significant scoliosis.

**Comment.** The large decrease in visual acuity during head movement (>4 lines) is consistent with a severe vestibular deficit. The presence of corrective saccades with slow head rotations as well as with rapid head thrusts also suggests a profound deficit. Although gentamicin was given systemically, her vestibular loss appears to be asymmetrical. The loss does not apper to have been compensated at this time, on the basis of the positive Romberg result as well as the poor VOR during slow head rotations. Her gait pattern may reflect fear as well as the vestibular deficit. Her history of depression may be a factor in the final level of recovery.

### *Caloric Test*

No response to either cool or warm irrigation of either ear. Iced-water irrigation of both the right and left ears resulted in nystagmus with peak slow-phase eye velocities of 8 deg/sec. The direction of the nystagmus reversed when the patient was moved from supine to prone position.

*(continued on following page)*

## CASE STUDY 1 *(continued)*

**Comment.** Iced water is a stronger stimulus than either cool or warm water. The nystagmus generated by iced water irrigation may represent either a response of the peripheral vestibular system or an alerting response to the extreme cold. If the nystagmus were due to excitation of the hair cells in the inner ear, the direction of the nystagmus would reverse when the patient was moved from supine to prone because the direction of endolymph flow would reverse. If the nystagmus were due to an alerting response, it would not reverse when the patient was moved from supine to prone position. The test results for this patient suggest some residual function in each ear.

### *Vestibular Rehabilitation*

The patient was seen 1 week later to institute a vestibular rehabilitation program. At that time she was still using a walker. She reported that her imbalance was induced by movement, occurred only while walking, and was worse in the dark. She denied having any falls but reported that she did stagger and sidestep while walking, and tended to drift to both the right and the left if she tried to walk without her walker.

### *Social History*

The patient lived with her husband. Her home had no stairs. She did not smoke or use alcohol.

### *Current Functional Level*

She was independent in self-care activities but was no longer driving. She had been inactive since the onset of this problem.

### *Subjective Complaints*

She stated that her symptoms disrupted her performance of both her usual work and outside activities (rated a 3 on the Disability Scale). 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. She rated her disequilibrium while walking as a 9.3 on an analog scale of 0 to 10 (10 worst). She rated her oscillopsia as a 5.7 on a similar scale.

### *Quantitative Dynamic Visual Acuity*

The patient's visual acuity during 120-deg/sec horizontal head movements was LogMAR = .450 (approximately 20/50-3; normal LogMAR = 0.000).

### *Gait*

When asked to walk without the walker, *her cadence was slow and she had a widened base of support*. Her step length was decreased but symmetrical. Arm swing and head and trunk rotation were markedly decreased, especially when she turned. She could not walk a straight path but had no sidestepping or foot crossover. Her gait pattern was affected by her scoliosis (arm swing asymmetry). She was able to walk more rapidly without a significant change in base of support or gait pattern. When asked to try to turn her head horizontally while walking, her cadence slowed significantly and she sidestepped occasionally.

### *Comment*

The patient's Disability Scale and Dizziness Handicap Inventory scores both indicate a moderate perception of disability/handicap. Shepard and colleagues have demonstrated that patients who score a 5 on the Disability Scale are less likely to experience improvement with vestibular rehabilitation than those who score 3 or lower, as this patient did (see Chapter 19). Although she has been using a walker, she was able to ambulate safely without it. Head movements (turning her head or turning herself around) increased her instability. Her dynamic visual acuity was consistent with that of other patients with BVL.

### *Goals*

The short-term goals for this patient were (1) to perform the vestibular adaptation exercises without a significant increase in symptoms, (2) to walk daily, and (3) to no longer use her walker at home. The long-term goals were (1) to return to all normal activities except possibly driving, (2) to improve visual acuity during head movements by two lines on the quantitative DVA test and to 20/50 on the clinical DVA test, and (3) to decrease symptoms by 50% during head movements.

*Plan*

The patient was started on a home exercise program, which included the *X*1 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 twice daily for 5 minutes each time, with rests, 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 ensure compliance. The total daily 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. The exercise program was changed to include the eye-head exercises both vertically and horizontally, and the *X*1 paradigm using a near target held in the hand was added. She was to practice walking and turning her head horizontally for 5 minutes twice daily. 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 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 *X*1 viewing paradigms, although her head movements were slow. The exercise program was modified to include performing the *X*1 paradigm using a target on a checkerboard, and the "imaginary 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 *X*1 viewing paradigm using a target placed against the wall was discontinued. Eventually, the patient was instructed in the *X*2 viewing paradigm.

One month after the initiation of her exercise program, the patient was walking with improved cadence and a narrower 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 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 0.143 (initial DVA was LogMAR 0.450), a three-line improvement. The plan at that time 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.

On her next visit, 1 week later, the patient came in complaining of increased difficulty with her balance and greater dizziness. She reported that the exercises were okay but she was dizzy while walking and even when she rolled over in bed.

*Comment*

The patient was making good progress, and both her balance and dynamic visual acuity had improved considerably when she suddenly had greater difficulty walking and more complaints of dizziness. Her complaint that she has increased dizziness when she rolls over in bed sounds like benign paroxysmal positional vertigo (BPPV).

Re-examination

Eye movements were recorded while the patient was moved into the right and left Dix-Hallpike positions. She developed an upbeating nystagmus, concurrent with complaints of vertigo, when moved into the left Dix-Hallpike position. The latency and duration of the nystagmus and vertigo were consistent with posterior canal canalithiasis. These results indicated that although the patient had a bilateral vestibular deficit, there was remaining function in the left posterior canal. The caloric test had previously shown remaining function in the left horizontal canal. The patient was treated for left posterior canal BPPV using the canalith repositioning maneuver. After successful remission of her positional vertigo, she resumed her exercises for the bilateral vestibular deficit.

## CASE STUDY 2

A 34-year-old woman with a history of diabetes and renal failure had been undergoing peritoneal dialysis for $1^1/_2$ years. She had been treated with gentamicin 9 months and 6 months previously for peritonitis. She had no complaints of disequilibrium after either of those drug courses. Two months after her previous treatment with gentamicin, she again developed peritonitis and again received IV gentamicin. After a few days, she complained of vertigo and tinnitus, experienced 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 work-up of the 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, 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 after even gentle head movements and vomited. She also had a positive Romberg test result. 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. In bilateral dysfunction due to 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 worsen for several weeks. Some patients have a partial reversal of symptoms with time. Often the vestibular symptoms are accompanied by hearing loss. Typically, however, the vestibular loss is symmetrical and patients do not have vertigo or spontaneous nystagmus, both of which are associated with either unilateral vestibular loss or asymmetrical bilateral vestibular deficits. Although gentamicin usually results in BVL, symptoms of asymmetrical effects on the vestibular and auditory systems have been reported.[6]

This patient's 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 the feet, which would contribute significantly to the vestibular problem. This sensory loss was probably due to diabetes. This finding was particularly important in developing her exercise program and in predicting her final level of recovery.

Both computed tomography and magnetic resonance imaging were normal. Audiogram showed an asymmetric sensorineural hearing loss, right worse than left (Fig. 21.4). Caloric tests showed a poor

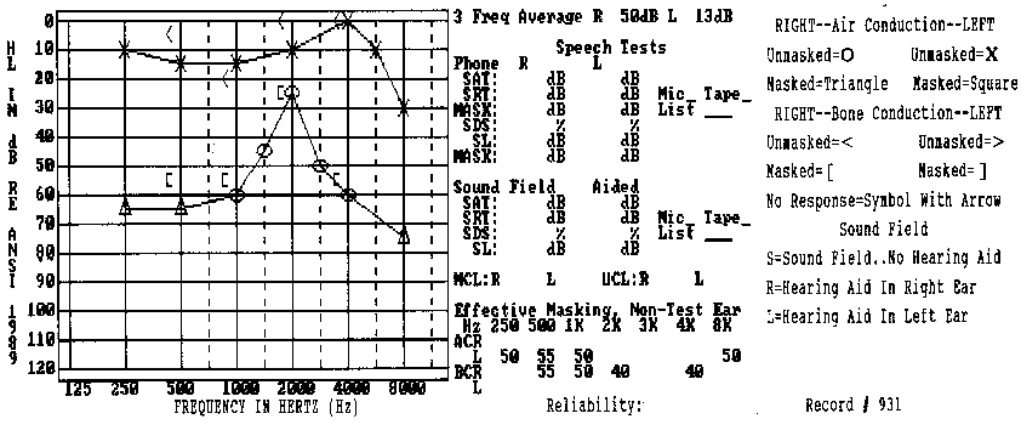


**Figure 21.4** Hearing test results from a patient with gentamicin ototoxicity. *Circles* and *triangles* indicate right ear, and *Xs* and *squares* indicate left ear. Note the asymmetry in hearing loss for this patient.

response bilaterally to warm or cool water, although iced 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 (Fig. 21.5). There was little optokinetic after-nystagmus, and the VOR time constant (Tc) was 2.4 sec to 60-deg/sec step rotations. At 240-deg/sec step rotations, some vestibular response was evident—the gain of the response was 0.15, and the Tc was 2.4 sec.

### *Treatment*

At this point, the patient was started on a vestibular rehabilitation program. She performed the X1 viewing paradigm exercise (see Fig. 21.3) first with horizontal head movements for 1 minute, and then with vertical head movements for 1 minute. Because head movement exacerbated her nausea, she rested for 10 minutes or more after each of these exercises. Initially she performed these exercises while sitting, up to 5 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. She was instructed on how to use a walker, and 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.

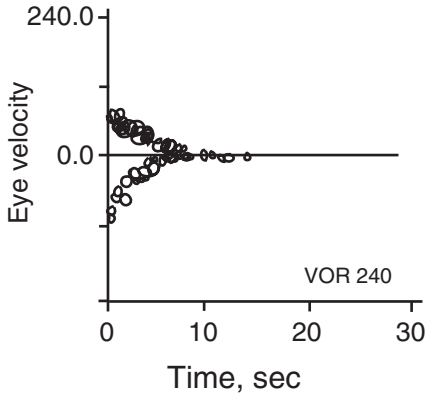


**Figure 21.5** Plot of the decay in slow-phase eye velocity with time during VOR-after nystagmus. Patient is first rotated in a chair in complete darkness for 2 minutes, and then the chair is stopped. The slow-phase eye movements that occur are due to the discharge of the velocity storage system and represent the function of the vestibular system. The results here show the poor peak slow-phase eye velocity (35 deg/sec) and the short time constant (<2 sec) in a patient with bilateral vestibular loss. A step rotation at 240 deg/sec was used. Eye movements were recorded with electro-oculography.

### *Comments*

Although this patient had a bilateral vestibular deficit, the caloric and rotary chair tests showed that she had remaining vestibular function (response to iced water caloric test on the left and the responses to 60- and 240-deg/sec step rotations). Her initial exercise program, therefore, consisted of vestibular adaptation exercises, because she had remaining vestibular function, and ambulation training. Her balance exercises were designed to gradually increase the difficulty of maintaining balance by slowly reducing her base of support, changing her arm positions (arms out, arms at sides, 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 in order to facilitate her ability to use the remaining somatosensory and vestibular cues.

### *Follow-Up*

The patient continued to be followed up as an outpatient. Exercises designed to facilitate the substitution of alterative 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 had to stop walking before turning around. Her Romberg test result was negative but she could not perform the Sharpened Romberg test. 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

*(continued on following page)*

## CASE STUDY 2 *(continued)*

retinal damage in the right eye from her diabetes, which essentially meant that she now had only partial visual, vestibular, and somatosensory cues for balance. As a result, she could no longer keep her balance even in well-lighted conditions. For 1 week she used either 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 test result was clinically normal but she could not perform a Sharpened Romberg test with eyes open. She was seeking part-time employment and was waiting for a kidney transplant.

## CASE STUDY 3

A 61-year-old man was referred by a neurologist for treatment of disequilibrium secondary to BVL. The patient had been hospitalized for a subarachnoid hemorrhage 18 months previously. During his hospitalization, he had several systemic complications, including renal failure, pulmonary infiltrates, and ventriculitis, and was treated with two courses of vancomycin, gentamicin, and ceftazidime.

The neurologist saw him 7 months after this hospitalization because of the patient's persistent disequilibrium. At that time, the patient complained that he stumbled occasionally and that he had increased difficulty walking on uneven surfaces, in the dark, or when he moved his head quickly. He denied having nausea, vertigo, or a rocking sensation, although he did state that he had a feeling of being "tilted" when he walked. He stated that the disequilibrium began after the hospitalization. He also had bilateral hearing loss but had no complaints of tinnitus, pressure, or fullness in the ears. The remainder of his history was noncontributory.

The neurological findings were normal except for the following: (1) visual acuity, as assessed using a wall chart, decreased from 20/20 with the head stationary to 20/100 during 2-Hz oscillations of the head, (2) right Horner's syndrome, (3) staircase saccades downward from the midposition, (4) decreased VOR gain based on visualization of the optic nerve head and on the presence of compensatory saccades during rapid head thrusts, (5) mild decrease in vibration sensation in his feet, right more than left, (6) inability to perform tandem walking, Sharpened Romberg test, or Fukuda's stepping test, and (7) bilateral hearing loss.

The caloric test showed a severe reduction in function bilaterally (Fig. 21.6). Quantitative rotary chair testing of the oculomotor system using step rotations showed low VOR gain (0.2 and 0.13 to the right and left, respectively) and short Tcs (2.2 sec bilaterally) to a 60-deg/sec step rotation and low gain (0.19 and 0.34) and Tcs (1.9 sec and 1.2 sec) to 240-deg/sec rotations. It was concluded that the patient had BVL, probably from the gentamicin, and he was referred for vestibular rehabilitation.

### *Treatment*

Prior to establishment of an exercise program, additional testing was performed. Dynamic posturography showed inability to maintain balance when both visual and somatosensory cues were altered and a decreased ability to maintain balance when visual cues were inappropriate (Fig. 21.7). Quantitative DVA testing showed a change in acuity from 20/20 (static) to 20/40 during head movements. Quantitative vibration threshold confirmed the moderate loss of vibration perception in the patient's feet. His gait was wide-based, and he frequently sidestepped while walking. He appeared to use excessive visual fixation to maintain balance during ambulation.

The patient was started on a program of exercises designed to (1) enhance remaining vestibular function, (2) develop alternative mechanisms to improve gaze stability, (3) improve static balance in the absence of visual cues, and (4) improve balance while ambulating.

### *Follow-Up*

Six weeks later, the patient's Romberg test result was normal but he still could not perform a Sharpened Romberg test with eyes open, nor could he perform Fukuda's stepping test with eyes closed. He continued to walk with a widened base of support. Quantitative testing of the oculomotor system showed no change

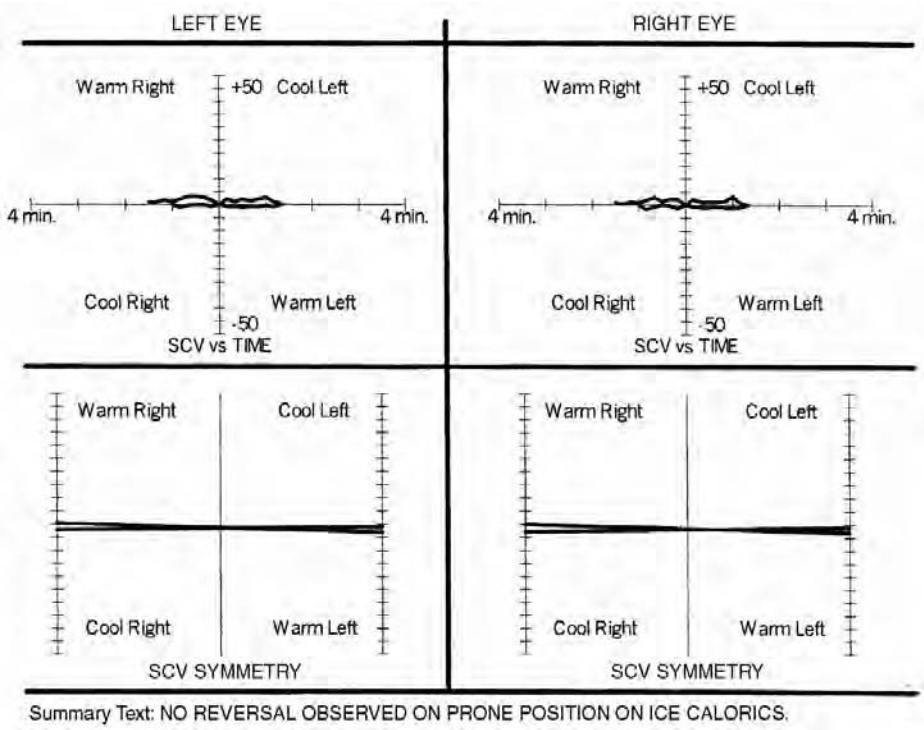


**Figure 21.6** Results from bithermal caloric irrigation of the external auditory canals in a patient with profound bilateral vestibular hypofunction. SCV = Slow component velocity.

from the previous test. Quantitative DVA testing showed an acuity of 20/30, a marked improvement over the previous value. The patient wished to return to driving, and the physical therapist suggested that should he decide to drive, he should start first in local traffic and even in an empty parking lot on a weekend. He was advised that driving at night and high-speed driving would still be hazardous.

One month later the patient reported that he had returned to driving during the day and that he was working part-time. He still could not walk in the dark or with his eyes closed without assistance. 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.

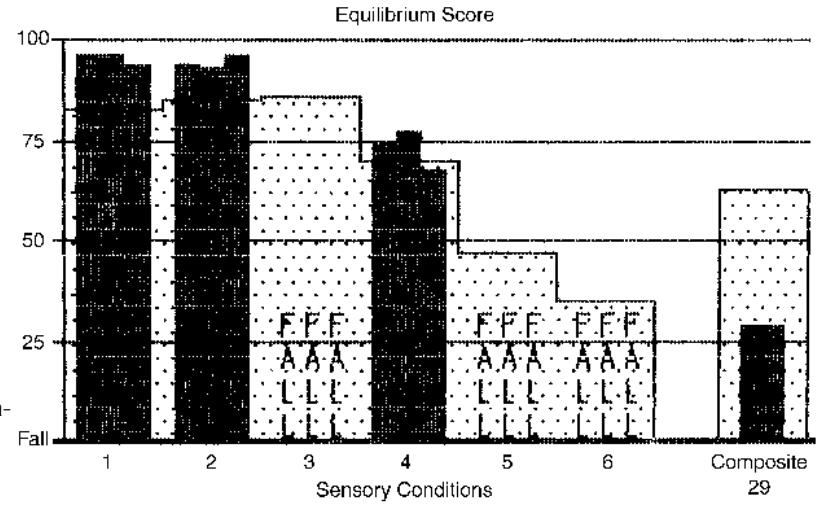


**Figure 21.7** Results from posturography test in a patient with bilateral vestibular loss. Test conditions were those shown in Figure 21.2.

## Summary

Patients with bilateral vestibular problems can be expected to return to many activities but will continue to have difficulties with balance in situations in which visual cues are absent or diminished. The level of disability partly depends on the degree of the vestibular loss but also reflects involvement of the visual and somatosensory systems. 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. Patients should be able to return to work, and most of them will be able to ambulate without the use of a cane or walker, at least when they are in well-lighted environments.

## Acknowledgment

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CHAPTER 22

# Management of the Pediatric Patient with Vestibular Hypofunction

*Rose Marie Rine, PT, PhD*

The vestibular system develops relatively early in gestation.[1–5] Investigations of the development and appropriate testing of this system have demonstrated that it is a dual-functioning system. The vestibular system is composed of vestibulo-ocular and vestibulo-spinal subsystems, each represented by distinct tasks and testing methods. Labyrinthine reflexes, which are mediated by the vestibulo-spinal system, provide postural tone necessary for the emergence of early motor milestones (e.g., rolling, sitting, standing).[6–8] The role of the vestibulo-ocular system in visual stabilization, acuity, and the development of visual spatial and perception abilities has been well documented.[9–12] 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.

The incidence of peripheral vestibular disorders in children (e.g., Ménière's disease, perilymphatic fistula, benign paroxysmal positional vertigo [BPPV]) similar to that in adults has been reported.[13–24] Furthermore, like adults with central nervous system impairment, children with traumatic brain injury or other central nervous system insult may be expected to have central vestibular deficits. However, investigations of vestibular dysfunction in children with these diagnoses are scarce.[20, 25–29] The functional integrity of the vestibular system is rarely tested in young children, and thus, any impairment is undetected and untreated. This situation may be in part due to children's inability to describe symptoms or even know that what they are experiencing is not "normal."[6, 30, 31] 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 in this population. This chapter reviews the incidence of vestibular deficits in young children, the development of postural control abilities, and appropriate testing and intervention for this population.

## Incidence of Vestibular Deficits in Children

There are numerous reports in the literature regarding vestibular dysfunction of various etiologies in young children and the consequent functional impairments.[13, 15, 17, 19, 21, 32, 33] Tsuzuku and Kaga[33] and others[17, 26, 27, 31, 34] have reported learning disabilities and delayed development of walking and balance abilities in children with vestibular system anomalies. Others have reported motor and balance deficits in young children with sensorineural hearing impairment (SNHI) and evidence of vestibular hypofunction (Figs. 22.1 and 22.2).[18, 35–38] In spite of these reports, children with SNHI or complaints of dizziness are rarely tested for vestibular function. Casselbrant[21] reported anomalies of postural control in children with otitis media, which was reversed after insertion of tympanostomy tubes. However, vestibular function tests were not

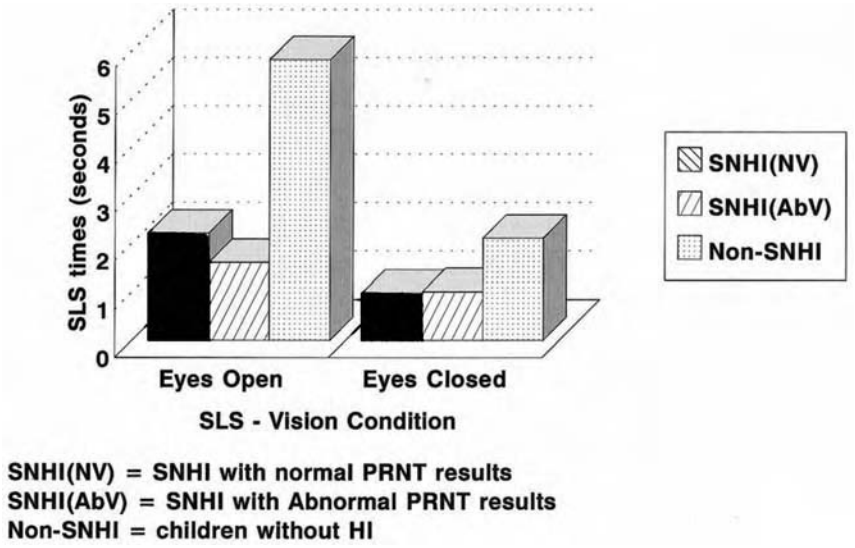


**Figure 22.1** Plot of mean single-leg stance times of children with and without sensorineural hearing impairment (SNHI). Children with SNHI and abnormal post-rotary nystagmus test (PRNT) results score lower than those with SNHI and normal PRNT results. (From Rine et al, 1996.[18])

reported. Schaaf[39] reported that the incidence of vestibular disorders was high in developmentally delayed preschoolers with a history of otitis media.

Furthermore, Eviatar and Eviatar[35] 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 is a maturational delay in vestibular system function, as evidenced by a reduced percentage of positive nystagmus response until 12 months of age. These investigators concluded that deviation of responses from the mean and standard deviation of the normative sample can be considered abnormal after the age of 12 months. In addition, although the incidence is lower than adults, there are recent reports of BPPV[13, 15, 40] in young children, as well as Ménière's disease[19] and vestibular neuritis[16] with consequent vertigo or impairments of balance abilities. Also, in spite of the documented functional relationship of the visual and vestibular systems and the effect that vestibular deficiency may have on gaze stability and oculomotor control,[12] children found to have 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 due to adaptation or plasticity.

Investigations of plasticity and recovery of function support the idea that age at the time of injury as well as age at the time of testing does affect results obtained on

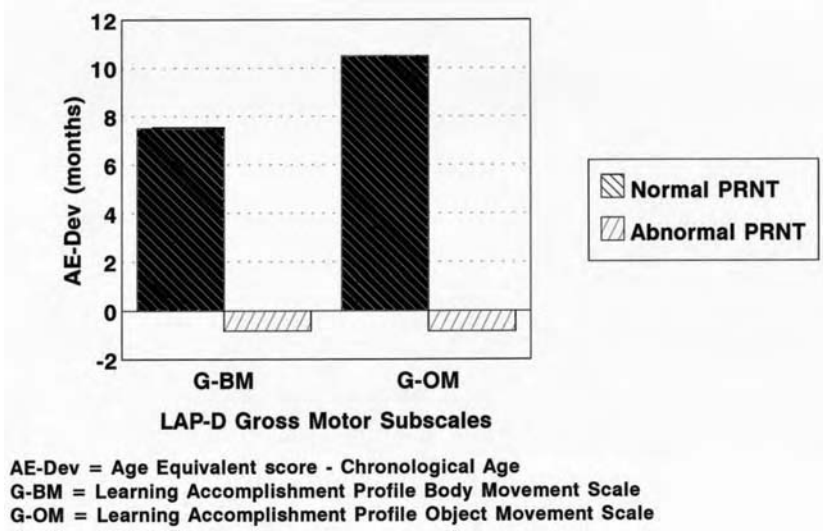


**Figure 22.2** Plot demonstrating the difference of the mean deviation of age-equivalent scores achieved on the Learning Accomplishment Profile (LAP) from chronological age between children with sensorineural hearing impairment (SNHI) and normal post-rotary nystagmus test (PRNT) results and those with SNHI and abnormal PRNT results. Those with abnormal PRNT results achieved scores below age level. (From Rine et al, 1996.[18])

tests of function.[41, 41] 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. On the basis of evidence that similar as well as different neural changes occur in individuals with nervous system damage at birth or during childhood in comparison with those who sustain damage later in life, it is logical to assume that different neural substrate changes occur in young children with vestibular deficits. Furthermore, these changes may mask the deficits typically noted with traditional testing methods. Rine et al[42] reported that the vestibular-related impairments evident on functional and post-rotary nystagmus test results were not identified by rotary chair testing in children diagnosed with SNHI since birth. The children with SNHI presented with delayed maturation of vestibular and vision ratios on posturography testing, hypoactive responses on post-rotary nystagmus testing, and balance abilities below age level (see Fig. 22.2). However, results of per-rotary nystagmus (rotary chair testing) were within normal limits for the majority of the children tested. In a separate study, Rine et al [17] reported evidence of a progressive delay in gross motor development in children with SNHI and concurrent vestibular hypofunction.

Horak et al[43] found that adults with bilateral vestibular deficits 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 than in either subjects without a deficit or those with 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 group but not the adult-onset group. In contrast, Rine and colleagues[38] noted delayed recovery responses on dynamic balance testing as well as deficient reweighting of sensory cues during posturography testing in young children with bilateral vestibular hypofunction since birth (Fig. 22.3). Mira and coworkers,[44] who examined children with benign paroxysmal vertigo, reported a high incidence of autonomic and neurologic signs (e.g., pallor, nausea, hyperhidrosis, phonophotophobia, blurred vision, migraine) accompanying the typical vertiginous attacks. Computed tomography (CT), electroencephalography (EEG), and audiometric test results were normal, as were nystagmus responses to rotational and caloric testing. 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.

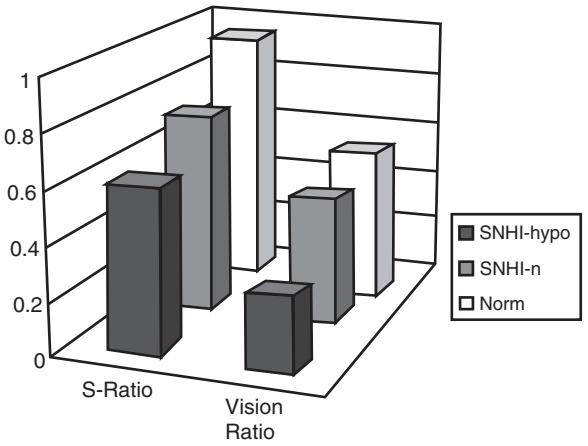


**Figure 22.3** Children with sensorineural hearing impairment (SNHI) and concurrent vestibular hypofunction (SNHI-hypo) achieved significantly lower somatosensory (S) and vision effectiveness ratios than those with SNHI without vestibular deficit (SNHI-n) and as well as those without deficit (Norm). This finding indicates deficient sensory organization for postural control.

Although vestibular and postural control deficits have been documented in adults with central nervous system involvement, similar investigations of children are scarce.[20, 25, 26, 28] It is logical to assume that like adults, children with traumatic brain injury or other central nervous system conditions present with similar vestibular deficits. In addition, owing to 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 [28] reported poor static postural control under various sensory environments in children with spastic cerebral palsy. Although it is expected that the muscle tone impairment involved is to a great deal responsible, postural sway results were significantly different between children with and children without impairment only on the swayed surface (SS), eyes closed–swayed surface (ECSS), and swayed vision–swayed surface conditions (SVSS), and thus were similar in all other conditions. This finding suggests central vestibular system and sensory organization impairment. Nashner et al[45] and others[25, 29] reported similar results, as well as a reversal of muscle activation patterns. However, in none of these studies was vestibular function formally tested. The identification of a vestibular deficit is important to the development of appropriate intervention as well as for prognostic purposes. Weiner-Vacher and colleagues[5] have reported that the acquisition of walking ability is related to measures of otolith, not

canal, vestibular function. Until recently, with the emergence of the use of vestibular-evoked myogenic potential tests, clinical testing of otolith function has not been possible. 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 vestibular dysfunction. For an understanding the implication of vestibular deficits early in life, as well as to enable the selection and interpretation of testing and the development of appropriate interventions for children, a review of the development of postural control and oculomotor control as they relate to vestibular function is warranted.

## Development of Postural and Oculomotor Control as Related to Vestibular System Function

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.[3, 46–50] Forssberg and Nashner[51] and others[3, 46, 47] 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) were significantly longer and matured at different rates. Haas and colleagues[46] claimed that the change in short-latency responses to adult-like levels by 5 years of age reflected improved nerve conduction velocity. The long-latency response, which is reportedly an indicator of vestibulospinal function,[52, 53] was not reduced to adult-like levels until 15 years of age. These investigators attributed this finding to acceleration of central polysynaptic transmission and myelinization. In addition, Shumway-Cook and Woollacott[54] reported that the attenuation of response seen in 7-year-old children and adults was not observed in younger children, with greater 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.[50, 55]

Woollacott and coworkers[55] reported that children without experience in sitting or standing did not demonstrate postural responses. Although muscle responses occurred in reaction to perturbation in all experienced subjects (either sitting or standing), the temporal organization of postural responses of the youngest experienced sitters (8 to 10 months old) and standers (14 months old) differed from those of adults and older children. Instead of the typical distal-to-proximal sequence of muscle activation, proximal muscles (neck) initiated the response.

Investigations by Forssberg and Nashner[51] showed that although children 3 through 10 years of age could maintain balance on all posturography test conditions, children younger than 7.5 years performed like vestibular-deficient adults in the posturography test conditions that required dependence on vestibular function (eyes closed and sway surface, and sway-referencing of both the visual surround and support surface). Furthermore, Foudriat and colleagues[50] reported that children 3 through 6 years of age had better stability in the swayed vision (SV) posturography condition than in the swayed surface (SS) test condition. Rine et al[49] reported increased variability of responses on posturography testing in children 4 to 6 years of age. These investigators[49] and others[48] also noted that somatosensory effectiveness was adult-like by 4 years of age (Fig. 22.4).

The following conclusions may be gleaned from these reports:

- Experience within a posture is critical for the development of postural control abilities;
- By 3 years of age, somatosensory effectiveness in postural control emerges, but in children younger than 7.5 years, visual and vestibular system effectiveness in postural control is immature;
- Maturation of postural responses occurs in a stagelike pattern. The period between ages 4 years and 6 years is a transition period in which mature patterns are emerging.

Investigators have suggested that these changes are due to maturation of the effectiveness of individual sensory systems and the sensory integrative mechanisms involved in postural control.[48, 49, 56, 57]

Reportedly, although vision is dominant in the early stages of learning to balance in a posture, shorter-latency proprioceptive responses are intact and more effectively used if visual information is removed.[55–57] Riach and Hayes[56] found that Romberg Quotients (RQs), the amount of sway with eyes closed expressed as a percentage of sway with eyes open, were less than 100 in young children because sway was greater with eyes open than with eyes closed. In adults, RQ is greater than 100. These results suggest that, although visual information domi-

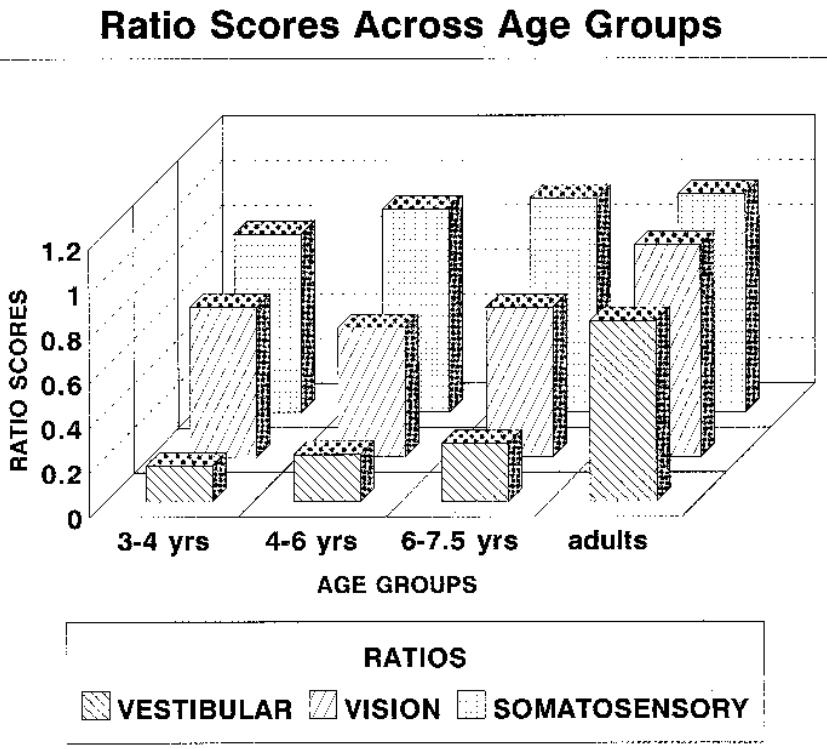


**Figure 22.4** Median of ratio scores attained by individuals of varying age without deficits. (From Rine et al, 1998.[49])

nates postural responses in young children, it also destabilizes.

Riach and Starkes[57] reported that like adults, children are able to reduce sway with visual fixation, but younger children (3 to 5 years old) had an increased number of saccades during attempted visual fixation than older children and adults. These investigators proposed that the increased saccades may be due either to an inability to visually fixate as well as adults or to an attempt to improve stability by increasing saccades. However, this conclusion is refuted by reports that the basic ability to stabilize visual flow with head and eye movements is acquired by 3 months of age.[12] Von Hofsten and Rosander[12] noted that although a substantial lag was evident in 1-month-old infants, the lag was diminished by age 3 months. This ability was attributed to maturity of both the visual and vestibular systems. It is possible that although infants can stabilize the visual environment, they are not able to coordinate and integrate this information with other sensory system information about vertical orientation.

Hirabayashi and Iwasaki[48] and Rine and colleagues[49] found immaturity of visual and vestibular system effectiveness in postural control through the age of 15 years. Because children younger than 7 years attain significantly different stability scores from those of adults in the eyes open and eyes closed conditions of posturography testing, scores in the other conditions cannot be used to depict sensory system function in postural control. To eliminate this problem, ratio scores were calculated to provide a measure of sensory system effectiveness in postural control.[48, 49] The investigators reported that somatosensory function was similar to that in adults by 3 to 4 years of age.[48, 49] Although adult values in visual function ratios were achieved by 15 years of age, this was not true of vestibular function ratios.[48] However, functional effectiveness of the vestibular system for gaze stabilization is reportedly adult-like by 3 years of age. Rine and Braswell[31] developed a clinical test of dynamic visual acuity (DVA) for use with young children and reported normative data. Test score is the difference between acuity with the head stable and acuity with the head moving at 2 Hz. Children as young as 3 years achieved scores similar to those in adults. Furthermore, it has been demonstrated that deficits in peripheral dysfunction not only affect DVA in young children, but results in reading acuity deficits [58, 59]

In summary, functional maturation of the vestibular and visual systems for postural control continues through 15 years of age, with the following observations:

- Although visual system effectiveness in postural control is less mature than that of the somatosensory system prior to age 7.5 years, it is the dominant source of information for postural control in standing.
- Vestibulo-ocular mechanisms are intact and mature by 1 year of age.
- Vestibular system effectiveness for gaze stabilization is mature by 3 years of age.
- The vestibulospinal mechanism, or the effectiveness of the vestibular system in postural control, continues to develop after 15 years of age.
- The sensory integrative capacity required for postural control evolves between 7 and 15 years of age.

The finding that development of postural control is affected by the integrity of all systems involved as well as by practice and experience in a posture suggests that deficits in any one system, in central processing capabilities, or in the ability to experience various upright postures will impede development. Furthermore, the finding that vestibular system effectiveness in oculomotor is adult-like on tests of DVA by 3 years of age, but that its effectiveness in postural control is not adult-like until adolescence, 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, which have been normed for this age group. This assessment is important for both (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.

## Evaluation of Children with Vestibular System Dysfunction

A comprehensive examination of vestibular function includes functional and diagnostic tests of VO and Vsp function.[7] Because diagnostic procedures are costly and may be uncomfortable, particularly for young children, screening for appropriate referrals for in-depth testing is warranted. On the basis of the literature, the following children should be tested with regard to motor and balance development and oculomotor control: those with SNHI, those with identified learning disability with evidence of Vsp or VO dysfunction, those with recurrent and chronic inner ear infections who also present with difficulties of coordination, reading, or developmental delay, and those with complaints of dizziness or visual stabilization difficulties (Table 22-1). Therapists can complete developmental functional testing of motor and postural control abilities, perceptual and oculomotor abilities, and screening of VO and Vsp function.[7, 31, 60] The purpose of these tests is threefold: (1) to establish that a functional balance deficit exists, (2) to isolate the contributions of the various components of the postural control system, and thus determine which component(s) is (are) problematic, and (3) to 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 Vsp or VO dysfunction should be referred for more comprehensive testing.

Functionally, balance represents postural control or vestibulospinal abilities, as does the aberrant persistence of labyrinthine reflexes. Clinical functional measures of balance that have been normed and standardized for use with children include the Functional Reach Test,[61] the balance subtests of the Peabody Developmental Motor Scales (PDMS),[62] the locomotor subtest of the second edition PDMS (PDMS-II),[63] and the Bruininks-Oseretsky Test of Motor Proficiency (BOTMP).[64] Care should be taken in the selection of tests, because not all are appropriate for all ages (Table 22-2). In addition to testing balance, the PDMS and BOTMP include visuomotor, perceptual-motor and eye-hand coordination subtests, which enable age-appropriate functional testing of the VO system. Tasks include target skills, tracing, bilateral coordination, and pencil-and-paper tasks. The BOTMP, PDMS, and the PDMS-II are valuable in documenting general gross and fine motor developmental status as well as providing more specific measures of balance and visuomotor development. The prone extension test component of the Sensory Integration and Praxis Tests (SIPT)[65] also provides a test of Vsp function. Other test components of the SIPT are used primarily to assess children with learning disabilities, focusing on form and space perception, praxis, and tactile discrimination.

For younger children, the Test of Sensory Functions in Infants[66] and the DeGangi-Berk Test of Sensory Integration[67] can be used to test ocular motor control and developmental reflex integration (e.g., symmetrical and asymmetrical tonic neck reflexes; Figs. 22.5 and 22.6). These tests of sensory integration dysfunction, which is reportedly correlated to vestibular dysfunction,[6] are appropriate for children with only mild motor delays. The use of these tests and interventions for motor planning, dyspraxia, and sensory integrative dysfunction are more appropriately and completely reviewed elsewhere[6] and are not further addressed here. Children who present with deficits on any of the complete tests noted previously or,

## ■ Table 22-1 SYMPTOMS OF VESTIBULAR DYSFUNCTION IN CHILDREN

| | |
|---|---|
| Peripheral disorders | 1. Nystagmus on head movement.<br>2. Visual instability on head movement; complaints of blurring or double vision; positive dynamic visual acuity test result.<br>3. Below–age level balance abilities (e.g., tandem, single-leg stance).<br>4. Complaints of spinning sensation or dizziness.<br>5. Below–age level vestibular ratios (also vision and somatosensory ratios, if since birth) on SOT; step or loss of balance on test conditions 5 & 6.<br>6. May or may not show increased latency on DPT.<br>7. Hypoactive or hyperactive responses on PRNT.<br>8. May or may not have hearing loss or tinnitus.<br>9. May be fearful of movement activities, or crave them; asymmetrical posturing in sitting or standing.<br>10. Complaint of incoordination. |
| Central disorders | 1. Delay or below–age level performance on gross motor tasks.<br>2. Delay or below–age level performance on visuomotor, visual perception tests; positive or negative dynamic visual acuity test result.<br>3. Persistence of tonic reflexes.<br>4. May manifest increased latency and amplitude of responses on posturography DPT.<br>5. Visual instability, particularly with head movement; failure on tests of dynamic visual acuity.<br>6. Normal, hypoactive, or hyperactive responses on PRNT, although majority either hypoactive or hyperactive.<br>7. Below–age level performance on dynamic posturography SOT with stepping or loss of balance on test conditions 4–6.<br>8. Below–age level vision and vestibular ratios on posturography SOT.<br>9. Possible sensory integrative dysfunction. |

DPT = dynamic perturbation test component of posturography test; PRNT = post-rotary nystagmus test; SOT = sensory organization test.

## ■ Table 22-2 ASSESSMENT OF VESTIBULAR DYSFUNCTION IN CHILDREN

| Assessment Tool | Test Type | Vestibular System Tested | Age Group | Items/Behaviors Tested |
|---|---|---|---|---|
| Peabody Developmental Motor Scales[62, 63] | Motor development<br>Balance ability/development<br>Visual perception<br>Balance, locomotor, nonlocomotor, visuomotor, and reflex subtests | Vsp<br><br><br>VO | Birth–80 mo | Developmental reflexes<br>Balance beam, EO & EC<br>Single-leg stance EO & EC<br>Hopping<br>Tandem stand and walk<br>Visual track, perception, tracing<br>Target activities |
| Bruininks-Oseretsky Test of Motor Proficiency[64] | Balance and gross motor ability<br>Balance, visuomotor, eye-hand, and bilateral coordination subtests | Vsp<br><br>Vsp, VO | 4–14 yr | Balance on beam in tandem and single-leg stance<br>Single-leg EO & EC<br>Target and tracing tasks<br>Romberg test, finger to nose, bilateral coordination tasks |

*(continued on following page)*

## ■ Table 22-2 ASSESSMENT OF VESTIBULAR DYSFUNCTION IN CHILDREN *(continued)*

| Assessment Tool | Test Type | Vestibular System Tested | Age Group | Items/Behaviors Tested |
|---|---|---|---|---|
| DeGangi-Berk Test of Sensory Integration[67] | Sensory integration and motor planning<br>Postural control, bilateral motor, and reflex integration subtests | Vsp, VO | 3–5 yr | Pivot prone and supine flexion<br>Postural tone<br>Dysdiadochokinesis<br>Tapping, jumping, and bilateral tasks<br>ATNR and STNR testing |
| Test of Sensory Function in Infants[66] | Sensory processing and reactivity in infants | VO, Vsp | 4–18 mo | Responses to tactile, vestibular, and visual ocular motor control |
| The Sensory Integration and Praxis Tests[65] | Sensory integration and motor planning abilities<br>Visual perception | Vsp, VO | 4–8 yr | Form and space perception<br>Praxis<br>Tactile discrimination<br>Vestibular integration |
| Southern California Post Rotary Nystagmus Test[70] | Vestibular ocular system test | VO | 5–11 yr | Nystagmus duration after rotation |
| Functional Reach[61] | Balance ability in standing | Vsp | 5–15 yr | Forward reaching, standing |
| Pediatric Clinical Test of Sensory Interaction for Balance[76] | Balance under varying sensory conditions | Vsp | 4–9 yr | Measures of sway in double-leg stance with EO & EC, while standing on foam or floor, or with conflict dome |
| Dynamic Posturography Testing [49, 73] | Functional test of balance<br>Effectiveness of vestibular, visual, and somatosensory systems | Vsp | Normative data 3 yr–adult on SOT | SOT: computerized measures of sway<br>DPT: measures EMG responses to perturbation, standing |
| Timed "Up and Go" test | Functional stand-and-walk test | Vsp | DPT component: 1.5 yr–adult | Time to rise from chair, walk 3 m, and return to sitting |
| Vestibular Auto-Rotation Test[69] | Visual stability (visuo-vestibular function) | VO | 3 yr–adult | Maintain eyes stable with head movement at various frequencies |
| Dynamic Visual Acuity Test[31] | Gaze stabilization | VO | 3 yr–adult | Visual acuity with head stable and moving at 2 Hz. |

ATNR = asymmetrical tonic neck reflexes; DPT = dynamic perturbation test; EC = eyes closed; EMG = electromyography; EO = eyes open; SOT = sensory organization test; STNR = symmetrical tonic neck reflexes; VO = vestibulo-ocular system; Vsp = vestibulo-spinal system.

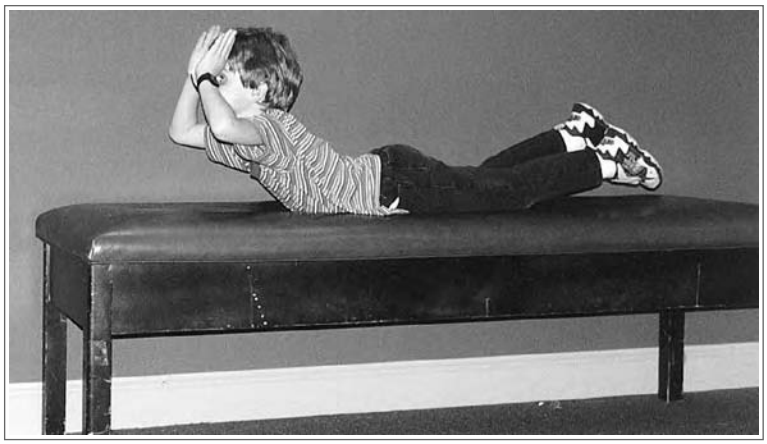

**Figure 22.5** The prone extension test examines the child's ability to assume and maintain a pivot prone position. Inability to do so is indicative of persistence of the tonic labyrinthine reflex.

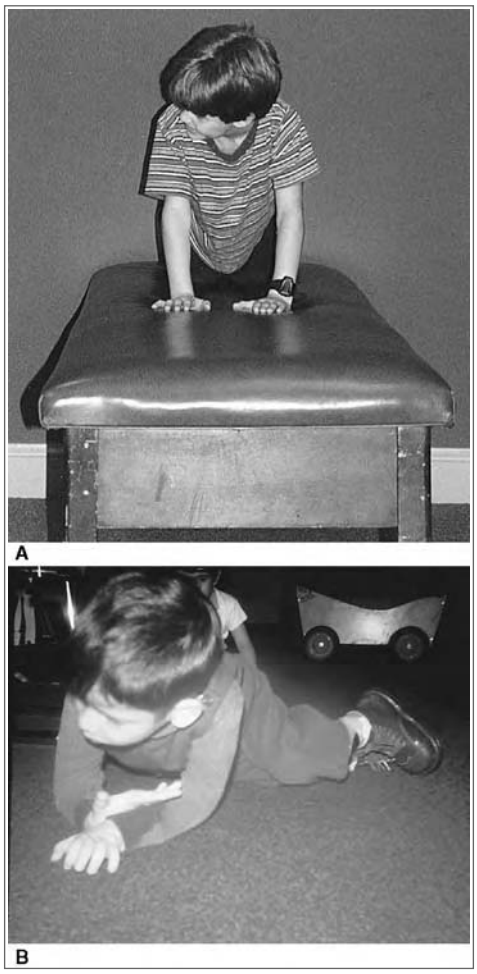


**Figure 22.6** One method of testing whether the asymmetrical tonic neck reflex is interfering with function is to request that the child maintain a quadruped position with elbows extended as he/she looks to the left or right. (*A*) The child without deficit is able to do so without difficulty. (*B*) In the child with a deficit, the arms collapse as the child attempts to look to his right.

more specifically, on the balance, reflexive, or visuomotor subtests, should be further examined for sensory (vision, somatosensory, and vestibular) and postural control system effectiveness, to include screening of the neurological and musculoskeletal systems.

The musculoskeletal system must be examined to determine whether restrictions in range of motion, pain, reduced strength, or limited endurance is present, as 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, BOTMP, and the sensory integration tests can provide screening of neurological status (e.g., Romberg testing with eyes open and closed, finger-to-nose test with eyes open and closed, test of developmental tonic reflex integration). Further neurological screening should include testing of deep tendon responses, cranial nerves and status of equilibrium reactions as well as screening of the sensory systems involved in postural control.[60]

Sensory screening should include an examination of the visual, somatosensory, and vestibular systems. Somatosensory screening involves testing the integrity of the tactile and proprioceptive senses, to include testing of touch sensitivity, localization and discrimination (graphesthesia), and proprioception or position sense in the lower extremities. Visual screening should include tests of visual tracking and visual field capabilities as well as an examination of the integrity of VO responses (see Table 22-2). VO mechanisms can be examined by noting the presence of abnormal responses such as gaze-evoked or positional nystagmus, corrective saccades on the head-thrust test,[68] and visual stabilization during head

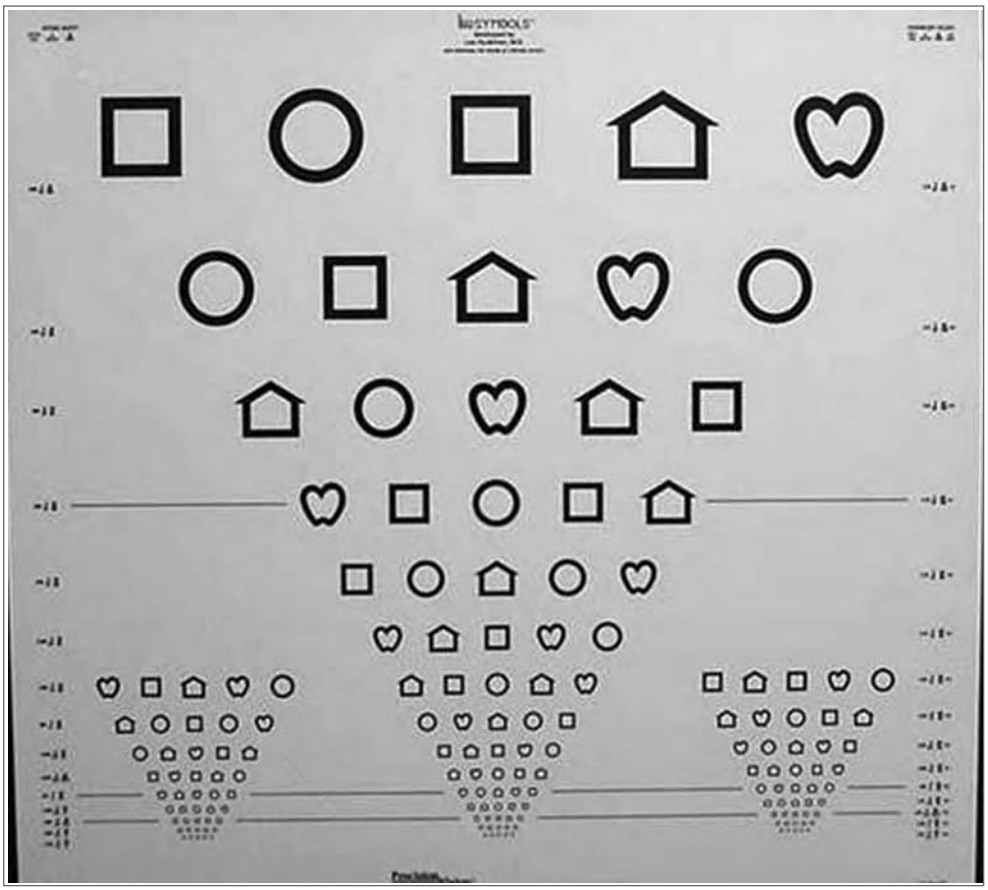

**Figure 22.7** For children who do not know letters, vision charts with symbols can be used. (Obtained from Lighthouse Low Vision Products, 36-02 Northern Blvd., Long Island City, NY 11101.)

rotation (DVA testing)[31] or vestibular autorotation testing (VAT).[69] For children, visual acuity charts are available with symbols (Fig. 22.7) to complete DVA testing, which has excellent sensitivity and specificity for the identification of vestibular hypofunction in children.[31] Additional screening tests of the VO system include the head-shake or post-rotary nystagmus tests.[69] The Southern California Post-Rotary Nystagmus Test (PRNT)[69, 70] has been normed for children through 9 years of age and is reportedly a reliable measure, having good correlation with rotary chair testing for the identification of hypofunction. If a balance or a visuomotor deficit is found concomitant with evidence of VO or Vsp deficit on tests previously described, further diagnostic testing of vestibular function is warranted.

To isolate and test canal vestibular function, caloric and rotary chair testing are used. Normative values are available for children as young as 1 year.[10, 35, 71] Otolith vestibular testing for children, to include vestibular-evoked myogenic potentials, off-vertical axis rotation, and subjective visual vertical, has also been reported.[5, 16, 32, 40] To test Vsp function, dynamic posturography testing is used.[38, 52, 72] Dynamic posturography testing involves both a sensory organization test (SOT) and dynamic perturbation test (DPT), which complement and expand upon the information provided by traditional clinical testing, and yield an objective, functional measure of postural control.[49, 73, 74] Although the sensory and motor tests noted previously 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. 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. Recently, normative values of SOT measures, to include ratio scores, have been reported for children 3 through 15 years of age.[48, 49] A clinical, less sophisticated form of the sensory organization test, which requires

inexpensive materials and is portable, has also been developed.[34, 75, 76] Westcott and colleagues[76] developed a pediatric version of this test, the Pediatric Clinical Test of Sensory Interaction for Balance (P-CTSIB), and reported fair to good reliability when combined sensory conditions scores were used. With 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.

The DPT component of posturography may be used to determine whether neurological dysfunction is evident, and at what level, and provides an indirect measure of Vsp function.[52, 77] Furthermore, maturation changes in the neuromotor component of postural control can be measured and monitored. Although Muller and colleagues [3] and Dichigans and associates[52] have reported that responses are evident with minimal maturation changes in the very early stages of learning to stand and walk, normative data for children have not yet been published. Harcourt[78] reported that although posturography testing may not be as 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.

## Treatment of Vestibular Dysfunction

### *Peripheral Disorders*

As in adults with peripheral dysfunction, once a diagnosis is complete, medical management and rehabilitation of the child with peripheral dysfunction should begin. This may include medication, surgery, or visual habituation and balance training activities (see previous chapters in this volume). Rine and colleagues[79] completed a wait-listed, 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 exercise intervention focused on substitution and habituation, three times weekly, under the direction of a physical therapist. 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[59] reported improvements of scores on DVA tests and subjective reports of gaze stability after exercise intervention in children with vestibular hypofunction. D'Agostino and coworkers[15] described a case of horizontal canal BPPV in a 10-year-old child, with "spontaneous" recovery after short-term hospitalization and repeated mobilization (e.g., roll side-to-side) in the supine position.

The balance and habituation training activities must be modified to the child's level of cognitive maturation and interest level, with particular consideration of the caregiver. Unlike the adult, who will be responsible for his or her own exercise regime, the child depends on parents and therapists to carry out the program and ensure compliance. To maximize the child's participation and cooperation, the use of toys, games, and other items to facilitate visual tracking, or the use of swings to provide the movement during visual stabilization activities, is important. Instead of letters, which are not motivating or fun, line pictures, moving ball or animal with symbols or letters, or even simple computer or video games may be used. If the child can interact with, attempt to grasp, or point to specific letters or symbols while sitting, swinging, and so on, the exercise becomes a game, is fun, and may minimize resistance and maximize effort and cooperation. This cooperation and effort are critical to the effectiveness of the exercise regimen.

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. It is critical to monitor progress and provide short-term training to be carried out at home with caregivers (e.g., visual tracking exercises, visual stabilization regimen, balance and movement exercises to enable resumption of age-appropriate levels of activity).

### *Central Vestibular and Postural Control Deficits*

As noted previously, 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 learning disabilities present with sensory organization and balance deficits,[28, 29] 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 or autism as well as in low-birth-weight, premature infants.[11, 27, 80–82] 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 visuomotor function should be evaluated for VO and Vsp function as well as differential diagnosis of the multiple factors involved. Treatment can then be developed to facilitate either the use and integration of systems intact but not used or 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 in varying sensory environments to encourage the use of intact systems and facilitate integration of information.

## CASE STUDY 1

A 7.5-year-old girl was admitted to the hospital with complaints of dizziness, nystagmus, spinning sensation, and inability to sit or stand. After being diagnosed with vestibular neuritis, she was 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; unable to walk on 3.5-inch balance beam in forward or sideways directions; able to stand in tandem 3 seconds, but not to walk without sidestepping.
Neurological screening: Deep tendon reflexes intact; finger-to-nose test result negative; rapid alternating upper and lower extremity movements intact.
Vision and VO testing: Upon testing, optokinetic and gaze-evoked nystagmus noted; tendency to keep head tilted to the right; favors left eye by semiclosure of the right during DVA test, which had normal result 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 (> 2 standard deviations below mean).

Parent was trained in visual stabilization training using flash cards the child enjoyed and was familiar with for single words:

1. Child to read card as it is moved to right and left at 1 Hz; with card stabilized at midline, child to read card as head is turned to the left and right at 1 Hz. Once child is able to do this without difficulties, speed to be increased to 2 Hz. Practice 2 minutes each, daily.
2. With 4 "word" cards taped on wall centered in front of child, child sitting 6 feet away, she practices reading cards as head is rotated right and left (binocularly and monocularly).
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.
4. Hopping games: hopscotch; jump rope swung slowly back and forth by Mom, and child to jump over it.

Due to difficulties in transportation, weekly monitoring of progress and adjustment of exercises were done via phone contact. In 2 months, all symptoms were relieved. The child was able to tandem stand and walk 5 feet. No difficulties were noted with reading or visual fixation with either card or head movement.

## CASE STUDY 2

Seven-year-old boy 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 lb). Sensorineural hearing impairment was diagnosed at 18 months. However, with use of hearing aids, audiological results were within normal limits, and the child did comprehend spoken language and spoke clearly. He also had a history of frequent ear infections, for which drainage tubes were inserted twice (bilaterally). Owing to vision and reading difficulties, this child had been recently put in a special education program for learning disability. He was referred for testing to establish vestibular functional status and to determine whether further testing or treatment was warranted. Parent reported the boy having typical activity levels until recently, including playing T-ball and basketball with peers (intramural sports). Review of records indicated treatment with gentamicin and amoxicillin during the past few years.

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 his "eyes do funny things"; when asked to demonstrate this, child replicated nystagmus. Difficulties with smooth pursuit evident, using corrective saccades and attempted head turns to maintain fixation on moving target. DVA score is normal. Although nystagmus was not exaggerated on head-shake test, child reported dizziness. Hyperactive response to PRNT with 35 seconds' duration of nystagmus and severe vertigo after rotation in either direction.
Motor and balance ability: Able to maintain single-leg stance for 6 seconds EO, 3 seconds EC. Able to walk 3.5-inch balance beam but not to balance standing across 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.
Neurological screening: Deep tendon reflexes within normal limits, as were rapid alternating movements, integration of labyrinthine reflexes, and finger-to nose test result.
Posturography (SOT) results: Patient relied on stepping strategy to maintain upright on conditions 5 and 6; posturing noted on condition 4; vision ratio of .29 (> 2 standard deviations below norm for age) and vestibular ratio of .45 (2 standard deviations below norm for age).

Patient was referred for further medical diagnostic testing for possible fistula, which was substantiated. Surgical correction was performed. Physical therapy after surgery focused on visual stabilization and balance retraining. Within 2 months, all vertigo and dizziness were 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, including playing basketball with peers.

## CASE STUDY 3

Nine-year-old girl with a diagnosis of spastic hemiplegia since birth presented with gait and balance deviations and delayed gross motor functioning. IQ score was within normal limits, so 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 extremities. Gait deviations included toe-heel contact on the right, knee flexion during midstance on right, lack of arm swing, and wide base of support. Child was unable to run or balance in single-leg stance on 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 position 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 with complaints of dizziness. Hyperactive nystagmus response on PRNT (duration of 90 seconds 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). Visual tracking intact in all directions with difference of two lines on DVA testing (head stationary versus rotating side-to-side).

Owing to the lack of dizziness or complaints of vertigo, 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 through the use of vestibular stimulation on scooter board as well as in net swing with and without visual fixation, with gradual increase in 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 at $<1$ Hz. Gradually this improved to her being able to 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, she could also do so in sitting position. 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 by 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. The patient was discharged from physical therapy services at school.

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CHAPTER 23

# Management of the Elderly Person with Vestibular Hypofunction

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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 due to 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. The vestibular insult or injury may be the same as in a younger individual, but the functional sequelae 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.[5–7] Functionally, VOR gain decreases, resulting in reduced visual compensation in response to head movement. These changes, however, appear to be frequency- and velocity-dependent.[8,9] Baloh and coworkers[10] found that older subjects had lower visual-VOR gain as velocity increased than younger subjects. Although these researchers examined this issue only at low-frequency rotation, there appears to be 35% less in visual-VOR gain in subjects younger than 75 years old than subjects aged 19 to 39 years. If the visual-VOR gain decreases with age, especially at higher velocities, it would lead to greater retinal slip and therefore poorer visual acuity during head movement. The changes in vestibular function have been compared to a progressive bilateral vestibular deficit[9–11] and may contri-

bute to the complaints of disequilibrium (an "off-balance" sensation) and to the gait ataxia, without any true vertigo, of many older people who are have nonsyncopal dizziness. Paige[12] suggests that this gradual, mild bilateral vestibular loss results in a mismatch in the visual and vestibular mechanisms. Interestingly, Nadol and Schuknecht[11] reported that degenerative changes can 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.[9] 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 drop in the vestibular and other systems' ability to compensate, the patient experiences symptoms of vestibular dysfunction. Thus, the problems are due to decompensation rather than to a new vestibular disorder.

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 6 and 21). The presence of visual and somatosensory changes with aging complicates the functional recovery for patient with a bilateral vestibular loss. Patients who present with instability of gait and functional mobility are more likely to require the use of a walker or cane and will always have difficulty walking in the dark.[13,14] 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 that he or she carry a flashlight at night so as never to have to walk in total darkness.[13] 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.

## 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 gluelike substance, and in all people there is a normal degeneration and regeneration of the crystals.[15] In addition, degenerative changes in the otoconia of the utricle and saccule increase with aging.[15] This process may contribute to the relatively high incidence of benign paroxysmal positional vertigo (BPPV) in the elderly. The incidence of idiopathic BPPV has been reported as peaking in the sixth and seventh decades of life.[16] Baloh and coworkers[10] reported that of 116 patients aged 70 or older that they saw for complaints of "dizziness," 25% had BPPV. Whitney and colleagues[17] 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[18] 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 was being seen for a balance or vestibular disorder. Gamiz and Lopez-Escamez[19] 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. Eighty-two percent of the adults in their survey older than 60 years also had a negative Dix-Hallpike test result at 30-day checkup, suggesting that older persons are successfully treated with canalith repositioning.

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 osteoporosis and osteopenia,[20] giant cell arteritis,[21] and higher rates of mild head trauma and diabetes.[22] All of these conditions are more common in older adults. Gizzi and colleagues[23] 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.[17] 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.[18] Because the Dix-Hallpike test result may be false-negative in some patients, Viirre and coworkers[24] 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[17] also suggest repeating the Dix-Hallpike maneuver as well as performing the test at different speeds with older adults in order to dislodge the otoconia. Also, the examiner is more likely to find evidence of BPPV earlier in the day.[17]

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 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 due to arthritis or other cervical disorders. Also, the patient should be screened carefully for vertebral basilar compromise. 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.

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.[25] 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 or who have had a cerebrovascular accident. We often perform the repositioning maneuver with two clinicians present to help the patient with position changes if they are severely impaired.

Another special group is adults with mental and/or physical disabilities. Having a family member present is helpful to ensure 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 due to a vestibular deficit without sophisticated testing (see Chapter 8). The results of these tests in older persons must be interpreted carefully and should be based on age-related normal values.[26] For example, in people in their 90s, caloric testing may document bilateral vestibular loss, but such people do not actually have a bilateral vestibular loss.

### *Visual Deficits*

Visual acuity, the ability to accommodate, and smooth pursuit normally decline with age.[27,28] These normal changes associated with aging can make adaptation after a vestibular insult more difficult.

An inability to adapt to the dark has been shown in the literature to be one of the reasons why older adults may fall.[29] Combining the dark adaptation disorder with vestibular dysfunction can make it dangerous for older adults with vestibular disorders to move from areas with ample light to darkened areas. This change in light has been shown to cause temporary blindness in older adults for more than a minute.[29]

In addition to dark adaptation, visual acuity and contrast sensitivity have been related to falls in older adults[30–34] 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.[35] 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.[35] Individuals with glaucoma also have difficulty with peripheral vision.[35] 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. Lord and associates[34] 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 glaucoma or other 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 23-1).[36] 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 to reduce hazards may be a very helpful intervention for people who have visual impairments.[35]

### *Somatosensory Changes*

Older adults appear to have a greater chance of reduced somatosensory function. Age-related electrophysiologi-

Box 23-1

## 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 2 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 |

**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 |

*(continued on following page)*

Box 23-1 *(continued)*

## HOME SAFETY CHECKLIST FOR DETECTION OF FALL HAZARDS

### 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 step-stool that you use to reach high cupboard and closet shelves? | yes | no |
| 33. Are ladders and step-stools in good condition? | yes | no |
| 34. Do you always use a step-stool 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 walk-ways, steps, and porches free of wet leaves and snow? | yes | no |
| 44. Do you sprinkle icy out-door areas with de-icers as soon as possible after a snowfall or freeze? | yes | no |
| 45. Do you have mats at door-ways 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 |

*(continued on following page)*

Box 23-1 *(continued)*

## HOME SAFETY CHECKLIST FOR DETECTION OF FALL HAZARDS

**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 thephone 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.)

cal and functional declines in the peripheral nervous system have been described in groups of healthy subjects 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.[37,38] Pathological changes such as axonal degeneration due to neuropathy result in abnormalities in distal somatosensation.[39,40] These somatosensory changes have functional implications. Ducic and colleagues[41] and Reid and associates[42] have reported that there are changes in balance as one has less sensation distally.

Older adults have more difficulty sensing vibration in the distal extremities as well as 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.[43]

### Musculoskeletal Deficits

Another potential difficulty in rehabilitating the older population 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.[44] 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.[45] Foot and ankle muscles appear to be very weak in many older patients, and strength training may be indicated. Waddington and coworkers[46] found that training with a wobble board enhanced older adults' ability to discriminate movement at the ankle.

## Postural Hypotension

Patients who show vestibular symptoms often complain of dizziness and/or imbalance. The dizziness needs to be differentiated from lightheadedness due to 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 5 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. Many drugs commonly taken by older adults can produce postural hypotension, including diuretics. Postural hypotension alone can put a person at risk for falling because of the significant dizziness that the patient experiences when changing positions quickly.

## Cerebellar Atrophy

Older adults may have disturbances in coordination and tend to move slowly. Patients with cerebellar disease appear to improve with balance therapy.[47] 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.[48–50] 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.

## Fear of Falling

Another common problem experienced by older individuals who have vestibular disorders is fear of falling.[51] Balance performance and confidence are related in community-living older people.[52] Older adults with vestibular loss often experience fear of falling[53] and, as a result, may reduce their activity level.[54–56] This fear of falling is extremely disabling to older adults and may actually prevent optimal functioning. Tinetti and associates[54] suggest that therapists work with patients to reduce their fear of falling, thus also enhancing their function.

When asking a patient about a fall, the therapist must make sure to be using the same definition of a "fall" as the patient. 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.[57–62] Shumway-Cook and associates,[57] 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 collegaues[58] 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.[58,62]

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.[58]

## 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.[63] 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.[64] Jacob[65] and Clark and associates[66] have suggested that anxiety is strongly related to vestibular dysfunction.

## 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.[67] Various studies have reported a reduction in falls rates after physical therapy intervention.[13,68,69]

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)[70–77] and the functional reach test.[78–83] The BBS has been used extensively to assess balance in older adults who have Parkinson's disease or stroke or who are frail (Box 23-2). It has been validated for use in persons with vestibular dysfunction.[77] 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.[76] Shumway-Cook and associates[76] determined that scores of 36 and lower on the BBS relate to 100% risk of falls in community living older adults.

Another tool that is helpful for assessing risk of falls is the functional reach test.[78] 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. In order 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[79] 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.[79] 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 also been shown to change over the course of rehabilitation.[83] Functional reach is sometimes administered to the patient who has dizziness,[80] although Wernick-Robinson and colleagues have suggested the test may have little value in persons with vestibular dysfunction.[84] 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 23-3).[85,86] 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.[87] The ABC scale has shown to be sensitive to change over the course of rehabilitation.[69,88,89]

The ABC scale has been compared to the Modified Falls Efficacy Scale.[56] 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[90] have reported that older adults who score 66% or less on the ABC are at high risk for falling. Myers and coworkers[85] 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.

The Vestibular Activities of Daily Living (VADL) Scale is another questionnaire that is helpful in determining the functional capabilities of people living with vestib-

*(text continued on page 387)*

Box 23-2

## 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 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 |

*(continued on following page)*

Box 23-2 *(continued)*

## BERG BALANCE SCALE

**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 finger tips 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 |

**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.

*(continued on following page)*

Box 23-2 *(continued)*

## BERG BALANCE SCALE

**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° safely in <4 sec each side | able to turn 360° safely one side only in <4 sec | able to turn 360° 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 |

**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 5–10 sec | able to lift leg independently; holds 3 sec | tries to lift leg; unable to hold 3 sec; remains standing independently | unable to; tries or needs assist to prevent falling |

**Total Score** _____

**Maximum Score** _____

Box 23-3

## 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 tip toes 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" vs "down" or "onto" vs "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.

ular disorders.[91,92] The VADL scale is further discussed in the chapter by Cohen on disability (see Chapter 24).

The modified Fast Evaluation of Mobility, Balance, and Fear Baseline (FEMBAF) questionnaire is extremely helpful in determining risk factors for falling (Box 23-4).[93,94] The therapist fills out "yes" or "no" answers to the risk factor questions either as he or she 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.

## Dizziness Assessment

The DHI is extremely helpful in determining what type of intervention will most benefit the patient with vestibular dysfunction.[53] This tool helps determine the self-perceived handicap of the individual completing the

Box 23-4

## 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:<br>• neurological (e.g., cancer, peripheral neuropathy, multiple sclerosis, lupus)<br>• cardiovascular (e.g., postural hypotension)<br>• musculoskeletal (e.g., total joint replacements, arthritis)<br>• sensory (e.g., visual impairment)<br>• other (amputation, Parkinson's disease, Alzheimer's disease) | _____ | _____ |
| 22. Takes medications that are potentially dangerous in regard to falls:<br>• hypotensives<br>• neuroleptics<br>• hypnotics/anxiolytics<br>• antiarrhythmics<br>• antiparkinsonians<br>• analgesics/anti-inflammatory drugs<br>• various vasoregulators | _____ | _____ |

Risk Factors (= total of "yes" answers): _____

*(continued on following page)*

Box 23-4 *(continued)*

## MODIFIED FAST EVALUATION OF MOBILITY, BALANCE, AND FEAR BASELINE QUESTIONNAIRE

**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 x 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.

form. The patient answers 25 questions related to dizziness and/or handicap. There are three subdivisions of the test, and subscores 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.[89] 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.[95,96]

## Typical Balance Tests

If the patient is complaining of balance problems, the single-leg stance (SLS), Romberg, and tandem Romberg tests are also performed. Bohannon and associates[97] have found that SLS times significantly decrease as people get older.[97] SLS, Romberg, and tandem Romberg tests help the therapist determine what kind of functional movements might be difficult for the patient.[97–101] 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.

## 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.[102]

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 to treat.

## 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 as well as 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.[88]

## What to Do Once the Risk Factor Has Been Identified

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 in order 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 contributer to falls.[32,33] 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 up to 6 weeks for improvement to be evident.[103–105] 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.[106] 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. If the patient has an extremely immobile foot, performing normal balance reactions will be difficult.

## CASE STUDY 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 to 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, normal. Rotational chair response, abnormal with moderately decreased gain and a mild directional preponderance. Positional testing, 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–.

*(continued on following page)*

## CASE STUDY 1 *(continued)*

The patient has already been diagnosed with bilateral BPPV. She is more symptomatic in the left Hallpike-Dix 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 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 gait.

## CASE STUDY 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 had two previous infectious events 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 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 12/100 to 5/100, 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 increased from 12 to 22. 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, and her composite sensory organization test score was 77/100. Her DHI score remained at 22/100, and her ABC score was 64%.

Mrs. M was given written ankle exercises, a 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 22/100. 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.

*(continued on following page)*

## CASE STUDY 2 *(continued)*

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 vestibular disorders require the physical therapist to think carefully before initiating an intervention program. Patient safety and encouraging compliance with the intervention are essential. Careful identification of the patient's functional limitations enables a therapeutic program to be devised to restore the older adult's function safely.

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CHAPTER 24

# Disability in Vestibular Disorders

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In the first published paper on vestibular rehabilitation, Cawthorne[1] wrote that any disorder of the labyrinth will be accompanied by balance disturbances that may be severe enough in extreme cases "to render the sufferer helpless and immobile." He also said that three weeks after an acute episode of "vestibular failure," presumably acute vestibular labyrinthitis or neuronitis, the patient begins to "creep about cautiously." When the patient has recurrent, momentary attacks of vestibular failure or with the head in certain positions—here he must have anticipated the diagnosis of benign paroxysmal positional vertigo (BPPV)—the result will be less "devastating." "Nevertheless, these recurrent attacks may engender such a feeling of insecurity that the sufferer feel impelled to limit his activities, and such people will frequently not venture out of doors unaccompanied."[1] Modern research has supported and extended his observations, but no other writers have summarized the problem so well.

## Evaluating Disablement

Knowing someone's symptoms may not help to understand that patient's functional limitations because symptoms and disablement are not necessarily related.[2–6] Disablement is sometimes more heavily influenced by the individual's life needs than by physiology. For example, the need to care for an elderly, disabled spouse may make someone with clinically significant vestibular weakness more functionally independent than someone with more social support, fewer life stresses, and more normal test results.[6] Furthermore, determining level of disablement is tricky. Self-ratings by patients may differ from ratings by their physicians and therapists, and from ratings by their significant others.[2] Rigby et al[4] pointed out that the patient is often more critical than the clinician.

A good assessment of disablement directly addresses the functional limitations caused by the disorder rather than the signs and symptoms of that disorder.[7] A variety of measures have been used to evaluate quality of life, disability, and outcomes of intervention, including subjective self-ratings of performance and objective ratings by health professionals. Some measures are specific to vestibular or balance disorders; others are more general. Assessments that are specific to vestibular disorders are sensitive to the subtle problems experienced by this population but do not allow comparison with other health conditions. General assessments allow comparison across health conditions but are insensitive to subtle but functionally significant problems experienced by people with vestibular disorders. The vast body of literature on evaluations for disability is beyond the scope of this chapter. For an excellent review of many general measures, as well as a discussion of the relative merits of general versus specific assessments, see the text by Dittmar and Gresham[8] as well as Reed's "Quick Reference" series.[9,10]

Three self-administered surveys of function have been developed specifically for vestibular disorders. The Dizziness Handicap Inventory (DHI)[3] is easy to use but omits some essential personal care skills and lacks sensitivity. It was developed and normed for patients with

Ménière's disease but has been used in other vestibular disorders. The Vestibular Disorders Activities of Daily Living Scale (VADL)[2,6] was developed for a wide range of vestibular disorders, is more specific to details of activities of daily living (ADLs), and is more sensitive than the DHI. A promising new self-assessment specifically for patients with Ménière's disease is the Ménière's Disease Outcomes Questionnaire, which addresses symptoms, some motor skills, and some social activities.[11] It could be improved with the addition of questions about problematic personal care activities, home management skills, and driving or other community mobility skills.

Self-ratings do not yield an unbiased assessment of performance skills but represent the patient's personal point of view. A widely used general self-rating is the Medical Outcomes Study Short-Form Health Survey Questionnaire (MOS SF-36).[12] The poor to moderate correlation of the DHI and MOS SF-36 suggests that they measure different domains.[13,14] The World Health Organization's International Classification of Functioning, Disability and Health (ICF)—the revised International Classification of Impairments, Disabilities and Handicaps—has the benefit of being an objective classification system but is cumbersome for even simple descriptions[15] and is not an assessment per se. Nevertheless, anyone interested in the topic of disablement should be familiar with the concepts embodied in the ICF.

Standardized tests allow for comparisons among subjects with similar diagnoses and among diagnostic groups. Investigators who use them, however, sometimes see the forest and forget the trees. The literature is replete with papers that report significant differences between average test scores but omit the important details as to what those test scores actually mean. Although many reports indicate that people with vestibular disorders have reduced functional independence and quality of life, few papers describe the tasks that are particularly problematic. That type of detailed information is essential for program development and treatment planning.

Several studies have used subjects with more than one diagnosis. In general, patients with vestibular disorders rate themselves as impaired in functional skills or having decreased quality of life.[16–20] Individuals with benign paroxysmal positional vertigo (BPPV) seem to be less impaired than others; patients with Ménière's disease and head trauma are relatively more impaired.[2,21,22] A study of a heterogeneous group of patients with vestibular disorders showed that, in general, inability to walk easily, restrictions in social life, and concerns about work are especially common.[23] Of particular concern is driving ability, because driving a car is potentially hazardous to the driver with a vestibular disorder as well as to other people. In industrialized countries, driving a motor vehicle is often essential for independent community mobility in the absence of adequate public transportation systems, which are lacking in much of the United States. Two studies have surveyed patients using instruments specific to the problem of driving.[24,25] Cohen et al.[26] used the Driving Habits Questionnaire, a well-normed instrument for doing a self-assessment by structured interview, and modified it by adding questions on some topics likely to be specifically problematic for patients with vestibular disorders.[24] These investigators found that 56% of patients with postoperative vertigo, 22% of patients with chronic vestibulopathy, and 15% of patients with Ménière's disease continued to drive despite their physicians' advice against it. Similarly, when Sindwani et al.[25] asked subjects whether they would continue to drive if their physician said it would be dangerous, 52% said they would. McKiernan and Jonathan[27] are rightly concerned about this problem.

## Benign Paroxysmal Positional Vertigo

Only a few papers mention disability and functional limitations in BPPV. Epley[28] noted that BPPV is at least mildly disabling for many patients. Some patients, he commented, must sleep sitting up. Some people with the disorder are nauseated much of the time and, if their jobs require upward pitch rotations of the head such as looking up (e.g., car mechanic), they may be unable to work.

Epley[28] also noted that some patients may have reduced ability to participate in active recreational activities. These observations were later supported by Cohen et al.[2] Herdman and Tusa[29] described complications that can occur after use of Epley's canalith repositioning maneuver. They noted that the patient in their Case 3 had such severe disequilibrium that she had difficulty walking. Disequilibrium so severe is unusual, but many patients feel that they must walk more slowly and carefully than usual. After treatment with repositioning maneuvers, most patients report that they can walk more normally, even though to the objective observer they may not have appeared to be ataxic before treatment. This discrepancy is easily explained. Many people are quite sensitive to subtle changes in their movement skills and may be aware of subtle differences that cannot be observed. Asking the patient to walk for several steps and report how he or she feels after treatment is an easy way to assess treatment success immediately. If the patient has some ataxia, then another trial of the maneuver may be indicated.

Cohen et al. have studied changes in ADLs in patients with BPPV in more detail. Using a five-point,

self-rated scale of ADL independence, Cohen and Jerabek[30] found that most patients with BPPV rated themselves as having to use safety guarding for many tasks before treatment, but rated themselves as independent after treatment. Similarly, using the VADL, Cohen et al.[2] reported that many patients with BPPV rated themselves as having mild discomfort or not performing tasks as well as usual, and these ratings improved after intervention. For tasks that involved pitch (up/down) rotations of the head, subjects rated themselves as having to be more careful.

This finding is consistent with the clinical descriptions given by patients. Some patients complain of difficulty making the bed, putting away grocery items on cupboard shelves, or changing a light bulb. Patients often report having discomfort when they look up or when they bend down to tie their shoes or reach for an object on the floor. Parents and grandparents of young children complain of being unable to bend down to pick up a child. More than one patient has complained of having vertigo when reclined in the dentist's chair. Occasionally, a female patient complains of having had vertigo while her hair was washed at the beauty salon. Although not usually handicapping, vertigo elicited by BPPV can be serious enough to affect job performance, especially when the occupational role involves significant vertical or pitch rotations of the head—that is looking up or down. A range of jobs can be affected, including car mechanic, truck driver, nurse, dentist, and any scientist who uses a light microscope.

Driving skills are of at least slight concern. In the driving study by Cohen et al,[24] many people with BPPV reported having to be cautious; 23% reported having difficulty under conditions of degraded visibility, spatial challenges, and busy traffic. By comparison, 14% of normals in the same survey reported having some difficulty. Some subjects with BPPV limited their driving to local roads and avoided highways, and some said they had stopped driving altogether. This decreased community mobility may influence the ability to go to work, run errands, care for dependent children and disabled adults, and otherwise participate in community life (Table 24-1).

## Chronic Vestibulopathy

The term *chronic vestibulopathy* is used at Baylor College of Medicine to refer to patients with chronic, recurrent, brief episodes of vertigo elicited by head movement. These patients have uncompensated labyrinthitis or vestibular neuronitis, and unilateral weakness on bithermal caloric testing or reduced vestibulo-ocular reflex (VOR) gains during low-frequency sinusoidal rotations in darkness.[31,32] They do not have Ménière's disease. They do have decreased independence in ADLs, as indicated by scores on both the VADL and DHI.[31] Decrements have been found in all subscores of the VADL: functional, ambulation, and instrumental or higher-level skills. Problematic skills span the range of ADLs, including bathing and dressing, ambulation on level and uneven surfaces, home management, and community mobility.[16,33] These patients often decrease their activities outside the home, forgoing participation in social activities such as church groups, daily walking programs outside the home, essential shopping trips, and other activities.[34] After 4 to 6 weeks of vestibular rehabilitation, these scores improve.

The psychological construct of locus of control—that is, one's sense of ability to influence one's own recovery—was weakly associated with improvements on the VADL and more strongly associated with improvements on the DHI. This finding indicates the important influence of psychological factors on functional recovery. This idea is supported by a retrospective report from a heterogeneous group of patients that showed a relationship between performance on tests of balance skills and self-perceived handicap[35] and by another report of the relationship between symptoms and handicap.[36]

Driving skills are a concern in patients with chronic vestibulopathy. They are often aware that they do not drive well.[2,16,33] In the driving study by Cohen et al,[24] 23% of subjects reported having difficulty driving under conditions of degraded visibility, spatial challenges, and busy traffic. Driving in the rain, at night, in freeway traffic, and in a ramped parking garage (where repeated turns may elicit vertigo) were the most problematic. This group reported having more driving problems than other patient groups.

## Bilateral Vestibular Impairment

Bilateral vestibular weakness causes disequilibrium and oscillopsia. In his autobiographical case report, JC[37] discussed the disabling effects of his own symptoms. In the initial stage he had to stabilize his head against the bed frame to reduce oscillopsia so he could read. Later, he was still unable to read street signs or see faces clearly while moving. Therefore, to avoid rudeness to his acquaintances, he used the strategy—considered socially inappropriate in taciturn New England—of greeting everyone he passed on the street. Patients with bilateral vestibular loss (BVL) are at significant risk of falling, and their scores on the DHI total score and subscores indicate perceived significant functional.[38]

No patients seen for vestibular rehabilitation at Baylor College of Medicine have been unable to perform

## ■ Table 24-1 PERCENTAGE OF SUBJECTS HAVING DIFFICULTY WITH PARTICULAR DRIVING CHALLENGES, BY DIAGNOSTIC GROUPS

| | *Percent of Subjects Having Difficulty* | | | | | | |
|---|---|---|---|---|---|---|---|
| **Driving Challenge** | **Normal (n = 51)** | **Postoperative (n = 9)** | **BPPV (n = 34)** | **CV (n = 27)** | **Ménière's Disease (n = 47)** | **Patients vs. Normal Subjects (*P* value)** | **Diagnostic Groups Differ (*P* value)** |
| Rain | 35 | 50 | 36 | 67 | 40 | 0.024 | 0.019 |
| Alone | 0 | 67 | 26 | 67 | 29 | < 0.001 | 0.001 |
| Parallel parking | 33 | 50 | 41 | 62 | 45 | 0.101 | 0.594 |
| Left turns across traffic | 4 | 57 | 15 | 46 | 30 | 0.001 | 0.086 |
| Freeway driving | 12 | 62 | 15 | 67 | 26 | 0.011 | < 0.001 |
| High-traffic local roads | 13 | 43 | 13 | 58 | 33 | 0.022 | 0.028 |
| Rush hour driving | 21 | 57 | 19 | 59 | 31 | 0.004 | 0.063 |
| Night | 22 | 43 | 37 | 73 | 57 | 0.002 | 0.108 |
| Parking spaces | 10 | 56 | 15 | 44 | 21 | 0.037 | 0.015 |
| Changing lanes | 12 | 50 | 18 | 59 | 30 | 0.007 | 0.006 |
| Staying in lane | 2 | 56 | 12 | 44 | 17 | < 0.001 | 0.005 |
| Traffic checks | 4 | 56 | 26 | 52 | 33 | < 0.001 | 0.105 |
| Ramped garages | 10 | 43 | 29 | 61 | 35 | 0.003 | 0.333 |

BPPV = benign paroxysmal positional vertigo; CV = chronic vestibulopathy.
From Cohen et al, 2003,[24] with permission of Elsevier.

personal care skills, but many of them have commented on having to use safety guarding while taking a shower as well as while doing household chores. Several people have said they had to modify the way they performed sports or have reduced their participation in athletics and other leisure activities outside the home. Shortly after developing bilateral vestibular weakness most people have difficulty walking over uneven surfaces and must be careful when walking on uneven sidewalks or across open ground. No data, however, are available on these points.

No studies have evaluated the driving performance of subjects with BVL per se, but these patients comment consistently that they do not see details clearly while driving. Of 34 patients with total or near-total BVL seen at Baylor College of Medicine for vestibular rehabilitation over a 10-year period, 4 were unable to drive for reasons unrelated to their vestibular impairment; 10 had stopped driving; and 20 could drive but complained of having to drive more slowly and carefully than usual, avoiding the highways whenever possible. All of the drivers complained of having difficulty reading the dashboard indicators and reading signs. This observation suggests that patients with BVL have significant oscillopsia. This idea is supported by two studies. In the first study, 7 of 10 patients with significant bilateral vestibular weakness had decreased ability to select information from an alphanumeric sign while being driven than while sitting still.[39] In the second study, patients with BVL had

decreased dynamic visual acuity (DVA) compared to normals both while walking and while standing still.[40] Although some of the patients in the second study were aware of decreased DVA while walking, none were aware of decreased DVA during quiet standing.

## Acoustic Neuroma

In one of the first studies of the effect of acoustic neuroma resection on functional skills, Weigand and Fickel[41] surveyed 541 members of the Acoustic Neuroma Association who had undergone surgery in the decade from the mid-1970s to the mid-1980s. Two percent to 5% of respondents had difficulty returning to self-care activities, housework, and shopping, and 10% to 15% reported some sexual dysfunction. In a later, more detailed examination of challenging tasks, routine self-care tasks were affected minimally but climbing a ladder was difficult for more than 60% of subjects who had undergone acoustic neuroma section.[42] Bateman et al.[43] subsequently reported that 6% of patients surveyed 1 to 3 years after acoustic neuroma surgery had some difficulty climbing stairs but that only 1% of respondents had difficulty gardening or doing household chores.

Cohen et al.[44] studied a group of subjects after they underwent acoustic neuroma resections. Subjects were studied as inpatients for up to 6 days and as outpatients for up to 3 months. Compared to results in a sham treatment group, vestibular rehabilitation given during the inpatient, postoperative week did not influence recovery during the inpatient period or during outpatient follow-up. As a whole, however, the sample showed clinically and statistically significant improvements in ADL independence over time. As indicated in Figure 24.1 showing previously unpublished data, Friedman repeated measures analysis of variance showed significant changes in personal care activities ($p \leq 0.01$) during the first postoperative week, which largely leveled off by Week 3. In higher level skills not performed as in-patients, Wilcoxon matched pairs signed rank tests indicated that skills improved from Week 3 to Week 7 ($p \leq 0.03$), but did not change significantly from Week 7 to Week 13. Thus, in the acute phase of recovery from acoustic neuroma resection patients have significant functional limitations in their ability to care for themselves independently.

Return to work is related to the length of time after surgery, the preoperative health status and motivation of the patient, and the nature of the patient's job. In two reports, some subjects were unable to return to work or had to change their jobs, but 70% to 79% of subjects could return to their usual work.[43,45] Another paper reported that 49% of patients who had undergone acoustic neu-

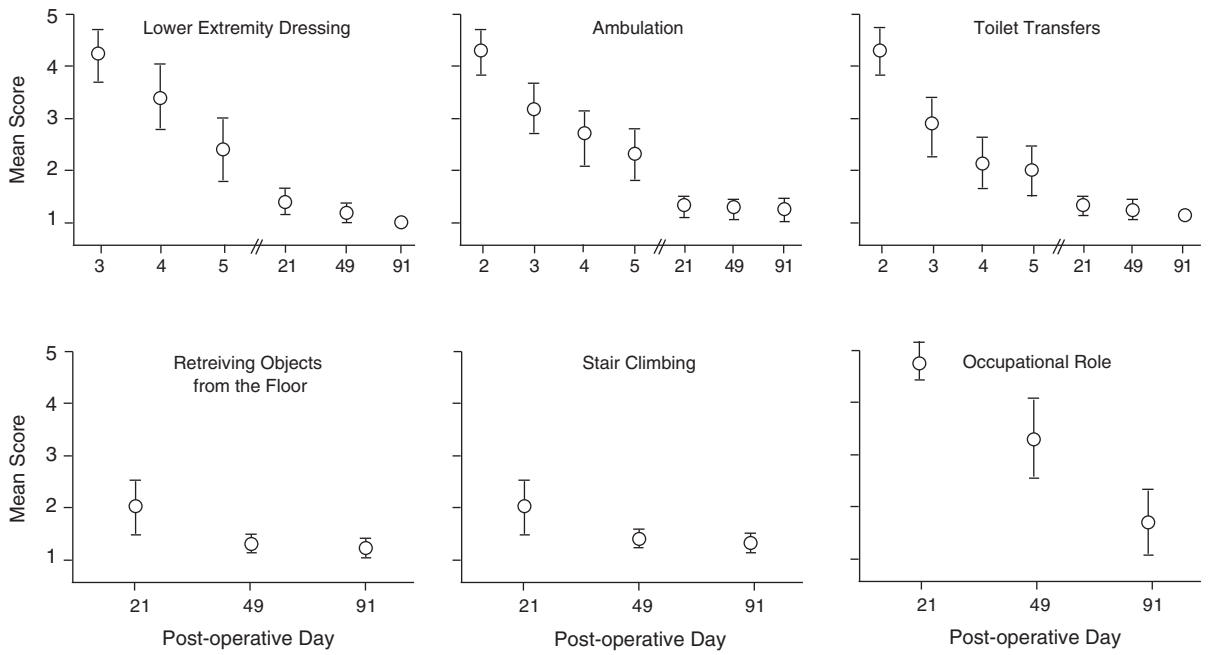


**Figure 24.1** Mean independence ratings for representative activities of daily living (1 = independent; 5 = dependent). Individual postoperative days are indicated in the first week; days 21, 49, and 91 are approximations. *Occupational* role refers to jobs or work. Error bars are 95% confidence intervals.

roma resection were able to return to work within 4 months and that 99% had returned to work within 1 year,[46] a finding that was confirmed by two other studies (see Fig. 24.1).[42,44]

These return-to-work data may be somewhat deceptive. Many patients complain that they need at least minor adaptations to their jobs, often because of hearing loss but sometimes due to blurred vision or disequilibrium. The job responsibilities of some people change.[42] Andersson et al.[47] pointed out that although only 12% of their 141 subjects were on sick leave or medical retirement after acoustic neuroma resection, at least some of the 38% of subjects who said they had retired for other reasons may have been influenced to retire by their postoperative status.

As with changes in measures of balance and vestibulo-ocular reflex,[44] changes in self-assessment of disability or handicap usually stabilize by 3 months after surgery.[48] A retrospective study of military personnel and civilians in high-risk jobs showed that of young to middle-aged adults who had cerebellopontine angle tumor surgery and who voluntarily worked in high-risk occupations, 86% returned to their previous level of work performance.[49] The investigators suggested that motivation to return to their chosen careers was an important factor for their subjects.

Driving a motor vehicle is essential for community mobility in many places. Therefore, despite their surgeons' instructions not to drive within the first postoperative month, many patients drive anyway.[24] As with the other diagnostic groups, driving is a challenging task for these patients, more challenging under conditions of degraded visual information. Not surprisingly, after acoustic neuroma resection, many people say they have difficulty driving at night; they do not have difficulty in daylight.[42] In a study conducted in Great Britain, 6% of patients reported being unable to drive 1 to 3 years after acoustic neuroma surgery.[43] In the driving study by Cohen et al.[24] 53% of postoperative acoustic neuroma subjects reported having difficulty driving under a variety of challenges, including conditions of degraded visibility, spatial orientation problems, and busy traffic.

Social activities are also sometimes affected. Weigand and Fickel[41] reported that 8% of people had difficulty returning to social activities after acoustic neuroma excision. Bateman et al.[43] reported that 15% of their patients had difficulty attending large social gatherings. The specific social context affects participation. People who had difficulty attending large gatherings may have been disturbed by their hearing loss or difficulty localizing sounds. Indeed, Bateman et al.[43] reported that 9.5% of their subjects had difficulty localizing sound sources, and 6% had difficulty following a conversation in a crowd. Postoperatively, patients have a small but significant reduction in the rate of seeing friends weekly, but more than 80% of subjects did see friends at least once a week before and after surgery.[42] Participation in sports was also decreased postoperatively although that change was not statistically significant.[42] That finding is supported by more recent work showing that 3% to 4% of respondents to a survey reported having decreased their participation in athletic activities.[43] Although these numbers are small, such changes may have had serious social ramifications for the individuals affected.

The effect of tumor size on overall quality of life is unclear. In one study patients with tumors smaller than 1.5 cm had a better postoperative quality of life than patients with larger tumors,[50] but other groups have found that tumor size is not significantly related.[42,48] Quality of life is not affected by age at time of surgery[42,44,51,52] or by participation in vestibular rehabilitation in the acute phase of recovery.[44,50–52] One group has reported that patients given vertigo habituation exercises took longer to resume independent ambulation than patients who did not have vestibular rehabilitation.[51]

The effect of facial nerve weakness after surgery is also unclear. One report indicated that for many patients, the effect on facial expressions and thus on socialization is a significant factor in disablement,[41] but another report indicated that facial nerve weakness is rarely disabling.[4] Postoperative facial nerve weakness is most likely to be an significant factor when eye closure is affected, because difficulty blinking or closing the eyelid can lead to serious eye deformities and decreased vision.

These data are interesting but do not tell the entire story. Descriptions of research findings are necessarily dry reading. Disablement, however, is anything but dry. The difficult work involved in coping with the consequences of surgery is often emotionally charged and frustrating. Several patients with facial nerve damage have commented that they no longer go out to restaurants with friends or relatives because they are embarrassed about drooling on the involved side. Some patients have said that difficulty localizing sound with just one ear in a noisy environment prevents them from going out to social events and other community activities.

An extreme example illustrates the problem and the multiple roles for therapists who do vestibular rehabilitation. A patient who lived alone complained of severe disequilibrium and falls 2 months after acoustic neuroma surgery. She also complained that her eye was sore. She

had not washed her clothes because she was unable to negotiate the stairs in her apartment building leading to the laundry facilities while carrying laundry. She was unable to clean up the piles of laundry and papers on the floors and work surfaces in her apartment and had fallen on some of the piles. She was too embarrassed to ask for help or to mention these problems to her physician. Instead, she purchased more clothing and put all the dirty laundry in bags. As well as doing balance therapy, the occupational therapist who saw her for vestibular rehabilitation 2 months after surgery informed her ophthalmologist about the patient's visual problems, and helped this patient develop strategies for hand-washing lingerie, carrying other laundry items, and keeping her home better organized and clutter-free.

The following paragraphs contributed by a patient who is also a staff member make the experience of recovery from acoustic neuroma resection more vivid. Emily K. Murphy, R.E.E.G.T., is a senior research technician in the Center for Balance Disorders, Bobby R. Alford Department of Otolaryngology–Head and Neck Surgery, Baylor College of Medicine. She runs the VIIth nerve intraoperative monitoring service and often staffs the kind of procedure she underwent. A creative and enthusiastic individual, she took responsibility for her own rehabilitation. Two years after her surgery, she drives her car in heavy traffic, wears high heels occasionally, goes biking and canoeing, does award-winning photography, and generally enjoys life. The following two paragraphs are her words:

> A couple of years ago, I was shocked to find out that I had an acoustic neuroma, a tumor whose removal I regularly monitor during surgery as part of my work. I remember thinking prior to this that it was, "No big deal. . . . You've got the tumor, you just get it out." It's a different ball game when it's in your field. Prior to surgery, I had bilaterally equal vestibular function. When I awoke from surgery, I felt as if I were in a centrifuge. By the second day, I had slowed down to 2 rpm, which was tolerable. The first month, with my head and eye movements relatively uncoordinated, I tired easily. I can remember sitting in the doctor's office, looking at a nebulous pattern on the carpet, and the whole floor seeming to shimmer—without my moving at all! Recovery was gradual (partly, I believe, because my "good" ear isn't that good). Doing things like jigsaw puzzles was good "brain training," and safe because I was sitting. It was hard work, and after 15 minutes I would break out in a sweat from the effort and have to take a break. After a rest, I would go at it again, gradually lengthening my tolerance. The ultimate challenge, though, was walking. Turning my head from side to side as I walked threw me off balance. If I were walking down the street and heard a car, I had to stop to turn and look.
>
> Even today, the hardest situation for maintaining my balance is leaving a movie theater near us. It has the unfortunate design of having small rows of lights at the edge of each step. In the dark, the bright lights are disorienting. With this decreased visual reference, I have almost fallen but was saved by my faithful husband. We have learned to let him go down first, and I keep my hand on his shoulder to steady myself. Even after over two years, I still have some problems with balance at night, walking with a flashlight, etc. However, the good news is there is nothing I don't do or try. I climb ladders (daytime!), use power tools, sail our catamaran, use a treadmill—anything I want to do. I just must be cautious when suddenly turning. I think attitude is extremely important. Instead of moaning about "why me?" one must look at each gain as a goal. The choice is being depressed about it for the rest of your life, or looking at it as a bump in the road of life. At some point you reach the top of the hill, and things start coming more easily.

## Ménière's Disease

A peculiarity of Ménière's disease is that the symptoms come and go. The intermittent nature of symptoms means that patients may feel normal or have a constant low level of discomfort and disability in between attacks or episodes but they feel significantly worse during exacerbations, with consequent greater disability. Patients with Ménière's disease have a significantly reduced quality of life on days when they have increased symptoms; on days when they have significant Ménière's "attacks," they are the most debilitated (Fig. 24.2).[53,54] Haye and Quist-Hanssen,[55] surveyed 111 patients with Ménière's disease seen between 1960 and 1970. They reported that Ménière's symptoms and fear of a Ménière's attack are incapacitating. They found that 23% to 46% of subjects had difficulty walking at dusk, were unfit for work or took early retirement, and had problems at work, at home, and among friends. Fifty-five percent of their subjects complained of nervousness.

Pharmacological and surgical interventions have improved considerably since the 1960s, so differences in reported percentages of disability from the mid-20th cen-

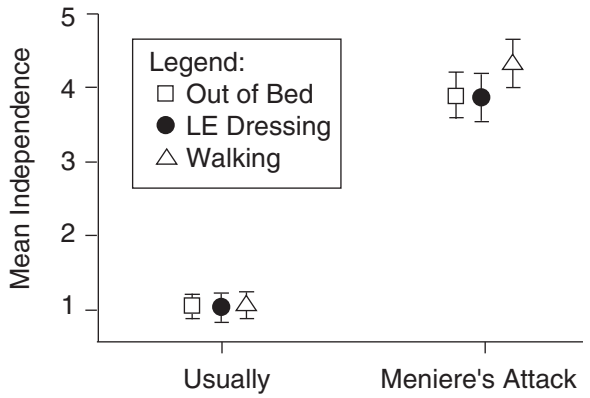


**Figure 24.2** Mean independence ratings for representative activities of daily living using a 5-point scale during quiescent periods when the patient feels relatively normal (left side of each pair) and during attacks of Ménière's disease (right side of each pair). Error bars are 95% confidence intervals; LE = lower extremity.

tury to the early 21st century are not surprising. Nevertheless, subsequent studies have consistently supported the findings of difficulties within the home, at work, to when socializing as well as concerns about having future, incapacitating Ménière's attacks.

In a study using the DHI, 53% of patients rated themselves as moderately to severely handicapped; unfortunately, details from the DHI subscales were not provided.[56] That study also showed that on the MOS SF-36, patients with Ménière's disease had lower scores on the emotional subscale than the physical subscales, from which the authors surmised that the psychological effects of the disorder are handicapping. A later report indicated that patients with Ménière's disease have lower scores on both physical and mental subscores of a short version of the MOS SF-36, the MOS SF-12, and on other measures,[54] supporting the earlier findings.[56]

Similarly, using a mail survey, another group asked subjects about their function in the preceding 6 months.[57] Subjects reported generally reduced life satisfaction with particular influence on leisure time and work. Unsteadiness, insomnia, anxiety, and lack of concentration were all significant complaints. Ménière's attacks were highly disruptive; 82% of subjects always or often discontinued their activities. Seventy-four percent of subjects avoided activities such as meetings, telephone calls, dinner, and physical activities. Thus, the majority of subjects avoided the kinds of activities involved in many jobs, social activities, and even volunteer work, child care, and home management.

Cohen et al.[53] surveyed 51 patients with Ménière's disease by mail to learn about their independence in self-care, mobility, and instrumental ADLs. These investigators found that during episodes of symptom exacerbation, Ménière's patients have difficulty using a telephone and doing other tasks that require good hearing; they also have particular difficulty with tasks that require good balance. At other times, they are independent (see Fig. 24.2).

In a study of patients after treatment with surgery or gentamicin, Soderman et al.[58] reported that quality of life, as measured by several scales, was generally good or very good. They found that vertigo, tinnitus, and hearing loss were all related to self-rated quality of life and all had equal influences. This difference from the Cohen study is attributed to successful treatment of vertigo by surgical or pharmacological intervention. When vertigo was successfully treated, patients were still bothered by the other symptoms. Vertigo had a greater influence on physical functional limitations, but hearing problems had a greater influence on psychosocial problems.[59] This differentiation explains earlier findings.[53,56] A subsequent study that used structured interviews with patients with Ménière's disease, however, suggests a more complicated relationship between disability and psychological factors. The interviews revealed that many patients with Ménière's disease have phobic reactions due to fear of having a Ménière's attack or to fear of ridicule for their ataxia, which might be mistaken for evidence of drunkenness.[60]

Two years after treatment with gentamicin, a group of patients with Ménière's disease had significant improvement on the DHI; that improvement was retained 5 years later.[61] In light of work showing different effects of vertigo and hearing impairment on quality of life measures, the omission of data from the DHI subscales in this study is unfortunate. In a study of patients with Ménière's disease treated with gentamicin, the pretreatment DHI scores were relatively high but dropped significantly over the 2-year follow-up period.[62] Interestingly in light of the findings reported by Soderman et al.,[59] scores on the Functional subscale decreased more than the Physical or Emotional subscales, although all subscales showed decreased scores.[61] Although vertigo also decreased, the relationship of changes in vertigo to changes on the subscales is unclear.

Return to work by patients with Ménière's disease varies. In the study by Cohen et al.,[53] most subjects who held paid employment reported that they continued to work, but 86% felt that their job performance suffered, and 70% had to adapt their jobs in some way. These data hint at the economic costs of this disorder (see Fig. 24.2). A survey of 133 patients with Ménière's disease 2 years after their initial diagnoses, unfortunately without statistical analyses of the data, suggested that many subjects who declined surgery but had received medication still had improvements in their self-ratings on a 4-point scale

of disability.[63] Initially, 23% rated themselves as moderately or totally disabled, but 2 years later that number decreased to 10%. In another large mail survey of 261 patients treated by the same surgeons, 99% of subjects who had been working were able to return to their jobs within 6 months, but 13% of subjects either had stopped working or were fired, and 7% of subjects had retired.[46] Thus, some individuals may have overestimated their abilities or found that the sequelae of surgery were too disabling. By contrast, in an interview study of 18 subjects, 9 had returned to work, 7 did not return to work, and 2 had retired preoperatively.[64]

The intensity of attacks in Ménière's disease varies, and the intensity of an attack affects the individual's ability to function while it occurs. Twelve percent of subjects surveyed by Cohen et al.[53] had no safe place to rest during an attack. The availability of a safe place to rest during an attack and the unpredictability of attacks were not correlated with whether or not the subject continued to work. Havia and Kentala[65] reported that 17% of their subjects were able to continue their activities during Ménière's attacks, but 56% of subjects had to lie down and 27% were completely incapacitated.

In rehabilitation, we often assess specific functional limitations but we may ignore the important psychosocial consequences. We may also ignore, or be unable to evaluate, individual coping styles. Nevertheless, personality factors strongly influence recovery in rehabilitation. As Emily Murphy wrote so clearly in the Acoustic Neuroma section of this chapter, a positive attitude is essential. Soderman et al.[58] made the important point that quality of life is related not only to symptoms but also to the ability to manage stress. Similarly, the interviews by Erlandsson et al.[60] revealed that stress and poor coping strategies exacerbate the disabling and isolating effects of the disorder. Thus, stress management interventions might be indicated to improve quality of life in some patients with Ménière's disease.

Driving an automobile can be but is not always problematic for patients with Ménière's disease. In a mail survey of Ménière's patients, 60% of respondents reported that driving was difficult, dangerous, or both.[53] In their structured interview study on driving problems, Cohen et al.[24] found that 33% of patients with Ménière's disease report having difficulty driving under conditions of degraded visibility, spatial challenges, and busy traffic. Also, patients with this disorder reported having had to pull off the road due to vertigo more often than any other patient group. Cohen et al. found no difference in crash rate between normals and patients with vestibular disorders, including patients with Ménière's disease.[24] Sindwani et al.,[25] however, found that 13% of patients with Ménière's disease reported having injured themselves. The circumstances of injury were not clear. These data suggest that having adequate public transportation systems is important so that patients with this disorder can avoid driving. Many metropolitan areas lack good public transportation. Therefore, having good shoulders on roads, where drivers can pull over in emergencies, is essential.

## Acknowledgments

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CHAPTER 25

# Assessment and Management of Disorders Affecting Central Vestibular Pathways

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Vestibular pathways run from the VIIIth 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 due to 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]

## Clinical Classification of Central Vestibular Disorders

The "elementary" neuronal network of the vestibular system is the di- or trisynaptic vestibulo-ocular reflex (VOR). There is evidence for a useful simple clinical classification of central vestibular syndromes according to the three major planes of action of the VOR (Fig. 25.1): 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. 25.2):

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

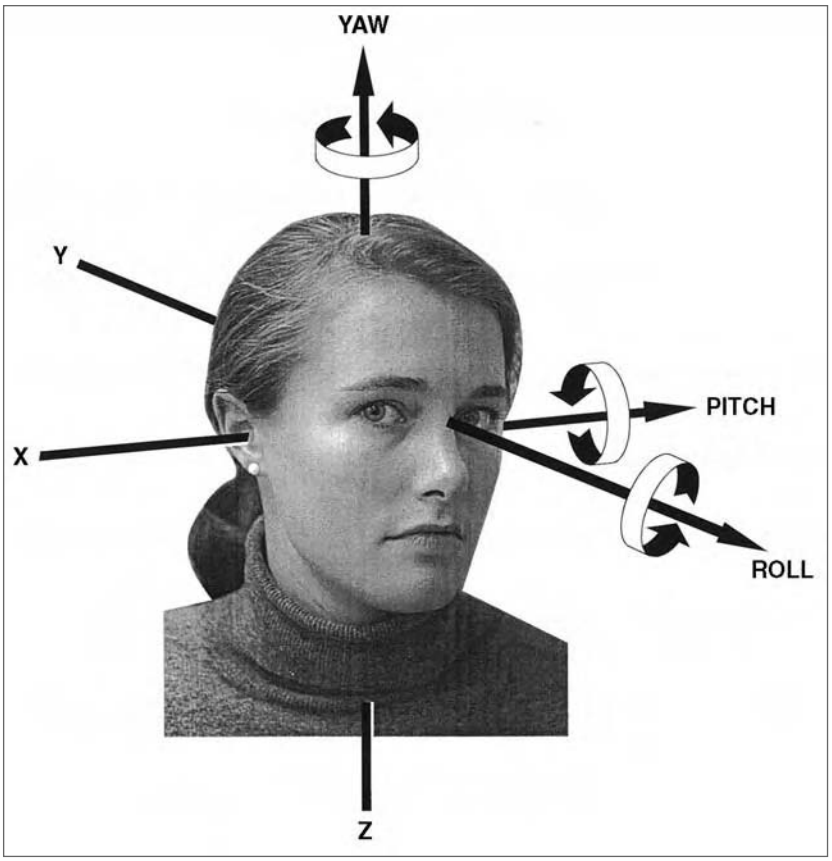


**Figure 25.1** Schematic representation of the three major planes of action of the vestibulo-ocular reflex. yaw = horizontal rotation about the vertical z axis; pitch = vertical rotation about the binaural y axis; roll = vertical rotation about the x axis ("line of sight"). (Courtesy of Alice Kniehase.)

Some vestibular disorders are characterized by a simultaneously 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. Dysfunction of this multisensory and sensorimotor cortex for spatial orientation and self-motion perception may be involved in spatial hemineglect and rare paroxysmal room-tilt illusions.

Most central vertigo syndromes have a specific locus (Table 25-1) but not a specific etiology. The etiology may, for example, be vascular, autoimmunological as in MS, inflammatory, neoplastic, toxic, or traumatic.

## 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[6] and the ocular motor nuclei[7,8] to subserve static and dynamic vestibular function in pitch (up and down in the sagittal plane) and roll (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:

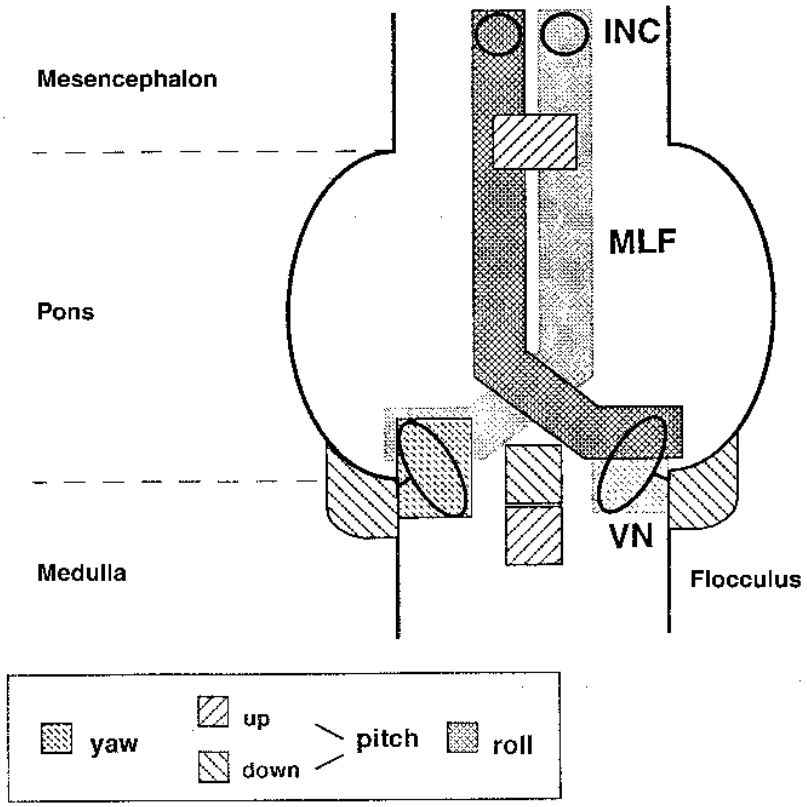


**Figure 25.2** Vestibular syndromes in roll, pitch, and yaw planes: Critical areas are schematically represented on the basis of our current knowledge of vestibular and ocular motor structures and pathways, a lesion of which causes a vestibular tone imbalance in one of the three major planes of action. 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]

- 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. 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.[9] Skew deviation was found in one third and a complete OTR in one fifth of these patients (Table 25-2).

## ■ Table 25-1 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 parietoinsular vestibular cortex |
| | 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, or frontal cortex lesions |
| | Tilt of perceived vertical with body lateropulsion (mostly contraversive) | Vestibular tone imbalance in roll with acute lesions of the parietoinsular vestibular cortex |
| 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 |
| Ponto-medullary 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 VIIIth nerve |
| | 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 |

*(continued on following page)*

## ■ Table 25-1 CENTRAL VESTIBULAR SYNDROMES *(continued)*

| Site | Syndrome | Mechanism/Etiology |
|---|---|---|
| | 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) |
| | 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<br>EA2 = autosomally dominant inherited calcium channelopathy |
| | Epidemic vertigo | Viral infection of cerebellum |

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

## ■ Table 25-2 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 |
| 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 | 111 | 94 | 47 | 36 | 31 | |

*Additional third nerve palsy.
†Slight torsion of about 2.8°.

Current clinical data support the following preliminary topographic diagnostic rules based on vestibular signs and symptoms in roll[3,4] (Fig. 25.3):

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.
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.
3. All tilt effects—perceptual, ocular motor, and postural—are ipsiversive (ipsilateral eye lowermost) and due to 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.

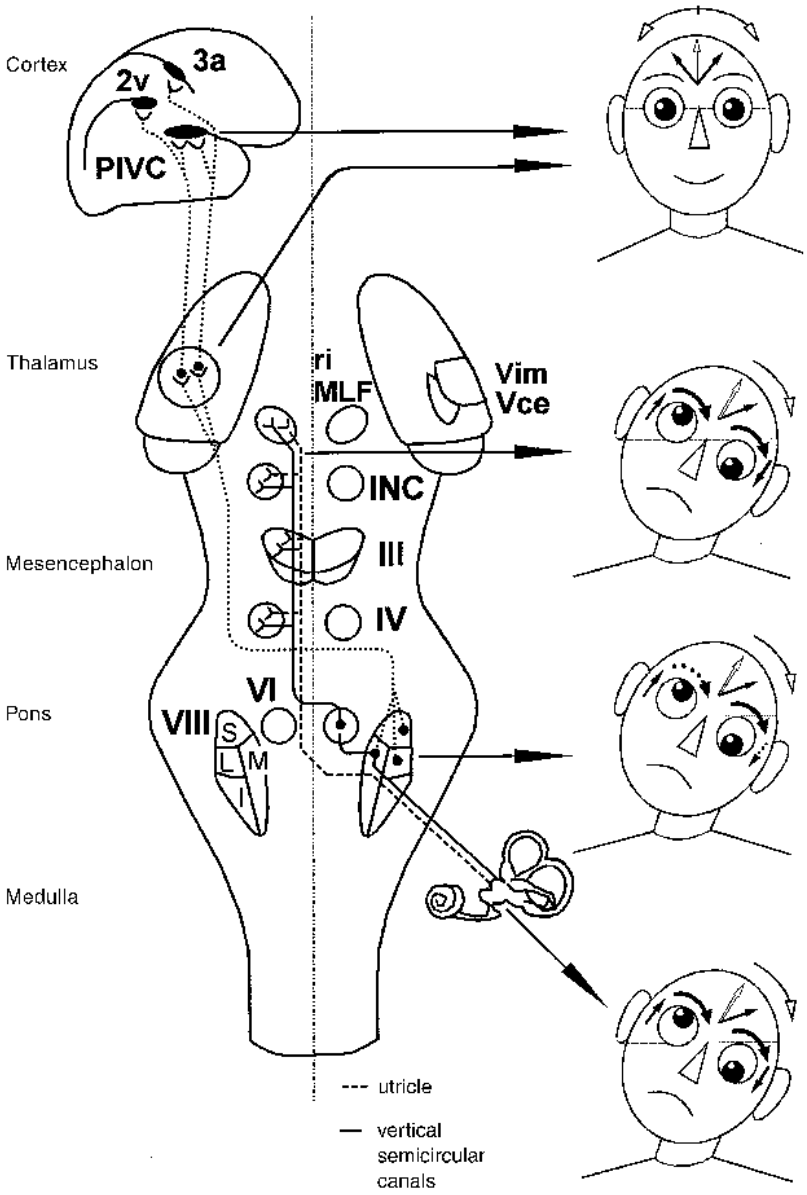


**Figure 25.3** Vestibular syndromes in roll plane: Graviceptive pathways from otoliths and vertical semicircular canals mediating vestibular function in roll plane. The projections from the otoliths and the vertical semicircular canals to the ocular motor nuclei (trochlear nucleus IV, oculomotor nucleus 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]

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. OTR with unilateral (ponto) medullary lesions (vestibular nuclei) indicates the "ascending" (reflexive) type of a tone imbalance of the VOR in roll.
6. OTR owing to rostral midbrain lesions (INC) reflects the "descending" type of tone imbalance involving the neural integration center for eye-head coordination in roll.
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.
9. OTR in unilateral paramedian thalamic infarctions (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).
11. Unilateral lesions of the PIVC cause moderate, mostly contraversive SVV tilts (temporal branches of the middle cerebral artery or deep perforators) and "cortical lateropulsion."
12. 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.[10]

Tilt effects caused by paroxysmal activation of "graviceptive" pathways point in the opposite direction to those caused by lesional inhibition, such as unilateral infarction.[11–14] 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 indicates 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.[11,15] We have also seen cases with unilateral thalamic hemorrhages or lower brainstem hemorrhages (cavernous angioma, lymphomas), after severe brainstem concussion, in MS, or associated with attacks of basilar migraine.

The paroxysmal OTR described in a patient with MS[13] may be a variant of the paroxysmal attacks assumed to arise from ephaptic spreading between adjacent demyelinated axons. We have observed repeated paroxysmal attacks of contraversive OTR with ipsiversive torsional nystagmus in the acute stage of Wallenberg's syndrome and in neurovascular cross-compression of the VIIIth nerve.

## 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 7 patients over a period of up to 1 to 3 years[16]) (Fig. 25.4). We found that repeated measurements of skew deviation, ocular torsion (OT), and tilts of SVV made during a single day showed consistent tilts.[9] Repeated measurements on subsequent days showed a gradual recovery, mostly within 30 days, both for OT and SVV (Fig. 25.5). Some patients, however, maintained a residual OT of a few degrees without a corresponding tilt of SVV for up to 2 years.

Recovery is probably 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. Physical therapy may facilitate this central compensation, but this possibility has not yet been proven in a prospective study.

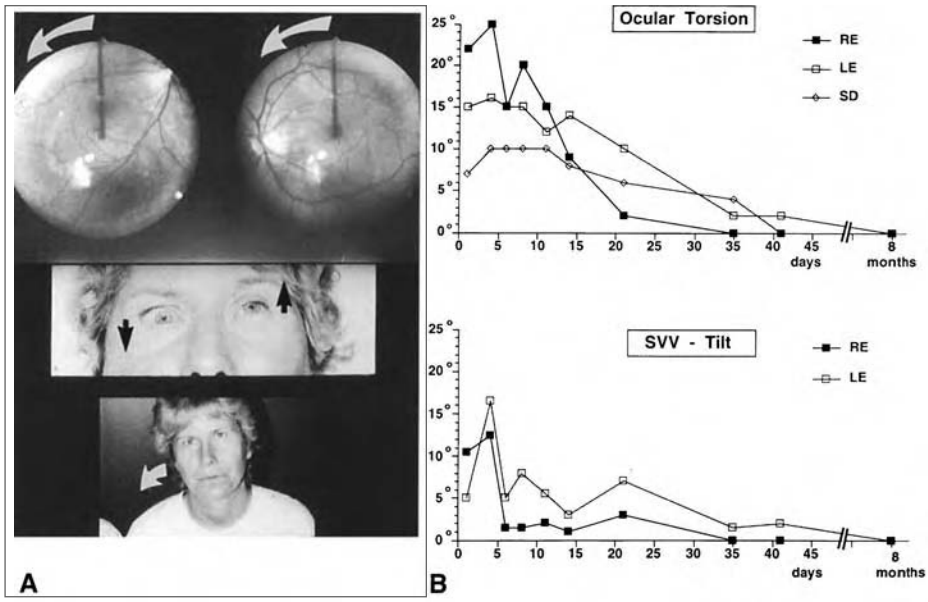


**Figure 25.4** (*A*) Patient with a left paramedian thalamic infarction presenting with a complete ocular tilt reaction (OTR) to the right. OTR consisted of contraversive head tilt of 20 degrees (bottom); skew deviation of 10 degrees, left eye over right eye; and ocular torsion of 15 to 20 degrees (counterclockwise 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.[16]

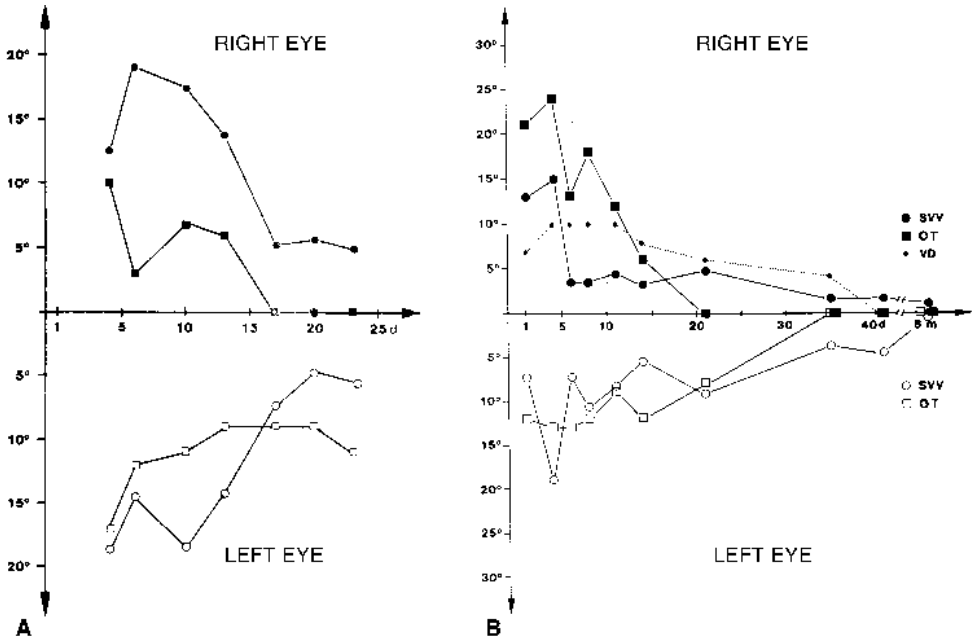


**Figure 25.5** Two representative time courses of deviations of subjective visual vertical (SVV) and ocular torsion (OT) (separate for the left and right eyes) in a patient with Wallenberg's syndrome on the left (*A*) and a patient with unilateral lesion of the region of the interstitial nucleus of 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 SV V 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]

Paroxysmal OTR in MS has been treated effectively with carbamazepine,[13] and baclofen has been reported to be of some therapeutic benefit in a patient with paroxysmal OTR and brainstem abscess.[14]

### Thalamic and Cortical Astasia Associated with Subjective Visual Vertical Tilts

An association of SVV tilts with falls is also typical for posterolateral (vestibular subnuclei) thalamic lesions. Thalamic astasia[17] 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.[18] According to our experience in some 30 patients with thalamic infarctions, the posterolateral type may cause contraversive or ipsiversive postural instability with SVV tilts, whereas the paramedian type (if it extends into the rostral midbrain) always causes contraversive falls. Masdeu and colleagues[19] have described astasia and gait failure with damage of the pontomesencephalic (locomotor) region. Although not discussed, it could also be explained in part by a vestibular 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,[20] lesions of the posterolateral thalamus cause an interruption of the vestibular pathways to the temporoparietal vestibular cortex areas of the affected side and also to the contralateral side.

Of 31 patients with cortical infarctions of the middle cerebral artery territory in one study, 21 showed significant, mostly contraversive, pathological SVV tilts.[21] The overlapping area of these infarctions centered on the posterior insula, which, according to functional imaging studies,[22–26] is homologous to the parietoinsular vestibular cortex in monkeys, and the adjacent superior temporal lobe.[27,28] 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, 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[29] is not surprising if one considers how these functions are corroborated. Our studies on OTR, lateropulsion, and SVV were concerned with static effects of vestibular dysfunction in roll.[3,4] 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.[30,31] 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. 25.2) have been associated with torsional nystagmus: for example, lesions of the vestibular nuclei,[31,32] the lateral medulla,[30,33] in rare cases the MLF (as indicated by an association with internuclear ophthalmoplegia),[34,35] the INC, and the riMLF.[36–38] 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.[38]

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 a (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 or metabolic disorders, 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.[39]

Clinically, DBN occurs more commonly than UBN and is often permanent (as in Arnold-Chiari malformation), 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). DBN and UBN can result from different pathomechanisms: UBN from a perhaps-vestibular tone imbalance of the VOR in pitch and a perhaps-non-vestibular one by a spontaneous downward ocular drift, and DBN from an asymmetry in the distribution of on-directions of vertical gaze velocity in the Purkinje cells.[40] Transitions between UBN and DBN have been frequently described in paramedian pontomedullary lesions. UBN occurs with paramedian plaques of the pontomesencephalic brainstem in MS, cerebellar degeneration, or drug intoxication. 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 25-1 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.[41] DBN is present in darkness as well as 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.[42,43] DBN may be episodic in Arnold-Chiari malformation[44] or paroxysmal.[32]

It has been reported that reversals from DBN to UBN can be provoked by upward gaze deviation,[45] convergence,[46] or transitions from sitting to supine position.[47] 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.[46,48,49]

Patients complain of a distressing illusory oscillation of the visual scene (oscillopsia) and postural imbalance. Both are obligatory but hitherto poorly studied symptoms of the syndrome. The retinal slip in DBN is misinterpreted as motion of the visual scene because the involuntary ocular movements that override fixation are not associated with an appropriate efference-copy signal. Oscillopsia is a permanent symptom, but the illusory motion is less than would be expected from the amplitude of the nystagmus; it increases with increasing amplitude; the mean ratio between the two is 0.37.[50]

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.[50,51] 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.

In spite of many clinical reports of DBN and UBN and multiple hypotheses about possible mechanisms, the pathophysiology is still not understood.[40,52,53] In the light of several clinical findings and experimental data, a general concept is that asymmetries in the cerebellobrainstem network that normally stabilize vertical gaze could lead to an imbalance in structures such as (1) the vertical cerebello-vestibular "neural integrator," (2) the

Box 25-1

## 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 due to 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**

The two most common causes are cerebellar ectopia and cerebellar degeneration, including alcoholic cerebellar degeneration

Other conditions: drugs (phenytoin, carbamazepine, lithium), multiple sclerosis, tumor, hematoma, vascular disease, encephalitis, magnesium depletion, vitamin $B_{12}$ deficiency (see Table 25-1)

**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

central connections of the vertical VOR, including both the SCC and the otolith responses, or (3) the vertical smooth pursuit system. In a review by Pierrot-Deseilligny and Milea,[54] DBN is explained by a floccular lesion that results in a disinhibition of the pathway from the superior vestibular nucleus via the ventral tegmental tract and thereby in a relative hyperactivity of the elevator muscles, inducing an upward slow phase.

## Etiology

The two most common causes of a downbeat nystagmus/vertigo syndrome are cerebellar ectopia (25%) (e.g., Arnold-Chiari malformation) and cerebellar degeneration (25%) (e.g., olivopontocerebellar degeneration). A further 10% to 20% of patients have a variety of conditions,[33,55] and in about 30%, an unequivocal diagnosis of the cause cannot be established.

In cerebellar degeneration and drug-induced DBN, an asymmetrical vestibulocerebellar disinhibition of the "Purkinje cell activity" of the flocculus on the vertical canal reflexes may be causative, evidence for which was found in a 2005 imaging study.[56] Nutritional cerebellar syndromes due to thiamine deficiency, in particular alcoholic cerebellar degeneration (see Box 25-1), not only cause a typical 3-Hz fore-aft oscillation of body sway[57] but also DBN.[55,58,59] Antiepileptic drugs, especially phenytoin[60] and carbamazepine,[61,62] can produce a reversible DBN with associated cerebellar signs, depending on the dosage of the drugs. Other causes are lithium toxicity,[55,63–65] felbamate intoxication,[66] and toluene abuse.[67] Severe magnesium depletion[68] and

vitamin $B_{12}$ deficiency[69] have also been reported to result in DBN.

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 25-1). Sometimes DBN is lithium-induced in preexisting Arnold-Chiari malformation[75] or is caused as an intermittent syndrome by head tilt due to vertebral artery compression[76] or by a vermian arachnoid cyst with associated obstructive hydrocephalus.[77] DBN is rare in children. It may be hereditary congenital[78] as a persisting syndrome or may occur during infancy and resolve naturally.[79]

## Management

DBN due to drugs, magnesium depletion, or vitamin $B_{12}$ deficiency is usually reversible when the intoxication or metabolic deficiency is reversed. DBN due to structural lesions in the posterior fossa is usually permanent, although a surgical suboccipital decompression in Arnold-Chiari malformation to relieve the compression of the herniating cerebellum against the caudal brainstem may lead to gradual improvement of some of the distressing symptoms.[80] Suboccipital craniotomy,[81] transoral removal of the odontoid process in the basilar impression[82] and of an osteophyte compressing the vertebral artery,[76] and surgical decompression of a syringomyelic cyst in the medulla[77,83] were able to resolve the syndrome only in individual patients. For one patient, base-out prisms were added to both spectacle lenses, because the convergence both dampened the nystagmus and decreased the oscillopsia.[42] In patients with DBN[85] and a stiff-person syndrome with glutamic acid decarboxylase antibodies[84] intravenous immunoglobulin was found to improve the DBN and the ataxia.

Target symptoms for symptomatic medical treatment are distressing oscillopsia and reduced visual acuity due to the fixation nystagmus. Postural imbalance is less prominent and less distressing. Some studies have demonstrated effects on the DBN based on the pathomechanism that spontaneous upward ocular drift and downbeat nystagmus are due to an asymmetry in the distribution of on-directions of vertical gaze velocity in the Purkinje cells.[40,85] Because the inhibitory influence of gamma-aminobutyric acid (GABA)–ergic Purkinje cells is assumed to be impaired in DBN, several agents that act on this receptor have been investigated. The GABA-A agonist clonazepam was found to improve DBN, but the studies reporting this finding were not controlled.[86,87] The GABA-A agonist baclofen is assumed to reduce DBN,[88] but only one of six patients had a response to baclofen in a double-blind crossover trial involving only a few patients.[89] Further, in the same trial, the alpha-2-delta calcium channel antagonist gabapentin was assumed to have a positive effect on DBN, but only one of six patients showed improvement.[89] Another study found that muscarinic antagonists, in particular the anticholinergic drug scopolamine, reduced nystagmus in five patients with acquired pendular nystagmus and in two patients with downbeat nystagmus; benztropine was less effective.[90]

On the basis of this assumed pathomechanism, the effects of aminopyridines were evaluated in patients with DBN and in a subsequent study on UBN. In 17 patients with DBN, the mean peak slow-phase velocity of DBN was measured before and 30 minutes after randomized oral ingestion of 20 mg of 3,4-diaminopyridine or placebo.[91,92] The mean peak slow-phase velocity decreased in 10 of 17 patients by more than 50%. Moreover, an improvement of postural imbalance could be found.[93] The assumed underlying mechanism is that aminopyridines increase the activity and excitability of the Purkinje cells— as found in animal experiments[94]—thereby augmenting the physiological inhibitory influence of the vestibulocerebellum on the vestibular nuclei. 4-Aminopyridine improves not only DBN but also smooth pursuit and the gain of the vertical VOR.[92,95]

## *Upbeat Nystagmus (Upbeat Nystagmus Syndrome)*

Box 25-2 summarizes the information given in this chapter about the upbeat nystagmus syndrome.

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[39] or a downward ocular drift by asymmetrical smooth pursuit commands.[95] 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. 25.6) 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

Box 25-2

## 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 due to 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**

Brainstem tumors, infarction, hematoma, cavernoma, multiple sclerosis, encephalitis, abscess, alcoholic degeneration (Wernicke's encephalopathy), drug intoxication, nicotine (see Table 25-1)

**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

vertical cerebello-vestibular "neural integrator," (2) the central connections of the vertical VOR, or (3) signals of the vertical smooth-pursuit system. UBM due to 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 to 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 0.145%.[96] The etiology of UBN is in general similar to that of DBN (see Box 25-2). 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[46,97] and in single cases of Fisher's syndrome,[98] central diabetes insipidus,[99] and Pelizaeus-Merzbacher disease,[100] and has even been associated with middle ear disease.[101] We have seen transient upbeat nystagmus associated with various intoxications, for example with antiepileptic drugs. UBN can on rare occasions be congenital.[102]

## Management

UBN may be associated with severe vertigo, ataxia, and nausea, particularly at first. Affected patients may require vestibular sedatives (e.g., dimenhydrinate or scopo-

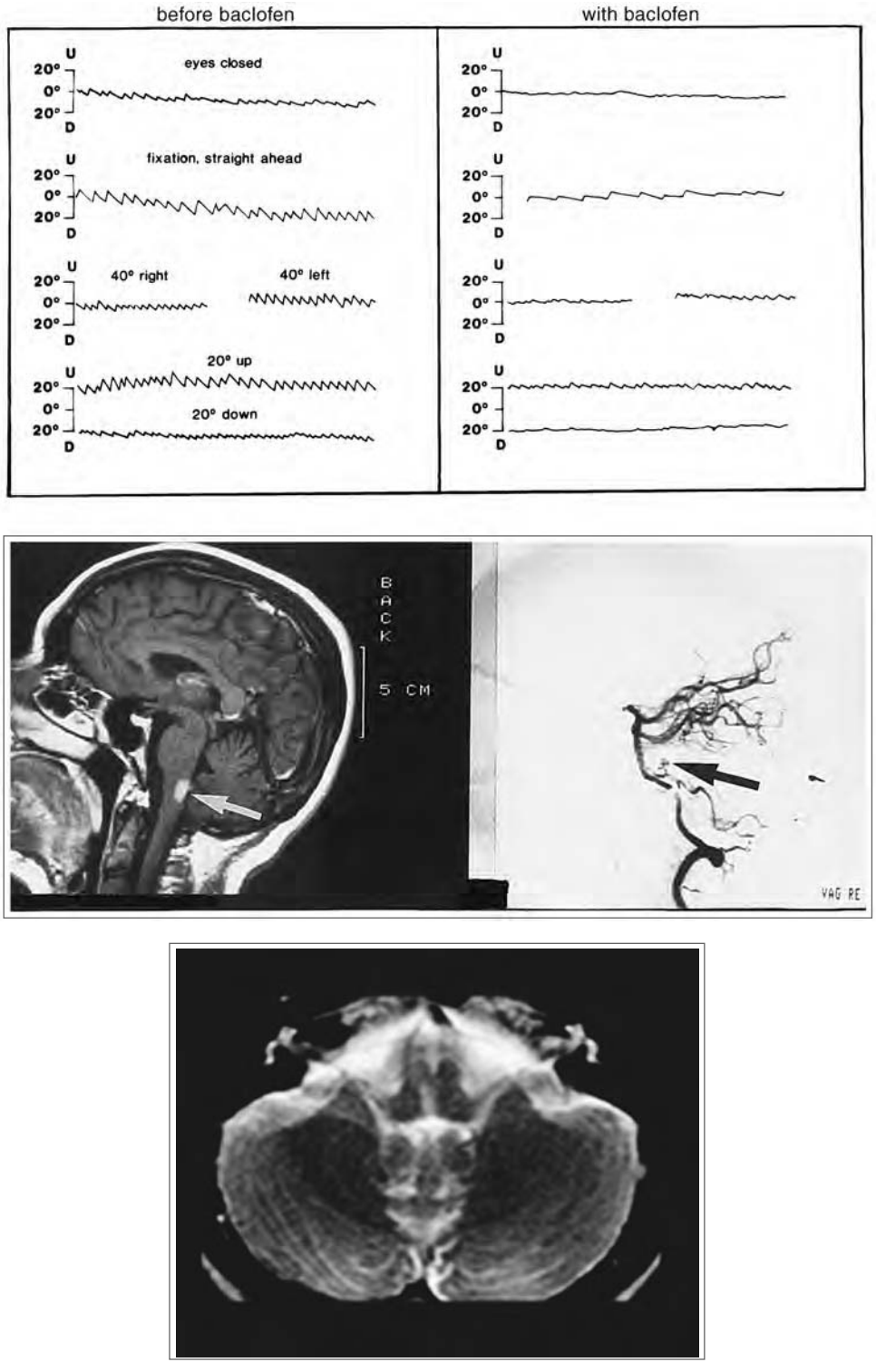


**Figure 25.6** Partial suppression of upbeat nystagmus by medical treatment with baclofen. (*Top*) Vertical electronystagmography recordings. (*Bottom*) Magnetic resonance image of a patient with unilateral upbeat nystagmus and a paramedian medullary infarction affecting the area of PMT neurons near the neurons of the perihypoglossal nuclei.

lamine) 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 (Fig. 25.6).[88] Carbamazepine was found to be effective in a single case of upbeat nystagmus due to multiple sclerosis.[103] In a single patient with UBN, Glasauer and colleagues[95] 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.[85]

## 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 due to imbalance in the vertical 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. 25.2). The larger extent of the latter area is due to 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. 25.2). There is a multisensory convergence within the parallel neural network of the vestibular nuclei,[104] 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.[105]

Some of the cases described as central variants of vestibular neuritis[106–110] 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 due to occlusion of the anterior inferior cerebellar artery (which may also supply the rostral vestibular nuclei) mimic vestibular neuritis.[106,111] The other main cause of confusion with disorders at the entry zone of the VIIIth nerve is multiple sclerosis.[108]

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 ("torsipulsion").[30,112]

Sometimes the clinical manifestation of a particular ocular motor abnormality such as OTR allows one to identify the SCC pathway affected (anterior or posterior). The different presentation of OTR in Wallenberg's syndrome with monocular dissociation of ocular torsion—either excyclotropia of the lower eye or incyclotropia of the upper eye—indicates involvement of the posterior or the anterior SCC pathways.[113] Lesions of the vestibular nuclei may also result in repetitive multidirectional paroxysmal nystagmus and vertigo, which have been reported to respond to treatment with carbamazepine.[44] The differential effects of a medullary lesion involving more than one VOR plane are reflected not only in the direction of eye movements but also in the preferred direction of increased body sway. Body sway histograms as measured by posturography are primarily diagonal in patients with Wallenberg's syndrome and moderate body lateropulsion (combination: roll and yaw) but primarily lateral in patients with severe body lateropulsion (roll

greater than yaw).[113] The involvement of roll and yaw planes is also reflected by the close correlation between the tilt of perceived visual vertical and the severity of body lateropulsion, both indicators—perceptual and postural—of a vestibular imbalance in roll.[114]

## 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.

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,[115–117] area 3aV (neck, trunk, and vestibular region of area 3a) in the central sulcus,[118] PIVC at the posterior end of the insula and retroinsular regions, [27,28] the periarcuate cortex area 6 pa,[119] area 7 in the inferior parietal lobule,[120] and the ventral intraparietal area (VIP) in the fundus of the intraparietal sulcus.[121–124] 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 extrapersonal space[5] but also of that generated when stationary human subjects perform optokinetic nystagmus.[125,126] 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.[5,127,128] 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) [20,25,119] and functional magnetic resonance imaging (fMRI).[22–24,26,125] 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, the parts of the inferior parietal lobule representing area 7 in the 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. 25.7). Simultaneous with these activations, deactivations of areas within the visual and somatosensory systems of both hemispheres were observed.[22,130] 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,[125,131] a reciprocal inhibitory cortical interaction between the sensory systems was assumed.[131]

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.[25] The determinants were (1) the subjects' 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 fast phase of vestibular caloric nystagmus.[20,25,132] Furthermore, in right-handed healthy volunteers, who performed allocentric visuospatial judgments (line bisection) with and without galvanic stimulation of the right or left vestibular nerve, the most relevant cortical area for the processing of vestibular information was located in the posterior insula bilaterally, right significantly more than left, including the PIVC.[133]

### *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

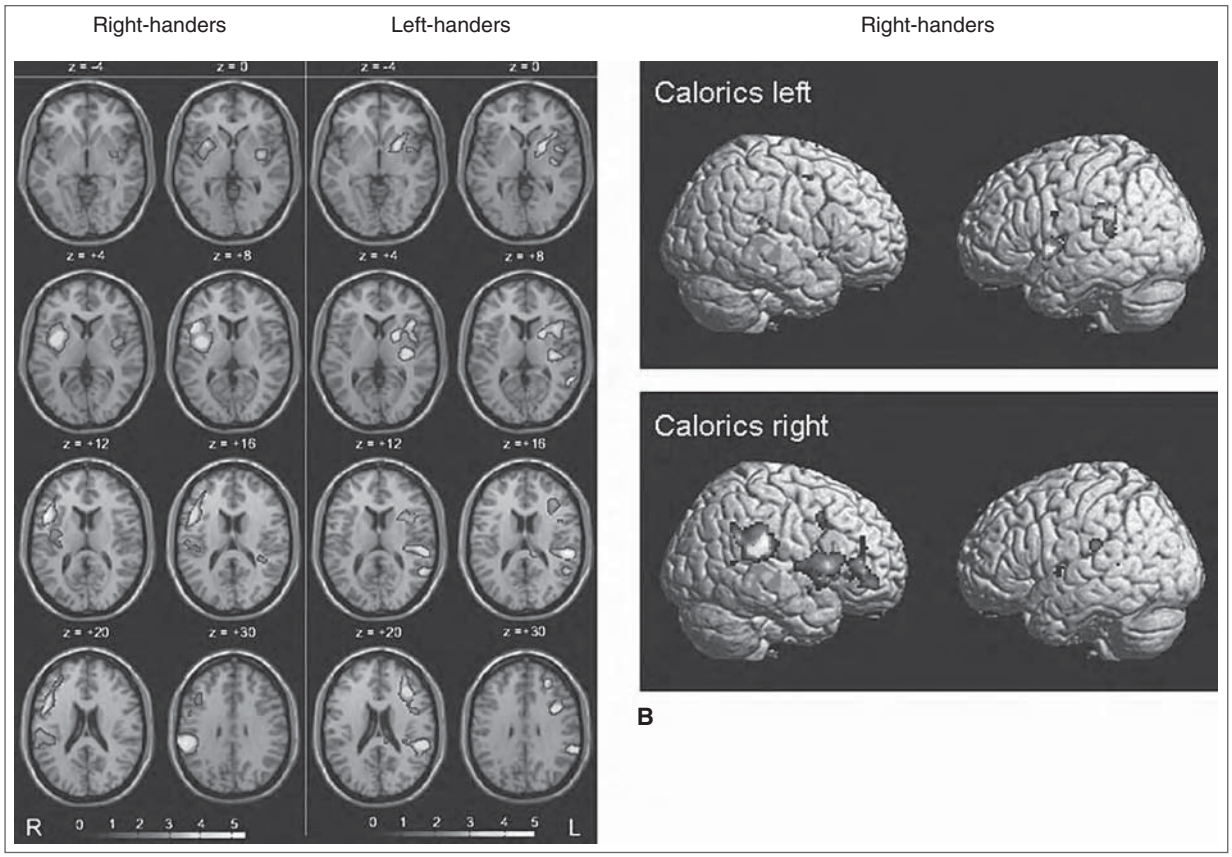


**Figure 25.7** (*A*) Areas activated during caloric stimulation (warm water at 44°C) of the right ear in right-handed, and of the left ear in left-handed, healthy volunteers (group analysis ; n = 12; P < .001; $^{15}O$-labeled $H_2O$ bolus, positron emission tomography). Activations were located in 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,[25] and Bense et al, 2003 [186]).

and head positions in order to deliver reliable input for adequate ocular motor and motor exploration of space.

Nature seems to have solved this impossible 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.[134,135] 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.[136] This function is most probably subserved by the temporal and posterior parietal cortex, a lesion of which produces a visuospatial hemineglect.

Karnath and associates[137,138] 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.[139]

In patients, an inappropriate vestibular input due to 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. 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 temporoinsular, inferior parietal, or frontal lobe lesions
- Vertical neglect below the horizontal meridian in bilateral parieto-occipital lesions
- Tilts of perceived vertical (mostly contraversive) and body lateropulsion in unilateral temporoinsular (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 cortex.

## 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 of the right hemisphere[140] and occurs less commonly with acute right or left lesions of the frontal premotor cortex.[141] 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.[142] 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.[143,144] Mesulam[145] 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)[140,146,147] as well as the inferior frontal gyrus[141,148] have been observed to correlate with spatial neglect. In addition, four studies have found the right superior temporal cortex and the right insula to be critically related to spatial neglect.[149–152]

Studies showing that vestibular (caloric) stimulation significantly improved spatial functioning have stressed the important role of the vestibular system in neglect.[153,154] 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.[136] 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[137,138] 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.[155] Spatial hemineglect also includes the back space of the body.[156]

## 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[157–159]; 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,[160] 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,[161,162] 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.[163] These sensations were similar to those induced by stimulation of the vestibular cortex.[157,158]

Vertigo has long been considered a manifestation of epileptic auras.[164,165] Most information on auras (vestibular epilepsy), including case descriptions, comes from older textbooks, for example, those by Bumke and Foerster[166] and Penfield and Jasper,[158] or from review articles.[167,168]

In a later study searching for the human representation of "vestibular cortex," Kahane and associates[169] 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[169] thus confirmed earlier findings by Penfield and coworkers,[158,170,171] 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.[172]

### Management

Vestibular seizures respond to antiepileptics. First-line drugs are gabapentin, sodium valproate, and lamotrigine.[173] If necessary and possible, surgical procedures may be considered.[174]

## *Paroxysmal Central Vertigo*

Nonepileptic paroxysmal vertigo or other vestibular syndromes may result from pathological excitation of various vestibular structures.[175] Most of them occur in multiple sclerosis, but others may be associated with a brainstem abscess (paroxysmal OTR[14]) or an arteriovenous malformation with previous bleeding (repetitive paroxysmal nystagmus and vertigo[32]) or brainstem infarction.

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

- Paroxysmal dysarthria, ataxia, and vertigo[176]
- Paroxysmal OTR[13]
- Paroxysmal room-tilt illusion[177]

### 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.[178–180] It has been attributed to muscle hypotonia or neglect and has also been observed after thalamic infarctions[181] and hemorrhages.[18,182] Masdeu and Gorelick[17] described 15 patients with "thalamic astasia," in the absence of motor weakness, sensory loss, and cerebellar signs, due to 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."[17] 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,[16,20] thalamic astasia and tilts of perceived vertical may both reflect a vestibular tone imbalance. Furthermore, what Masdeu and colleagues[19] 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[183] or hemisensory loss and hemiparesis.[184] The lesions involve the ventral lateral nucleus of the thalamus and the adjacent posterior limb of the internal capsule and the mid- to posterior thal-

amus containing the dentatorubrothalamic and ascending pathways.[183,184]

### 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.[9,113,185] 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.[15,113]

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.[15,113] This hypothesis could explain why patients fall without vertigo or warning signals from the multisensory spatial orientation system. 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,[21] making it most likely that the cortical lateropulsion is also due to a vestibular tone imbalance in the roll plane.

Lateropulsion in both dorsolateral medullary lesions and posterior insular lesions spontaneously recovers within days to weeks.[113] 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.

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CHAPTER 26

# Non-vestibular Dizziness and Imbalance: From Disuse Disequilibrium to Central Degenerative Disorders

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*Non-vestibular imbalance*, defined as imbalance that is not due to an inner ear or vestibular nerve disorder, 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 imbalance that respond to rehabilitation. Disuse dysequilibrium 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 dysequilibrium and therefore respond to physical therapy (PT). Some individuals with these disorders do not have disuse dysequilibrium 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.

It is best to have any patient with chronic imbalance first assessed by a physician in order 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 for four visits. At each visit, outcomes scores are re-assessed. The patient is then reevaluated by the physician with the physical therapist present, 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 useful in patients who have progressive problems (spinocerebellar degeneration, progressive peripheral neuropathy, and progressive supranuclear palsy).

## Disuse Disequilibrium and Fear of Fall

### *Description*

In elderly individuals, there is progressive decline in muscle bulk, joint range of motion, and reflex time.[1] 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 dysequilibrium.[2] Fear of fall can occur as a result of disuse dysequilibrium and can also exacerbate disuse dysequilibrium by reducing the patient's willingness to participate in a home exercise program.[3]

Figure 26.1 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 patient. The *squares* indicate the percentage of patients seen within each decade with fear of fall (FOF) and disuse dysequilibrium (DD) (*right axis*).

### Useful Outcome Tests

Imbalance in individuals with disuse disequilibrium and fear of fall readily responds to gait and balance PT. Outcome measurements that are useful in patients with disuse dysequilibrium are shown in Table 26–1. In each column of this table, the PT fills in the date and the results of each of the outcome measurements. A full description of these outcome measurements can be found in Chapter 19. They are as follows:

- 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, Dysequilibrium Visual Analogue Scale (VAS)
- Others: Activities of Daily Living (ADLs)

A description of posturography SOT can be found in Chapter 8 (pg 135 and Figure 8.17A).

### Management

A daily home exercise program that increases endurance, balance, and lower extremity strength frequently resolves the problem.[4] Success depends on compliance by the patient and support from the family or friends. Case Study 1 in this chapter describes a patient with disuse dysequilibrium 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.[5–7] In severe cases, patients with these disorders have gait apraxia, gait initiation defects, and severe retropulsion.

The etiology of leukoaraiosis (ischemic white matter disease) is believed to be significant small vessel disease.[8] Age, hypertension, diabetes mallitus, 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 in the corona radiata and sometimes in the pons on T2-weighted magnetic resonance imaging (MRI) of the head.

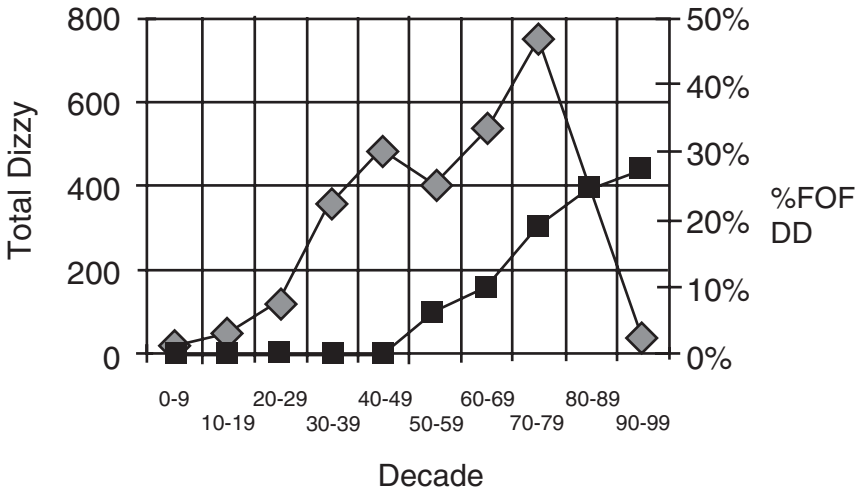


**Figure 26.1** Age distribution of disuse disequilibrium and fear of fall in patients seen in a dizziness and balance clinic. See text for explanation.

## ■ Table 26-1 USEFUL OUTCOME MEASUREMENTS FOR IMBALANCE

| ***Treating P.T.:*** | ***Patient Name:*** | | ***Age:*** | ***Gender:*** | ***Diagnosis:*** | | ***History :*** | |
|---|---|---|---|---|---|---|---|---|
| Date: | | | | | | | | |
| Tinetti Fall Risk Assessment Balance=25 Gait=12 | | | | | | | | |
| 8ft Up & Go Normal=8.5s | | | | | | | | |
| Posturography: Sensory Organization Test (SOT) Composite Score | | | | | | | | |
| Posturography: Number of falls on SOT | | | | | | | | |
| Activities of Daily Living (ADLs) Max=15 | | | | | | | | |
| Assistive device inside/outside | | | | | | | | |
| Activities-Specific Balance Confidence Score (ABC) (nl≥80%) | | | | | | | | |
| Disequilibrium Visual Analogue Scale (0–10, 10 worse) | | | | | | | | |
| Dynamic Gait Index Score (DGI) | | | | | | | | |
| Gait speed (ft/sec) | | | | | | | | |

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.[6] 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. Some patients with NPH may benefit from insertion of a permanent shunt system to drain the hydrocephalus.[9] 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.

## Useful Outcome Scores

Patients with imbalance and cognitive decline should undergo outcome measures similar to those listed in Table 26–1, with additional 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 26–2.

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 Timed Up and Go test and the Walk While Talk 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 (WWT) experienced a subsequent fall.[10] The same prediction is also applicable to nondemented, community-dwelling older adults.[11] 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 in a chair and is instructed to stand up and walk as quickly and as safely as for 3 meters, turn around, walk back to the chair, and sit down. Then the patient is instructed to perform the test while counting backward 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) > 4.5 sec indicates fall risk

In the Walk While Talk test, subjects are timed while they walk at self-selected speed 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): | > 18 s at risk for falls |
| WWT-simple: | > 20 s at risk for falls |
| WWT-complex: | > 33 s at risk for falls |

Table 26–2 lists two 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

Improvement rates in cognition and balance in patients with NPH after insertion of a shunt vary from 65% to 77%.[12] Unfortunately, there is no standard for outcome assessment, and randomized, prospective clinical trials are lacking.

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

■ Table 26-2 **OUTCOME MEASURES USED IN PATIENTS WITH IMBALANCE AND DEMENTIA**

***Pt Name: Age: Gender: Diagnosis: History :***

| | Pre-drain Date | 2hrs post-drain Date |
|---|---|---|
| Mini-Mental State Exam (MMSE) | | |
| Times gait task (TUG) | | |
| TUG cognitive | | |
| TUG manual | | |
| diff TUG (TUG manual–TUG) | | |
| Average gait speed (ft/s) | | |
| Walk While Talk (WWT) (40 ft) | | |
| WWT-simple | | |
| WWT-complex | | |
| Fall history | | |
| Dynamic Gait Index (DGI) | | |
| Posturography: SOT composite score | | |
| Retropulsion on Clinical Exam | | |

cognitive and balance decline.[13] 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 2 describes a patient with NPH, and Case Study 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 due to 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 retropul sion test, the examiner has the patient stand with feet slightly apart and instructs the patient to take no more than one step backward 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 backward 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 due to 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 very heterogeneous group of disorders that can affect different

portions of the cerebellum and structures outside the cerebellum, including peripheral nerve and brainstem structures. 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) owing to a defect in the cerebellar vermis; this inability can cause greater motion sensitivity during head movements.

## Useful Outcome Scores

Most patients with the neurological disorders discussed here do not have cognitive decline at onset, so we usually use the outcome measures listed in Table 26–1. Patients with PSP and some with Parkinson's disease demonstrate cognitive decline as the disease progresses, so the outcome measures in Table 26–2 then become more relevant.

## Management

Patients with these degenerative disorders often need a longer course of treatment than those who have disuse dysequilibrium. 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 dysequilibrium and require PT.

There is a treatment overlap between patients with PSP and those with Parkinson's disease. A 4-week trial of low-dose Sinemet should be tried to improve gait and balance in patients with PSP, but most cases do not respond to medication. There is no surgical treatment for this disorder. Gait and balance PT should be prescribed. Patients undergoing PT often need a rollator to prevent backward falls.

Patients with proprioception defects due to peripheral neuropathy or dorsal root/dorsal column disease are encouraged to use visual cues while standing and walking.

Patients with SCA and disuse disequilibrium or VOR defects have a good response to PT. Some patients with VOR cancellation defects do well with habituation exercises. Case Study 4 describes a patient with SCA.

## CASE STUDY 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. 26.2). This assessment is excellent for patients at risk for fall.[14]

*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.

## TINETTI FALL RISK ASSESSMENT

**Patient MT 64-57-08** **Date 03/26/99**

| | |
|---|---|
| Sitting Balance | (2) Steady, stable<br>(1) Holds onto chair to keep upright<br>(0) Leans, slides down in chair |
| Arising from chair | (2) Steady without holding on<br>(1) Uses arms of chair<br>(0) Unable without help or multiple attempts |
| Immediate standing balance | (2) Steady w/o support<br>(1) Steady, w/ support<br>(0) Unsteady (grabbing moves feet, etc) |
| Standing | (2) Steady w/ feet together<br>(1) Steady, feet apart<br>(0) Unsteady or holds on |
| Balance w/eyes closed | (2) Steady, feet together<br>(1) Steady, feet apart<br>(0) Unsteady; holds on |
| Sternal nudge light pressure 3 times | (2) Steady<br>(1) Moves feet by keeps balance<br>(0) Begins to fall |
| Turning (360) | (2) Continuous steps no grabbing or staggering<br>(1) Discontinuous, puts foot down completely before raising the other<br>(0) Unsteady or holds on |
| Neck turning | (2) Horizontal and vertical (at ceiling), steady<br>(1) Decreased ability but no unsteadiness or pain or dizziness<br>(0) Unsteady or is symptomatic |
| One-legged stance, eo | (1) Able 5 sec w/o holding on<br>(0) Unable |
| Back extension ask to lean backwards | (2) Good extensions w'o holding on, staggering<br>(1) Tries but decreased ROM or holds on<br>(0) Will not attempt or staggers |
| Reaching up high | (2) Able to take down object w/o holding on or becoming unsteady<br>(1) Able to reach but needs to hold on<br>(0) Unable or unsteady |
| Bending down | (2) Able, single attempt, doesn't hold on<br>(1) Able, single attempt but holds on<br>(0) Unable or multiple attempts |
| Sitting down | (2) Able, one smooth motion<br>(1) Needs to use arms, not smooth<br>(0) Falls into chair, misjudges distance |
| **Gait** | |
| Initiation of gait | (1) Begins immediately, single smooth motion<br>(0) Hesitates, multiple attempts, not smooth |
| Step height | (1) Completely clears, 1-2 inches, right<br>(1) Completely clears, 1-2 inches, left<br>(0) Does not clear, right<br>(0) Does not clear, left |
| Step length | (1) Right foot passes left foot by foot length<br>(0) Right foot does not pass left by full foot length<br>(1) Left foot passes right foot by foot length<br>(0) Left foot does not pass right by full foot length |
| Step symmetry | (1) Same or nearly same on both sides<br>(0) Varies, or advances with same foot on every step |
| Step continuity | (1) No breaks or stops in stride<br>(0) Stops between steps, step length varies |
| Path deviation | (1) Foot follows straight line<br>(0) Foot deviates side to side or in one direction |
| Trunk stability | (2) Normal<br>(1) Knees or back flexed, arms not abducted to assist<br>(0) Marked sway |
| Walk stance | (1) Normal base of support<br>(0) Widened base of support |
| Turning around while walking | (1) Normal, continuous<br>(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 26.2** Example of the use of the Tinetti Fall Risk Assessment.

## CASE STUDY 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. 26.3). No obstruction of CSF flow was found in any of the ventricles, consistent with a diagnosis of communicating hydrocephalus. Table 26–3 lists the outcome measures as discussed in Table 26–2 in the text. The left column of Table 26–3 lists the measure and the normal range, column two (Pre LP) lists the measure before a lumbar puncture, and column three (2 Hrs LP) lists the measures after removal of 40 cc of CSF by lumbar puncture. For example, a mini-mental (MMSE) of <24 is considered cognitively impaired. This test was 19 in Mr. G before the LP was 19 and after the LP was 21. Therefore, his cognition did improve after the LP.

*Comment*

On the basis of history, lumbar puncture, and head MRI findings, this patient has NPH. His outcome scores before the LP 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, 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.

■ Table 26-3 **OUTCOME SCORES FOR A PATIENT WITH NORMAL-PRESSURE HYDROCEPHALUS AFTER LARGE-VOLUME CSF DRAINAGE PROCEDURE (CASE STUDY 2)**

| Pt Name: Mr. G<br>Age: 85 Gender: M Dx:NPH | **Pre Drain<br>Date: 03/31/06** | **2 Hrs Post-Drain<br>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 + 2SD) | 17.83 | 17.30 |
| diff TUG (TUG manual – TUG)<br>(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 |
| 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 |

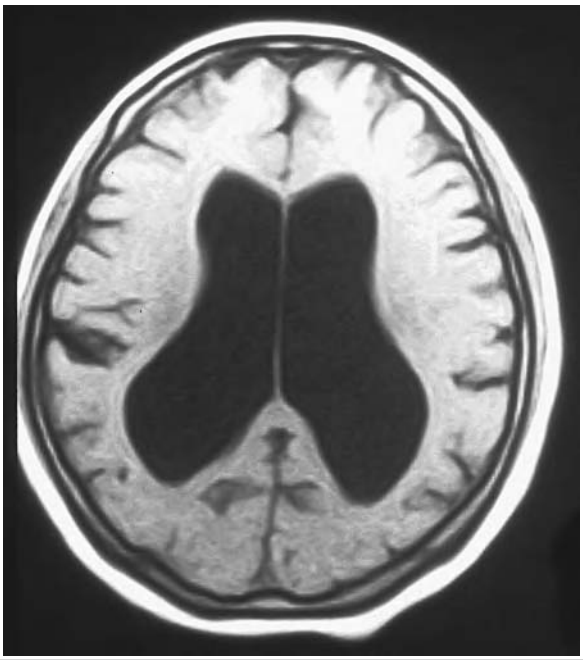

**Figure 26.3** Magnetic resonance image of the head in the horizontal plane illustrating hydrocephalus.

## CASE STUDY 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. 26.4). 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.

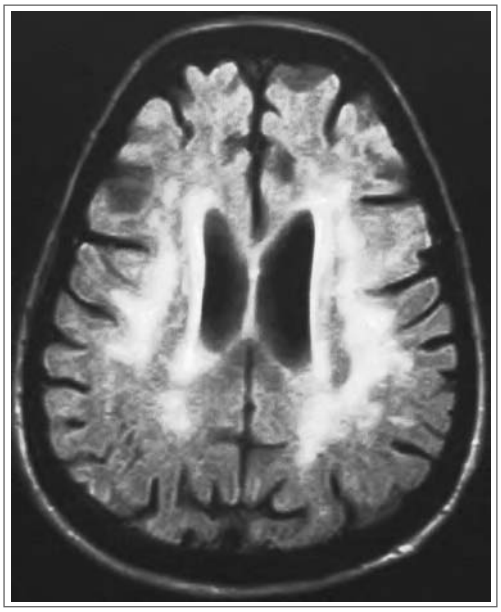

**Figure 26.4** Magnetic resonance image of the head in the horizontal plane showing extensive white matter ischemic changes otherwise known as leukoaraiosis.

## CASE STUDY 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. 26.5). 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 is 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 as well as 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 *X*1viewing (see Chapter 20) while sitting, moving her head both horizontally and vertically and using both near and distant targets She performed these 3–5 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 26–4 shows the improvement in outcome scores with PT for Ms. P. This table is the same as Table 26–1. See page 432 for description.

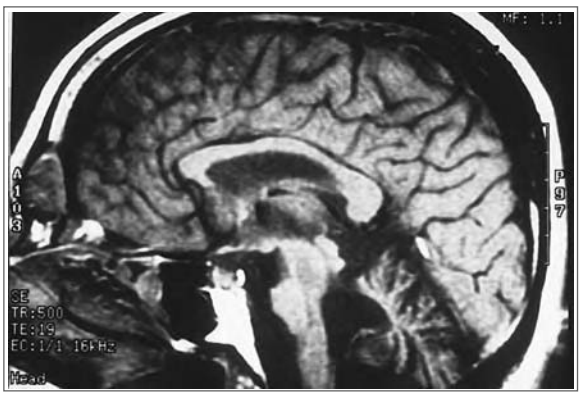


**Figure 26.5** Magnetic resonance image of the head in the sagittal plane in a patient with spinocerebellar atrophy type 3.

■ Table 26-4 **OUTCOME SCORES IN A PATIENT WITH CEREBELLAR ATAXIA (CASE STUDY 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 |

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CHAPTER 27

# Assessment and Management of the Patient with Traumatic Brain Injury and Vestibular Dysfunction

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This chapter discusses the array of vestibular disorders associated with traumatic brain injury (TBI). Many of these vestibular deficits occur spontaneously or in association with viral or vascular lesions that have been discussed in other chapters. Because of the confounding influence of TBI, however, we assess and treat patients with it differently from patients with peripheral vestibular system pathology alone. TBI also affects ultimate recovery from vestibular deficits. What distinguishes the patient with TBI from other patients with peripheral vestibular disease is the mechanism of vestibular injury and the high incidence of other neurological deficits complicating the recovery process.

Recognizing and treating symptoms of vestibular dysfunction, including dizziness and imbalance, and distinguishing the etiology of those symptoms from other non-vestibular causes are essential parts of TBI rehabilitation. Many therapists involved in rehabilitating patients with TBI are familiar with approaches to treating imbalance but lack strategies to treat complaints of dizziness. As a result, therapists often try to avoid provoking dizziness when treating patients with TBI. However, avoiding the movements that provoke dizziness can actually delay recovery from vestibular system dysfunction, resulting in persisting symptoms.[1,2] Persisting symptoms complicate recovery and contribute to long-term loss of functional independence.[3–5]

The objectives of this chapter are to discuss mechanisms of traumatic injury producing peripheral vestibular system disorders in TBI. Strategies for assessing and treating symptoms of peripheral vestibular system dysfunction are reviewed. In addition, the chapter discusses the effect of other traumatically induced sensory, motor, and cognitive deficits on strategies for treating the patient with TBI and associated vestibular system pathology.

## Vestibular Pathology

As many as 30% to 65% of patients with TBI suffer symptoms of traumatic vestibular pathology at some point during their recovery.[4–9] Symptoms of peripheral vestibular system dysfunction can include vertigo, eye-head dyscoordination affecting the ability to stabilize gaze during head movements, and imbalance affecting the ability to maintain stability when standing and walking.[10] The specific constellation of symptoms found in individual

patients varies according to the type and extent of injuries to both vestibular and central neural structures. Understanding peripheral vestibular pathology and its effect on gaze, postural, and perceptual functions is essential because treatment varies with the patient's symptoms and diagnosis.[11]

## Mechanism of Injury and Resulting Vestibular Disorders

Trauma can induce vestibular-related symptoms through damage to the inner ear itself, to the vestibular nerve, or to central structures and pathways (Table 27-1). These mechanisms, discussed in detail in Chapter 6, are summarized briefly here. Many of the vestibular deficits associated with TBI also occur spontaneously or are associated with viral or vascular lesions that have been discussed in other chapters. Other deficits are specific sequelae of the head trauma itself. The actual mechanism of labyrinthine damage may be the concussive force itself, rupture of the membranous labyrinth, or hemorrhage. Temporal bone fracture can disrupt the bony or membranous labyrinth or the vestibular nerve. Contusions and hemorrhage can damage the brainstem, cerebellum, or cerebral hemispheres. There can be pain and dysfunction of the neck resulting in dizziness (see Chapter 29). Finally, a variety of psychological responses may cause or increase dizziness and may contribute to a person's level or restriction of participation.

### *Concussion*

Inner ear concussion injury is the most common vestibular sequela of TBI.[8,12,13] Symptoms of concussive-type VIIIth cranial nerve or labyrinthine injury can include high-frequency sensorineural hearing loss, benign paroxysmal positional nystagmus/ vertigo (BPPN/V), postural dyscontrol, and gait ataxia. BPPV in TBI is believed to occur because of the intense acceleration of the utricular otolithic membrane, which results in displacement of otoconia to the posterior semicircular canal. Displaced otoconia, adhering to the cupula or free-floating in the long arm of the posterior canal, may result in displacement of the cupula in response to gravity in specific posi-

■ Table 27-1 **SITES AND MECHANISMS OF HEAD TRAUMA–INDUCED "DIZZINESS"**

| Site | Syndrome | Mechanism |
|---|---|---|
| Inner ear | Benign paroxysmal positional vertigo | Dislodging of otoliths, cupulolithiasis or canalithiasis |
| | Post-traumatic endolymphatic hydrops | Decreased endolymphatic flow |
| | Perilymphatic fistula | Rupture of round or oval window or of membranous labyrinth |
| | Labyrinthine concussion | Endolymphatic hemorrhage |
| | Temporal bone fracture | Disrupts bony or membranous labyrinth |
| Vestibular nerve | Temporal bone fracture | Disruption of VIIIth cranial nerve |
| Brainstem or vestibulocerebellum | Downbeat, upbeat, and torsional nystagmus<br>Central positional vertigo<br>Ocular tilt response<br>Postconcussive syndrome | Contusion or hemorrhage of brainstem or cerebellum |
| Cerebral cortex | Tornado epilepsy | Post-traumatic seizures |
| Neck | "Whiplash" | Flexion-extension injury |
| Psychological | Panic disorder; chronic anxiety<br>Depression<br>Somatization<br>Compensation neurosis | Psychogenic |

tions.[14] BPPV produces a transient positional nystagmus and vertigo in a characteristic head-dependent position. In addition, patients with BPPV complain of transient vertigo when lying down and rolling over to the affected side, or when looking up or down (see Chapter 17).

Changes in intracranial pressure can produce ruptures in the round or oval window and a perilymph fistula (PLF) between the middle and inner ear.[8,11,15–17] PLF can result in fluctuating hearing loss, episodic vertigo, and gait and balance disturbances. In addition, patients with PLF may have a number of cognitive symptoms, including memory, concentration, and attention deficits.[18] The incidence of PLF in patients with head injury is uncertain.[16,18–20]

Vascular injuries, including hemorrhage into the membranous labyrinth, can injure the endolymphatic system, producing a post-traumatic hydrops or Ménière's disease–type syndrome, with corresponding symptoms of tinnitus, hearing loss, vertigo, and imbalance.[8,11]

### *Fractures*

Traumatic fractures of the temporal bone can produce unilateral or bilateral vestibular hypofunction. Longitudinal fractures account for approximately 80% of all temporal bone fractures and are associated primarily with blows to the parietal and temporal regions of the skull.[12,21] Anatomic damage associated with longitudinal fractures primarily occurs to middle ear structures, leading to conductive hearing loss. Associated vestibular symptoms are considered secondary to a concussive injury to the membranous labyrinth.

Transverse fractures account for approximately 20% of all temporal bone fractures and are reported to result most frequently from blows to the occiput.[12,21] Transverse fractures of the temporal bone produce a unilateral loss of vestibular function that can be either partial or complete. Functional effects of a unilateral loss of vestibular function include spontaneous nystagmus and vertigo (acute stage only), head movement–provoked vertigo, problems with gaze stabilization, and disequilibrium affecting the ability to maintain stability in sitting and especially in standing and while walking.[22] Functional effects of a bilateral loss of vestibular function include oscillopsia (inability to stabilize gaze during head movements) and severe imbalance affecting stance and gait.

### *Central Vestibular Lesions*

Traumatic head injury can also damage central vestibular structures.[3,13,23] Multiple petechial hemorrhages in the brainstem that damage central vestibular structures have been reported in both the mildly (no loss of consciousness) and moderately head-injured patient. Symptoms include spontaneous nystagmus and oculomotor problems including ocular dysmetria, cogwheeling during smooth-pursuit eye movements, and marked optokinetic asymmetries. Spontaneous and/or provoked vertigo may or may not occur in the patient with central vestibular lesions.

## Vestibular Rehabilitation

Vestibular rehabilitation is a comprehensive approach to assessing and treating symptoms of vestibular system pathology.[19,20,24] The overall goal of assessment is to document the patient's functional problems and the many sensory, motor, and cognitive limitations contributing to loss of functional independence. Specific evaluation of vestibular system pathology, summarized in Box 27-1, focuses on assessment of vertigo, control of eye-head coordination for stabilizing gaze, and musculoskeletal and neural components of postural control underlying the ability to maintain stability and orientation when sitting,

Box 27-1

**VESTIBULAR REHABILITATION ASSESSMENT**

**Vertigo**

Spontaneous

Provoked:

Positional

Movement

**Eye-Head Coordination**

Oculomotor control—saccade and smooth pursuit

Gaze stabilization during head movements

Gaze fixation suppression

**Postural Control in Sitting, Standing, and Walking**

Functional status

Underlying impairments:

- Biomechanical constraints
- Neuromuscular constraints
- Sensory/perceptual constraints

**Other Limitations**

Pain

Cognitive and behavioral constraints

standing, and walking. Sorting out the relative contribution of vestibular system pathology to overall loss of function can be difficult, because functional deficits after TBI are usually due to a combination of many interacting factors.

Vestibular rehabilitation uses exercise to decrease dizziness, improve gaze stabilization, and retrain sensory and motor aspects of postural control. In most instances exercises are designed to facilitate central nervous system (CNS) compensation rather than alter underlying vestibular disease. The exception is treatment of BPPV, which is aimed at altering the underlying mechanism that produces the symptoms rather than facilitating CNS adaptation. Strategies for treating vestibular system dysfunction are individualized to each patient's problems and focus on both remediating underlying impairments and improving functional skills.[19,20] Vestibular rehabilitation programs are often modified in the patient with TBI owing to the frequency of both physical and cognitive problems after trauma. Modifications include the following:

- Providing physical assistance to accommodate movement problems
- Ensuring greater supervision to accommodate cognitive and behavioral problems
- Progressing through the stages of exercises more slowly because of the multiplicity of the patient's problems

The following sections discuss clinical strategies for assessing and treating symptoms of vestibular system disorders, including vertigo, eye-head coordination, and postural dyscontrol, in the patient with TBI.

## *Vertigo*

### Assessment

When assessing vertigo, the therapist notes whether dizziness symptoms are spontaneous or provoked and determines the situations and conditions that precipitate complaints of dizziness.[19,20,25] Characteristics of vertigo are noted, such as latency of onset, duration, intensity, and the effect of repeating the movement. Associated autonomic symptoms, such as nausea, sweating, and pallor, are noted. The presence and type of nystagmus are recorded. Head movements are repeated in sitting, standing, and walking. In addition to subjective complaints of dizziness, episodes of staggering and disequilibrium associated with complaints of dizziness are recorded, particularly in standing and walking.

The patient with unilateral vestibular hypofunction (partial or complete) experiences both spontaneous and provoked vertigo in the acute stage. Often by the time a patient with TBI and concomitant unilateral loss of vestibular function enters a rehabilitation program, spontaneous complaints have resolved. Patients continue, however, to complain of vertigo, lasting from seconds to minutes, that is provoked by head movements in all planes. Vertigo in the patient with BPPV lasts 30 to 45 seconds and is provoked by placing the patient in the Dix-Hallpike position. Patients with BPPV may also report having vertigo while lying down and rolling to the affected side, while looking up, or while leaning over.[26,27]

### Treatment

Habituation exercises are used to diminish dizziness in patients who have movement-provoked symptoms of vertigo. Habituation exercises involve repeating the movements that provoke vertigo between 5 and 10 times, two to three times a day. When a patient is residing in an inpatient rehabilitation facility, habituation exercises are incorporated into twice-daily physical therapy treatments, usually at the end of each exercise session. As mentioned previously, habituation exercises are routinely modified to accommodate both movement and cognitive limitations in patients with TBI. Written exercise sheets are used, and patients are given logs to record exercise sessions. Because of the high frequency of behavioral and cognitive problems, including attention and memory deficits, in patients with TBI closer supervision and physical assistance are more often required in the rehabilitation of vestibular dysfunction in such patients than in patients with other types of vestibular dysfunction.

The canalith repositioning maneuver (CRM) developed by John Epley[26] has been shown to be an effective treatment for patients with BPPV (see Chapter 19). An alternative to the CRM is the liberatory maneuver described by Semont and colleagues[27] for management of BPPV. When the CRM has been used in an inpatient who is undergoing rehabilitation, the nursing staff must be given information regarding the necessary follow-up precautions in order to ensure that the patient is consistent in adhering to these precautions (see Chapter 17). Inability or failure to adhere to follow-up precautions can make the procedure ineffective.[28]

## *Eye-Head Coordination*

### Assessment

Eye movements from both the visual and vestibular systems used to keep gaze stable during voluntary and involuntary movements of the head are also examined.[20,24] Gaze stabilization is tested with the patient seated, stand-

ing unsupported, and walking. Visually generated eye movements, including smooth-pursuit and saccadic eye movements, are assessed. The patient's ability to maintain a stable gaze during horizontal and vertical head motions of varying speed is examined. Finally, the patient's ability to keep gaze fixed on an object moving in phase with the head is used to test visual suppression of vestibular-driven eye movements. Subjective complaints of dizziness, blurred vision, and oscillopsia are noted.

Following TBI, eye-head dyscoordination can result from (1) damage to the vestibular system that disrupts vestibulo-ocular reflex (VOR) function, (2) deficits within the visual system, including loss of ocular motility, visual acuity/field deficits, and visual perceptual deficits, (3) orthopedic injuries that limit cervical motion, and (4) damage to cerebellar structures resulting in loss of visual suppression of the VOR.[29,30]

### Treatment

Exercises are used to improve gaze stabilization when the head is still and in motion. Saccadic and smooth-pursuit tracking exercises are performed initially if the patient has dizziness during these eye movements. Next the patient is asked to keep the gaze fixed on a central target while moving the head either horizontally or vertically for a progressively longer period of time. Finally, patients are given exercises to improve visual modulation of vestibulo-ocular responses (see Chapter 20).[16,24,31] The patient repeats these exercises first in a supported sitting position and then progresses to performing them while standing and walking.

Because so many of the exercises used to treat symptoms of vestibular system disorders involve movement of the head and neck, treatment of cervical complaints is essential to recovery of function. Vestibular rehabilitation exercises combined with physical modalities and orthopedic manual skills have produced excellent results.[32]

## *Postural Control Underlying Stability*

The ability to maintain a stable position is critical to independence in most functional skills. *Stability* is defined as the ability to maintain the center of body mass within limits determined principally by the extent of the support base.[33] Stability requires a continuous interaction between the individual and the environment, and involves many bodily subsystems collectively referred to as the *postural control system* (see Chapter 3).

## *Assessment*

Because postural control is complex, involving the interaction of many systems, its assessment must be multidimensional. Assessment focuses on documenting the ability to perform functional skills requiring posture control as well as investigating the underlying impairments that constrain the maintenance of postural stability. Impairments are limitations within the individual that restrict sensory and motor strategies for postural control.[34] As shown in Figure 27.1, impairments affecting postural stability after TBI can be musculoskeletal, neuromuscular, sensory/perceptual, or cognitive.

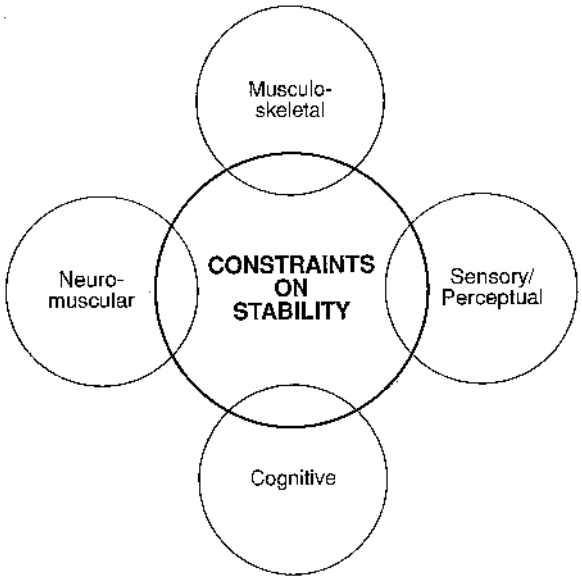


**Figure 27.1** Categories of constraints on stability after traumatic brain injury.

The information gained through assessment is used to develop a comprehensive list of problems, establish short- and long-term goals, and formulate a plan of care for retraining posture control. A thorough assessment must include review of the patient's medical and social history as well as of current symptoms and concerns.[34] Procedures for assessing the multiple levels of postural control have been described in detail elsewhere[34–36] and so are only briefly reviewed here.

### Functional Status

A number of tests are available to measure functional skills related to postural control. Our outpatient program uses three tests to document functional balance and mobility skills. The Berg Balance Scale, shown in Box 27-2, rates performance from 0 (cannot perform) to 4 (normal performance) on fourteen different tasks, including the ability to safely and independently sit, stand, reach, lean over, turn and look over each shoulder, turn in a complete circle, and step.[37] The total possible score on this scale is 56, indicating excellent balance. The Berg

Box 27-2

## 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) Able to stand, no hands and stabilize independently.

(3) Able to stand independently using hands.

(2) Able to stand using hands after several tries.

(1) Needs minimal assistance to stand or to stabilize.

(0) Needs moderate or maximal assistance to stand.

**2. Standing Unsupported**

Instruction: Stand for 2 minutes without holding on.

Grading: Please mark the lowest category that applies:

(4) Able to stand safely 2 minutes.

(3) Able to stand 2 minutes with supervision.

(2) Able to stand 30 seconds unsupported.

(1) Needs several tries to stand 30 seconds unsupported.

(0) Unable to stand 30 seconds unassisted.

If subject is able to stand 2 minutes 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) Able to sit safely and securely 2 minutes.

(3) Able to sit 2 minutes under supervision.

(2) Able to sit 30 seconds.

(1) Able to sit 10 seconds.

(0) Unable to sit without support, 10 seconds.

**4. Standing to Sitting**

Instruction: Please sit down.

Grading: Please mark the lowest category that applies:

(4) Sits safely with minimal use of hands.

(3) Controls descent by using hands.

(2) Uses back of legs against chair to control descent.

(1) Sits independently but has uncontrolled descent.

(0) 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) Able to transfer safely with only minor use of hands.

(3) Able to transfer safely with definite need of hands.

(2) Able to transfer with verbal cueing and/or supervision.

(1) Needs one person to assist.

(0) Needs two people to assist or supervise to be safe.

**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) Able to stand 10 seconds safely.

(3) Able to stand 10 seconds with supervision.

(2) Able to stand 3 seconds.

(1) Unable to keep eyes closed 3 seconds but stays steady.

(0) Needs help to keep from falling.

**7. Standing Unsupported with Feet Together**

Instruction: Place your feet together and stand without holding on.

Grading: Please mark the lowest category that applies:

(4) Able to place feet together independently and stand 1 minute safely.

*(continued on following page)*

Box 27-2 *(continued)*

## BERG BALANCE SCALE

(3) Able to place feet together independently and stand for 1 minute with supervision.

(2) Able to place feet together independently but unable to hold for 30 seconds.

(1) Needs help to attain position but able to stand 15 seconds feet together.

(0) Needs help to attain position and unable to hold for 15 seconds.

The following items are to be performed while standing unsupported:

**8. Reaching Forward with Outstretched Arm**

Instruction: 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) Can reach forward confidently >10 inches.

(3) Can reach forward >5 inches safely.

(2) Can reach forward >2 inches safely.

(1) Reaches forward but needs supervision.

(0) Needs help to keep from falling.

**9. Pick Up Objects from the Floor**

Instruction: Pick up the shoe/slipper placed in front of your feet.

Grading: Please mark the lowest category that applies:

(4) Able to pick up slipper safely and easily.

(3) Able to pick up slipper but needs supervision.

(2) Unable to pick up but reaches 1–2 inches from slipper and keeps balance independently.

(1) Unable to pick up and needs supervision while trying.

(0) Unable to try/needs assistance to keep from falling.

**10. Turning to Look Behind/Over Left and Right Shoulders**

Instruction: Turn to look behind you over/toward left shoulder. Repeat to the right.

Grading: Please mark the lowest category that applies:

(4) Looks behind from both sides and weight shifts well.

(3) Looks behind one side only; other side shows less weight shift.

(2) Turns sideways only but maintains balance.

(1) Needs supervision when turning.

(0) 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) Able to turn 360 degrees safely in <4 seconds each side.

(3) Able to turn 360 degrees safely one side only in <4 seconds.

(2) Able to turn 360 degrees safely but slowly.

(1) Needs close supervision or verbal cueing.

(0) 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 4 times.

Grading: Please mark the lowest category that applies:

(4) Able to stand independently and safely and complete 8 steps in 20 seconds.

(3) Able to stand independently and complete 8 steps in >20 seconds.

*(continued on following page)*

Box 27-2 *(continued)*

## BERG BALANCE SCALE

(2) Able to complete 4 steps without aid, with supervision.

(1) Able to complete >2 steps; needs minimal assistance.

(0) Needs assistance to keep from falling/unable to try.

**13. Standing Unsupported, One foot in Front**

Instruction (demonstrate to subject): Place one foot directly in front of the other. If you feel that you cannot place your foot directly in front, try to step far enough ahead that the heel of your forward foot is ahead of the toes of the other foot.

Grading: Please mark the lowest category that applies:

(4) Able to place foot tandem independently and hold 30 seconds.

(3) Able to place foot ahead of other independently and hold 30 seconds.

(2) Able to take small step independently and hold 30 seconds.

(1) Needs help to step but can hold 15 seconds.

(0) Loses balance while stepping or standing.

**14. Standing on One Leg**

Instruction: Stand on one leg as long as you can without holding on.

Grading: Please mark the lowest category that applies:

(4) Able to lift leg independently and hold >10 seconds.

(3) Able to lift leg independently and hold 5–10 seconds.

(2) Able to lift leg independently and hold >3 seconds.

(1) Tries to lift leg; unable to hold 3 seconds but remains standing independently.

(0) Unable to try or needs assistance to prevent fall.

(From Berg, 1993.[37])

Balance Scale has been shown to have excellent interrater reliability and relatively good concurrent validity. It has shown to be an effective predictor of fall risk in community-dwelling older adults.[38] Its ability to predict fall risk in patients with TBI is unknown.

Self-selected walking speed has been used as a measure of a patient's ability and confidence in walking.[39] There is a paucity of information on what constitutes independent functional mobility. One study reported that in order to perform instrumental activities of daily living (ADL) skills, the average person must be able to (1) walk a minimum of 1000 feet, (2) achieve a velocity of 80 meters per minute for 13 to 27 meters in order to cross a street safely, (3) negotiate a 7- to 8-inch curb independently, and (4) walk and turn the head without loss of balance.[40] In our clinic, two methods are used to assess functional mobility. The first is the Three-Minute Walk Test, which requires subjects to walk at their preferred pace for 3 minutes over a 524-foot indoor course.[34] The course is carpeted and involves four different turns. Distance walked in the 3 minutes is recorded, as is the number of times the patient moves outside a 15-inch walking path. The second test of functional mobility is the Dynamic Gait Index, which is used to evaluate the ability to adapt gait to changes in task demands such as changing speeds, head turns in the vertical or horizontal direction, stepping over or around obstacles, and stair ascent and descent. The test involves eight tasks that are scored from 0 to 3, for a total of 24 points.[34]

New research suggests that dual-task methods of assessing balance and mobility may be more sensitive indicators of functional mobility problems than tests that focus on single-task performance.[41,42] The use of dual-task measures may be particularly important in patients with TBI owing to the frequency of cognitive impairments such as disorders in attention, memory, and judgment. Kerns and Mateer[43] report an example of a patient with TBI who, when given the task of grating cheese, was noted to progressively slump down and forward until his forehead touched the counter. This patient was unable to successfully divide attention between two tasks, maintaining a vertical posture and grating cheese.[44] We have

found that in patients with mild to moderate head injury, postural control is particularly compromised during the simultaneous performance of cognitive tasks requiring visual processing.[43] The development of dual-task methods of assessing functional balance and mobility skills is just beginning.[41] However, their usefulness in documenting functional limitations in patients with TBI is very promising.

### *Impairments Limiting Postural Stability*

Evaluation of motor impairments involves an assessment of muscle tone, strength, range of motion, cerebellar coordination, static postural alignment in sitting and standing, and examining coordinated multijoint movements used to recover stability after perturbations of different size, amplitudes, and directions.[34] Although not performed routinely in the clinic, electromyography and kinematic analysis of associated body movements can be used to quantify dyscoordination in stance and gait as well as improvements during recovery.

Weakness, particularly hemiparesis, is frequently a primary neuromuscular impairment affecting balance in the patient with TBI. Traumatic injury to the cerebellum or deep hemorrhagic lesions in the basal ganglia can affect the timing and scaling of muscles working synergistically for postural control. Clinical indicators of muscular dyscoordination during postural tasks include asymmetrical use of limbs for movement control and excessive movements at the joints, including excessive flexion of the hip and loss of knee control.

Assessment of sensory impairments involves evaluating sensation (vibratory sense, stereognosis, and peripheral visual acuity), determining the patient's ability to remain oriented under different sensory conditions, and evaluating whether perceptions relevant to stability are accurate.[20,23] Moving platform posturography is one approach to examining the organization and selection of senses for postural control.[45] Posturography uses a moving force-plate in conjunction with a moving visual surround to determine the patient's ability to correctly select from among visual, somatosensory, and vestibular inputs, the most appropriate sense for orientation (see Chapter 3).

Alternatively, the Clinical Test for Sensory Interaction in Balance (CTSIB) tests the patient's use of alternative sensory cues for orientation, using procedures similar to those of posturography.[46] This test, originally described in 1986, used six sensory conditions to examine a person's ability to maintain stability under altered sensory contexts. These six sensory conditions are shown in Figure 27.2. In our clinic, we have modified this test and now use only four sensory conditions, eliminating the dome conditions (3 and 6) but maintaining the two foam conditions (4 and 5). The dome was originally created to identify patients with visual motion sensitivity—a heightened sensitivity to motion in the environment, such as is found in crowded shopping centers. The dome has not proved to be sensitive or specific enough in identifying visual motion sensitivity.[47] Compliant foam (medium-density Sunmate Foam) is used to reduce the effectiveness of support surface somatosensory inputs for orientation. Procedures for administering the CTSIB are detailed elsewhere.[48]

Sensory impairments affecting orientation after TBI can result from pathology within the peripheral vestibular system or within central structures that organize sensory information for postural orientation. Although both posturography and the CTSIB test can identify and quantify functional problems in selecting sensory inputs for postural control, they cannot determine the anatomic location of the injury producing these functional problems.

## Treatment

Treatment of instability involves remediating impairments and improving sensory and motor strategies essential to ensuring stability in functional tasks performed in sitting, standing, and walking.[19,20,24] Biomechanical limitations and movement disorders are a particular concern in the patient with a vestibular deficit because they limit the patient's ability to move in ways that are necessary for compensation of the deficit.

Treatment of biomechanical limitations involves physical modalities, such as heat and ultrasound, as well as exercises to improve range of motion, joint flexibility, and body alignment.[34,49] Treatment of neuromuscular problems varies according to the nature of the problem. Strengthening exercises are used to improve impaired force generation. Therapy for muscular dyscoordination consists of functional electrical stimulation, electromyographic biofeedback, and neuromuscular facilitation exercises.[34]

Exercises to improve sensory function for orientation require the patient to maintain balance during progressively more difficult movement tasks while the therapist varies the availability and accuracy of one or more senses for orientation.[19,20] For example, during exercises to decrease sensitivity to visual motion cues, the patient performs balance and movement tasks while visual cues are reduced or absent (with eyes closed or while wearing blinders or a blindfold) or are inaccurate

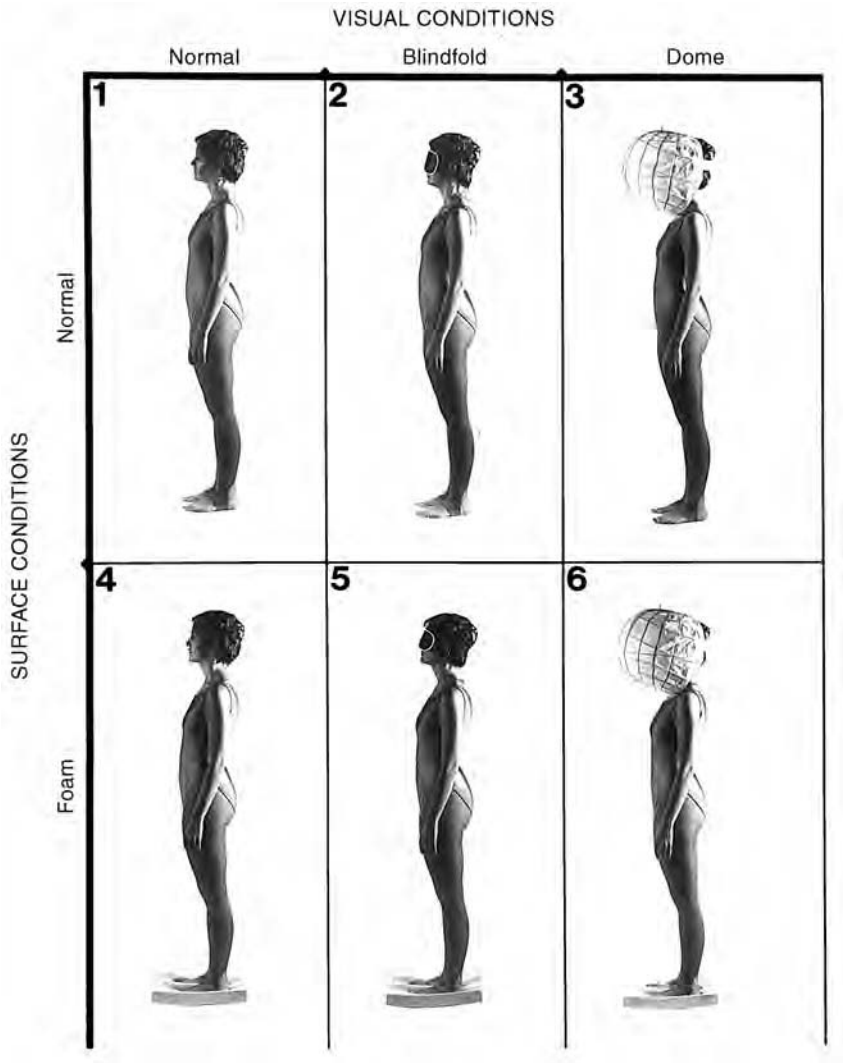


**Figure 27.2** The six positions used to test sensory interaction in balance. (From Shumway-Cook and Horak, 1986.[46])

for orientation (use of prism glasses or optokinetic stimuli, and within a large moving visual surround).[50] During these exercises, accurate orientation cues from the surface are essential.

In contrast, exercises to improve use of vestibular inputs for postural control in the patient with partial loss of vestibular function involve decreasing the availability of both visual and somatosensory input for orientation. An example would be asking the patient to reach for an object while wearing blinders and standing unsupported on a piece of foam or a thick piece of carpet.

Patients who have had a complete bilateral loss of vestibular function are taught to rely on visual and/or somatosensory cues for postural control. Because these patients have no residual vestibular function available to them, the goal of therapy is sensory substitution for, rather than enhancement of, remaining vestibular function.

In summary, treatment of postural dyscontrol in the patient with TBI and associated vestibular system pathology is directed at helping the patient reestablish effective sensorimotor strategies for balance control. Therapy focuses on practicing functional tasks such as standing unsupported while reaching, leaning, or turning, and moving from sit to stand. In addition, treatment seeks to remediate specific deficits in musculoskeletal and sensorimotor systems underlying postural dyscontrol.

## Time Course for Recovery

The time course for recovery from traumatic vestibular lesions is different from that in vestibular lesions of other etiologies.[51] In the patient with TBI and vestibular system pathology, concomitant CNS lesions impair the compensatory process itself. As a result, recovery from traumatic

vestibular lesions is often protracted. Pfaltz and Kamath[51] compared recovery in patients with a unilateral loss of vestibular function due to various causes, including trauma, Ménière's disease, labyrinthectomy, and other diseases. At 6 months, only one third of the patients with unilateral loss due to trauma were symptom free; the majority of the other patients had achieved compensation and were symptom free. At 18 months, many of the patients with traumatic vestibular loss continued to show persisting symptoms of vestibular system disorder.

Berman and Fredrickson[3] studied 321 mildly and moderately head-injured patients with vestibular system pathology. Vertigo was reported in 34% of mildly head-injured patients and in 50% of moderately head-injured patients. Sixty percent to 70% of patients with central vestibular dysfunction had persisting symptoms 5 years after injury. Almost half of these patients never returned to work.

These and other studies suggest that the typical time course for recovery from vestibular system disorder in patients with TBI is protracted, requiring one to three times longer than in patients with vestibular system disorders from other causes. Many patients with traumatic vestibular dysfunction show persisting symptoms of dizziness and disequilibrium several years after trauma. The extent to which specific exercise interventions can influence the time course of recovery from vestibular system dysfunction in the patient with TBI is an important theoretical and clinical question currently under study.

## CASE STUDY

### Case Report

S.D. is a 31-year-old man admitted for rehabilitation 1 month after a closed-head injury. He fell 40 feet, striking the left side of his head, and experienced a brief (< 5 minutes) loss of consciousness. He was admitted to the hospital with a Glasgow Coma Scale score of 13. Computed tomography showed multiple left parietal-occipital cerebral contusions. Associated trauma included three fractured ribs and a contusion of the left hip. There was no previous history of medical or neurological problems.

Otological evaluation involved both a clinical examination and electronystagmography, including tests for gaze and positional nystagmus, oculomotor function (saccade, smooth-pursuit, and optokinetics), and VOR function using rotational chair testing. Results of this examination indicated the presence of post-traumatic BPPV. Other test results were within normal limits, suggesting no evidence for vestibular hypofunction or a central vestibular lesion.

#### *Assessment*

On assessment, the patient was found to have moderate to severe imbalance (score 46 out of 56 on the Berg Balance Scale). The patient had difficulty maintaining stability when moving from sit to stand, when standing with eyes closed or with a reduced base of support, and when reaching, leaning, or turning while standing. The patient used a cane and required standby assistance for safety when walking. On the Three-Minute Walk Test, he walked 135 feet and had four significant path deviations. Gait pattern abnormalities included an equinovarus foot position at foot strike, circumduction of the right leg during the swing phase of gait, and hyperextension of the right knee during the stance phase of gait. He had significant problems with adapting his gait to changing task demands, scoring 13 out of 24 on the Dynamic Gait Index. He required minimal assistance to maintain his balance when walking with head turns (horizontal or vertical) and when stepping over obstacles.

Sensorimotor constraints were as follows:

1. Positional vertigo in the left Hall pike position as well as vertigo during gait with vertical head motions.
2. Postural dyscontrol, including:
   - Right hemiparesis and dyscoordination.
   - Limitation of range in right ankle.
   - Asymmetries in weight-bearing, primarily in stance and during gait.
   - Difficulty organizing sensory inputs for balance and effectively using vestibular inputs for orientation in the absence of visual and somatosensory cues.
3. Other problems:
   - Pain due to fractured ribs and contusions on the left hip
   - Moderate cognitive impairments including memory and attention deficits
   - Decreased awareness of his limitations, raising a number of safety issues.

*Treatment*

Treatment focused on eliminating positional vertigo, balance retraining to improve ability to perform functional skills in sitting and standing, and gait retraining to improve gait pattern, endurance, and adaptive capability when walking.

The CRM was used to eliminate BPPV. Balance retraining involved exercises to improve strength, range of motion, and coordination in the right extremities as well as postural sway biofeedback to reduce weight-bearing asymmetries in standing and improve range of center of mass movements over the hemiparetic leg. Manual cues and feedback were used to improve joint coordination at the knee and hip while the patient practiced voluntary sway in standing, while moving from sit to stand, and during gait. The patient also practiced maintaining balance during such functional tasks as reaching, leaning, and turning in the standing position. In order to improve his ability to use vestibular inputs for postural control, the patient practiced a variety of stance balance tasks while standing on foam or various grades of carpets while wearing blinders or with eyes closed.

Outcomes after 4 months of rehabilitation (1 month as an inpatient, 3 months as an outpatient) were as follows:

The BPPV was successfully eliminated by the CRM. Postural control underlying stability in sitting, standing, and walking had improved; the patient was transferring and walking independently and no longer needed a cane to walk. Results of posturography testing examining sensory aspects of postural control in S.D. before and after rehabilitation are shown in Figure 27.3.

Figure 27.3 compares S.D.'s performance on balance tasks under altered sensory conditions at 1 month and 6 months after his TBI. Peak-to-peak center-of-gravity angle, in degrees, is plotted for individual trials in the six conditions, conditions 3 through 6 have three trials each (see Chapter 3). Encompassing individual trials is a larger histogram that shows the upper limits of normality, established using the 95th percentile from 250 normal control subjects ages 8 to 70.[52] At one month, S.D. lost balance (indicated in Fig. 27.3 by "test stopped") when either visual or surface cues for orientation were reduced or inaccurate (conditions 3, 4, 5, and 6). At 6 months after injury, he fell on the first trial only when deprived of both visual and surface cues simultaneously (conditions 5 and 6).

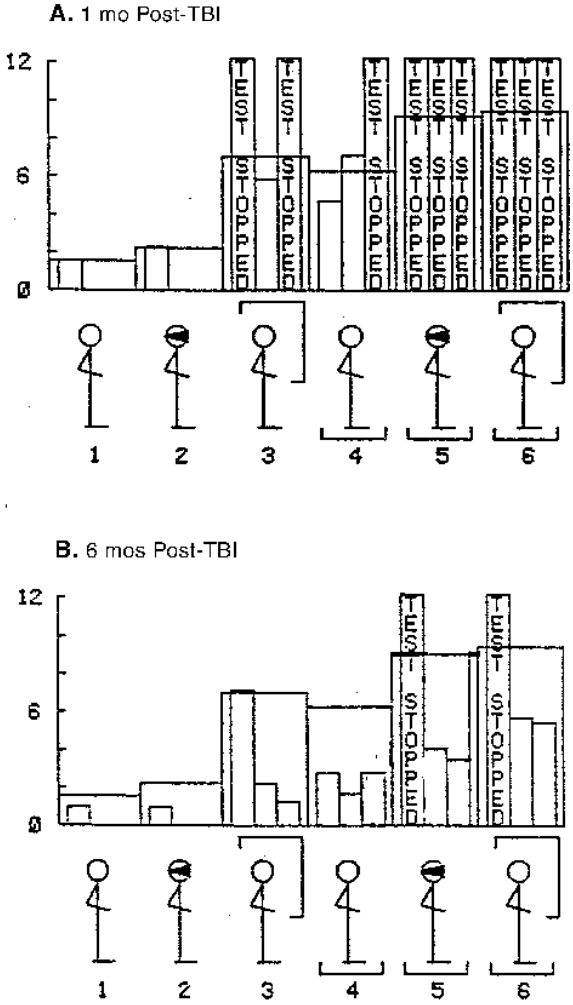


**Figure 27.3** Recovery of sensory organization aspects of postural control is shown by comparing body sway during altered sensory conditions at 1 month and 6 months after traumatic brain injury (TBI). See text for details.

## Summary

Assessment and treatment of vestibular system pathology are essential parts of rehabilitating the patient with TBI. However, after TBI, injuries to other parts of the CNS can complicate recovery from vestibular system dysfunction in the following ways:

1. Associated trauma can produce pain and restrict movements.
2. Musculoskeletal and neuromuscular problems can affect the patient's ability to move in ways that are necessary to achieve CNS compensation.
3. Damage to visual and somatosensory systems may limit the availability of these senses as alternatives to lost vestibular inputs.
4. Cognitive and behavioral problems make compliance with a vestibular exercise program difficult.
5. Intracranial injury can damage neural structures important to the compensatory process, resulting in persisting symptoms and a protracted recovery.

A comprehensive treatment plan that incorporates vestibular rehabilitation exercises that have been modified to adjust for the preceding limitations is an effective way to enable patients with TBI to gain functional independence and minimize persisting and disabling symptoms of vestibular system pathology.

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CHAPTER 28

# Non-vestibular Dizziness and Imbalance: Suggestions for Patients With Migraine and Mal de Débarquement Disequilibrium

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## Definition of Non-vestibular Dizziness

There are numerous causes for patients to complain of symptoms related to vertigo, disequilibrium, lightheadedness, and/or unsteadiness and combinations thereof. Although the majority of these complaints arise in association with direct insults limited to the peripheral vestibular apparatus, other sites of origin with specific disorders can manifest symptoms of a similar nature. Vestibular and balance rehabilitation programs are symptom-driven for both entrance criteria and the development of the treatment plans, not specific to the site of lesion or the diagnosis. Therefore, management of the patient with balance disorder by means of vestibular and balance rehabilitation programs to control and/or eliminate the symptoms is potentially applicable to a wide variety of etiologies and lesion sites.[1–3] Scattered results in the older literature has suggested that the approach utilized for the stable peripheral vestibular insult (i.e., no complaints of spontaneously occurring symptoms) may be applied with success to the patient with non-vestibular balance problems.[4]

There is now a growing literature addressing the use of vestibular and balance rehabilitation therapy (VBRT) techniques in patients with other than direct peripheral or central vestibular lesions.[5–12] The purpose of this chapter is, therefore, to review the outcomes of patients with non-vestibular balance disorders to whom VBRT techniques have been applied. This will be accomplished via review of published population studies for migraine and anxiety/panic disorders. Our experience of patients with mal de débarquement is also presented. The bulk of the chapter concentrates on migraine and anxiety-associated dizziness. These two disorders individually or as comorbidities are known to be commonly direct causes of dizziness and balance complaints and frequent cause of lack of progression in otherwise stable unilateral vestibular lesions.

The three groups of patients discussed here have been identified as having symptoms suggesting that a VBRT program may be applicable. Inclusion in therapy programs of this type is considered appropriate if the patient's symptoms are:

1. Provoked by head or visual motion.
2. Constant and exacerbated by head or visual motion.
3. Functional deficits identifiable for balance and/or gait even though head or visual motion does not interfere.

These criteria describe a stable condition, one that does not vary in its overall character with time, as would occur with patients describing symptoms that occur only in a spontaneous manner, without provocation or warning. Those patients with complaints of spontaneous events of disequilibrium, with or without vertigo, are considered either inappropriate or have a poor prognosis for the use of vestibular and balance rehabilitation secondary to presence of their spontaneous events.[2] Of the three groups of patients reviewed, those with migraine are the group that is likely to experience spontaneous events of vertigo or lightheadedness/imbalance symptoms. Therefore, in evaluating the effectiveness of VBRT for this group, it is important to recognize whether the study subjects otherwise met the general criteria previously listed for use of VBRT.

## Mal de Débarquement

Most individuals exiting from a ship (and, occasionally, a train or after long car excursions) after a cruise of a day or longer experience a continuing sensation of rocking and/or other forms of linear shipboard movement. These perceptions are especially prominent when the individual is sitting or standing still and typically abate after 12 to 36 hours.[5,13] For a small percentage of otherwise healthy persons, the feeling of movement does not resolve after the usual interval; they are said to have mal de débarquement (MDD). This perseveration of dominantly linear movement sensations when the patient is still or is involved with ambulation has been suggested to result from a variety of causes, with the specific theme that of the central system being slow in readapting to a nonmoving support surface for standing, sitting, or lying.[5,14] On the basis of a case study, Lewis[14] suggests that the frequency and acceleration characteristics of the stimulating vehicle are mimicked in the expression of symptoms by the patient with MDD.

In general, balance function testing of a traditional nature does not show results outside normal ranges in the patient with MDD. Specific testing of the otolith organs has only recently become more accessible to the routine balance center, and as yet, no studies have been reported on the directly measured status of the otolithic organs in MDD. The implication is that this disorder does not involve identifiable malfunction of the peripheral mechanism, especially that of the semicircular canals. Although postural control abilities are usually within normal limits from a clinical perspective, Nachum and colleagues[15] have shown, in a prospectively designed control-group study of navy crew members, that postural control abilities were poorer in the group susceptible to MDD.

The natural history of this disorder is spontaneous resolution within 6 to 12 months, although some patients continue to have symptoms for considerably longer. There is a female preponderance in the general MDD population.[5] Because the majority of the patients do not have specific head motion–provoked symptoms but do have the perception of balance deficits, the therapy programs provided to the these patients involve balance activities.

Patients with MDD are a small population, one of the largest series, that of Hain and colleagues,[5] reporting on 27 subjects. Therefore, we describe a small group of patients from our own experience—6 patients seen in our clinic during a 4.5-year interval—to look at the efficacy of applying VBRT in this population. Each of the patients met the previously listed criteria regarding their symptom presentation for use of a VBRT program. In order to rule out the possibility of unstable peripheral system involvement, we excluded any patients with complaints of spontaneous events. Site-of-lesion determination was based on a full series of studies including, but not limited to, electronystagmography (ENG), extensive ocular motor evaluation, rotational chair testing, and dynamic posturography.[16] No indications were obtained that would imply peripheral system involvement. The patients in our small sample were not treated with medications during the time they were undergoing VBRT. Therapy outcome was indicated by two global measures, disability score and post-therapy symptom score. Additionally, a single specific measure of sensitivity to head movement, the Motion Sensitivity quotient (MSQ), and dynamic posturography for postural control were used.[2] Results of the specific measure of dynamic posturography were normal and unchanged in all patients before and after therapy. We assigned disability scores on the basis of questioning each patient as to the effect of the symptoms on daily activities. The disability scores are defined as follows:

| Score | Description |
|---|---|
| 0 | No disability; negligible symptoms. |
| 1 | No disability; bothersome symptoms. |
| 2 | Mild disability; performs usual work duties but symptoms interfere with outside activities. |
| 3 | Moderate disability; symptoms disrupt performance of both usual work duties and outside activities. |
| 4 | Recent severe disability; on medical leave, or had to change job because of symptoms; off work for less than 12 months. |
| 5 | Long-term severe disability; unable to work for more than 1 year or has established permanent disability with compensation payments. |

A post-therapy symptom score was assigned to each patient at the time he or she was started on a maintenance program. The score was determined by responses to a series of specific questions asking the patient to compare symptoms at the end of active therapy with those prior to therapy. The scores are defined as follows:

| Score | Description |
|---|---|
| 0 | No symptoms remaining at the end of therapy. |
| 1 | Marked improvement in symptoms; mild symptoms remaining. |
| 2 | Mild improvement; definite persistent symptoms remaining. |
| 3 | No change in symptoms relative to pre-therapy period. |
| 4 | Symptoms worsened with therapy activities on a persistent basis relative to pre-therapy period. |

The findings from this small sample, shown in Table 28-1, contrast with those reported by Hain and colleagues.[5] Fifteen of their 27 patients undertook a VBRT program whose results were described as being of limited productivity. The outcome measures shown in Table 28-1 indicate consistent improvement in symptoms and disability from before to after therapy in our 4 patients with follow-up. As with any disorder in which the natural history is that of spontaneous resolution, caution must be exercised in drawing conclusions from a 4-person sample of improvement. With the lack of head movement–provoked symptoms and of any problems with vestibulo-ocular reflex function, habituation and adaptation activities were not used. Instead, the program concentrated on walking with head movements and the slow balance movements like those used in tai chi.

■ Table 28-1 **THERAPY OUTCOME PARAMETERS FOR FOUR OF SIX PATIENTS WITH MAL DE DÉBARQUEMENT**

| | Disability Score Change [Pre-Therapy Score] | Change in MSQ | Post-Therapy Symptom Score |
|---|---|---|---|
| Case 1 | 1 [2] | 3.3 | 1 |
| Case 2 | 2 [3] | | 1 |
| Case 3 | 3 [4] | | |
| Case 4 | 1 [2] | −0.8 | 1 |

The difference between our experience and that of Hain and colleagues[5] may relate to the fact that the larger study used a questionnaire, so that the use of VBRT was simply reported on by the participants and not a controlled event. The participants came from various areas of the United States, so details of the focus of their VBRT programs are not available. In general, the application of traditional vestibular rehabilitation techniques of adaptation and habituation would not be expected to affect this population. Those with MDD are basically without symptoms when in motion at a reasonable speed. It is when they are still or moving slowly that they experience the symptoms. Therefore, although only anecdotal, the evidence shown in Table 28-1 suggests that activities related to balance in a slow format, such as that of tai chi, may be of some use in patients with MDD while they await the natural resolution process. Additionally, the study by Hain and colleagues[5] suggests that the use of benzodiazepines separately or with a customized VBRT program have a positive effect on symptoms of MDD, but traditional vestibular-suppressive medication (meclizine or scopolamine) are not helpful.

## Migraine-Associated Dizziness

Epidemiological studies demonstrate a significantly greater prevalence of vestibular symptoms in patients with migraine,[17,18] and a significantly higher incidence of migraine in patients with dizziness complaints than is found in the general, unscreened population.[19] It is reported that in 30% to more than 50% of patients with migraine, symptoms of dizziness are inconsistently associated with a headache or are completely independent of

head pain.[17,20–23] The frequency of abnormal test findings on ENG, rotational chair testing, and posturography reported by numerous studies is summarized by Furman and colleagues.[24] They found an average of 9.9% of a total of 534 patients with spontaneous nystagmus and up to 54.2% with abnormal asymmetry on rotary chair testing. A number of studies indicate a sensorineural loss of auditory sensitivity in patients diagnosed with migraine-associated dizziness (MAD).[25] In general, the losses of sensitivity were typically in the mild to moderate range and of a variety of configurations. From these statistics it is apparent that the relationship between migraine and dizziness is significant.

There are two major ways that migraine can cause problems with balance and dizziness. First, migraine can cause the symptoms of dizziness and imbalance in a patient who does not have a peripheral or central vestibular deficit, fitting our definition of non-vestibular dizziness. Second, migraine may be comorbid with a peripheral vestibular lesion. In this second situation, the migraine condition may well cause a slowing of the central compensation process and result in reduced effectiveness with the use of a VBRT program. Although this second situation is not strictly a non-vestibular condition, interaction of a non-vestibular condition that causes a prolongation of symptoms of dizziness, and the common occurrence of migraine in conditions of dizziness, make consideration of this situation worthwhile in this context. We now consider the role of vestibular and balance rehabilitation in both situations.

Whitney and associates[6] conducted a retrospective study of 14 patients with MAD (no indications of peripheral or central vestibular involvement) and 25 patients with peripheral vestibular involvement and self-reported history of migraine to investigate the effect of customized VBRT over 1 to 11 months of active therapy. Multiple outcome measures were used that involved physical ability for gait (Dynamic Gait Index [DGI]); self-perceived symptom intensity (scale from 0 to 100), self-perceived disability (Dizziness Handicap Inventory [DHI]), with three subscales for emotional, physical and functional impact of symptoms; and confidence scaling in performing activities of daily living (Activities-Specific Balance Confidence Scale [ABC]) (detailed discussions of these tools are found elsewhere in this text). A composite score, using a weighted combination of the ABC, DHI and DGI scores, was used to give a global measure of performance. This process was validated, showing a strong correlation with the disability score of Shepard and Telian.[26] Overall, there was a significant improvement ($P < .05$) for all outcome measures from before to after therapy. However, within the MAD group, although the physical measure of gait ability (DGI), perceived physical ability (functional and physical subscales of DHI), and composite score showed significant changes in perceived intensity of symptoms, the emotional impact of symptoms (emotional subscale of DHI) and, to a lesser extent, confidence in performance of daily activities (ABC; $P = .06$) did not show significant improvement after therapy. In contrast, the group with peripheral disorders and a history of migraine showed significant post-therapy improvement or a trend for significant improvement on all outcome measures (physical and functional subscales of the DHI; $P = .06$).

Whitney and associates[6] approached the issue of medication use by comparing outcomes for all patients undergoing antimigraine medication therapy (7 of the MAD group and 10 of the migraine history group) with those not taking concomitant medication. This comparison showed statistically greater improvement for the medication group on composite score, DHI, and DGI. However, when this same comparison was performed within the MAD and migraine history groups, only the MAD group showed more improvement in those on medication. Medication use had no significant effect on the migraine history group.

Collectively, the results of this study suggest that for patients with MAD, change in physical performance does not result in perceived improvement unless the underlying causative disorder of migraine is being successfully addressed. The findings also imply that a simple history of migraine with a peripheral disorder does not necessarily constitute a comorbidity of significance that would influence either compensation or the effectiveness of VBRT.

To address the implication that a simple history of migraine may not be an influential factor in effectiveness of VBRT, the same group of investigators conducted a second retrospective study involving 31 patients with peripheral or central vestibular diagnosis (other than that of MAD) and a history of migraine.[7] Historical controls were selected via chart review to match with the experimental group on the basis of age $\pm 5$ years, diagnosis, and vestibular function test values but no indications of migraine. Both groups underwent customized VBRT with outcome measures as described for the previous study, with the addition of the Timed "Up and Go" Test, which quantifies the speed at which a person is able to stand, walk 3 meters, turn, and return to the seated position (see discussion elsewhere in this text). The mean change in all of the outcome measures showed statistically significant improvement ($P < .05$) after therapy for both groups. A significantly greater change was noted in the DHI for the control group than in the migraine group, with all other outcomes showing no significant difference between groups. When a comparison of the number of

patients from each group who had what they defined as a clinically significant improvement in the outcome measures, statistical differences were noted for ABC and DGI results, showing that more patients in the control group improved than in the migraine group. The other outcome measures again did not demonstrate a significant difference, although for all outcome measures, more of the control group had clinically significant improvements than the migraine group. Although it has been demonstrated that in a group of patients with vestibular problems but without history of migraine or anxiety, their perception of handicap is related in a correlative manner to their functional impairment.[27] This was not the case with the cohort of migraine patients in the second study reported by the Whitney group.[7]

In summary, VBRT programs do appear to have a role to play in the treatment of dizziness produced by the migraine condition or in situations in which the comorbidity of vestibular system impairment and migraine may interact synergistically. It seems clear that although the VBRT programs are of positive assistance, the overall results in the patients with MAD or with a simple associated history of migraine are not as effective as in patients without migraine complications. This appears to be partly due to the reduced correlation between improvement in physical ability and the patients' perception of disability, perhaps caused by ongoing headaches even though the dizziness component may have been reduced or eliminated. The implication is that the underlying disorder of migraine must be treated simultaneously with the use of a VBRT program for maximum results. Given that patients with migraine have a significant sensitivity to motion (referred to as space motion discomfort),[24] the use of active head movement exercises in a patient with active migraines may be counterproductive, and this sensitivity may limit the use of various VBRT techniques.

Treatment of the migraine condition with behavioral and/or pharmacological methods has the opportunity to enhance the effectiveness of a simultaneous VBRT program. To more definitively address these implications, a prospectively designed study of patients with MAD, identified as meeting strict definitional criteria for MAD,[19] involving a controlled comparison between simultaneous treatment of the migraine condition during VBRT, to use of VBRT without the migraine treatment is needed.

## Primary Anxiety and Panic

The association of psychiatric disorders in general, and anxiety, panic, and agoraphobia in particular, has prevalence in an unselected population of dizzy patients that is significantly greater than in the general population.[28–30] The interested reader is referred to summaries of the documentation of this relationship and possible mechanisms of cause and effect that are beyond the scope of this chapter.[31–33] As with migraine, anxiety, panic, and phobic avoidances can be related to dizziness as a direct cause or as a means of interfering with the natural compensation process.[34–36] Several mechanisms by which psychological disorders may stall the compensation process, even with the use of VBRT efforts to promote the process, have been proposed.[34] First, and likely the most common, would be the natural avoidance of situations and movements that provoke symptoms of dizziness or disorientation that are currently produced or were initially produced by a lesion of the peripheral vestibular system. These behavioral reactions to an initial set of symptoms are counterproductive to the activities needed for natural central vestibular compensation. The behavioral changes are fueled by a fear of danger from the symptoms experienced as well as the embarrassment of appearing out of control when symptoms occur. If the behavioral changes continue in an uninterrupted manner, they can develop into actual psychiatric illness not unlike agoraphobia and/or lead to a self-generation of a portion of the original symptoms even though the initiating disorder, such as benign paroxysmal positional vertigo, has resolved.[32,34]

A second consequence of the natural avoidance of situations and movements is a subconscious reduction in the reliance on information from the vestibular and proprioceptive system leading to an obligatory use of visual information. This change in the cognitive, motor, and perceptual responses to symptoms then leads to further incidences of disequilibrium in conditions in which visual information is inappropriate for the maintenance of stance or orientation. This vicious circle leads to further avoidance of and hypersensitivity to visual motion as a provoking event for symptoms of imbalance and disorientation.[11,34] This hypersensitivity to normal daily inputs may lead to yet another mechanism of interference with the compensation process. In this case, the heightened arousal leads to conditioned autonomic responses from the anxiety produced by the daily exposure to normal visual, proprioceptive, and vestibular stimuli. The somatic expressions of the anxiety (e.g., nausea, perception of abnormal sway when standing, fear response) fuel the already altered perceptual, motor, and avoidance changes.[34]

In the counseling of patients as to the possibility that a major portion or all of their ongoing symptoms may be related to anxiety disorder, it is clearly helpful to explain the mechanisms by which this could occur secondary to an initial neurotological insult to the vestibular system. This issue is also clarified by the information summa-

rized in the review works relating the direct anatomic pathways between the vestibular system and those portions of the brain known to be involved with anxiety, panic, and fear avoidance reactions.[31–33] Although little is known as to the direct effects of neurotransmitters and other chemical actions in the compensation process, some evidence has been found that simple stress alone interferes with compensation in unilateral labyrinthectomized rats that may be partially accounted for by changes in the concentration of a steroid biosynthesis enzyme.[35] Information of this nature helps patients accept the concept that anxiety, panic, and avoidance behaviors have a physical basis and that the problem is not "just in the head," relieving concerns about the stigma that accompanies psychiatric disorders.

The preceding discussion dealt primarily with psychiatric disorders as secondary developments of an insult to the peripheral or central vestibular system, producing a situation in which the compensation process is retarded or the anxiety disorder is conditioned on the initiating symptoms and then produces symptoms modeled after those from the insult. Although these two situations may be considered "non-vestibular," more germane to this chapter would be the situation in which a psychiatric disorder is the primary disorder that manifests in the form of complaints of vague dizziness and imbalance.

A study by Staab and Ruckenstein [36] approached the question of how often this latter situation occurs compared with development of psychopathology secondary to a neurotological insult. These investigators also explored whether the psychological disorders have different general characteristics and risk factors. This retrospective review of 132 patients found that the subjects could be divided into three groups according to what was believed to be the initiating cause of the symptoms of dizziness. First were those patients in whom anxiety disorder was the only cause of the symptoms of dizziness. Second was the group for whom an identifiable neurotological disorder of the peripheral vestibular system or diagnosis of MAD, considered a neurotological disorder in this study, exacerbated a preexisting anxiety disorder. Third was the group in whom a peripheral vestibular insult or MAD provoked an anxiety disorder that interfered with recovery from the initial insult (the types of patients discussed previously). One of the surprising findings was that of this total cohort of patients, all of whom were referred initially for the complaints of dizziness, a full third of the patients met the diagnostic criteria set by Staab and Ruckenstein for anxiety disorder or panic as the sole cause of the complaints of dizziness. In the other two thirds, the exacerbation or development of anxiety disorder was stimulated by a primary neurotological disorder, each in equal proportion. Therefore, each group accounted for a third of the total cohort. This finding alone supports the contention that anxiety and dizziness are a two-way communicating relationship,[33] with each primary disorder—anxiety or neurotological insult—able to stimulate the other. The other dominant finding was that patients with prodromal anxiety conditions were much more likely to experience full panic attacks and/or major anxiety disorders. Those without the anxiety risk factors before the vestibular insult or development of MAD were more likely to have mild anxiety disorders and minor avoidance behaviors.[36] The recognition of this difference from a diagnostic prospective may be of assistance in the development of a treatment plan for patients with these problems.

Considerations of treatment of patients with combinations of dizziness and anxiety, independent of whether anxiety or an insult to the peripheral vestibular system is the primary lesion, start with the tools available for this population. Medications have been shown to be modestly effective in controlling both the dizziness and the psychiatric condition, primarily serotonin reuptake inhibitors.[33,37] These same medications are also suggested in the patient with anxiety, dizziness, and migraine together.[33,38] From a therapy standpoint, the development of cognitive and behavioral therapies has been shown effective for mild depressive and anxiety disorders when compared with medication.[32] The cognitive techniques work to help a patient recognize thought processes that would initiate, amplify, or sustain an anxiety or phobic behavior. The use of exposure and desensitization were part of the initial techniques for behavioral therapy. The two therapies have clear overlap and complement each other to the extent that they are often used together.[32]

VBRT techniques involve a significant amount of exposure activity, similar to that used in behavioral therapy. Although the use of VBRT is proposed primarily for promoting the compensation process, it may well serve as a desensitization technique and a form of behavioral therapy, especially in patients with mild primary anxiety disorder resulting in dizziness complaints.[4,32] The combination of cognitive, behavioral, and vestibular rehabilitation therapy techniques would appear to be a logical manner in which to deal with the complex symptom complaints in this patient group.

An attempt to test this contention was shown in a prospective, randomized control group study that used cognitive-behavior therapy combined with VBRT.[9] The cognitive-behavioral techniques were added to see whether it was feasible to combine such techniques in a group of elderly patients who were otherwise suitable candidates for a vestibular rehabilitation program. There

were significant improvements in the treatment versus no treatment control group for walking time and DHI value. Unfortunately, no changes were seen in measures of anxiety or depression. Although this study showed no detrimental effect of the combination of the therapy techniques, it is difficult to evaluate any effectiveness of the addition of cognitive-behavioral aspects because the comparison was to a no-treatment control.

A second study separated the effects of behavioral therapy and of VBRT by treating the patients with behavioral therapy first and then treating them with VBRT in an effort to see what each therapy had to offer patients with agoraphobia and vestibular dysfunction.[10] The principle outcome was an improvement in a global measure of intensity of symptoms with the behavioral therapy and no change in adjunctive indicators of anxiety and mobility. Yet after VBRT, further change in the global indicator was seen along with the adjunctive measures. This result supports the use of VBRT in patients with anxiety disorders, even though the indications for direct peripheral or central vestibular involvement are minimal.[10]

Indications to support use of general VBRT in populations of patients with the comorbidities of anxiety, panic, agoraphobia, and dizziness at the primary care level by a therapist or by a primary care nurse have been shown in two large, prospectively designed, randomized, controlled studies.[8,12] In both studies, significant changes were noted for the treatment group in physical measures and measures of well-being, including those of anxiety and depression, in comparison with the control groups.

Lastly, the form that a vestibular rehabilitation program takes may make a significant difference in the effectiveness of VBRT in patients with significant perceptual-motor changes such as those previously discussed—related to the obligatory reliance on visual cues. In a prospectively designed, controlled study, two groups of patients, all of whom had undergone a conventional form of VBRT without improvement, were started on customized vestibular rehabilitation programs. The experimental group received supplemental therapy work directed at desensitization of visual stimuli through direct exposure to a variety of provocative visual motion events under differing support conditions ranging from sitting to walking.[11] The results showed that both groups improved on all measures of outcome. However, the experimental group had a greater improvement as well as a statistically greater improvement on measures related to visual sensitivity. Although both groups improved on measures of anxiety, these improvements were seen to correlate with improvement in visual sensitivity, explaining the greater improvement in direct anxiety measures seen in the experimental group.[11] The implication of this study is that incorporation of techniques that may be difficult to develop or control in a traditional, customized home program may require a repeat-visit format for specialized stimulus exposure in specific patients, especially those with comorbid anxiety and a clear over-reliance on visual stimuli.

In summary, although the entire arena of treatment of dizziness comorbid with psychiatric disorders is just in its infancy, there appears to be reasonable support for the use of VBRT in some manner. As was the situation for migraine-related dizziness, attention in some form has to be paid to the comorbid psychiatric condition, which must be addressed directly for effective resolution of the symptoms of dizziness being expressed. It is possible that the use of VBRT alone may serve this role when it acts as a form of behavioral therapy for mild conditions of anxiety or minimal avoidance without complications of panic or agoraphobia.

## Methodological Considerations for Assessment and Treatment Development

Although current data on the use of vestibular rehabilitation techniques in the conditions discussed as "non-vestibular" are just appearing in the literature in well-performed studies, there are no indications at present that the assessment process used to develop VBRT programs should be altered for the situations discussed. The assessment process used for unilateral and bilateral vestibular hypofunction (described in this text) has focused on determination of provocative activities in the form of eye/head and/or visual motion stimuli that would cause symptoms and identification of functional deficits of balance and gait. This process also recognizes deficits of balance and gait that are the primary complaint, without the presence of transient or constant symptoms of vertigo or lightheadedness. Therefore, the use of a consistent format for assessment of these patients is appropriate in general. What does need special attention in the conditions described previously is the taking of the patient's history in whom evidence for MDD, MAD, or psychiatric disorders would first appear. Being familiar with the profile for MDD, having structured questioning for suspected MAD,[19,24] and touching on the general historical and characteristic behaviors for anxiety, panic, and phobic avoidance [31,32] will allow the therapist to further customize the overall management plan to coordinate with other recommended treatment activities

(see other discussion in this text on these disorders). It is not unreasonable, on the basis of referral patterns, that the therapist may be the first to recognize MDD or the comorbidity of migraine and/or a psychiatric condition as a significant contributor to the patient's symptoms. This recognition should prompt discussion with the referring physician for a broadening of the overall treatment plan.

Identification of patients for whom the symptom complaints are not the direct result of an insult to the balance system per se does not prevent use of a vestibular and balance rehabilitation program as an adjunct treatment for those patients. It does, however, require counseling of the patient for realistic goals and appropriate emphasis on what should be the primary treatment modality. For disorders with a natural history of spontaneous resolution, such as MDD, one must be cautious about assuming the effects of a therapy program.

Although VBRT is being shown effective, further efforts are needed to develop a better understanding of how VBRT can be integrated with other treatment methods for patients with non-vestibular system disorders. Changes in treatment programs could benefit the patients with multisensory deficits, such as those with disequilibrium associated with aging or with head trauma considered elsewhere in this text. Also possible is that new combined treatment strategies may have improved outcomes for the traditional patient with a peripheral vestibular lesion.

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CHAPTER 29

# Non-vestibular Diagnosis and Imbalance: Cervicogenic Dizziness

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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 as well as 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 due to 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 in 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 (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.[5,13]

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 utility, 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. 29.1). In theory, once the central nervous system (CNS) has adapt-

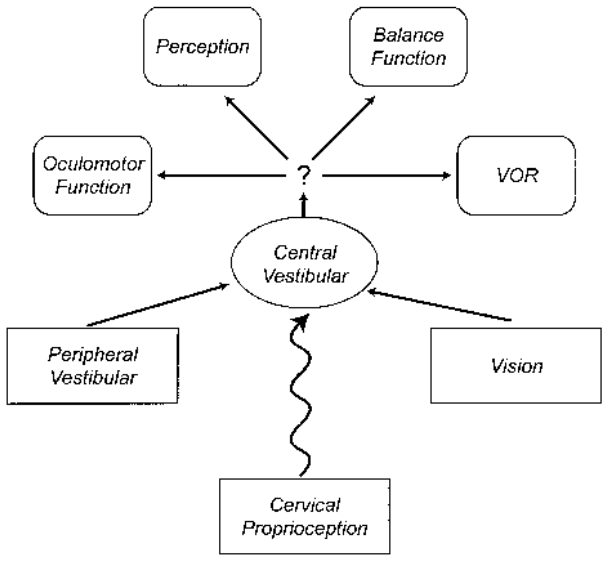


**Figure 29.1** Schematic diagram of the multisensory mismatch hypothesis. Peripheral inputs (vestibular, cervical, and visual) converge on central vestibular structures and affect oculomotor function, balance function, vestibulo-ocular reflex (VOR), and perceptions of dizziness. Cervical dysfunction (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.

ed 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.[14] 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.[15] McCouch and colleagues[15] 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.[16–21]

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.[22] Rubin and associates[23] 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.[24] 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 awake, 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), a diverse set of symptoms (including headache, dizziness, paresthesia, anesthesia, as well as cervical, thoracic, lumbar and upper extremity pain) that occur after a flexion-extension cervical injury.[25] 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[26] reported on the oculomotor effects seen after both local anesthetic injections and 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 post-injection in 10 animals by means of a rotating optokinetic stimulus (1 deg/sec$^2$ acceleration up to 200 deg/sec). Igarashi and coworkers[26] reported bidi-

rectional 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 post-injection. 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 to 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 due to 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 stimulus 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 a have an asymmetrical affect 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[26] 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.

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,[27,28] Karlberg and Magnusson[29] 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[29] conducted their study in normal humans, the results may support the findings of Igarashi and coworkers[26] after unilateral C1 and C2 dorsal root section. Recall that these 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[29] 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 cervicogenic dizziness 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[30] 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 can not 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[31] 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 disease is 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 to 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 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.[32–34] 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 to 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[32] and Treleaven and colleagues[34] noted that patients with WAD and dizziness had greater complaints of pain than patients with WAD but no dizziness. Treleaven and colleagues[34] 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 [$\rho$] = 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 changes in SPNT difference are greatest in patients with WAD, there are significant changes in the SPNT difference in individuals with other causes of cervical pain, such as cervical spondylosis and fibromyalgia, compared with normal individuals.[33] 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, but the studies have design flaws that raise questions about their validity. Tjell and Rosenhall[32] determined the sensitivity and specificity of the SPNT using 2 standard deviations from the mean of the SPNT 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, as 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%. One of the problems with this study is that it used a post hoc analysis of sensitivity and specificity, as the subjects had been diagnosed as being normal or having WAD with dizziness, and WAD but no dizziness prior to the actual testing. To appropriately determine the sensitivity and specificity of the SPNT, the test should be conducted in a blinded fashion on a mixed population that has not been previously selected and categorized as to diagnosis. In addition, in calculations of sensitivity and specificity of a test, the threshold level for normal/abnormal test results should be determined independent of data from the tested population. The SPNT test, therefore, suggests that there is some correlation between severity of WAD and changes seen in the test, although the exact nature of this correlation is not known at this time. The SPNT test shows promise, but it has not been adequately validated for use in identifying individuals with cervicogenic dizziness.

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[35] 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.[36] 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.

In summary, while 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.

### *Balance and Proprioceptive 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[35] 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 as well as 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 due to a disruption of the proprioceptive inputs.

Alund and colleagues[37] measured standing balance in patients with chronic cervical pain and associated symptoms of vertigo or imbalance, which were compared to 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 dynamic posturography (Equitest, Neurocom International Inc, Clackamas, OR). 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 as well as 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[38,39] 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 sex-matched controls, the individuals with upper quarter pain had greater velocity and increased variance of sway with eyes closed when either site was vibrated.[39]

In a subsequent study, Karlberg and colleagues[38] 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[38] 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[40] 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 0.32–0.41, $P < 0.01$) 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, as vestibular lesions were not ruled out in the subjects of the study except by patient history.

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[41] 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[42] showed a decrement in cervical kinesthesia in patients with chronic cervical pain. In addition, they found that patients enrolled in a kinesthetic retraining program in combination with medical analgesic therapy experienced improvements in kinesthetic sense and cervical rotation range as well as 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[43] 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 due to 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[43] 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[44] also noted greater cervical repositioning errors for rotation as well as 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,[45] knee,[46,47] and shoulder.[48] 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.

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.[49]

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.[50] 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 informa-

tion 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[51,52] 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.[52] 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.[53]

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 neurologic 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 neurologic causes of dizziness may also cause restricted cervical motion and neck pain owing to 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 consistently indicative of a cervical cause of the dizziness, and the clinical decision making process should first rule in or rule out otologic and neurologic causes of the symptoms. If the symptoms are due to an otologic or neurologic cause, this fact should become apparent with a comprehensive clinical examination, vestibular function tests, or radiological evaluation. If there are no apparent neurologic or otologic causes for the symptoms, one should conduct a more detailed evaluation of the upper quarter. This evaluation should include assessment of active and passive cervical range of motion, tests for instability in the upper cervical spine, neurologic examination of the upper extremities (strength, sensation, and reflexes), palpation of the cervical spine musculature, and segmental mobility testing of the cervical spine. A detailed description of these examination procedures is beyond the scope of this chapter; two specific test maneuvers, however, are addressed here.

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[54] and Childs and colleagues[55] 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.[56] 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 due to 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.[57] 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).[58] 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 in the seated position first and no signs or symptoms are noted, subsequent Dix-Hallpike test findings can be attributed to BPPV.

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 a clinical variation of the neck torsion nystagmus test,[59–61] whereby one looks not for nystagmus but for provocation of symptoms, as individuals without 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. 29.2*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. 29.2*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. It is important to perform the complete sequence in order to determine whether the symptoms originate from the cervical spine, the vestibular system, or both. A similar approach can be utilized to differentiate between vestibular and cervical causes of dizziness induced by vertical head movements (cervical flexion-extension).

Another method of assessing whether or not 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. While 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.

## 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 (due to joint restrictions or muscle tightness), hypermobility, increased muscle tone, trigger

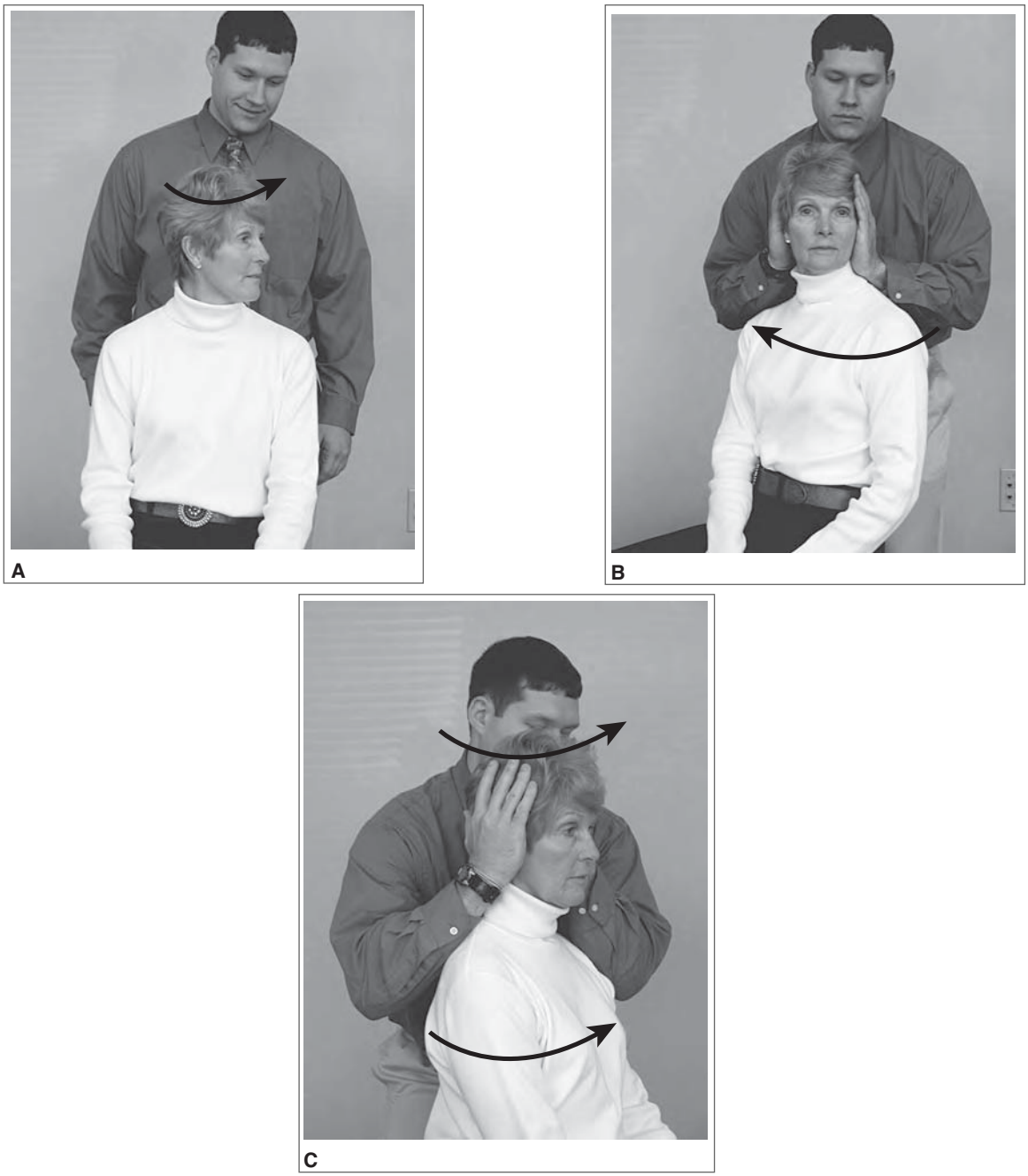


**Figure 29.2** (*A*) The patient complains of symptoms of dizziness with head rotation to the left. (*B*) To assess the cervical contribution to her symptoms, her head is stabilized in space, and she turns her body to the right. (*C*) To assess the vestibular contribution to her symptoms, she is rotated *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.

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.

Owing to 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 their hands such that the superior nuchal line of the patient's skull is resting on the therapist's fingertips (Fig. 29.3*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. A second method of performing the traction is for the therapist to cup the patient's occipital region in the palm and fingers (Fig. 29.3*B*). The therapist applies traction by leaning away from the patient.

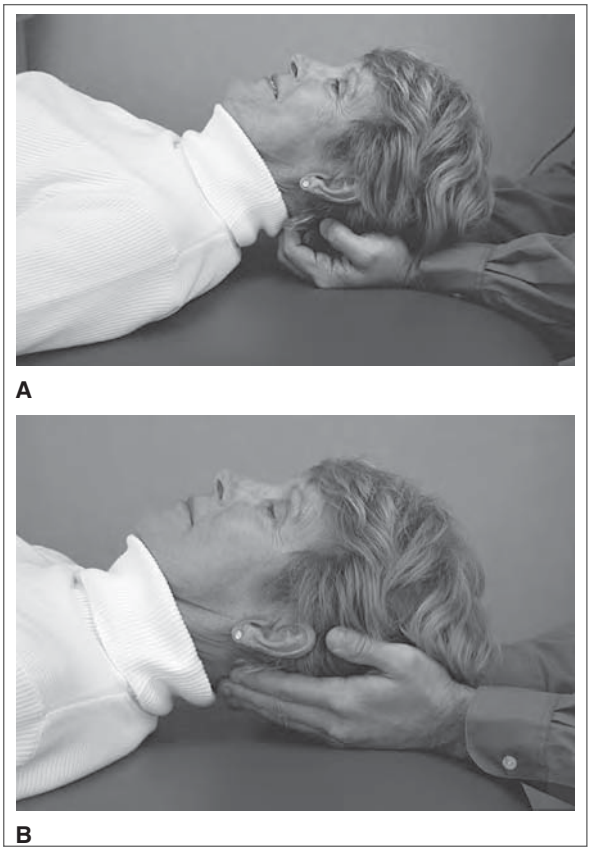


**Figure 29.3** Methods of cervical traction: (*A*) The patient rests the head on the therapist's fingertips, and the therapist can lean away from the patient, providing traction. (*B*) A variation of this technique has the therapist supporting the patient's occipital region in the palms and fingers. By leaning away from the patient, the therapist can apply traction to the upper cervical spine.

The second technique is designed to increase rotation between C1 and C2 (Fig. 29.4). 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 10–15 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 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[42] have been shown to be effective. The basic components of this exercise approach are as follows:

**Figure 29.4** Mobilization of C1–C2 to increase cervical rotation to the left.

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 utilize glasses in which all but foveal vision is blocked (Fig. 29.5*A*). Since 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 in order to accomplish the task. In order to maintain gaze on a fixed target while the trunk is rotated, head motion must be disassociated from trunk motion (Fig. 29.5*B*).

Finally, the patient can practice angle repositioning while using the foveal glasses (Fig. 29.6). 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 in order 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.

There is evidence that treatment of cervical dysfunctions can lead to decreased symptoms of dizziness and improvements in postural stability.[62] In one of the few randomized, controlled trials on this issue, Karlberg and colleagues[63] 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 owing to the variety of symptoms and physical findings. Prior to 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

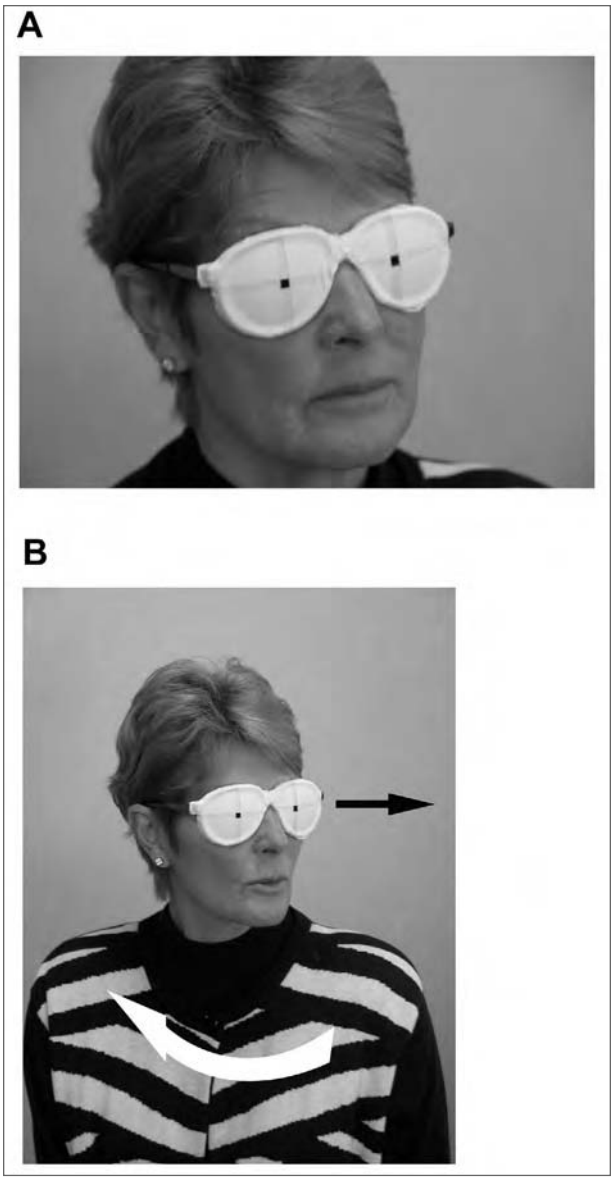


**Figure 29.5.** Cervical kinesthesia training exercises. (*A*) Foveal glasses. (*B*) Eyes maintain gaze on a fixed target (indicated by *straight arrow*) while the trunk is rotated to the right. The foveal glasses necessitate disassociation of head and trunk movements. Similar exercises can be performed with hip and trunk flexion and extension in the sitting position.

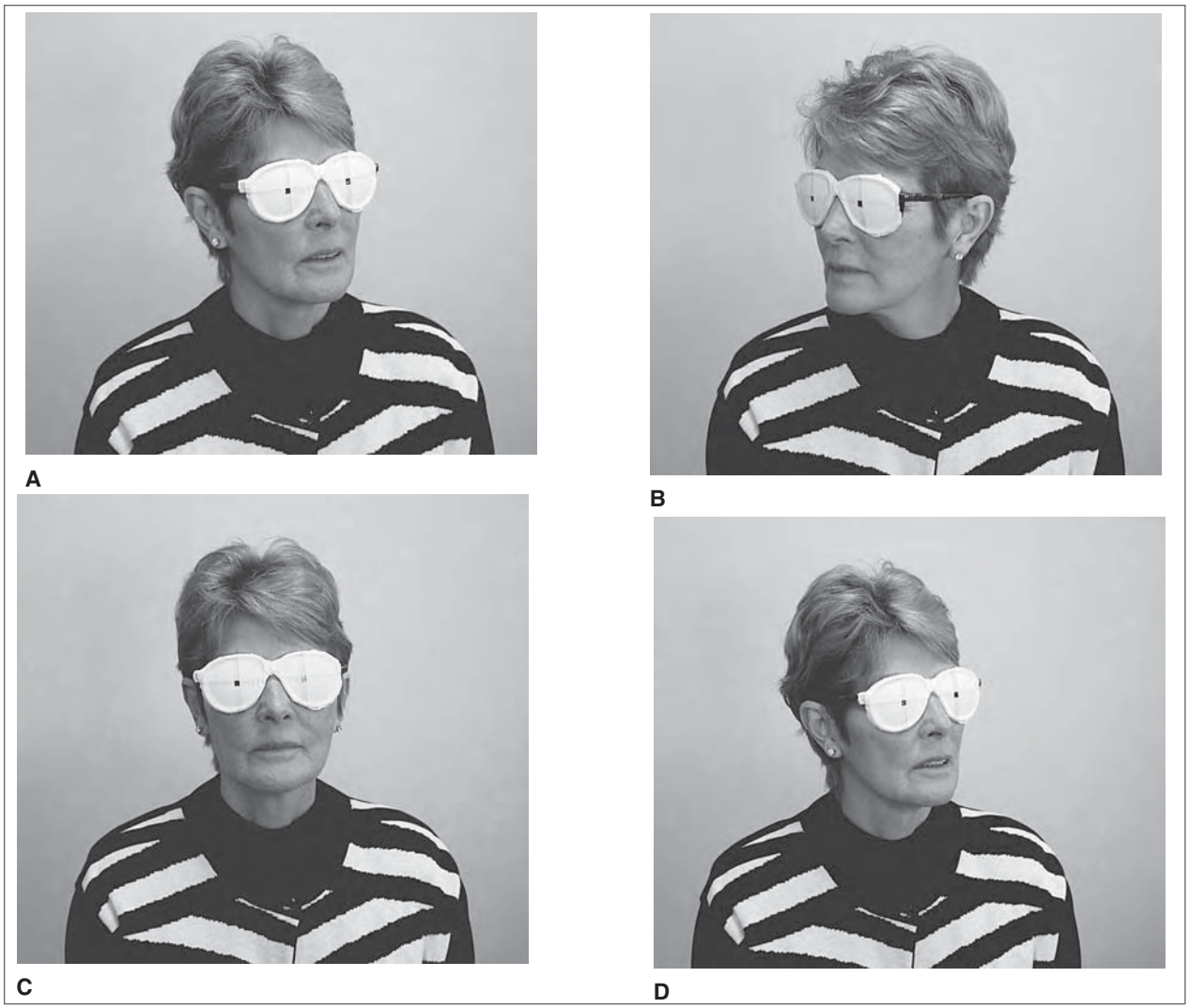


**Figure 29.6** Cervical repositioning using foveal glasses. (*A*) Gaze is fixed on a target, and head position is memorized. (*B*) The patient closes the eyes and fully rotates the neck in one direction while the eyes remain closed. (*C*) The patient attempts to return the head to the starting position while the eyes remain closed. (*D*) The patient opens the eyes and, if necessary, adjusts the head position 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.

velocities when tested with vibration applied to the posterior cervical musculature. This study and other nonrandomized controlled trials in the literature suggest that treatment of the identified cervical dysfunction in individuals with suspected cervicogenic dizziness may lead to resolution of the dizziness as well as the cervical signs and symptoms.

## 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.

CASE STUDY

## 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 prior to 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 and exertion and after sitting in one position for a prolonged period (e.g., driving or reading). Associated with the increased symptoms of disequilibrium are cervical and shoulder symptoms. 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 positive since 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.

*(continued on following page)*

## CASE STUDY *(continued)*

*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″ 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 4–6 weeks. Since 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.

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# APPENDIX A

## Questionnaire for History and Examination

**02/13/07** **QUESTIONNAIRE** **PHYSICIAN NAME, MD**

**NAME:**_____ **AGE:**_____ **DATE:** _____

**PRESENT OCCUPATION:**_____

**Name** and **address** of physician(s) you wish our report to be sent:

_____

_____

_____

**Please answer these questions to the best of your ability. PLEASE BRING QUESTIONNAIRE WITH YOU – DO NOT MAIL IT BACK. Please give necessary details for yes answers. We realize that this form is long, but when it is filled out carefully, it allows us to devote more time to your specific problem, rather than asking you related questions during your visit. If you need to CHANGE or CANCEL an appointment, please contact our office.**

Describe your major problem or the reason why you are seeing us.

Please describe in detail the circumstances and date in which the problem began 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.

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
      ( ) Poor balance
      ( ) Falls
   b. Sense of movement of the environment or of one's own body
      ( ) Rotation (spinning, tumbling or cartwheeling)
      ( ) Linear movement or pulling
      ( ) Tilt
   c. Sensations not associated with movement of the environment
      ( ) Lightheadedness or impending faint
      ( ) Floating
      ( ) Swimming
      ( ) Giddiness
      ( ) Rocking
      ( ) Spinning inside of head
      ( ) Fear or avoidance of being in public places

d. Associated symptoms
   ( ) Sweating
   ( ) Nausea
   ( ) Vomiting
   ( ) Queasiness
e. Impaired vision
   ( ) Double vision
   ( ) Blurred vision
   ( ) Flashes of light
   ( ) Jumping of vision when walking or riding in a car

**2. To what extent is your dizziness or imbalance brought on by:**

| **(Check one answer for each question.** | **None** | **Some** | **Severely** |
|---|---|---|---|
| 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** |
|---|---|---|
| Can you bring on your dizziness voluntarily? If answered yes, please describe. | | |
| Do or did you have moderate-severe motion (car or boat) sickness. If yes, when did it start? | | |
| Do or did you avoid situations in which you were tumbled or spun (amusement rides, merry-go-rounds)? When did you begin? | | |
| Has anyone observed jerking of your eyes with dizzy spells? | | |

**4. Have you ever had: (If yes, please give details.)**

| | YES | NO |
|---|---|---|
| Infections of ears | | |
| Difficulty with your hearing | | |
| Pain, fullness, popping or pressure in ear | | |
| Pain, pins/needles, numbness, twitching, or weakness of face | | |
| Crossed eyes or lazy eye. | | |
| Ringing in ears (called tinnitus)<br>If you answered yes to tinnitus, please answer the following questions. | | |
| State the frequency and duration of the tinnitus during the past 6 months | | |
| Please circle the correct answers. The tinnitus is primarily | | |
| in the left, right or both ears. It is steady, pulsating. | | |
| It is high, low pitched. | | |

**5. REVIEW OF SYSTEMS (If yes, give details.)**

| **Within the last 6 months have you noted:** | YES | NO |
|---|---|---|
| Significant loss in strength | | |
| Significant loss of energy | | |
| 10 lb. or more weight change (if yes, up__or down | | |
| Significant memory loss (amnesia) | | |
| Significant change in hand writing | | |
| Pins and needles, numbness in arms or legs | | |
| Muscle or joint aches (If yes, which muscles or joints_____) | | |
| Urinary incontinence (leakage of urine) | | |
| Problems with sleeping | | |
| Shortness of breath | | |
| Trouble chewing_____ or swallowing_____ or speaking_____ | | |
| Incoordination | | |
| Palpitations (irregular or fast beating) of the heart | | |
| Headaches | | |
| If you answered yes to headaches, please answer the following: | | |
| Approximate age they began _____; | | |
| Number per month _____; Pain intensity (1–10 with 10 the most severe) _____ | | |
| Since the onset of headaches have you had at least 5 headaches that: | | |
| 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. PAST MEDICAL HISTORY (If yes, give details.)**

Have you had any injuries due to trauma? (If yes, please describe the injury and when it occurred.)

Have you had any surgery? (If yes, please describe the surgery and when it occurred.)

| **Have you been exposed to any of the following? (If yes please, describe the exposure and when it occurred.)** | **YES** | **NO** |
|---|---|---|
| Child abuse | | |
| Intravenous antibiotics | | |
| Loud noises (guns, machinery, loud music) | | |
| Drug therapy for cancer (if yes, what type) | | |

| **Have you had any of the following infections? (If yes, give details.)** | **YES** | **NO** |
|---|---|---|
| Syphilis or venereal disease | | |
| Lyme disease | | |
| Meningitis | | |
| Other infections | | |

| **Has your past or present health been affected by: (If yes, give details.)** | **YES** | **NO** |
|---|---|---|
| Heart problems | | |
| Diabetes | | |
| Thyroid disorders | | |
| Treatment by a psychiatrist_____ or counselor_____ | | |
| Depression_____, anxiety_____, severe stress_____, phobias_____ | | |
| High cholesterol | | |
| High_____ or low_____ blood pressure | | |
| Pain in back of jaw (TMJ), grinding | | |
| Loss of consciousness (faints) | | |
| Seizures or convulsions | | |
| Arthritis | | |
| Neck pain | | |

List all major illnesses, injuries, and surgeries not described above

| 7. SOCIAL HISTORY | YES | NO |
|---|---|---|
| Do or did you use alcohol? How much? | | |
| Do or did you ever smoke: | | |
| If so how many packs/day? | | |
| What age did you start? | | |
| If you quit at what age? | | |
| How many cups of caffeinated drink per day (coffee, tea, soda)? | | |

**8. FAMILY HISTORY**

Which family members (dad, mother, children) have or had:

| |
|---|
| Headaches |
| Meniere's syndrome |
| Hearing loss |
| Vertigo or dizzines |
| Balance problems or tremor |
| Diabetes |
| Cancer or brain tumors |
| Stroke |
| Heart disease |
| High blood pressure |
| Psychiatric disorders |
| Other neurologic diseases |

If your parents are alive, what are their ages?

If your parents have died, at what age and from what cause?

**9. ALLERGIES TO MEDICATIONS and please note if drug causes rash or difficulty breathing.**

**10. 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.

| 11. 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?) | | | |

**12. MULTIDIMENSIONAL DIZZINESS INVENTORY: SECTION A**

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 |

**MULTIDIMENSIONAL DIZZINESS INVENTORY: 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 | |

# Index

An "f" following a page number indicates a figure; a "t" indicates a table, and a "b" indicates a box.

## C

**D**

**W**



**Y**