VESTIBULAR SYSTEM

Vestibular System

Balance

Work
Life

Moderation in all things as you seek the middle path

'BHealthy Balance'

For a person to live well they must find the balance of NOT tooooo much versus NOT toooo little
WHAT IS A VESTIBULAR DISORDER?

The vestibular system includes the parts of the inner ear and brain that process the sensory information involved with controlling balance and eye movements. If disease or injury damages these processing areas, vestibular disorders can result. Vestibular disorders can also result from or be worsened by genetic or environmental conditions, or occur for unknown reasons.

The most commonly diagnosed vestibular disorders include *benign paroxysmal positional vertigo* (BPPV), *labyrinthitis or vestibular neuritis*, *Ménière’s disease*, *secondary endolymphatic hydrops*, and *perilymph fistula*. Vestibular disorders also include *superior canal dehiscence*, *acoustic neuroma*, *ototoxicity*, *enlarged vestibular aqueduct*, and *mal de débarquement*. Other problems related to vestibular dysfunction include *migraine associated vertigo* and complications from *autoimmune disorders*.
PREVALENCE AND INCIDENCE

Because of difficulties posed by accurately diagnosing and reporting vestibular disorders, statistics estimating how common they are, how often they occur, and what social impacts they have range widely. Yet even the lowest estimates reflect the fact that vestibular disorders occur frequently and can affect people of any age.

One recent large epidemiological study estimates that as many as 35% adults aged 40 years or older in the United States—approximately 69 million Americans—have experienced some form of vestibular dysfunction.¹

According to the National Institute on Deafness and Other Communication Disorders (NIDCD), a further 4% (8 million) of American adults report a chronic problem with balance, while an additional 1.1% (2.4 million) report a chronic problem with dizziness alone.² Eighty percent of people aged 65 years and older have experienced dizziness,¹ and BPPV, the most common vestibular disorder, is the cause of approximately 50% of dizziness in older people.³ Overall, vertigo from a vestibular problem accounts for a third of all dizziness and vertigo symptoms reported to health care professionals.⁴

"I can't do things I used to enjoy. How can a problem with my ear affect my mood and concentration?"

Symptoms of chronic dizziness or imbalance can have a significant impact on the ability of a disabled person to perform one or more activities of daily living such as bathing, dressing, or simply getting around inside the home, affecting 11.5% of adults with chronic dizziness and 33.4% of adults with chronic imbalance.⁵ The painful economic and social impacts of dizziness are significantly underestimated.⁶

Vestibular disorders not only profoundly affect adults, but also children. Once thought to be exceptionally rare, pediatric vestibular disorders are receiving increasing attention from clinicians as an overlooked problem.⁷ In addition to impairments of motor development and balance, vestibular deficits may cause poor gaze stability that inhibits children from learning to read. Despite new awareness of pediatric vestibular disorders, children are currently not typically screened for them, and as a result frequently fail to receive medical treatment.
Labyrinthitis and Vestibular Neuritis

INFECTIONS OF THE INNER EAR

Vestibular neuritis and labyrinthitis are disorders resulting from an infection that inflames the inner ear or the nerves connecting the inner ear to the brain. This inflammation disrupts the transmission of sensory information from the ear to the brain. Vertigo, dizziness, and difficulties with balance, vision, or hearing may result.

Infections of the inner ear are usually viral; less commonly, the cause is bacterial. Such inner ear infections are not the same as middle ear infections, which are the type of bacterial infections common in childhood affecting the area around the eardrum.

INNER EAR STRUCTURE AND FUNCTION
The inner ear consists of a system of fluid-filled tubes and sacs called the labyrinth. The labyrinth serves two functions: hearing and balance.

The hearing function involves the cochlea, a snail-shaped tube filled with fluid and sensitive nerve endings that transmit sound signals to the brain.

The balance function involves the vestibular organs. Fluid and hair cells in the three loop-shaped semicircular canals and the sac-shaped utricle and saccule provide the brain with information about head movement. Signals travel from the labyrinth to the brain via the vestibulocochlear nerve (the eighth cranial nerve), which has two branches. One branch (the cochlear nerve) transmits messages from the hearing organ, while the other (the vestibular nerve) transmits messages from the balance organs.

The brain integrates balance signals sent through the vestibular nerve from the right ear and the left ear. When one side is infected, it sends faulty signals. The brain thus receives mismatched information, resulting in dizziness or vertigo.

Neuritis (inflammation of the nerve) affects the branch associated with balance, resulting in dizziness or vertigo but no change in hearing. The term neuronitis (damage to the sensory neurons of the vestibular ganglion) is also used.

Labyrinthitis (inflammation of the labyrinth) occurs when an infection affects both branches of the vestibulocochlear nerve, resulting in hearing changes as well as dizziness or vertigo.

**BACTERIAL AND VIRAL INFECTIONS**

Inner ear infections that cause vestibular neuritis or labyrinthitis are usually viral rather than bacterial. Although the symptoms of bacterial and viral infections may be similar, the treatments are very different, so proper diagnosis by a physician is essential.
**BACTERIAL**

In *serous labyrinthitis*, bacteria that have infected the middle ear or the bone surrounding the inner ear produce toxins that invade the inner ear via the oval or round windows and inflame the cochlea, the vestibular system, or both. Serous labyrinthitis is most frequently a result of chronic, untreated middle ear infections (*chronic otitis media*) and is characterized by subtle or mild symptoms.

Less common is *suppurative labyrinthitis*, in which bacterial organisms themselves invade the labyrinth. The infection originates either in the middle ear or in the cerebrospinal fluid, as a result of bacterial meningitis. Bacteria can enter the inner ear through the cochlear aqueduct or internal auditory canal, or through a fistula (abnormal opening) in the horizontal semicircular canal.

**VIRAL**

Viral infections of the inner ear are more common than bacterial infections, but less is known about them. An inner ear viral infection may be the result of a systemic viral illness (one affecting the rest of the body, such as infectious mononucleosis or measles); or, the infection may be confined to the labyrinth or the vestibulo-cochlear nerve. Usually, only one ear is affected.

Some of the viruses that have been associated with vestibular neuritis or labyrinthitis include herpes viruses (such as the ones that cause cold sores or chicken pox and shingles), influenza, measles, rubella, mumps, polio, hepatitis, and Epstein-Barr. Other viruses may be involved that are as yet unidentified because of difficulties in sampling the labyrinth without destroying it. Because the inner ear infection is usually caused by a virus, it can run its course and then go dormant in the nerve only to flare up again at any time. There is currently no way to predict whether or not it will come back.

**SYMPTOMS AND ONSET OF VIRAL NEURITIS OR LABYRINTHITIS**
Symptoms of *viral neuritis* can be mild or severe, ranging from subtle dizziness to a violent spinning sensation (vertigo). They can also include nausea, vomiting, unsteadiness and imbalance, difficulty with vision, and impaired concentration.

Sometimes the symptoms can be so severe that they affect the ability to stand up or walk. *Viral labyrinthitis* may produce the same symptoms, along with tinnitus (ringing or noises in the ear) and/or hearing loss.

**ACUTE PHASE**

Onset of symptoms is usually very sudden, with severe dizziness developing abruptly during routine daily activities. In other cases, the symptoms are present upon awakening in the morning. The sudden onset of such symptoms can be very frightening; many people go to the emergency room or visit their physician on the same day.

**CHRONIC PHASE**

After a period of gradual recovery that may last several weeks, some people are completely free of symptoms. Others have chronic dizziness if the virus has damaged the vestibular nerve. Many people with chronic neuritis or labyrinthitis have difficulty describing their symptoms, and often become frustrated because although they may look healthy, they don’t feel well. Without necessarily understanding the reason, they may observe that everyday activities are fatiguing or uncomfortable, such as walking around in a store, using a computer, being in a crowd, standing in the shower with their eyes closed, or turning their head to converse with another person at the dinner table.

Some people find it difficult to work because of a persistent feeling of disorientation or “haziness,” as well as difficulty with concentration and thinking.

**DIAGNOSIS AND TREATMENT**

No specific tests exist to diagnose vestibular neuritis or labyrinthitis. Therefore, a process of elimination is often necessary to diagnose the condition. Because the symptoms of an inner ear virus often mimic other medical problems, a thorough examination is necessary to rule out other causes of dizziness, such as stroke, head injury, cardiovascular disease, allergies, side effects of prescription or nonprescription drugs (including alcohol, tobacco, caffeine, and many illegal drugs), neurological disorders, and anxiety.

**TREATMENT DURING THE ACUTE PHASE**

When other illnesses have been ruled out and the symptoms have been attributed to vestibular neuritis or labyrinthitis, medications are often prescribed to control nausea and to suppress dizziness during the acute phase. Examples include Benadryl (diphenhydramine), Antivert (meclizine), Phenergen (promethazine hydrochloride), Ativan (lorazepam), and Valium (diazepam). Other medications that may be prescribed are steroids (e.g., prednisone), an antiviral drug (e.g., Acyclovir), or antibiotics (e.g., amoxicillin) if a middle ear infection is present. If nausea has been severe enough to cause excessive dehydration, intravenous fluids may be given.

If treated promptly, many inner ear infections cause no permanent damage. In some cases, however, permanent loss of hearing can result, ranging from barely detectable to total. Permanent damage to the vestibular system can also occur. Positional dizziness or BPPV (Benign Paroxysmal Positional Vertigo) can also be a secondary type of dizziness that develops from neuritis or labyrinthitis and may recur on its own chronically. Labyrinthitis may also
cause endolymphatic hydrops (abnormal fluctuations in the inner ear fluid called endolymph) to develop several years later.

**TESTING AND TREATMENT DURING THE CHRONIC PHASE**

If symptoms persist, further testing may be appropriate to help determine whether a different vestibular disorder is in fact the correct diagnosis, as well as to identify the specific location of the problem within the vestibular system. These additional tests will usually include an audiogram (hearing test); and electronystagmography (ENG) or videonystagmography (VNG), which may include a caloric test to measure any differences between the function of the two sides. *Vestibular evoked myogenic potentials* (VEMP) may also be suggested to detect damage in a particular portion of the vestibular nerve.

Physicians and audiologists will review test results to determine whether permanent damage to hearing has occurred and whether hearing aids may be useful. They may also consider treatment for tinnitus if it is present.

If symptoms of dizziness or imbalance are chronic and persist for several months, vestibular rehabilitation exercises (a form of physical therapy) may be suggested in order to evaluate and retrain the brain’s ability to adjust to the vestibular imbalance. Usually, the brain can adapt to the altered signals resulting from labyrinthitis or neuritis in a process known as compensation. Vestibular rehabilitation exercises facilitate this compensation.

In order to develop effective retraining exercises, a physical therapist will assess how well the legs are sensing balance (that is, providing proprioceptive information), how well the sense of vision is used for orientation, and how...
well the inner ear functions in maintaining balance. The evaluation may also detect any abnormalities in the person’s perceived center of gravity. As part of assessing the individual’s balancing strategies, a test called computerized dynamic posturography (CDP) is sometimes used.

After the evaluation, personalized vestibular rehabilitation exercises are developed. Most of these exercises can be performed independently at home, although the therapist will continue to monitor and modify the exercises. It is usually recommended that vestibular-suppressant medications be discontinued during this exercise therapy, because the drugs interfere with the ability of the brain to achieve compensation.

The exercises may provide relief immediately, but a noticeable difference may not occur for several weeks. Many people find they must continue the exercises for years in order to maintain optimum inner ear function, while others can stop doing the exercises altogether without experiencing any further problems. A key component of successful adaptation is a dedicated effort to keep moving, despite the symptoms of dizziness and imbalance. Sitting or lying with the head still, while more comfortable, can prolong or even prevent the process of adaptation.
"When the body electric kicks in and does it's job why should we be so surprised?? And call it spontaneous remission"

Desire' Dubounet

It's not spontaneous it's only educator
SCIO Disease Dictionary

TREATMENT SUGGESTED

**Color** - set patient's favorite if desired, or choose color by chakra that is deficient

**Cosmic**: set 1 for physical body, 2 for astral, 3 for etheric, 4 for mental, 5 for cosmic, 6 for other

**Magnetic Method** - 1+10 is universal, 7 for detox, 8 for regrowth of new tissue, 3 for injury, 2 for metabolic correction, 5 for inflammation, 6 for infection, 9 for psych stress, 2 for energy stimulation

**Frequency** -

Scalar for 30 min once a month in early stages once a week in later stage

Auto Trivector for 30 min once a month in early stages once a week in later stage
Title:

Part of the Following:

Large Scale Study of the Safety and Efficacy of the SCIO/Eductor Device

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Abstract:

This study demonstrates the safety and effective qualities of the SCIO device used in a large scale study. A large scale study of over 100,000 patients with over 300,000 patient visits reported their diseases. Many of them reported vestibular disease. And the results of their therapy are reported in this study. 58 were treated 53% improvement was seen.

Introduction:

Over View:

This Large scale research was designed to produce an extensive study of people with a wide variety of diseases to see who gets or feels better while using the SCIO for stress reduction and patient monitoring. The SCIO is an evoked potential Universal Electro-Physiological Medical apparatus that gauges how an individual reacts to miscellaneous homeopathic substances. The device is registered in Europe, America, Canada, S Africa, Australia, S. America, Mexico and elsewhere. The traditional software is fully registered. Some additional functions where determined by the manufacturer to be worthy of evaluation. Thus a study was necessary to determine safety and efficacy. (As a result of these studies these additional functions are now registered within the EC)

A European ethics committee was officially registered and governmental permission attained to do the insignificant risk study. Qualified registered and or licensed Biofeedback therapists where enlisted to perform the study. Therapists were enrolled from all over the world including N. America, Europe, Africa, Australia, Asia, and S. America. They were trained how to study and how to attain informed consent and transmit the results to the ethics committee, IRB (Institutional Review Board).

2,569 therapists enlisted in the study. There were 101,201 patients. 69% had more than one visit. 43% had over two visits. There were over 300,000 patient visits recorded. The therapists were trained and supervised by medical staff. They were to perform the SCIO therapy and analysis. They were to report any medical suspected or confirmed diagnosis. Therapist’s personnel are not to diagnose outside of the realm of their scope of practice. Then the therapist is to inquire on any reported changes during the meeting and on follow-ups any measured variations. It must be pointed out that the Therapists were free to do any additional therapies they wish such as homeopathy, nutrition, exercise, etc. Therapists were told to not recommend synthetic drugs. Thus the evaluation was not reduced to just the device but to the total effect of seeing a SCIO therapist.

Part 1. The emphasis was on substantiating safety followed by efficacy of the SCIO.

Part 2. Proving the efficacy of the SCIO on diseases (emphasis on degenerative disease)

Part 3. Proving the efficacy of the SCIO on the Avant Garde therapies of Complementary Med

Part 4. QQC standardization
**Methods and Materials:**

**SCIO Device:**

The SCIO is an evoked potential Universal Electro-Physiological Medical device that measures how a person reacts to items. It is designed to measure reactions for allergy, homeopathy, nutrition, sarcoodes, nosodes, vitamins, minerals, enzymes and many more items. Biofeedback is used for pre-diagnostic work and or therapy.

The QXCI software will allow the unconscious of the patient to guide to repair electrical and vibrational aberrations in your body. For complete functional details and pictures, see appendix.

**Subspace Software:**

The QXCI software is designed for electro-physiological connection to the patient to allow reactivity testing and rectification of subtle abnormalities of the body electric. If a patient is not available a subspace or distance healing link has been designed for subspace therapeutics. Many reports of the success of the subspace have been reported and thus the effectiveness and the safety of the subspace link is part of this test. Many companies have tried to copy the subspace of Prof. Nelson and their counterfeit attempts have ended in failure.

**SOC Index:**

The SCIO interview opens with a behavioral medicine interview. This is called the SOC Index. Named after the work of Samuel Hahnemann the father of homeopathy, he said that the body heals itself with its innate knowledge. But the patient can suppress or obstruct the healing process with some behavior. Hahnemann said that the worst way to interfere with the healing natural process was Allopathy or synthetic drugs. Theses upset the natural healing process by unnatural intervention and regulation disturbance. Other ways to Suppress or Obstruct the Cure are smoking, mercury amalgams, stress, lack of water, exercise and many others. This behavioral survey then gives an index of SOC.

The scores relate to the risk of Suppression and Obstruction to the natural Cure. The higher the scores the more the Suppression and or Obstruction. The scores of 100 or lower are ideal. A copy of the SOC index questions appear in the appendix.

**Study Technicians:**

The study technicians were educated and supervised by medical officers. The study technicians were to execute the SCIO therapy and analysis. All were trained to the standards of the International Medical University of Natural Education. Therapists from all over the world including N. America, Europe, Africa, Australia, Asia, S. America and elsewhere were enlisted to perform the study according to the Helsinki study ethics regulations.

They were to chronicle any medical suspected or confirmed diagnosis. Therapists personnel are not to diagnose outside of the realm of their scope of practice. Then the study technician is to inquire on
any disclosed observations during the test and on follow-ups report any measured changes.

To test the device as subspace against the placebo effect, two of the 2,500+ therapists were given placebo SCIO devices that were totally outwardly the same but were not functional. These two blind therapists were then assigned 35 patients each (only 63 showed). This was to assess the double blind factor of the placebo effect as compared to the device. Thus the studied groups were

A. placebo group,  B. subspace group,  and  C. attached harness group.

Cross placebo group manipulation was used to further evaluate the effect.

**Important Questions**: these are the key questions of the study

1. Define Diseases or Patient Concerns
2. Percentage of Improvement in Symptoms
3. Percentage of Improvement in Feeling Better
4. Percentage of Improvement Measured
5. Percentage of Improvement in Stress Reduction
6. Percentage of Improvement in SOC Behavior
7. What Measured + How (relevant measures to the patient’s health situation)
8. If Patient worsened please describe in detail involving SOC_

After the patient visit is was complete the data was e-mailed to the Ethics Committee or IRB for storage and then analysis. This maneuver minimized the risk of data loss or tampering. Case studies were reported separately in the disease analysis.

**MEDICAL DETAILS**

The most commonly diagnosed vestibular disorders include **benign paroxysmal positional vertigo (BPPV)**, **labyrinthitis or vestibular neuritis**, **Ménière’s disease**, **secondary endolymphatic hydrops**, and **perilymph fistula**. Vestibular disorders also include **superior canal dehiscence**, **acoustic neuroma**, **ototoxicity**, **enlarged vestibular aqueduct**, and **mal de débarquement**.
Results:

Before we review the direct disease improvement profiles, we need to review the overall results. The first most basic question in the results is the basic feedback of the generic patient conditions.

1. Percentage of Improvement in Symptoms
2. Percentage of Improvement in Feeling Better
3. Percentage of Improvement Measured
4. Percentage of Improvement in Stress Reduction
5. Percentage of Improvement in SOC Behavior

The SOC index gives us great insight to this study. Each disease has a different cut off where the ability of the SCIO to help was compromised. As a general index scores of 200 + where much less successful.

Urinary Incontinence

This disease group total number of patients was 59.

Subspace Treatment 4 patients, 55 SCIO Harness Patients

OVERALL ASSESSMENT

A. Subspace Treatment 7 patient visits

There were 0 cases of patients who reported a negative Improvement.

None of these cases reported any major difficulty.

There were

0 cases reporting no improvement of Symptoms, .001% of Subgroup
3 cases reporting no improvement in feeling better, .001% of Subgroup
3 cases reporting no improvement in stress reduction .001% of Subgroup

41%--- Percentage of Improvement in Symptoms
41%--- Percentage of Improvement in Feeling Better
40%---Percentage of Improvement Measured
43%-- Percentage of Improvement in Stress Reduction
B. SCIO Harness Treatment 55 patient visits

There were 0 cases of patients who reported a negative Improvement.

None of these cases reported any major difficulty.

There were

1 case reporting no improvement of Symptoms, 0.01% of Subgroup
4 cases reporting no improvement in feeling better, 0.02% of Subgroup
1 case reporting no improvement in stress reduction 0.01% of Subgroup

55%--- Percentage of Improvement in Symptoms
59%--- Percentage of Improvement in Feeling Better
53%--- Percentage of Improvement Measured
62%-- Percentage of Improvement in Stress Reduction
39%----Percentage of Improvement in SOC Behavior
Acoustic Neuroma

WHAT IS AN ACOUSTIC NEUROMA?

An acoustic neuroma (also known as vestibular schwannoma or acoustic neurinoma) is a benign (nonmalignant), usually slow-growing tumor that develops from the balance and hearing nerves supplying the inner ear. The tumor comes from an overproduction of Schwann cells—the cells that normally wrap around nerve fibers to help support and insulate nerves.

HOW DOES IT DEVELOP?

As the acoustic neuroma grows, it compresses the hearing and balance nerves, usually causing unilateral (one-sided) hearing loss, tinnitus (ringing in the ear), and dizziness or loss of balance. As it grows, it can also interfere with the facial sensation nerve (the trigeminal nerve), causing facial numbness. It can also exert pressure on nerves controlling the muscles of the face, causing facial weakness or paralysis on the side of the tumor. Vital life-sustaining functions can be threatened when large tumors cause severe pressure on the brainstem and cerebellum. Unilateral acoustic neuromas account for approximately eight percent of all tumors inside the skull; one out of every 100,000 individuals per year develops an acoustic neuroma. Symptoms may develop in individuals at any age, but usually occur between the ages of 30 and 60 years. Unilateral acoustic neuromas are not hereditary.

HOW IS IT DIAGNOSED?

Early detection of an acoustic neuroma is sometimes difficult because the symptoms related to its early stages may be subtle, if present at all. Diagnosis can be complicated because similar symptoms are common for many middle and inner ear problems.
Once the symptoms appear, a thorough ear examination and hearing test (audiogram) are essential for proper diagnosis. Computerized tomography (CT) scans, enhanced with intravenous dye for contrast, and magnetic resonance imaging (MRI) are critical in the early detection of an acoustic neuroma. These tests are helpful in determining a tumor’s location and size and in planning its microsurgical removal.

**HOW IS IT TREATED?**

Early diagnosis of an acoustic neuroma is key to preventing its serious consequences. The three treatment options are surgical removal, radiation, and monitoring. Typically, the tumor is surgically removed. The exact type of operation involved depends on the size of the tumor and the level of hearing remaining in the affected ear.

If the tumor is very small, hearing function may be preserved and accompanying symptoms may improve. As the tumor grows larger, however, surgical removal becomes more complicated because the tumor may have damaged the nerves that control facial movement, hearing, and balance, and may also have affected structures of the brain. When the tumor has affected these nerves, its surgical removal can worsen a person’s symptoms because sections of the nerves themselves may also need to be removed. In this case, vestibular rehabilitation may help promote central nervous system compensation for the inner-ear deficit.

As an alternative to conventional surgical techniques, radiosurgery with a gamma knife or linear accelerator may be employed to reduce the size or limit the growth of the tumor. Alternately, radiation therapy is sometimes the preferred option for elderly patients, patients in poor health, patients with bilateral acoustic neuroma (a tumor affecting both ears), or patients whose tumor is affecting their only hearing ear. In some cases, usually involving elderly or medically infirm patients, it may be preferable to “watch” the tumor with repeated MRIs to monitor the tumor for any growth.

**Age-related dizziness and imbalance**

**BALANCE AND FALL PREVENTION FOR SENIORS**

One of the leading health concerns for people over the age of 60 is falling, which is often related to balance problems. Each year, between 20 and 40 percent of adults over 65 who live at home fall. The consequences of falls can be disastrous; between 12 and 67 percent of elderly adults who fracture a hip die within one year. As a result, major scientific efforts are devoted to determining the causes of falling in older adults in an attempt to reduce this significant health hazard.

**CAUSES OF IMBALANCE IN OLDER PEOPLE**
Balance in walking and standing is dependent on many factors. Good balance requires reliable sensory input from the individual’s vision, vestibular system (the balance system of the inner ear), and proprioceptors (sensors of position and movement in the feet and legs). The elderly are prone to a variety of diseases that affect these systems, including cataracts, glaucoma, diabetic retinopathy, and macular degeneration, which all affect vision; diabetic peripheral neuropathy, which affects position sense in the feet and legs; and degeneration of the vestibular system.

Balance is also dependent on good muscle strength and joint mobility. A sedentary lifestyle and arthritis or diseases of bones and muscles can compromise strength and mobility.

A tendency to fall and symptoms of dizziness should not be dismissed as unavoidable consequences of aging but may be important signs of a disease that might be cured or controlled. Because balance is a complex function, there is often no single identifiable cause of falls in an elderly person. However, older people with chronic dizziness or imbalance are two to three times more likely to fall in comparison with older people who do not experience these problems.1

Symptoms of a sense of lightheadedness or disorientation (dizziness) and/or a mild to violent spinning sensation (vertigo) can have a variety of causes: vestibular (inner ear) disorders, central nervous system disorders (such as stroke), cardiac problems (including low or high blood pressure), low blood sugar, infection, hyper-ventilation associated with anxiety attacks, medication side effects or interactions between drugs, or an inadequate or poorly balanced diet. A thorough evaluation by a physician is usually necessary to help sort out these different possible causes and arrive at a correct diagnosis. This task can be even more complicated when multiple problems are present. In such cases, the trouble in any one system may not be severe, but the combined effects may be enough to cause a serious problem with balance. For example, an elderly individual with arthritis in the ankle joints and a mild degeneration in vestibular function may be able to balance adequately until undergoing an operation to remove cataracts. The disturbance in vision during the healing process and the adjustment to the new glasses or contacts may then be sufficient to result in imbalance and falls.

THE AGING VESTIBULAR SYSTEM

Most people are familiar with the problems associated with the aging of senses such as vision and hearing. However, the vestibular system is another sensory system that can also begin to function poorly with age, leading to a diminished quality of life.

The vestibular system is a complex structure of fluid-filled tubes and chambers that constitutes part of the inner ear. Specialized nerve endings inside these structures detect the position and movement of the head and also detect the direction of gravity. The signals sent from the nerves of the vestibular system are critically important to the brain’s ability to control balance in standing and walking and also to control certain types of reflexive eye movements that make it possible to see clearly while walking or running.

Anatomical studies have shown that the number of nerve cells in the vestibular system decreases from about age 55. Blood flow to the inner ear also decreases with age. Idiopathic bilateral (occurring on both sides) vestibular loss becomes more severe as age progresses. When the vestibular system is damaged by any cause, an individual may
experience dizziness and balance problems. However, the gradual, age-related loss of vestibular nerve endings can result in severe balance problems without any associated dizziness. This type of slow loss of vestibular function may be first noticed as difficulty walking or standing, especially in the dark while on soft or uneven surfaces (such as thick carpet or a forest path).

**SPECIFIC VESTIBULAR DISORDERS IN OLDER ADULTS**

Of all vestibular disorders, benign paroxysmal positional vertigo (BPPV) is one of the most common in older adults. BPPV causes vertigo, dizziness, and other symptoms due to debris that has collected within a part of the inner ear. This debris, called otoconia, is made up of small crystals of calcium carbonate (sometimes referred to colloquially as “ear rocks”). With head movement, the displaced otoconia shift, sending false signals to the brain and causing dizziness or vertigo.

Symptoms of BPPV are almost always precipitated by a change in head position. Getting out of bed and rolling over in bed are two common “problem” motions. Some people feel dizzy and unsteady when they tip their heads back to look up.

Ménière’s disease is another vestibular disorder that causes dizziness. Ménière’s disease produces a recurring set of symptoms as a result of abnormally large amounts of a fluid called endolymph collecting in the inner ear. These symptoms typically include spontaneous, violent vertigo, fluctuating hearing loss, ear fullness, and/or tinnitus.

The incidence of Ménière’s disease (number of new cases per year) is difficult to assess. Estimates vary widely, in part because of the variability in diagnostic criteria across studies. The prevalence, however (all cases within a population), is generally known to increase with age.

Other vestibular disorders that may occur in older adults include vestibular neuritis (inflammation of the vestibular branch of the vestibulo-cochlear nerve, resulting in dizziness or vertigo but no change in hearing) and ototoxicity (exposure to certain chemicals that damage the inner ear or the vestibulo-cochlear nerve, which sends balance and hearing information from the inner ear to the brain). Ototoxicity can result in temporary or permanent disturbances of hearing, balance, or both.

**PRECAUTIONS**

Although the problem of imbalance in older persons can be complex, there are a few simple precautions that everyone can follow to help ensure an active old age. Balance in standing and walking is at least partly a skill that older adults can learn to maintain and/or improve, and it is dependent on good general physical condition. Therefore, sound nutritional and health habits—including regular exercise, such as walking or participating in Tai Chi—can go a long way toward preventing balance trouble.

In older people, a regular physical examination by a doctor familiar with the problems of aging can help identify and correct potential problems before a serious fall. In addition, making sure that the elderly person’s environment is
safe (with good lighting, secure footing, clear walkways, handrails and anti-skid devices in bathrooms, etc.) can help prevent falls and their attendant injuries.

The elderly have a higher risk of contracting many different kinds of diseases. As a result, the average elderly person is more likely to have a disease that interferes with balance than a younger person. A tendency to fall and symptoms of dizziness should not be dismissed as unavoidable consequences of aging but may be important signs of a disease that might be cured or controlled. The vestibular system should not be ruled out as a source of these symptoms.

The ability to move about freely is an important factor in the quality of life for both younger and older people, and a healthy vestibular system is vitally important to freedom of movement.

Reference

1. Ko CW, Hoffman HJ, Sklare DA. Chronic Imbalance or Dizziness and Falling: Results from the 1994 Disability Supplement to the National Health Interview Survey and the Second Supplement on Aging Study. Twenty-ninth MidWinter Meeting of the Association for Research in Otolaryngology (ARO); National Institute on Deafness and Other Communication Disorders (NIDCD). Feb. 2006.

Autoimmune Inner Ear Disease

WHAT IS AUTOIMMUNITY? HOW IS IT CONNECTED TO VESTIBULAR DISORDERS?

Parts of the immune system, working constantly and behind the scenes, patrol the body in search of foreign invaders and relentlessly attack them once found. On rare occasions, in some people the immune system runs amok, identifies the body itself as foreign, and launches a lethal attack. This self-attack is referred to as an autoimmune reaction.

The immune system can attack just the ear, attack the ear and some other body part like the eye, or attack the entire body (including the ear). An autoimmune reaction also creates debris. Even if the ear is not being directly attacked, it can end up with debris transported from distant locations and deposited by the circulation. This debris in the ear can cause problems.

Some autoimmune disorders that can affect the ear include Cogan’s syndrome, relapsing polychondritis, polyarteritis nodosa, Wegener’s granulomatosis, systemic lupus erythematosus, ulcerative colitis, Sjogren’s syndrome, and rheumatoid arthritis.

Hearing loss has been viewed historically as the main inner ear effect of an autoimmune problem, but the vestibular system can also be attacked. Several factors determine the type of vestibular symptoms that may be experienced. Those factors include the speed with which the vestibular loss occurred, the degree of loss, whether one side or both
sides are affected, and whether the damage has triggered a problem with fluctuating function (for example, if endolymphatic hydrops developed from the autoimmune reaction). The symptoms of autoimmune problems can be similar, even indistinguishable, from other vestibular disorders.

Diagnosing an autoimmune disorder as the cause of inner ear symptoms can be difficult. To succeed, a physician must have training and experience in these disorders. Most otolaryngologists are not trained or experienced in autoimmune disorders in general, and a rheumatologist trained in autoimmune disorders is unlikely to be highly familiar with vestibular function. Thus gaps exist in diagnosis and treatment. In addition, if vestibular symptoms occur as part of a body-wide problem, simultaneous non-vestibular symptoms may make the diagnosis difficult.

No slam-dunk sort of diagnostic test exists for this type of ear problem. The best tests, such as the 68-kD antigen, are expensive and not widely available. Most tests can easily be positive when there isn’t an autoimmune problem and negative when there is; the tests aren’t as accurate as one would like. Sometimes the diagnosis is made only if a favorable response is seen to drug treatment.

In general, autoimmune disorders occur more frequently in women than men and less frequently in children and the elderly. When the ear is attacked, the progression of damage and functional loss is rapid, occurring over weeks to months and usually progressing rapidly to the second ear.

Autoimmune inner ear disease (AIED) is the name used to describe the variety of disorders in which the ear is the sole target of an inappropriate attack by the immune system. This disorder differs from other vestibular disorders because medical treatment can succeed when given early and aggressively. (“Early” means days to weeks or months.) An early diagnosis is important because treatment can not only stop the disease progression but in some cases can reverse the damage.

The standard treatments for autoimmune reactions are drugs that reduce immune function (immuno-suppression), and they have body-wide effects. Their use requires diligence on the part of the patient and the provider. They include steroids, chemotherapy agents, anti-transplant rejection drugs, and the newer anti-tumor, necrosis-factor drugs. The physician must be both knowledgeable and experienced with these drugs and their side effects, and the patient must follow the physician’s instructions closely. Because a rheumatologist has extensive training and experience in these treatments, nearly all otolaryngologists and neurotologists have them manage the drug treatment.

Drugs and their dosages cannot be tinkered with; side effects must be understood and watched for and never ignored. It is imperative that life on an immunosuppresser drug be understood before treatment is begun. With the proper precautions, treatment can be relatively safe, and the results can be outstanding.

General immune system research and study of autoimmune disorders is underway in many locations. Scientists are also conducting research related to autoimmune disorders and the inner ear. Most inner ear research has centered on finding an identifiable inner ear "marker" or chemical that can be tested for so that diagnosis can be faster and more accurate. Other research examines treatments. For example, the American Academy of Otolaryngology-Head and
Neck Surgery in conjunction with the National Institutes of Health is studying the treatment of autoimmune inner ear disease.

Although drug companies are not directly studying treatments for inner ear problems caused by autoimmune reactions, they are heavily involved in seeking pharmaceutical treatments of many more common and well-known autoimmune maladies.

Autoimmunity isn’t involved in most cases of vestibular disorders; however, it can cause large losses when continuing unchecked. Effective treatments are available when the diagnosis is timely.

References

Benign Paroxysmal Positional Vertigo (BPPV)

BPPV IS THE MOST COMMON VESTIBULAR DISORDER.

Benign paroxysmal positional vertigo (BPPV) is the most common disorder of the inner ear’s vestibular system, which is a vital part of maintaining balance. BPPV is benign, meaning that it is not life-threatening nor generally progressive. BPPV produces a sensation of spinning called vertigo that is both paroxysmal and positional, meaning it occurs suddenly and with a change in head position.

WHY DOES BPPV CAUSE VERTIGO?

The vestibular organs in each ear include the utricle, saccule, and three semicircular canals. The semicircular canals detect rotational movement. They are located at right angles to each other and are filled with a fluid called endolymph. When the head rotates, endolymphatic fluid lags behind because of inertia and exerts pressure against the cupula, the sensory receptor at the base of the canal. The receptor then sends impulses to the brain about the head’s movement.

BPPV occurs as a result of otoconia, tiny crystals of calcium carbonate that are a normal part of the inner ear’s anatomy, detaching from the otoletic membrane in the utricle and collecting in one of the semicircular canals. When the head is still, gravity causes the otoconia to clump and settle (Figure 1). When the head moves, the otoconia shift. This stimulates the cupula to send false signals to the brain, producing vertigo and triggering nystagmus (involuntary eye movements).
Figure 1: Inner ear anatomy. Otoconia migrate from the utricle, most commonly settling in the posterior semicircular canal (shown), or more rarely in the anterior or horizontal semicircular canals. The detached otoconia shift when the head moves, stimulating the cupula to send false signals to the brain that create a sensation of vertigo. © Vestibular Disorders Association. Image adapted by VEDA with permission from T. C. Hain.

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In addition to vertigo, symptoms of BPPV include dizziness (lightheadedness), imbalance, difficulty concentrating, and nausea. Activities that bring on symptoms can vary in each person, but symptoms are precipitated by changing the head’s position with respect to gravity. With the involvement of the posterior semicircular canal in classic BPPV, common problematic head movements include looking up, or rolling over and getting out of bed.

BPPV may be experienced for a very short duration or it may last a lifetime, with symptoms occurring in an intermittent pattern that varies by duration, frequency, and intensity. It is not considered to be intrinsically life-threatening. However, it can be tremendously disruptive to a person’s work and social life, as well as pose a health hazard due to an increased risk of falls associated with dizziness and imbalance.
CAUSES

BPPV is the most common vestibular disorder; 2.4% of all people will experience it at some point in their lifetimes. BPPV accounts for at least 20% of diagnoses made by physicians who specialize in dizziness and vestibular disorders, and is the cause of approximately 50% of dizziness in older people.

The most common cause of BPPV in people under age 50 is head injury and is presumably a result of concussive force that displaces the otoconia. In people over age 50, BPPV is most commonly idiopathic, meaning it occurs for no known reason, but is generally associated with natural age-related degeneration of the otolithic membrane. BPPV is also associated with migraine and ototoxicity. Viruses affecting the ear (such as those causing vestibular neuritis) and Ménière’s disease are significant but unusual causes. Occasionally BPPV follows surgery as a result of the trauma on the inner ear during the procedure combined with a prolonged supine (laying down face-up) position. BPPV may also develop after long periods of inactivity.

Figure 2a: Canalith repositioning procedure (CRP) for right-sided BPPV. Steps 1 & 2 of CRP are identical to the Dix-Hallpike maneuver used to elicit nystagmus for diagnosis. The patient is moved from a seated supine position; her head is then turned 45 degrees to the right and held for 15-20 seconds.

DIAGNOSIS

BPPV is diagnosed based on medical history, physical examination, the results of vestibular and auditory (hearing) tests, and possibly lab work to rule out other diagnoses. Vestibular tests include the Dix-Hallpike maneuver (see Figure 2a) and the Supine Roll test. These tests allow a physician to observe the nystagmus elicited in response to a change in head position. The problematic semicircular canal can be identified based on the characteristics of the observed nystagmus.

Frenzel goggles, especially of the type using a TV camera, are sometimes used as a diagnostic aid in order to magnify and illuminate nystagmus. If electronystagmography (ENG) is employed to observe nystagmus with position changes, it is important that the equipment used is capable of measuring vertical eye movements. A physician may also order radiographic imaging such as a magnetic resonance imaging scan (MRI) to rule out other problems such as a stroke or brain tumor, but such scans are not helpful in diagnosing BPPV. In addition, a
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Recommended treatment for most forms of BPPV employs particle repositioning head maneuvers that move the displaced otoconia out of the affected semicircular canal. These maneuvers involve a specific series of patterned head and trunk movements that can be performed in a health care provider’s office in about 15 minutes.

MANEUVERS FOR POSTERIOR CANAL BPPV

Particle repositioning head maneuvers are considered to be more effective than medication or other forms of exercise-based therapy in treating posterior canal BPPV. However, even with successful treatment with such maneuvers, BPPV recurs in about one-third of patients after one year, and in about 50% of all patients treated after five years. The canalith repositioning procedure (CRP) is the most common and empirically proven treatment for posterior canal BPPV. Also called the Epley maneuver or the modified liberatory maneuver, CRP involves sequential movement of the head into four positions, with positional shifts spaced roughly 30 seconds apart (Figure 2a and 2b). Differing opinions exist about the benefits of using mastoid vibration during CRP, with a recent evidence-based research review suggesting that it probably does not benefit patients. Occasionally, when CRP is being performed, neurological symptoms (e.g., weakness, numbness, and visual changes other than vertigo) occur, caused by compression of the vertebral arteries. In this case, persisting with the maneuver can lead to stroke. However, medical professionals can modify the exercises or use special equipment so that the positions are attained by moving body and head simultaneously, thereby avoiding the problematic compression.

The Semont maneuver involves a procedure whereby the patient is rapidly moved from lying on one side to lying on the other. Although many physicians have reported success treating patients with the Semont maneuver and support its use, more studies are required to determine its effectiveness.

Figure 2b: Canalith repositioning procedure (CRP) for right-sided BPPV (continued). In Step 3 of the CRP, the head is turned 90 degrees until the unaffected left ear is facing the floor. The patient turns her body to follow her head, and the position is held for 15-20 seconds; afterwards, she returns to a seated position. The mirror image of these maneuvers can be performed for left-sided BPPV.
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Because of the relative rarity of horizontal canal BPPV, there are no best practices established for treatment maneuvers; however, the most widely studied is the Lempert maneuver. This maneuver entails moving the head through a series of 90° angles and pausing between each turn for 10 to 30 seconds. Other techniques such as the Gufoni maneuver and the Vannucchi-Asprella liberatory maneuver have also been used to treat horizontal canal BPPV, but additional well-supported clinical studies are needed to assess their effectiveness.

MANEUVERS FOR ANTERIOR CANAL BPPV

There is no definitive treatment for anterior canal BPPV and no controlled studies of it have yet been completed. However, there is a logical modified maneuver for the anterior canal that is essentially a deep (exaggerated) Dix-Hallpike. Other proposed treatments employ reverse versions of the maneuvers used for posterior canal BPPV; for example, the reverse Semont (starting nose down and turned to the unaffected side), or the reverse Epley (again starting nose down). These treatments are geometrically reasonable, but require additional study to prove their efficacy.

POST-TREATMENT CONSIDERATIONS

After successful treatment with particle repositioning maneuvers, residual dizziness is often experienced for up to three months. Whether post-treatment activity restrictions are useful has not been adequately studied. Nevertheless, many physicians recommend that their patients sleep in an elevated position with two or more pillows and/or not on the side of the treated ear, wear a cervical collar as a reminder to avoid quick head turns, and avoid exercises that involve looking up or down or head rotation (such as freestyle lap swimming). Such precautions are thought to help reduce the risk that the repositioned debris might return to the sensitive back part of the ear before it either adheres or is reabsorbed.

OTHER BPPV TREATMENT OPTIONS

If head maneuvers don’t work, other treatment options include home-based exercise therapy, surgery, medication, or simply coping with the symptoms while waiting for them to resolve.

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Exercises performed at home are sometimes recommended. Brandt-Daroff exercises (Figure 3) involve repeating vertigo-inducing movements two to three times per day for up to three weeks. After receiving training from a doctor or physical therapist, a patient can perform the exercises at home, but they are more arduous than office treatments. With adherence to the prescribed schedule, Brandt-Daroff exercises have been reported to reduce vertiginous responses to head movements in 95% of cases. Patients performing Brandt-Daroff exercises may develop multicanal BPPV as a complication and so should note any symptom changes to their physicians. Another home exercise method is daily self-administration of particle repositioning head maneuvers. One potential problem with this method is that it may cause the condition to worsen or initiate problems in another semicircular
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![Canalith repositioning procedure (CRP) for right-sided BPPV (continued). In Step 3 of the CRP, the head is turned 90 degrees until the unaffected left ear is facing the floor. The patient turns her body to follow her head, and the position is held for 15-20 seconds; afterwards, she returns to a seated position. The mirror image of these maneuvers can be performed for left-sided BPPV.](image)

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Another home exercise method is daily self-administration of particle repositioning head maneuvers. One potential problem with this method is that it may cause the condition to worsen or initiate problems in another semicircular canal. Although one study has reported a cure rate as high as 95% for this strategy, insufficient evidence exists to recommend or refute its use.

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Figure 3: Brandt-Daroff exercises. The patient sits upright, turns her head 45 degrees to the left, then lies down quickly on her right side for 10 seconds. After returning to an upright seated position, the patient turns her head 45 degrees to the right, lies down quickly on her left side for 10 seconds, then returns to an upright seated position.
SURGERY

If head maneuvers and vestibular rehabilitation exercises are ineffective in controlling symptoms, surgery is sometimes considered. The goal of surgery is to stop the inner ear from transmitting false signals about head movement to the brain. Several surgical approaches are possible; however, a procedure called posterior canal plugging, also called fenestration and occlusion of the posterior canal, is preferable to other methods. These include removing the balance organs with a labyrinthectomy; severing the vestibular portion of the vestibulo-cochlear nerve with a vestibular nerve section, thus terminating all vestibular signals from the affected side; or severing the nerve that transmits signals from an individual canal with a singular neurectomy.

Canal plugging stops the movement of particles within the posterior semicircular canal with minimal impact on the rest of the inner ear. This procedure should not be considered until the diagnosis of BPPV is certain and all maneuvers or exercises have been attempted and found ineffective. The surgery poses a small risk to hearing; some studies show it to be effective in 85% to 90% of individuals who have had no response to any other treatment, although further research is recommended.

MEDICATION

Motion sickness medications are sometimes helpful in controlling the nausea associated with BPPV and are sometimes used to help with acute dizziness during particle repositioning maneuvers. Otherwise, medications are rarely considered beneficial. Medication that suppresses vestibular function in the long term can interfere with a person making necessary adaptations to symptoms or remaining physically active because of side-effects such as drowsiness. The American Academy of Otolaryngology—Head and Neck Surgery recommends against using vestibular suppressant medications, including anti-histamines and benzodiazapines, to control BPPV. Likewise, the American Academy of Neurology reports that there is no evidence supporting the routine use of medication to treat the disorder.

WAIT-AND-SEE

Sometimes, adopting a “wait-and-see” approach is used for BPPV. Physicians often choose to monitor patients with BPPV before attempting treatment because it frequently resolves without intervention. This may also be the approach taken with rare variants of BPPV that occur spontaneously or after maneuvers and exercises. Coping strategies during this wait-and-see phase can involve modifying daily activities to help minimize symptoms. For example, this may involve using two or more pillows while in bed, avoiding sleeping on the affected side, and rising slowly from bed in the morning. Other modifications include avoiding looking up, such as at a high cupboard shelf, or bending over to pick up something from the floor. Patients with BPPV are also cautioned to be careful when positioned in a dentist’s or hairdresser’s chair, when lying supine, or when participating in sports activities.

FINDING DIAGNOSIS AND TREATMENT FOR BPPV

A list of vestibular disorder specialists is available from on our website. This provider directory is annotated to indicate those specialists who are trained to perform canalith repositioning maneuvers.
Some helpful documents available from VEDA:

- **Inner Ear Surgeries Meant to Control Vertigo/Disequilibrium**
- **Vestibular Rehabilitation: An Effective, Evidenced-Based Treatment**

**References**


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For horizontal canal BPPV that does not respond to head maneuvers, a home treatment called forced prolonged positioning may be recommended. This requires a patient to rest in bed for at least 12 hours with the head turned toward the unaffected ear, permitting the canaliths to gradually move out of the canal. Finally, some physicians suggest that after office treatment, patients might perform a daily self-canalith repositioning exercise at home to support the treatment’s continued effectiveness. However, such home treatment probably does not affect the reoccurrence rate of posterior canal BPPV.\textsuperscript{16}

\textbf{MANEUVERS FOR HORIZONTAL CANAL BPPV}

Because of the relative rarity of horizontal canal BPPV, there are no best practices established for treatment maneuvers; however, the most widely studied is the Lempert maneuver.\textsuperscript{1} This maneuver entails moving the head through a series of 90° angles and pausing between each turn for 10 to 30 seconds. Other techniques such as the Gufoni maneuver and the Vannucchi-Asprella liberatory maneuver have also been used to treat horizontal canal BPPV, but additional well-supported clinical studies are needed to assess their effectiveness.\textsuperscript{1}

\textbf{MANEUVERS FOR ANTERIOR CANAL BPPV}

There is no definitive treatment for anterior canal BPPV and no controlled studies of it have yet been completed. However, there is a logical modified maneuver for the anterior canal that is essentially a deep (exaggerated) Dix-Hallpike.\textsuperscript{13} Other proposed treatments employ reverse versions of the maneuvers used for posterior canal BPPV; for example, the reverse Semont (starting nose down and turned to the unaffected side), or the reverse Epley (again starting nose down). These treatments are geometrically reasonable, but require additional study to prove their efficacy.

\textbf{POST-TREATMENT CONSIDERATIONS}

After successful treatment with particle repositioning maneuvers, residual dizziness is often experienced for up to three months. Whether post-treatment activity restrictions are useful has not been adequately studied.\textsuperscript{1} Nevertheless,
many physicians recommend that their patients sleep in an elevated position with two or more pillows and/or not on the side of the treated ear, wear a cervical collar as a reminder to avoid quick head turns, and avoid exercises that involve looking up or down or head rotation (such as freestyle lap swimming). Such precautions are thought to help reduce the risk that the repositioned debris might return to the sensitive back part of the ear before it either adheres or is reabsorbed.

OTHER BPPV TREATMENT OPTIONS

If head maneuvers don’t work, other treatment options include home-based exercise therapy, surgery, medication, or simply coping with the symptoms while waiting for them to resolve.

VESTIBULAR REHABILITATION HOME EXERCISES

Exercises performed at home are sometimes recommended. Brandt-Daroff exercises (Figure 3) involve repeating vertigo-inducing movements two to three times per day for up to three weeks. After receiving training from a doctor or physical therapist, a patient can perform the exercises at home, but they are more arduous than office treatments. With adherence to the prescribed schedule, Brandt-Daroff exercises have been reported to reduce vertiginous responses to head movements in 95% of cases. Patients performing Brandt-Daroff exercises may develop multicanal BPPV as a complication and so should note any symptom changes to their physicians.

Another home exercise method is daily self-administration of particle repositioning head maneuvers. One potential problem with this method is that it may cause the condition to worsen or initiate problems in another semicircular canal. Although one study has reported a cure rate as high as 95% for this strategy, insufficient evidence exists to recommend or refute its use.

For horizontal canal BPPV that does not respond to head maneuvers, a home treatment called forced prolonged positioning may be recommended. This requires a patient to rest in bed for at least 12 hours with the head turned toward the unaffected ear, permitting the canaliths to gradually move out of the canal. Finally, some physicians suggest that after office treatment, patients might perform a daily self-canalith repositioning exercise at home to support the treatment’s continued effectiveness. However, such home treatment probably does not affect the reoccurrence rate of posterior canal BPPV.

Figure 3: Brandt-Daroff exercises. The patient sits upright, turns her head 45 degrees to the left, then lies down quickly on her right side for 10 seconds. After returning to an upright seated position, the patient turns her head 45 degrees to the right, lies down quickly on her left side for 10 seconds, then returns to an upright seated position.
SURGERY

If head maneuvers and vestibular rehabilitation exercises are ineffective in controlling symptoms, surgery is sometimes considered. The goal of surgery is to stop the inner ear from transmitting false signals about head movement to the brain. Several surgical approaches are possible; however, a procedure called posterior canal plugging, also called fenestration and occlusion of the posterior canal, is preferable to other methods. These include removing the balance organs with a labyrinthectomy; severing the vestibular portion of the vestibulo-cochlear nerve with a vestibular nerve section, thus terminating all vestibular signals from the affected side; or severing the nerve that transmits signals from an individual canal with a singular neurectomy.

Canal plugging stops the movement of particles within the posterior semicircular canal with minimal impact on the rest of the inner ear. This procedure should not be considered until the diagnosis of BPPV is certain and all maneuvers or exercises have been attempted and found ineffective. The surgery poses a small risk to hearing; some studies show it to be effective in 85% to 90% of individuals who have had no response to any other treatment, although further research is recommended.

MEDICATION

Motion sickness medications are sometimes helpful in controlling the nausea associated with BPPV and are sometimes used to help with acute dizziness during particle repositioning maneuvers. Otherwise, medications are rarely considered beneficial. Medication that suppresses vestibular function in the long term can interfere with a person making necessary adaptations to symptoms or remaining physically active because of side-effects such as drowsiness. The American Academy of Otolaryngology—Head and Neck Surgery recommends against using vestibular suppressant medications, including antihistamines and benzodiazepines, to control BPPV. Likewise, the American Academy of Neurology reports that there is no evidence supporting the routine use of medication to treat the disorder.

WAIT-AND-SEE

Sometimes, adopting a “wait-and-see” approach is used for BPPV. Physicians often choose to monitor patients with BPPV before attempting treatment because it frequently resolves without intervention. This may also be the approach taken with rare variants of BPPV that occur spontaneously or after maneuvers and exercises. Coping strategies during this wait-and-see phase can involve modifying daily activities to help minimize symptoms. For example, this may involve using two or more pillows while in bed, avoiding sleeping on the affected side, and rising slowly from bed in the morning. Other modifications include avoiding looking up, such as at a high cupboard shelf, or bending over to pick up something from the floor. Patients with BPPV are also cautioned to be careful when positioned in a dentist’s or hairdresser’s chair, when lying supine, or when participating in sports activities.

FINDING DIAGNOSIS AND TREATMENT FOR BPPV

A list of vestibular disorder specialists is available from on our website. This provider directory is annotated to indicate those specialists who are trained to perform canalith repositioning maneuvers.
ADDITIONAL RESOURCES

Some helpful documents available from VEDA:

- **Inner Ear Surgeries Meant to Control Vertigo/Disequilibrium**
- **Vestibular Rehabilitation: An Effective, Evidenced-Based Treatment**

References


Reasons for Facial Nerve Damage

1. Reasons for Facial Nerve Damage

   NF2 results in hearing and balance issues in 90% of people. This is the result of VS (Vestibular Schwannoma), Schwannoma tumors on the Vestibular Nerve, often till the nerves are destroyed.

   The Vestibular Nerve is very close to the Facial Nerve and many individuals with VS damage also have Facial Nerve damage they are likely to have some degree of Facial Paralysis.

   Damage to the Facial Nerve can happen as a result of:

   - Growth of a Vestibular Schwannoma
   - Surgery / Microsurgery
   - Radiation Treatment

   For individuals with NF2, medical treatments are often necessary because a tumor is life threatening, other treatments are necessary to help individuals maintain a normal life, an example of that would be for the prevention of hearing loss. Since there is currently no treatment to destroy all tumors developed as a result of NF2, medical treatment is only a matter of tumor management.

   The facial nerve can easily be broken or otherwise weakened, during treatment management of Vestibular Schwannoma. There are different treatment techniques surgery, radiation and chemotherapy trials for different approaches to manage tumors. Options are often based on; exact tumor location, tumor size, a tumors shape and rate of growth, in addition to; an individual's age, height, weight and general health. Some of the different treatment options can ultimately lead to damage to the facial nerve.

2. About the Facial Nerve
Vestibular Shwanoma, Shwanoma tumors along Cranial Nerve 8, are an issue that develops in many individuals with NF2.

Cranial Nerve 8 travels a similar path as Cranial Nerve 7 (the Facial Nerve), extending out of the brainstem right next to each other. The purpose of CN8 is hearing and balance.

Learn more about Hearing and Balance: [CN8 - Vestibulocochlear Nerve](#).
Major Nerve Branches Outside of the Skull

CN7 - Facial Nerve
Paths of the 5 Exterior Branches

The facial nerve branches off to smaller nerves and muscles that go to 5 different parts of the face. Therefore, when the nerve is damaged those smaller veins are not supplied with enough blood for circulation which is necessary for muscles on the different areas of the face to move. Each nerve branch affects the movement of different muscles.

The image to the right and the description shows how far the Facial Nerve reaches.

1. Temporal Branch
2. Zygomatic Branch
3. Buccal Branch
4. Marginal Mandibular Branch
5. Cervical Branch
1. Temporal Branch - (Frontal Branch): This Nerve Branch affects the muscles in the Forehead.
2. Zygomatic Branch - (Malar Branches): This Nerve Branch affects the Upper Cheek.
1&2. Temporal & Zygomatic Branch: Together these Nerve Branches affect the muscles control opening and closure of the Eye.
3. Buccal Branch - (Infraorbital Branches): This Nerve Branch affects the Cheek and Above the Mouth Muscles.
4. Marginal Mandibular Branch: This Nerve Branch affects the Chin Muscles.
5. Cervical Branch: This Nerve Branch some of the Neck Muscles.

The nerves in the picture on the right show where on the face, outside of the skull, that the nerve branch extends out. The X in the image is pointing is the approximate location where the nerve extends outside of the skull, before branching to the different parts of the face.

3. Facial Nerve Damage Issues

- **Eyes:** The nerves from the Zygomatic Branch results in eyelid problems. This nerve controls the ability or lack thereof to blink or produce tears. Normally blinking coats the eye with a tear each time, without the ability to do so different eye creams and eye protection is needed, or the cornea will crack and permanently damage vision in the eye. Learn about [Facial Nerve Damage and Dry Eyes](#).
- **Eating:** Without the ability to move the Buccal branch and the Marginal Mandibular Branch, holding food in your mouth becomes very frustrating, and awkward. Drinking with a straw is often necessary.
- **Talking:** The same nerves that make eating difficult can also make properly or clearly pronouncing certain letters hard to do. Letters like - B, P, M and W.
- **Droopy Face:** Lack of complete eyelid closure and a fallen smile.
- **Nasal Issues**

4. Common Questions

**Question 1:** Why is the Facial Nerve Damaged during VS (Vestibular Schwannoma) surgery?

Tumors that grow on the nerves that effect hearing and balance tend to push on the Facial Nerve which follows a similar path inside the brain. During different tumor management techniques used to preserve hearing and balance the Facial Nerve is lost.

**Question 2:** How can Facial Nerve damage be prevented?

Focused radiation treatments typically damage other nerves in the surrounding area, but different Microsurgery surgery techniques to remove VS are less likely to damage the Facial Nerve. However, if the tumor is already affecting the Facial Nerve, the chances of saving the nerve regardless of treatment choice is minimal.

**Question 3:** In addition to Dry Eyes, what eye issues does Facial Nerve damage result in?

Diplopia (Double Vision or Lazy Eye). Learn about [NF2 Eye Issues](#).
**Question 4: Why are other face controls not mentioned here?**

The Facial nerve does not control all of the functions of the face and are controlled by **Cranial Nerve 5, the Trigeminal Nerve** which includes the following 3 branches:

- **Ophthalmic Nerve (V1):** Muscles for the eyelids, eyebrow, forehead and nose.
  This branch affects the glands for tear production and mucous membrane of the nasal cavity.
- **Maxillary Nerve (V2):** Sensation from the maxillary, nasal cavity, sinuses and taste.
- **Mandibular Nerve (V3):** Face Sensations and Muscles for biting, chewing, and swallowing.

**Question 5: Is pain a side effect of Facial Nerve Damage?**

No. Pain or any other feeling, numbness or lack of feeling in the face is the result of damage of **CN5, the Trigeminal Nerve.**

5. **Healing Time**

Nerves that are a part of the Central Nerve System (CNS) do not heal as easily as nerves in other areas of the body. There are things that can be done to help the nerve heal that should be considered when a nerve might or has been damaged.

Natural healing that might take place will not visually be seen till about 4 to 6 months after the damage occurs. Everyone is different, so it might take more or less time for different people, but the majority of the healing that will occur on its own will be in the first year. The nerve damage will unlikely be completely restored on its own but there are some options to consider to help maximize the amount of healing.

6. **Things that Can Aid in Healing**

A few things can help encourage the most amount of healing:

- Facial Nerve Physical Therapy
- Acupuncture
- Food & Herbs or Supplements
- Facial Reanimation Surgery

Non-surgical options when possible should be considered before surgical options, but if after time passes those do not do enough for you Facial Reanimation is an option you may want to consider.

7. **Facial Nerve Physical Therapy**

Facial nerve physical therapy can consist of different types of treatments which should be discussed with a physical therapist.

3 forms of possibly physical therapy include:

- **Muscle Exercise Training** - It is suggested to visit a physical therapist as soon as possible after the nerve is damaged for the best and most amount of results, including the prevention
of Synkinesis (Movement that results in an involuntary contraction of other facial muscles. Example - the eye muscles causing the eye to squint when smiling)

- **Massage Therapy**
- **Electrotherapy** - There are different forms of electrical stimulation and it is usually not considered until well after neuropathy has set in. The stimulation jolts all the facial nerves at the same time and can make it harder to control different parts of the face individually, which is required for natural facial expressions. Methods on this might include; Electrical Stimulation (ES), Electromyography Biofeedback (EMG Bio), Ultrasound, Laser, or Short-Wave Diathermy (SWD)

8. Acupuncture

Acupuncture helps to stimulate blood to flow back to the different nerves in the face to encourage movement. For maximum results, starting it within a few months after a nerve is damaged is ideal; each weekly treatment will help encourage the blood flow back into the face to help regain control little by little.

**Acupuncture Warnings!**

- It is important to get Acupuncture from someone who is a doctor.
- Many offices that do Acupuncture also do Back Massage to ease back pain and is very dangerous for individuals with spine tumors and should not be done to any of us with NF2.
- It is very important to go to a medical professional, a certified doctor, for acupuncture. Needles placed in the wrong location can cause serious harm or even kill the patient being treated. - [Read More]

9. Food and Supplements

Antioxidants help encourage blood flow by helping to send more oxygen to veins; as a result they increase the circulation through the entire body.

A few glasses of water will also help with overall circulation. If you drink anything that dehydrates you like coffee or alcohol, you need to drink more than the standard amount of water for your body size.

Remember, all things should be taken in moderation.

Cayenne Pepper, Ginkgo Biloba, Garlic and Hawthorn Berry.

**Neuro-Otology - Vestibular Disorders**

**History**

Vertigo, strictly defined, refers to an hallucination of movement. When the symptom complex is one of spinning or rotation, the cause is almost always the inner ear or peripheral vestibular system. Although some patients experience a definite sense of environmental spin or self-rotation, most do not present solely with true spinning vertigo. The most
common complaint is *dizziness*, a term that represents a variety of symptoms (Table 7.1). The examiner should elicit an exact description of what the patient is experiencing. Is it a spinning sensation that could be characterized as vertigo, pointing to the peripheral vestibular apparatus? Is it a sensation of falling without rotation? Is it a sensation of unsteadiness or imbalance? Is there a particular direction in which the patient tends to fall? When the patient's complaint is actually incoordination or clumsiness, the cause may be cerebellar dysfunction or peripheral neuropathy. When the symptom complex is "lightheadedness" or "swimmy-headedness," the examiner should think of presyncope or syncope and consider systemic factors such as postural hypotension, vasodepressor syncope, or cardiac arrhythmia.

<table>
<thead>
<tr>
<th>Table 7.1. SYMPTOMS ENCOMPASSED BY THE TERM DIZZINESS</th>
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<tbody>
<tr>
<td>Vertigo</td>
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<tr>
<td>Unsteadiness</td>
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<tr>
<td>Imbalance</td>
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<tr>
<td>Spinning</td>
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<tr>
<td>Floating</td>
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<td>Fainting</td>
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<td>Lightheadedness</td>
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<td>Swaying</td>
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<td>Twisting</td>
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<td>Blurring vision</td>
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<tr>
<td>Disorientation</td>
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<td>Poor equilibrium</td>
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After trying to define the true qualitative nature of the symptom complex, one must proceed to a consideration of temporal factors. Is the patient's experience a continuous one? Are there episodes of severe symptomatology with symptom-free intervals? If the symptoms are episodic, do they occur only when the patient is upright?

Patients often have difficulty describing their symptoms. Initially, it is important to have patients provide their own descriptions before the examiner biases the outcome by suggesting descriptive phrases. Some patients are asked to describe their symptoms without using the word *dizziness* cannot further characterize the symptoms.

In addition to determining whether the symptom complex is episodic, one should define duration, length of symptoms, and any associated complaints such as tinnitus, hearing loss, double vision, slurred speech, numbness, or paralysis. A history of episodic disequilibrium accompanied by diplopia, slurred speech, perioral numbness, dimming of vision, and occasional drop attacks would suggest transient vertebrobasilar ischemia. Are there associated symptoms such as headache, and have these occurred at earlier times? If the patient experienced severe episodes of imbalance in early life, followed by occipital or generalized headaches, especially throbbing, the history would suggest basilar artery migraine. Did the dizziness follow head trauma, a systemic illness accompanied by aminoglycoside antibiotic therapy, or a mild upper respiratory infection? Episodic positional vertigo with brief episodes of spinning while turning over in bed suggests a common condition, benign paroxysmal positional vertigo (BPPV). Did the symptom complex occur following ear surgery or infection, deep-sea diving, or a concussive blow to the ear? Such a history, with or without hearing loss, would suggest a perilymph fistula.
There are a significant number of patients whose balance disorders are aggravated or even caused by anxiety. If the disequilibrium or dizziness is of long duration, it is often difficult to tell whether the symptom complex is caused by anxiety or depression or whether the anxiety or depression are secondary to the dizziness. One always tries to make a positive diagnosis of a neurosis or chronic anxiety disorder on the basis of other symptomatology and historic information. There may be a history of previous episodes of serious depression or anxiety attacks, and these should be elucidated before concluding that dizziness is secondary to anxiety.

Neurologists and neuro-otologists follow a large number of patients with chronic vertiginous sensations who remain undiagnosed. These patients complain of constant or intermittent disequilibrium, often aggravated by position change, as well as by visual stimuli such as moving traffic and patterned wallpaper or by passing food displays in supermarkets. Many of these patients have become agoraphobic; they hesitate to leave their homes and particularly fear driving a car that will be passed by other automobiles. Some of these persons have had a single attack of acute peripheral vestibulopathy but have never made appropriate central compensation or adapted to their peripheral abnormality. Although mechanisms for compensation remain unclear, most patients, particularly those younger than 30, rapidly recover from an acute peripheral vestibulopathy. Elderly patients or patients with a previously existing intrinsic brainstem abnormality will rarely make adequate compensation for an acute peripheral vestibulopathy. These patients continue to complain of severe disequilibrium and have exacerbated symptoms with a variety of visual inputs. They often have completely normal examinations and vestibular tests.

Figures 7.1 and 7.2 illustrate what might happen following an acute peripheral vestibular abnormality. In some individuals, as diagramed in the right-hand panel of Figure 7.2, there is decreased ability to compensate for peripheral vestibular abnormality. One possibility would be a congenital inability to make CNS compensation, but others include (a) an acquired central inability to compensate due to CNS lesions, as from multiple sclerosis or previous brainstem stroke; (b) a fluctuating peripheral vestibular problem, as in Ménière’s disease; (c) relative inactivity without much afferent input; and (d) a peripheral vestibular apparatus providing inaccurate, although nonfluctuating, afferent information. Careful history taking may reveal childhood meningitis, a remote head injury, or particular susceptibility to motion sickness in childhood. During history taking, these possibilities should be explicitly sought.
Figure 7.1. Left, Vestibular afferent input during normal horizontal head rotation to the right. Increased firing rate from right peripheral vestibular apparatus. Ocular deviation shows slow-phase deviation to the left. VN, vestibular nuclei. (Adapted from Baloh 1984 and Daroff 1977). Right, Acute left peripheral vestibulopathy with resultant acute vertiginous sensation simulating head rotation to the right. Slow-phase ocular deviation to the left (small arrow) and fast phase of nystagmus to the right (bold arrow) and away from the side of the peripheral vestibular injury.
Examination of the Dizzy Patient

GENERAL EXAMINATION

Every patient with a disorder of equilibration or true vertigo should have a screening general physical examination. Patients who exhibit symptoms suggesting presyncope or actual syncope must have particular attention paid to their cardiovascular systems. Not only should patients have their blood pressure measured in the resting, sitting, and standing position, but they also should have their blood pressure measured at 1-minute intervals up to 5 minutes after assuming the upright position, as delayed postural hypotension is not uncommon. Exercise-induced hypotension is an important observation and should lead to consideration of conditions such as the Shy-Drager syndrome, diabetic autonomic neuropathy, and cardiac defects such as aortic stenosis and obstructive cardiomyopathy. Whenever episodic symptomatology is associated with a question of alteration of consciousness or lightheadedness, particular attention should be paid to the possibility of cardiac dysrhythmia. Most patients with cardiac dysrhythmias do not report associated sensations of irregular heartbeat, thumping in the chest, or fluttering; however, examination may reveal an irregular cardiac rhythm or cardiac murmur.
During the general examination, attention should be paid to systemic conditions that could give rise to a general feeling of malaise or weakness interpreted by the patient as a disorder of balance. Conditions leading to sudden syncope may be revealed on the general physical examination. Patients with suspected extracranial vascular disease not only should have the head and neck auscultated for bruits, but also should have a general examination of the peripheral vascular system, including the cranial and carotid pulses and evaluation for significant varicose veins that might lead to venous pooling and hypotensive episodes.

The neurologic examination should be directed by the patient's history. In patients with clear-cut episodic vertigo, the neurologic examination will usually be normal, with the exception of the ocular motor findings to be described. However, when the patient's symptom complex is more vaguely defined and includes disequilibrium or unsteadiness, particular attention must be paid to examination of the motor system, reflexes, sensation, and cerebellar function.

All patients with undiagnosed disorders of equilibration, however described, should have a complete neurologic examination. Portions of the neurologic examination are described briefly below, followed by suggestions of which entities might cause abnormality.

**MENTAL STATUS EXAMINATION**

Signs of diffuse alteration in consciousness may suggest overmedication, metabolic encephalopathy, or an acquired dementing process. Focal disturbances in intellectual function, such as a subtle aphasia, may lead to the consideration of multi-infarct dementia with accompanying brainstem infarctions or of a mass lesion in the dominant hemisphere.

**CRANIAL NERVE EXAMINATION**

Alterations in visual sensory function can be a primary or exacerbating cause of disequilibrium. Even the recent addition of a new refractive correction, particularly lenses for presbyopia, may be an added or primary cause of imbalance. Visual field defects such as unsuspected bitemporal or homonymous field defects from infarcts or tumors may cause patients to run into objects or feel disoriented in space. The presence of papilledema or absent venous pulsations on funduscopy should be an immediate clue to raised intracranial pressure. Altered corneal sensation can be the clue to a previously unsuspected cerebellopontine angle mass. Simple auditory screening tests may reveal a previously unsuspected hearing loss and should always lead to formal audiologic testing. Abnormalities on examination of cranial nerves IX through XII raise the differential diagnosis of multiple cranial neuropathies caused by collagen vascular disease, tumors of the base of the skull, or nasopharyngeal carcinoma.

**OCULAR MOTOR EXAMINATION**

The presence of spontaneous or induced nystagmus is of critical importance in the diagnosis of peripheral, central, or systemic causes of imbalance. Nystagmus types of particular note are described in the section on the directed neurootologic examination. Defective downward gaze is often the first condition often accompanied by disequilibrium. The presence of asymmetric slowing of the adducting eye indicating an internuclear ophthalmoplegia is a subtle but important clue to the presence of brainstem multiple sclerosis, brainstem infarction, or mass lesion of the posterior fossa.

**MOTOR SYSTEM EXAMINATION**
The examination of motor function can reveal focal or diffuse weakness indicating CNS or neuromuscular disorders. A subtle hemiparesis may be the true cause of the patient’s balance complaint. Diffuse hyperreflexia reflects cerebral or spinal cord dysfunction and, in combination with cerebellar abnormality, might lead to the diagnosis of a spinocerebellar degeneration.

SENSORY EXAMINATION

Examination of sensation can reveal a significant peripheral neuropathy leading to a diagnosis of diabetes or toxic neuropathy. Selective loss of sensory modalities conveyed by the posterior column, such as proprioception and vibration, may indicate that the patient has vitamin B12 deficiency or early tabes dorsalis. Such patients are relatively steady during the Romberg test with eyes open but rapidly lose balance and fall in any direction when visual compensation is eliminated by eye closure.

CEREBELLAR SYSTEM EXAMINATION

Obvious limb or body ataxia should be an immediate clue to the CNS abnormality as the cause for the patient’s imbalance. Unsteadiness during Romberg testing with eyes open and only slight exaggeration on eye closure indicates a cerebellar abnormality. Cerebellar dysfunction is usually accompanied by abnormality during gait testing or even difficulty maintaining balance while seated. Patients with symptomatic peripheral vestibulopathy tend to fall toward the side of the abnormality during eye closure with the head straight ahead. Unilateral limb ataxia is almost always an indicator of focal posterior fossa abnormality, such as infarct, demyelination, abscess, or tumor.

DIRECTED NEURO-OTOLOGIC EXAMINATION

A directed neuro-otologic examination should be performed, particularly when there are abnormalities of the auditory, ocular motor, and vestibular systems. Audiologic testing is discussed below. During the neurologic examination, there may be subtle signs of peripheral vestibular dysfunction indicated by nystagmus. On external examination, the nystagmus fast phase is away from the ear with the vestibular abnormality. During the funduscopic examination, particular attention should be paid to the movement of the optic disc. A rhythmic, subtle, horizontal, slow- and fast-component nystagmus is frequently present in patients with new peripheral vestibular dysfunction. The nystagmus is brought out by reducing fixation during the funduscopic examination. For example, with the patient staring at a dimly lit target in the distance, the presence of a slow ocular drift to the left and a fast phase to the right of the optic disc should indicate to the examiner that the patient has a subtle left beating nystagmus in the primary position. The findings indicate a right peripheral vestibular abnormality. Fast, upward, rhythmic, vertical movement of the optic disc seen during funduscopic examination signifies the presence of downbeat nystagmus. The examiner should then search carefully for the presence of downbeat nystagmus during examination of oblique and downward gaze. The need to search for presence of any type of nystagmus during the directed neuro-otologic examination cannot be overemphasized. The directed neuro-otologic examination should include a detailed otoscopic examination of the external auditory canal and the tympanic membrane. The presence of a retracted or scarred eardrum suggests prior middle ear infection. The presence of a blue mass behind the tympanic membrane points to a glomus jugular tumor.

The patient should be tested for balance during standing, walking, and turning, and for the presence of past-pointing. Past-pointing is a tendency for the repetitively elevated and lowered outstretched fingers to drift unidirectionally. Past-pointing is a clear indication of tonic imbalance in the vestibular system. If, during Romberg testing, the patient tends to fall in a certain direction, can this direction be altered by changing head position? The ability to alter the direction of the fall during Romberg testing by head turning indicates a peripheral vestibular abnormality. For example, a patient with an acute left peripheral vestibulopathy will tend to fall to the left during eye closure with the head straight ahead,
but will fall backward (toward the abnormal ear) with the head turned left, and will fall forward during eye closure when the head is turned to the right.

The physician should test for the presence of an intact vestibulo-ocular reflex (VOR) and observe whether the patient is able to maintain steady ocular fixation during funduscopic examination as the head is gently rotated from side to side. The patient with an intact VOR can still maintain fixation on distance objects during head turn. The absence of this ability produces an apparent nystagmus, most easily observed during funduscopic examination, which is good evidence for a defective VOR. A different test of vestibulo-ocular control is for the patient to fix on his or her own thumb while rotating the head in the same direction. During this maneuver, the patient must suppress the vestibulo-ocular response to permit combined head and eye tracking. The loss of this ability may be a subtle clue to cerebellar system dysfunction. The patient should also be examined for the presence of nystagmus when visual fixation is reduced by wearing Frenzel glasses, which blur the patient's vision. The lenses also magnify the eye, allowing better detection of low-amplitude nystagmus.

If the patient has no cervical problems, a head-shaking test can be performed. The patient is asked to shake the head rapidly 20 times back and forth while wearing Frenzel glasses. Then the patient is observed to determine whether there is any primary position horizontal nystagmus. The maneuver may bring out a latent nystagmus indicating vestibular imbalance. The fast phase of the nystagmus would be away from the ear with the peripheral vestibulopathy.

**Differential Diagnosis of Dizziness**

Because ongoing or episodic conditions accompanied by vertigo, unsteadiness, or presyncope are produced by multiple and often subtle causes, it is not surprising that a significant number of patients cannot be readily diagnosed. A major differential diagnostic classification would include broad categories such as 

(a) peripheral vestibulopathy, 
(b) central neurologic disorders, and (c) systemic or medical conditions. There is some ambiguity in the use of the term central, which has been used by otolaryngologists to include causes that are central or proximal to the vestibular end-organ and therefore include the vestibular portion of the eighth nerve. Neurologists, however, consider conditions that affect the vestibular nerve, such as tumors, as peripheral in location because they are on a cranial nerve and are extraaxial. Because masses or neoplasms can enlarge to involve other structures in the cerebellopontine angle, particularly the brainstem, conditions that affect the eighth nerve are discussed for convenience in the central category.

**Peripheral Causes of Vertigo**

Peripheral causes result from dysfunction of vestibular end-organs (semicircular canals, utricle, and saccule) (Table 7.2).
Peripheral vestibulopathy encompasses terms such as vestibular neuronitis, labyrinthitis, and viral neurolabyrinthitis. Such terms imply an inflammatory mechanism, which is unproved. Vestibular neuronitis, strictly speaking, is characterized by single or recurrent sudden episodes of true vertigo lasting from hours to days and often associated initially with vomiting. When the condition is associated with hearing loss, the entire labyrinth is assumed to be involved, and the term labyrinthitis is used. Despite this technical distinction, many neuro-otologists, otologists, and neurologists use the terms vestibular neuronitis and labyrinthitis interchangeably, whether or not auditory symptoms are present. In such patients the vertiginous sensation may be provoked by head movement, but not necessarily by a particular head position.

Whether isolated viral involvement of the vestibular nerves is a cause of acute or episodic vertigo is controversial. Many prefer the term acute or recurrent peripheral vestibulopathy. In the acute phase, many patients present with sudden severe vertigo, nausea, and vomiting, without any hearing disturbance or facial weakness. The acute symptoms usually resolve in a few days to a week but may recur in weeks or months. If true vertigo is part of the symptom complex, the condition is most likely to be associated with some disorder of the peripheral end-organ. However, patients with either acute peripheral vestibulopathy or, more commonly, recurrent attacks may experience only a sensation of lightheadedness or floating or a feeling of "walking on tennis balls." Even if the patient has had hundreds of episodes, it is important to determine whether any of them were associated with spinning vertigo. Over time, the nature of the patient's symptom complex may change, even with peripheral vestibulopathy, from vertiginous sensations to those of pure unsteadiness or disequilibrium.

Epidemic and seasonal outbreaks of acute vertigo have suggested an infectious origin due to viral disease, but this largely remains unproved. Viral labyrinthitis can also be part of a systemic viral infection such as mumps, measles, infectious mononucleosis, or upper respiratory tract viral infections. Isolated viral infections of the labyrinth are also believed to cause the sudden onset of hearing loss and/or vertigo in both children and adults. Otitic herpes zoster is

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**Table 7.2. **PERIPHERAL CAUSES OF VERTIGO*

| 1. | Peripheral vestibulopathy (includes labyrinthitis, vestibular neuronitis, acute and recurrent peripheral vestibulopathy) |
| 2. | Benign positional vertigo (includes benign positional nystagmus, benign paroxysmal vertigo) |
| 3. | Post-traumatic vertigo |
| 4. | Vestibulotoxic drug-induced vertigo |
| 5. | Meniere’s syndrome |
| 6. | Other focal peripheral diseases (includes local bacterial infection, degeneration of hair cells, genetic anomalies of labyrinth, cupulolithiasis, tumor of VIII nerve, otosclerosis, fistula of labyrinth, and rarely, focal ischemia and others) |

*Hearing loss often
an infection characterized by pain in the ear, followed in 1 to 110 days by a vesicular eruption in the external ear. When the seventh and eighth nerves are affected, there is a combination of facial weakness, hearing loss, and vertigo known as the Ramsay Hunt syndrome. Whenever vertigo is associated with severe ear pain or facial pain, one must consider this possibility. A dysesthetic area of skin may precede by many days the appearance of the skin eruption.

**BENIGN POSITIONAL VERTIGO**

Benign positional vertigo refers to a symptom complex classically described as indicating benign peripheral (end-organ) disease. These symptoms, differentiated from central neurologic symptoms, are outlined in Table 7.3. The signs and symptoms of benign positional vertigo are transient and rarely last longer than 40 seconds. They frequently occur when a certain position is assumed, such as lying down or turning in bed. Other causes of vertigo are also intensified by position change. Depending on whether the symptom (vertigo) or sign (nystagmus) is being emphasized, this condition is termed paroxysmal positional vertigo (BPPV) or benign paroxysmal positional nystagmus (BPPN). In a major review of 240 cases, Baloh and associates described the clinical and eye movement-recording features in patients with BPPV. In each case, after a rapid position change from sitting to head-hanging position, a stereotyped torsional paroxysmal nystagmus was observed. The time to onset of the nystagmus, the latency, was from 0 to 40 seconds, with an average of approximately 8 seconds. The initial phase of the nystagmus was rotary and upward, with the upper pole of the eye beating toward the ground on visual inspection. From the examiner’s observation, the nystagmus should appear counterclockwise, with the left ear down. The average age of onset was 54 years, and the most common identifiable causes were head trauma (17%) and viral neurolabyrinthitis (15%).

<table>
<thead>
<tr>
<th>Symptom or Sign</th>
<th>Peripheral</th>
<th>Central</th>
</tr>
</thead>
<tbody>
<tr>
<td>Latency (time to onset of vertigo or nystagmus)</td>
<td>0-40 seconds (mean 7.8)</td>
<td>No latency</td>
</tr>
<tr>
<td>Duration (signs and symptoms of single episode)</td>
<td>less than 1 minute</td>
<td>Begins immediately</td>
</tr>
<tr>
<td>Fatigability (Habituation)</td>
<td>Yes 87%</td>
<td>Symptoms may persist</td>
</tr>
<tr>
<td>(lessening signs and symptoms with repetition of provocative maneuver)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Nystagmus direction</td>
<td>Direction fixed, torsional, up, upper pole of eyes toward ground</td>
<td>No</td>
</tr>
<tr>
<td>Intensity of signs and symptoms</td>
<td>Severe vertigo, marked nystagmus, systemic symptoms such as nausea</td>
<td>Direction changing, variable</td>
</tr>
<tr>
<td>Reproducibility</td>
<td>Inconsistent</td>
<td>Usually mild vertigo, less intense nystagmus, rare nausea</td>
</tr>
</tbody>
</table>

Troost and Patton described historic factors that should lead to the consideration of BPPV: (a) symptoms associated with certain head positions; (b) rotational vertigo of episodic nature; (c) an antecedent episode of severe rotary
vertigo, with or without nausea and vomiting, associated with upper respiratory infection that suggests prior viral neuro-labyrinthitis; (d) a history of head trauma before attacks of vertigo; (e) most severe symptomatology early in the day, with lessening symptoms as the day progresses; and (f) a relative absence of spontaneous symptoms without head movement or positional change. Physical examination findings include (a) vertical-rotary BPPN produced by provocative maneuvers (Fig. 7.3); (b) latency to onset of symptoms once precipitating head position is achieved; (c) short-duration nystagmus (3 to 30 seconds); and (d) adaptation of nystagmus and symptoms (i.e., disappearance with repeated maneuvers). Additionally, BPPN is not a constant feature of the physical examination, being present on some examinations but absent at other times. Typical nystagmus upon assumption of certain head postures is considered the single most important physical finding in making the diagnosis of BPPV (Fig. 7.4).

Figure 7.3. Provocative maneuvers for positional vertigo and nystagmus. The patient is abruptly moved from a seated position to one with the head hanging 45° below the horizontal and rotated 45° to one side. He or she is then observed for positional nystagmus. The maneuvers are repeated with the head straight back and turned to the other side. (From Troost BT, Patton JM. Exercise therapy for positional vertigo. Neurology 1992;42:1441-1444.)
Figure 7.4. In benign paroxysmal positional nystagmus, the nystagmus fast phase is horizontal-rotary directed toward the undermost ear when gaze is directed toward the undermost ear (upper panel). The nystagmus fast phase is upward toward the forehead when gaze is directed to the uppermost ear (middle panel). With the eyes in the central orbital position, the nystagmus fast phase is vertical upward and rotary toward the down ear (bottom panel). (From Troost BT, Patton JM. Exercise therapy for positional vertigo. Neurology 1992;42:1441-1444.)
Posttraumatic vertigo immediately follows head trauma in most cases, implying end-organ damage in the absence of other CNS signs. The interval between injury and onset of symptoms can, however, be days or even weeks. The mechanism for the delay of symptoms is uncertain but includes hemorrhage into the labyrinth, with later development of serous labyrinthitis. Another mechanism for delayed posttraumatic positional vertigo is cupulolithiasis, in which the calcareous deposits (otoconia) of a damaged organ of the labyrinth are displaced to a sensitive region of the posterior canal, making it more susceptible to stimulation in certain head positions. In posttraumatic vertigo, the symptoms may be those of general peripheral vestibulopathy or benign positional vertigo. Generally, the prognosis is good, with symptoms gradually resolving within weeks to months. As pointed out by Baloh and colleagues, disabling persistent positional vertigo unresponsive to medical therapy occurs more commonly than was previously recognized. Many patients respond to exercise therapy, as described below, and rarely need selective section of the nerve to the posterior semicircular canal.

DRUG TOXICITY

Patients with dizziness produced by vestibulotoxic drugs are presumed or documented to have persistent injury to the peripheral end-organ. Among the agents causing such end-organ injury are the aminoglycosides. Streptomycin and gentamicin are most detrimental to the vestibular end-organ; kanamycin, tobramycin, and neomycin cause more damage to the auditory end-organ. Patients usually report progressive unsteadiness, particularly when visual input is diminished, as happens at night or in a darkened room. Vestibular testing documents a progressive bilateral loss of vestibular function. The aminoglycosides are concentrated in the endolymph and perilymph; thus the hair cells are exposed to high concentrations of the drugs. Extreme caution should be used in patients with renal disease, because most of these agents are primarily eliminated by the kidney. This type of end-organ toxicity should be contrasted with that produced by the large group of drugs with widespread reversible central and peripheral nervous system effects (Table 7.4). These drugs cause transient disequilibrium that subsides with cessation of the medication.

- **Table 7.4.**  SYSTEMIC CAUSES OF VERTIGO AND DIZZINESS

1. Drugs (including anticonvulsants, hypnotics, antihypertensives, alcohol, analgesics, tranquilizers)
2. Hypotension, presyncope (including primary cardiac causes and postural hypotension from a wide variety of causes)
3. Infectious diseases (including syphilis, viral and other bacterial meningitides, and systemic infection)
4. Endocrine diseases (including diabetes and hypothyroidism)
5. Vasculitis (including collagen-vascular disease, giant cell arteritis, and drug-induced vasculitis)
6. Other systemic conditions (including the hematological disorders, polycythemia, anemia and dysproteinemia, sarcoidosis, granulomatous disease, and systemic toxins)

MÉNIÈRE’S SYNDROME

Ménière’s syndrome is characterized by attacks of severe vertigo, tinnitus, fluctuating hearing loss, and ill-described aural sensations of fullness, with spontaneous recovery in hours to days. Initially, the patient develops a sensation of
fullness and pressure along with decreased hearing and tinnitus in a single ear. This is followed by severe vertigo, which reaches peak intensity within minutes and slowly subsides over hours. A sense of disequilibrium persists for days after an acute episode. Occasionally, sufferers from Ménière’s syndrome experience such severe attacks that they suddenly fall to the ground. Consciousness is not lost in such episodes, although awareness of surroundings may be altered by the intensity of the accompanying sensation and nausea, which has been called Tumarkin’s crisis. The most consistent pathologic finding in Ménière’s syndrome is an increase in the volume of the endolymphatic fluid and distention of the canals, hence the term endolymphatic hydrops. Although some specific causes such as bacterial, viral, and syphilitic infections may lead to the same pathologic changes and symptoms, most cases are idiopathic. Ménière’s disease usually develops between the ages of 30 and 50 and is slightly more common in women than in men. The prognosis is for progressive reduction in hearing along with increasing frequency of attacks. Some patients stabilize with no subsequent attacks of severe vertigo, but they are left with residual hearing loss. Fifty percent of Ménière’s patients become bilateral. The hearing loss often progresses to a moderate degree of deficit and then stabilizes.

OTHER PERIPHERAL VESTIBULAR CONDITIONS

Many other disorders affect the peripheral labyrinth, including acute otitis media, chronic ear infection, hereditary degenerative disorders of the end-organ, and local tumors. Conditions such as a vertebrobasilar transient ischemic attack (TIA) or focal ischemic stroke of the end-organ, particularly in an elderly patient, are often cited as a cause of vertigo. Such isolated involvement is difficult to document, and vertebrobasilar insufficiency should not be diagnosed without associated brainstem symptoms and signs.

Central Causes of Vertigo

Central pathologic causes of vertigo result from dysfunction of the vestibular portion of the eighth cranial nerve, the vestibular nuclei within the brainstem, and their central connections (Table 7.5). Neural connections with the central vestibular nuclei include interaction with the vestibular portions of the cerebellum (primarily the cerebellar flocculus), the visual sensory system, and afferent connections from muscle, joint, and tactile receptors. Normal persons will experience physiologic vertiginous sensations when visual and vestibular inputs are in conflict or when they are initially exposed to heights. Central pathologic causes of vertigo are less common than either peripheral or systemic causes, the vertiginous symptoms are usually less prominent, and additional neurologic signs are usually present on examination.
BRAIN-STEM ISCHEMIA AND INFARCTION

The posterior circulation supplies blood to the brainstem, cerebellum, and peripheral vestibular apparatus, in addition to other structures. It is not surprising that vertebrobasilar insufficiency may be accompanied by vertigo. In general, brainstem TIAs should be accompanied by neurologic symptoms or signs in addition to vertigo or dizziness before a clear-cut diagnosis is entertained. However, it is clear that isolated episodes of vertigo lasting many minutes may be due to posterior circulation dysfunction. Symptoms include transient clumsiness, weakness, loss of vision, diplopia, perioral numbness, ataxia, drop attack, and dysarthria. Common signs of vertebrobasilar ischemia include disorders of motor function such as weakness, clumsiness, or paralysis. A crossed defect (a motor or sensory deficit on one side of the face and the opposite side of the body) is good evidence of brainstem dysfunction. If the occipital lobes are the site of ischemia, transient visual loss in the form of complete or partial homonymous hemianopia will occur. Ataxia, imbalance, unsteadiness, or disequilibrium not necessarily associated with spinning vertigo may occur because of labyrinthine or cerebellar ischemia.

However, it is incorrect to believe that dizziness must be present before a TIA of the posterior circulation can be diagnosed. Isolated symptoms like those described may occur without dizziness. On the other hand, it has been overemphasized that such symptoms must always accompany dizziness when the vertiginous symptoms are due to brainstem TIA. In elderly patients with no laboratory evidence of peripheral vestibulopathy or systemic disease, episodic disequilibrium or dizziness may be due to vertebrobasilar disease.

Sudden hearing loss with moderate dizziness may be due to infarction in the distribution of the internal auditory artery. In isolation, this symptom complex is uncommon in elderly patients with atherosclerotic verte-basilar disease and is more suggestive of diseases affecting small and intermediate-diameter arteries, such as syphilis, systemic lupus erythematosus, or periarteritis nodosa. In the atherosclerotic patient, such symptoms are usually accompanied by other signs of brainstem or cerebellar dysfunction, which allow a more certain diagnosis. If actual brainstem infarction occurs, neurologic signs are often present on examination. Such signs may not be obvious and should be carefully sought. They include nystagmus of the central type, hyperreflexia, internuclear ophthalmoplegia,
homonymous visual field defects, dysarthria, vertebral bruits, and ataxia. Symptoms of dizziness are also quite common in proximal extracranial occlusion of the vertebral arteries and in the subclavian steal syndrome.

Up to this point, the emphasis has been on the accompanying signs and symptoms that almost always occur with vertebrobasilar disease. However, acute severe vertigo, mimicking labyrinthine disease, is an early symptom of acute cerebellar infarction in the distal territory of the posterior inferior cerebellar artery. To differentiate this condition from labyrinthine disease, particular attention is directed to the type of nystagmus that is present. Acute peripheral vestibulopathy usually causes unidirectional nystagmus, with the fast phase in the opposite direction. This is similar to the situation described by the mnemonic COWS (Cold, Opposite, Warm, Same) for remembering the direction of the nystagmus fast phase during thermal irrigation of the ear. The fast phase is away from the side of the cold water irrigation. Cold water mimics a peripheral destructive lesion of the labyrinth, and almost all lesions are destructive. Therefore, with a peripheral labyrinthine disturbance, the nystagmus fast phase is in the opposite direction, or away from the involved ear. The nystagmus increases during gaze in the direction of the fast phase, or contralateral to the peripheral vestibulopathy. Swaying or falling occurs toward the side of the lesion (opposite the nystagmus fast phase). The nystagmus direction is said to be fixed in that it tends to be unidirectional, away from the side of the peripheral vestibulopathy, and tends to remain horizontal on upward gaze.

In certain syndromes of the posterior circulation, the initial presentation with acute vertigo suggests peripheral vestibulopathy. With incipient cerebellar infarction, the sway or fall is toward the side of the lesion. The accompanying nystagmus may be variable in direction but is most prominent during gaze toward the lesion. In other words, with central lesions, the fast phase of the nystagmus is in the direction of gaze (direction changing nystagmus) but becomes more prominent when gaze is directed ipsilateral to the lesion. Ocular motor findings are often present in brainstem disease, such as limitation of vertical gaze, upbeat or downbeat nystagmus, or disconjugate nystagmus.

Multiple sclerosis should only be diagnosed following the documentation of disseminated CNS lesions such as optic neuritis, transverse myelitis, internuclear ophthalmoplegia or other brainstem signs, and magnetic resonance imaging (MRI) changes. Occasionally, signs and symptoms suggesting multiple sclerosis, including disequilibrium and dizziness, may be mimicked by an intrinsic brainstem tumor in a young patient.

CEREBELLOPONTINE ANGLE TUMORS

Tumors of the cerebellopontine angle rarely present solely with episodic vertigo. The most common tumor in this location results from a proliferation of the Schwann cells, hence the name schwannoma. Most of these tumors arise on the vestibular portion of the eighth cranial nerve within the internal auditory canal. They progressively enlarge, deforming the internal auditory meatus and compressing adjacent neural structures such as the acoustic portion of the eighth nerve, the facial nerve, the trigeminal nerve, the brainstem, and the cerebellum. Other tumors occurring in the cerebellopontine angle include meningiomas, epidermoids, and metastases.

The most common symptoms associated with eighth nerve tumors are progressive hearing loss and tinnitus. Vertigo occurs in approximately 20%, but a symptom of imbalance or disequilibrium is more common. Rarely, a patient with a vestibular nerve tumor may present with subtle hearing loss, tinnitus, and episodic vertigo. All those with progressive unilateral hearing loss, and particularly those with any vestibular symptoms, should be carefully examined for additional neurologic signs such as a depressed corneal reflex.

CRANIAL NEUROPATHY
Multiple or isolated cranial neuropathies occur in focal or systemic disease, including vasculitis, granulomatous disease, and meningeal carcinomatosis. The cause is often elusive. Evidence of systemic involvement is elicited by history, physical examination, and laboratory evaluation. Cogan's syndrome may be considered with cranial neuropathies. The condition is characterized by nonsyphilitic keratitis associated with vertigo, tinnitus, ataxia, nystagmus, rapidly progressive deafness, and systemic involvement.

POSTERIOR FOSSA LESIONS

Posterior fossa lesions in a variety of locations are unusual causes of isolated vertigo. The symptoms are usually positional vertigo of the central type (Table 7.3). MRI with coronal and sagittal reconstructions permits identification of small tumors close to the tissue-bone artifact in computed tomography (CT) scans.

Acquired disease of the brainstem and cerebellum produces a variety of types of nystagmus, which sometimes present as a complaint of oscillopsia, an illusion of environmental movement characterized by bouncing or jiggling of objects. Although oscillopsia is a common complaint with bilaterally reduced vestibular function, as from ototoxicity, the present of vertical oscillopsia should alert the physician to look for primary-position upbeat or downbeat nystagmus. These nystagmus types are reliable indicators of CNS abnormality due to structural intrinsic midline cerebellar disease or drugs.

SEIZURE DISORDERS

Seizure disorders, especially complex partial epilepsy, are rare causes of dizziness or vertigo. The history almost always reveals additional symptoms such as loss of awareness, automatic behavior, or generalized seizure activity following an aura of vertigo. However, rare seizure patients, documented by additional history and EEG, have isolated auras of the symptoms listed in Table 7.1.

Systemic or Medical Causes of Vertigo

Systemic causes have been given a separate category to include more widespread conditions that secondarily affect peripheral and/or central vestibular structures to produce vertigo or dizziness (Table 7.6).
**Table 7.6. SYSTEMIC CAUSES OF VERTIGO AND DIZZINESS**

| 1. Drugs (including anticonvulsants, hypnotics, antihypertensives, alcohol, analgesics, tranquilizers) |
| 2. Hypotension, presyncope (including primary cardiac causes and postural hypotension from a wide variety of causes) |
| 3. Infectious diseases (including syphilis, viral and other bacterial meningitides, and systemic infection) |
| 4. Endocrine diseases (including diabetes and hypothyroidism) |
| 5. Vasculitis (including collagen-vascular disease, giant cell arteritis, and drug-induced vasculitis) |
| 6. Other systemic conditions (including the hematological disorders, polycythemia, anemia and dysproteinemia, sarcoidosis, granulomatous disease, and systemic toxins) |

**DRUGS**

Side effects of drug ingestion frequently cause dizziness in the broadcast definition of the term. Vestibulotoxic drugs, as previously described, can produce true vertigo. The dizziness produced by other drugs is more a sense of weakness, disequilibrium, or "fuzzy headedness." The agents listed in Table 7.6 are among the most common offenders. Every attempt should be made to determine the type and quantity of medication being taken by the dizzy patient. Frequently, the elimination or reduction of medication such as a mild tranquilizer will produce improvement. The dizzy patient may have been treated with a variety of medications that themselves can add to disequilibrium or dizziness.

**HYPOTENSION**

The multiple causes of presyncope or postural hypotension are often responsible for complaints of vertigo or dizziness. Again, careful historic review and documentation of physical findings such as postural hypotension or cardiac arrhythmia direct further investigation and therapy. Presyncope is described as lightheadedness, among other phrases, and is actually a common mechanism for dizziness or even vertiginous sensations. Postural hypotension is a common side effect of antihypertensive agents, diuretics, and dopaminergic agents. When the symptom is intermittent, a history of light-headedness following change from recumbent or sitting posture to an erect position, but not the reverse, is more helpful than blood pressure measurements. In adolescents, a hyposensitive carotid sinus reflex during the growth spurt is not rare, and transient symptoms of postural dizziness might be explained by this mechanism.

**ENDOCRINE DISORDERS**

Among the endocrinopathies that cause disorders of equilibration are diabetes and hypothyroidism. The mechanism in diabetes is probably the autonomic neuropathy and orthostatic hypotension that may accompany the disease. Though much less common as a specific cause, hypothyroidism should be considered when the symptoms of vertigo remain undiagnosed. Indeed, dizziness is not an infrequent presenting complaint in patients with thyroid deficiency.
The remaining systemic conditions rarely present with isolated vertigo but are included as additional primary or secondary causes.

MULTIPLE AFFERENT SENSORY LOSS

The vestibular system functions to provide (a) spatial orientation at rest or during acceleration, (b) visual fixation during head and/or body movement (the vestibulo-ocular reflex), and (c) feedback control of muscle tone to maintain posture. These functions and their control mechanisms are interconnected in a complex fashion. Thus, the symptoms of episodic vertigo reflect disturbances in more than one system. The combination of multiple sensory deficits can produce disorientation or disequilibrium that is interpreted as dizziness or vertigo. This often occurs in the elderly, in whom vision (cataracts), hearing (presbycusis), and proprioception (peripheral neuropathy) may all be impaired. There is an entity known as presbylibrium, or imbalance resulting from aging, which may be due to a selective progressive deterioration of the peripheral vestibular apparatus or a combination of sensory deficits.

Even an intact person is easily confused by afferent sensory information, as exemplified by the sensation of spinning or true vertigo experienced during full-field optokinetic stimulation. Almost every individual while quietly seated will experience a compelling illusion of rotation while viewing a moving environment of optokinetic stripes (the circular-vection illusion). Thus, it is not surprising that patients with subtle abnormalities of peripheral or central vestibular mechanisms experience definite momentary periods of disorientation while viewing a moving patterned environment. Some experience episodic vertigo during vehicular travel.

The age-related degeneration of vestibular receptors, presbylibrium, contributes to vertigo. Although most younger patients readily compensate for unilateral peripheral vestibular damage, older patients frequently cannot or have very gradual improvement, indicating either bilateral peripheral vestibular dysfunction or a separate central abnormality that decreases their ability to compensate.

DIZZINESS IN CHILDHOOD

The most common causes of vertigo and dizziness in childhood and infancy are similar to those in the adult: acute peripheral vestibulopathy, trauma, and infection. Vertigo following air travel is more common in children than in adults because of the frequency of accompanying middle ear infection and effusion. Migraine is a significant cause of episodic dizziness or vertigo in childhood and should be considered even when the symptoms of headache are minimal.

Benign paroxysmal vertigo in childhood is a variety of vestibular neuronitis. Although unaccompanied by loss of consciousness, children may fall during the course of an attack. The episodes may last minutes to hours or recur for many weeks or even months, gradually decreasing in severity. The preservation of consciousness during an attack distinguishes the condition from temporal lobe seizures with a vestibular component and from vestibulogenic epilepsy in which an attack is triggered by labyrinthine stimulation. Congenital anomalies of the inner ear and brainstem are rare causes as is vascular disease or tumor in childhood. Rarely, typical signs and symptoms of Ménière's disease occur in childhood, the youngest reported patient being age 3.

Laboratory Evaluation of Dizziness

The primary techniques for evaluating vestibular function are electronystagmography (including caloric, specific ocular motor, and rotational testing) and posturography. Various screening tests are required with undiagnosed vertigo, and neuroradiologic imaging is indicated when a central cause is suspected.
ELECTRONYSTAGMOGRAPHY (ENG)

ENG is the most readily available test for assessing the vestibular system. Eye movements are recorded by means of the corneoretinal potential by surface electrodes, with the results printed on strip-chart recording paper or analyzed by a computer. A primary function of the ENG is to determine whether there is unilateral weakness or decreased caloric responses bilaterally. Each ear is irrigated separately with warm and cool stimulation, produced by either water or by air. The resulting nystagmus is analyzed manually or by computer to determine the slow phase velocity (SPV) of the induced nystagmus. Peak SPV resulting from the warm and cool stimulation of one ear is compared with that from the other ear. The most important finding during ENG is a significant reduction in the response on one side compared with the other. A difference of more than 20 to 25% in one ear, compared with the other, is a clear indication of hypofunction in one peripheral vestibular apparatus. The ear with the weaker response is said to have a reduced vestibular response or unilateral weakness. A bilateral weakness is defined as an SPV below 8% to 10% for both warm and cool stimulation. Typical ENG recordings are shown in Figure 7.5.

![ENG recordings](image)
Electronystagmogram (ENG). Typical bitemporal electrode recording using AC coupling. A. Calibration. Upward sweep of trace indicates eye movement to right; decay in position of trace is due to AC dal. A DC-coupled recording, standard in some laboratories, would show maintenance of this position before the eye returns to midline. B. Smooth-pursuit tracking eye movement trace shows sinusoidal side-to-side movement interspersed with minor saccadic interruption. C. Right-ear cold caloric test demonstrating left-beating nystagmus. D. Left-ear cold caloric testing demonstrating right-beating nystagmus. E. Right-ear warm caloric testing demonstrating right-beating nystagmus. F. Left-ear warm caloric testing demonstrating left-beating nystagmus. G. Optokinetic testing with tape moving to right demonstrating rightward-beating nystagmus. The electronystagmogram would be interpreted as showing minor reduction in right ear responses because of slightly reduced responses in right-ear warm caloric testing. The asymmetry would be less than 30% and therefore not of clinical significance.

Equally important information gained from the use of the ENG includes (a) documentation of spontaneous and induced nystagmus, (b) quantitation of fast eye movements, (c) smooth pursuit tracking, (d) optokinetic responses, and (e) gaze testing. These are briefly discussed below.

a. Positional nystagmus induced by certain head movements may be documented by the ENG, including the latency to onset. There is usually a delay in onset of the nystagmus or latency with peripheral types of positional nystagmus. The ENG may document a primary position horizontal or vertical nystagmus. Vertical primary position nystagmus suggests central nervous system disease. One type of induced nystagmus is positional nystagmus provoked by certain head movements.

b. The average speed of the fast eye movement is recorded. Slow saccades indicate CNS disease, such as degeneration in the brainstem.

c. When smooth pursuit tracking eye movements are interrupted by a series of small saccades (a nonspecific abnormality known as cogwheel, or saccadic, pursuit), it may be caused by drowsiness, drugs, or CNS disease.

d. A major asymmetry in the optokinetic response indicates unilateral parieto-occipital CNS dysfunction.

e. Nystagmus produced during ocular excursions, in any direction, is known as gaze-evoked nystagmus. Gaze-evoked nystagmus can result from drugs such as sedatives or anticonvulsants or from cerebellar and brainstem abnormality.

ROTATIONAL TESTING
The patient is rotated in a chair controlled by a computer and eye movements are measured. Patients are rotated in the dark with eyes open while performing mental tasks assigned to distract them from mental imagery that might suppress eye movement. During a chair rotation to the right, the eyes move to the left and then recenter with a fast phase. Thus, the slow component (phase) is in the direction opposite the spin, and the fast component of the resultant nystagmus is in the direction of the rotation. The fast components are eliminated by computer, and a slow phase is reconstructed and compared with the speed of the chair rotation. In this way, a gain (slow eye movement speed ÷ chair rotation speed) at different frequencies is obtained. Measurement is made of symmetry, which compares the response to rotation in one direction with that in the opposite direction. Also measured during rotational testing is the time relationship between the slow eye movement and the slow movement of the chair. This difference is called the phase lag. Various phase lags are also plotted against the frequency of rotation of the chair. Thus, both gain and phase plots are produced during rotational testing. Rotational testing provides little information about the site of the lesion, as opposed to caloric testing in the ENG. However, it is quite beneficial in quantitating bilaterally reduced vestibular function such as occurs with ototoxicity. Rotational testing, therefore, is helpful in determining response patterns in patients with bilateral vestibular loss. A symmetric response of a person with a previous unilateral peripheral vestibular abnormality indicates vestibular compensation, and abnormal phase-lag is a nonspecific marker indicating some degree of prior peripheral vestibular abnormality.

POSTUROGRAPHY

Posturography is a means of quantifying the Romberg test. Changes in body sway during Romberg testing with feet directly together, both with eyes open and eyes closed, are measured by means of a computer. Most recently, a dynamic posture platform has been introduced. The patient is surrounded by a movable visual field and stands on a posture platform that is mobile. By manipulating the visual field, visual cues that help maintain posture may be eliminated. Similarly, by moving the posture platform in response to movement of the feet, attempts are made to remove proprioceptive cues. The test results in all conditions are reported, and an interpretation is made on the basis of which systems are defective. Posturography, a promising technology currently in use and under evaluation for assessment of balance disorders, may be useful in rehabilitation.

ADDITIONAL DIAGNOSTIC TESTS

Patients with undiagnosed vertigo should have metabolic screening tests including a blood count, electrolytes and glucose determinations, and thyroid function testing. Many physicians involved in the evaluation of dizzy patients also perform lipid screens for hypercholesterolemia or increased triglycerides. The laboratory investigation, like the physical examination, is directed particularly by the patient’s history. If there is a history of presyncope or syncope, the patient must have a cardiac evaluation, including at least an electrocardiogram and rhythm strip. A more suggestive history would lead to a Holter 24-hour monitor or an event monitor, during which the patient wears a battery-powered apparatus that can be activated at times of symptoms. This device then records the cardiac rhythm. The presence of auditory symptoms requires a complete audiologic evaluation as described below. Multiple or recurrent cranial neuropathy would lead to a variety of screening tests for collagen vascular disease or skull-based pathology or meningitic processes.

Neuroradiologic Investigation

In the past, the primary neuroradiologic techniques for determining CNS abnormality and, in particular, cerebellopontine angle tumors included tomography of the temporal bone, CT scanning, and posterior fossa myelography with air or other contrast material. Currently, the high resolution obtainable on CT scanning has largely eliminated the need for tomography of the temporal bone. MRI has largely supplanted CT scanning for
cerebellopontine angle tumors. For general neurologic screening, a CT scan, with and without contrast, is appropriate in patients suspected of having a CNS disorder on the basis of history or physical examination. The workup must include an MRI (Figs. 7.6, 7.7, 7.8) when there are persistent symptoms suggesting a CNS disorder. The best available images of the cerebellopontine angle and brainstem are clearly afforded via MRI.

Figure 7.6. MRI scan showing marked atrophy of cerebellum in a patient who had progressive unsteadiness.
Figure 7.7. Typical MRI scan of patient with multiple sclerosis showing periventricular white matter abnormality (arrow). White matter lesions extending in a perpendicular fashion from the ventricle are virtually pathognomonic for multiple sclerosis.
Figure 7.8. High-resolution MRI scan of the posterior fossa demonstrating cranial nerves VII and VIII to the viewer's right and a large cerebellar pontine angle vestibular schwannoma on the viewer's left (arrows).

Therapy for Peripheral Vestibular Disorders

The emphasis in this section is on medical and, to a lesser extent, surgical treatment of peripheral vestibular dysfunction and vertigo.

MEDICAL TREATMENT

Therapy is outlined in Table 7.7 for symptomatic treatment of dizziness presumed to be of peripheral origin. When a definitive diagnosis such as vestibular schwannoma, autoimmune disorder, perilymph fistula, or systemic vasculitis has been made, the therapy must be directed to the underlying disorder.
Although most of the drugs used for dizziness are loosely referred to as vestibular suppressants, their mechanism of action may not be defined, and it is often unclear which agents will be effective in a given patient. The primary vestibular afferent system could be suppressed directly or indirectly through the inhibitory portion of the vestibular efferent system. An important effect of some agents may be to act on other sensory systems such as proprioceptive or visual inputs to the vestibular nuclei of the brainstem.

Few controlled studies have investigated the response of patients with presumed peripheral vestibular dysfunction. Most of the drugs used are empirical, based on studies of the prevention of motion sickness in normal subjects or on the various regimens used by otologists for Ménière's syndrome. Each drug class is discussed separately below.
Antihistamines are among the most commonly employed agents in the treatment of dizziness. The initial drug usually employed is meclizine hydrochloride in doses up to 50 mg three times per day. Since the main side effect of antihistamines is drowsiness, the smallest dose should be used initially, even as low as 12.5 mg two to three times per day.

For dizziness, antihistamines in the H₁ antagonist group are used. Possibly the blockers, effective in motion sickness, act by central antagonism of acetylcholine (ACh), as does scopolamine. An excellent drug as a second choice is Promethazine, a phenothiazine with the strongest ACh-blocking action. The usual starting dose is 25 mg three times per day, but if this amount produces drowsiness and still has a positive effect, the drug dosage may be reduced to 12.5 mg three times a day.

Anticholinergics that block the muscarinic effect of ACh have been widely used and studied for the prevention of motion sickness. Atropine acts centrally to stimulate the medulla and cerebrum, but the closely related alkaloid scopolamine is more widely used. Transdermal delivery of scopolamine may prevent or mitigate the nausea and vomiting associated with motion sickness, but not the dizziness. In general, transdermal scopolamine is not useful in patients with acquired vestibulopathy. Frequent side effects are blurred vision and dry mouth, in addition to occasional confusion. Some patients have significant difficulty when they try to discontinue scopolamine patches. A side effect of low-dose scopolamine or atropine is the transient bradycardia (4 to 8 beats fewer per minute) associated with the peak action of oral scopolamine at 90 minutes and diminishing thereafter.

Sympathomimetics have been used in the treatment of motion sickness, particularly in combination with anticholinergics. The sole agent in this class that may have an application in combination with other drugs is ephedrine. Tolerance may develop after a few weeks of treatment.

Antiemetics may be used when prominent nausea is an accompanying feature of the patient's complaint. Many of the antihistaminic and anticholinergic drugs listed here are also used for their antiemetic actions. Prochlorperazine (Compazine) should be used with caution, particularly by the intramuscular route, because of the high incidence of dystonic reactions. Because promethazine (Phenergan) has a significant antiemetic effect, this drug is particularly useful when there is prominent nausea.

Tranquilizers is the general name for drugs from different classes with central and probably peripheral effects. They include benzodiazepines, butyrophenones, and phenothiazines. Diazepam is one of the most widely prescribed drugs for the treatment of dizziness. Many believe it should not be the first choice, primarily because of the significant potential for habituation and depression and because it can be the actual cause of dizziness. Nonetheless, it does remain the first choice of many otoneurologists or otologists. Other longer-acting benzodiazepines may be helpful in certain patients, but no study has substantiated their effectiveness. Haloperidol in small oral doses (0.5 mg three times a day) is effective in many patients with peripheral vestibular dysfunction who are not helped by other antidizziness medications.

Combination preparations and other agents include those listed in Table 7.6 and are frequently useful, particularly the combination of ephedrine and promethazine. Some other agents and regimens used primarily in the medical management of Meniere's disease are listed. Low-sodium diets and diuretics have been helpful with some patients. In the belief that in some cases an effect on blood supply to the peripheral end-organ might be a factor, agents such as cyclandelate have been used. The general approach to the patient with an acute or chronic vestibulopathy is to first use an antihistamine such as meclizine hydrochloride. If this is not helpful, the next step is to use promethazine (Phenergan), and if this is ineffective, low doses of haloperidol or low-dose Valium, always keeping in mind the potential for habituation with benzodiazepines.
EXERCISE THERAPY

It is important to recognize that BPPV is responsible for at least 50% of all causes of vertigo, and exercise therapy may be curative in up to 90% of patients. The primary therapeutic option is one form or other of exercise therapy. The severity of the individual attacks and accompanying nausea may be lessened by medical therapy; however, this does not prevent future attacks.

Exercise therapy is indicated for all patients with BPPV. There are two general approaches to therapy: (a) a single treatment session in an outpatient office setting and (b) a series of exercises performed by the patient at home. Each is briefly described below.

OFFICE SINGLE-TREATMENT APPROACH. Among the single-treatment approaches are the canal repositioning maneuver (CRP) and its modifications (Fig. 7.9). One standard protocol is described below. This technique works best for patients in whom a specific head position produces attacks of vertigo, such as with the left ear down. Often, the examiner notices a characteristic rotary vertical nystagmus accompanying the vertigo when the head is placed in the offending position (Figs. 7.3 and 7.4).

Figure 7.9. Positioning sequence for left PSC shows orientation of left labyrinth and gravitating canaliths. S, Start, patient is seated with operator behind patient. Oscillation is started. 1, Head is placed over end of table, 45° to left, with head extended. (Canaliths gravitate to center of posterior semicircular canal.) 2, Head is rotated 45° to right; head is kept well extended while coming from...
position 1. (Canaliths reach common crus.) 3. Head (and body) are rotated until facing downward 135° from supine. (Canaliths traverse common crus.) 4. Patient is brought to sitting position; head is kept turned to right while coming from position 3. (Canaliths enter utricle.) 5. Head is turned forward with chin down about 20°. (From Epley JM. Fine points of the canalith repositioning procedure for treatment of BPPV. Insights Otolaryngol 1994;2(9):7.)

Treatment Protocol for the Left Ear

1. The patient is moved quickly from a seated position back over the end of the examination table, with the head extended and turned approximately 45° with the left ear down. In each position, there may be nystagmus induced as a result of change from the prior head position. The patient is kept in the position until the nystagmus or symptoms subside, typically 10 to 15 seconds.
2. The head is slowly rotated so that the right car is now turned 45° down, keeping the head extended.
3. The head and body are rotated to the right until the patient is facing downward.
   This position is maintained for approximately 15 seconds.
4. The patient is then brought gradually up to a seated position with the head turned to the right.
5. The head is turned forward with the chin slightly depressed.

Over the next 24 to 48 hours, some recommend that the patient remain upright as much as possible. Another variation is to apply a hand-held mechanical oscillator to the head in each position. The overall success of this single treatment is reported to be 50 to 75%.

HOME EXERCISE THERAPY. The patient is first instructed carefully about the type of exercise to be performed (Fig. 7.10).

Figure 7.10. Exercise therapy. The patient begins in the seated position and then leans rapidly to the side, placing the head on the bed or table. The patient remains there until the vertigo subsides and then returns to the seated upright position, remaining there until all symptoms subside. The maneuver is repeated toward the opposite side, completing one full repetition. Ten to 20-repetitions
Treatment Protocol for Either Ear

1. In a seated position, on the edge of a couch or bed, the patient is asked to quickly lie on one side, placing the worst ear (if one can be discovered) down first (Fig. 7.9). The patient then moves rapidly from the sitting position and rests the head on a pillow or other support, without moving forcefully enough to produce a neck injury.

2. The patient then returns rapidly to an upright seated position and remains there for 30 seconds or until symptoms subside.

3. The patient rapidly lies down on the other side and remains there for approximately 30 seconds or until the symptoms subside.

4. The patient then returns to the upright seated position. This constitutes a single repetition.

Twenty repetitions should be performed two times per day. Each session lasts approximately 30 minutes. Some patients have intense symptoms at the onset of the BPPV, including vomiting. It is clear that patients who experience extreme discomfort during the maneuvers are not likely to pursue them on their own outside of an office or hospital setting. These patients may need hospital admission or hydration in an outpatient setting, with the concurrent administration of vestibular suppressant medications. Most patients are willing to perform exercises at home. This protocol is particularly useful for BPPV patients who have the following:

1. Bilateral BPPV
2. Uncertainty as to which ear is involved
3. Failure of single office treatment protocols

Recovery can be quite rapid occurring during the first few days of exercise therapy. Others progressively improve over weeks and months, suggesting that the vestibular system may adapt to whatever abnormal perturbation is causing the symptoms.

Approximately 50% of patients who have well-defined vertigo and nystagmus in certain head positions will have improvement following the single-treatment maneuver. Variations include the use of a hand-held oscillator or longer durations in each single position. The home set of maneuvers, known as the Brandt-Daroff maneuvers, may take days, weeks, or even months to produce a cure, but progressive improvement of symptoms should be noticed by the patient within the first few weeks. It is estimated that approximately 20% of patients have recurrences within the first year, and either of the maneuvers described above may be repeated with high expectation of further improvement. The overall success rate of exercise therapy approaches 90%, even with patients who have been symptomatic for years.

SURGICAL THERAPY

Surgical therapy of chronic peripheral vestibular dysfunction includes exploration for fistulas, endolymphatic shunts, and destructive end-organ surgery. The details of these procedures may be found in standard otology texts. In patients with severe Ménière's disease for whom no medical therapy such as that described earlier has been effective and who have severe recurrent disabling attacks, a labyrinthectomy may be performed. Unfortunately, Ménière's disease may become bilateral, eventually resulting in the need for labyrinthectomy or vestibular nerve section on the contralateral side. A medical labyrinthectomy may be performed by the use of aminoglycoside drugs, those
particularly destructive to the peripheral vestibular hair cells. Surgical or medical labyrinthectomy is usually a last resort in patients who have clearly defined severe attacks of peripheral vestibulopathy, presumably from Ménière’s disease.

Various shunting procedures have been used in the treatment of Ménière’s disease or endolymphatic hydrops. Although some patients can benefit, the long-term success with such shunting procedures, which include shunts to the mastoid region and to the subarachnoid space, has been only modest.

Some patients with benign paroxysmal positional vertigo do not have a benign course. Patients who experience classic but disabling symptoms persisting over months are candidates for exercise therapy as described earlier. On rare occasions, the exercise therapy is unsuccessful; such patients are candidates for section of the nerve from the posterior semicircular canal.

Management of Central and Systemic Vestibular Disorders

MEDICAL TREATMENT

Clearly the management of central vestibular disorders depends on the diagnosis. A simple separation into peripheral and central vestibular dysfunction is not always possible, as alluded to above. Some patients have inadequate central compensation for a peripheral vestibular abnormality and thus remain symptomatic. In such patients, medical therapy for peripheral vestibular dysfunction, as described above, may prove quite effective. When a specific diagnosis (e.g., postural hypotension secondary to diabetic peripheral neuropathy) is made, attention should be directed to treatment of the primary condition. Severe postural hypotension is notoriously difficult to manage. In general, the approach is to use agents that increase vasoconstriction, others that prevent vasodilation, or drugs that might increase cardiac output. Plasma volume may be increased by the use of mineralocorticoids such as fludrocortisone acetate, but they should be prescribed cautiously.

The patient who is diagnosed as having primary CNS disease, whether it be brainstem infarction or spinocerebellar degeneration, must be managed as a patient without the accompanying symptoms of disequilibration would be. Medical therapy of vertebrobasilar ischemia is directed at preventing new infarctions, primarily with antiplatelet agents and, on rare occasions, anticoagulation. Cerebellar dysfunction not caused by tumor may be treated symptomatically. Vestibular Nupressant medication may add a modicum of improvement, and agents helpful in the therapy of essential tremor, such as beta-blocking drugs or primidone, may result in modest symptomatic improvement.

Therapy for systemic conditions producing vertigo also depends on the diagnosis. If systemic drug therapy, as with benzodiazepines, is actually the cause of disequilibration, then of course alteration in the medical regimen may prove efficacious. Withdrawal of all drugs, be they anticonvulsants or benzodiazepines, must proceed with caution to avoid precipitating the effects of withdrawal.

Cervicogenic Dizziness

DIZZINESS MAY RESULT FROM NECK PAIN
Neck pain often accompanies dizziness, but it may be difficult to tell whether the dizziness and the neck pain are related or just coincidental. The influence of head position on equilibrium has been known since the mid-1800s. However, a clinical syndrome relating neck pain and/or injury to dizziness and disequilibrium was not discussed until the 1950s.

Ryan and Cope described a syndrome of disequilibrium and disorientation in patients with many different diagnoses of neck pathology including cervical spondylosis, cervical trauma, and cervical arthritis. They introduced the syndrome as cervical vertigo. As true spinning vertigo is rarely associated with this syndrome, cervicogenic dizziness is a more correct name for this syndrome and will be used here.

**CERVICOCENIC DIZZINESS**

Cervicogenic dizziness tends to be a controversial diagnosis because there are no diagnostic tests to confirm that it is the cause of the dizziness. Cervicogenic dizziness is a diagnosis that is provided to people who have neck injury or pain as well as dizziness and in whom other causes of dizziness have been ruled out.

People with cervicogenic dizziness tend to complain of dizziness (a sensation of movement of the self or the environment) that is worse during head movements or after maintaining one head position for a long time. The dizziness usually occurs after the neck pain and may be accompanied by a headache. Often the dizziness will decrease if the neck pain decreases. The symptoms of dizziness usually last minutes to hours.

People with cervicogenic dizziness may also complain of general imbalance that may increase with head movements and with movement in the environment. Although no formal studies have been completed, true cervicogenic dizziness is thought to be rare.

An evaluation for cervicogenic dizziness involves a thorough medical evaluation because the symptoms are similar to other causes of dizziness. Testing of inner ear function is usually requested to ensure that the peripheral or central vestibular system is intact. A health care practitioner may perform a maneuver in which the body is turned while the head is held fixed to see if it causes nystagmus (eye movements) or dizziness to confirm the suspected diagnosis. The results of this test need to be correlated with subjective symptoms and the clinical findings because the test can also be positive in healthy individuals.

Cervicogenic dizziness often occurs as a result of whiplash or head injury and is often seen in conjunction with brain injury or injury to the inner ear. It is often difficult to distinguish between cervicogenic dizziness and other medical problems. Cervicogenic dizziness that occurs in conjunction with brain injury or another form of dizziness will be more difficult to diagnose and treat. It is important to be patient while health care professionals sort through the problems and treat them in the most logical order.

The majority of patients with cervicogenic dizziness improve with only treatment of the neck problem. Several studies have reported that approximately 75 percent of patients improve with conservative treatment of the neck such as medication, gentle mobilization, exercise, and instruction in proper posture and use of the neck. For other patients, improvement involves treatment of the neck problem in addition to vestibular therapy. Vestibular rehabilitation is directed at what problems are found on evaluation and may include eye exercises, balance exercises, walking, and graded exposure to environments that make you dizzy.
SUMMARY

Cervicogenic dizziness is a syndrome of neck pain accompanied by an illusory sense of motion and disequilibrium; it is a diagnosis provided to people once all other causes of dizziness are ruled out.

Cervicogenic dizziness will usually resolve with treatment of the neck problem but may require vestibular rehabilitation for complete resolution of symptoms. In general, the prognosis for patients with cervicogenic dizziness is good, with 75 percent of patients having improvement of symptoms.

References

Cholesteatoma

WHAT IS A CHOLESTEATOMA?

A cholesteatoma is a skin growth that occurs in an abnormal location, the middle ear behind the eardrum. It is usually caused by repeated infection that causes an ingrowth of the skin of the eardrum. Cholesteatomas often take the form of a cyst or pouch that sheds layers of old skin that builds up inside the ear. Over time, the cholesteatoma can increase in size and destroy the surrounding delicate bones of the middle ear. Hearing loss, dizziness, and facial muscle paralysis are rare but can result from continued cholesteatoma growth.

HOW DOES IT OCCUR?
A cholesteatoma usually occurs because of poor eustachian tube function as well as infection in the middle ear. The eustachian tube conveys air from the back of the nose in the middle ear to equalize ear pressure ("clear the ears"). When the eustachian tubes work poorly, perhaps due to allergy, a cold, or sinusitis, the air in the middle ear is absorbed by the body, resulting in a partial vacuum in the ear. The vacuum pressure sucks in a pouch or sac by stretching the eardrum, especially in areas weakened by previous infections. This sac often becomes a cholesteatoma. A rare congenital form of cholesteatoma (one present at birth) can occur in the middle ear and elsewhere, such as in the nearby skull bones. However, the type of cholesteatoma associated with ear infections is most common.

WHAT ARE THE SYMPTOMS?

Initially, the ear may drain, sometimes with a foul odor. As the cholesteatoma pouch or sac enlarges, it can cause a full feeling or pressure in the ear, along with hearing loss and tinnitus. An ache behind or in the ear, especially at night, may cause significant discomfort. Dizziness, or muscle weakness on one side of the face (the side of the infected ear) can also occur. Any, or all, of these symptoms are good reasons to seek medical evaluation.

IS IT DANGEROUS?

Ear cholesteatomas can be dangerous and should never be ignored. Bone erosion can cause the infection to spread into the surrounding areas, including the inner ear and brain. If untreated, deafness, brain abscess, meningitis, and rarely death can occur.

TREATMENT

An examination by an otolaryngologist — head and neck surgeon can confirm the presence of a cholesteatoma. Initial treatment may consist of a careful cleaning of the ear, antibiotics, and eardrops. Therapy aims to stop drainage in the ear by controlling the infection. The extent or growth characteristics of a cholesteatoma must also be evaluated.

Large or complicated cholesteatomas usually require surgical treatment to protect the patient from serious complications. Hearing and balance tests, X-rays of the mastoid (the skull bone next to the ear), and CAT scans (3-dimensional X-rays) of the mastoid may be necessary. These tests are performed to determine the hearing level remaining in the ear and the extent of destruction caused by the cholesteatoma.

Surgery is performed under general anesthesia in most cases. The primary purpose of the surgery is to remove the cholesteatoma and infection, and achieve an infection-free, dry ear. Hearing preservation or restoration is the second goal of surgery. In cases of severe ear destruction, reconstruction may not be possible. Facial nerve repair or procedures to control dizziness are rarely required. Reconstruction of the middle ear is not always possible in one operation; and therefore, a second operation may be performed six to twelve months later. The second operation will...
attempt to restore hearing and, at the same time, will allow inspection of the middle ear space and mastoid for residual cholesteatoma.

Admission to the hospital is usually done the morning of surgery, and if the surgery is performed early in the morning, discharge may be the same day. For some patients, an overnight stay is necessary. In rare cases of serious infection, prolonged hospitalization for antibiotic treatment may be necessary. Time off from work is typically one to two weeks.

Follow-up office visits after surgical treatment are necessary and important, because cholesteatoma sometimes recurs. In cases where an open mastoidectomy cavity has been created, office visits every few months are needed in order to clean out the mastoid cavity and prevent new infections. In some patients, life-long periodic ear examinations are required.

SUMMARY

Cholesteatoma is a serious but treatable ear condition that can only be diagnosed by medical examination. Persisting earache, ear drainage, ear pressure, hearing loss, dizziness, or facial muscle weakness signals the need for evaluation by an otolaryngologist.

Enlarged Vestibular Aqueduct Syndrome (EVAS)

WHAT IS AN ENLARGED VESTIBULAR AQUEDUCT?

The vestibular aqueduct is a tiny, bony canal that extends from the inner ear endolymphatic space toward the brain. It is shielded by one of the densest bones in the body, the temporal bone, which also houses the sound-sensing cochlea and motion-sensing vestibular organs vital to our ability to hear and maintain balance. Inside the vestibular aqueduct is the endolymphatic duct, a tube that connects the endolymph (a fluid) in the inner ear to the endolymphatic sac. The function of the endolymphatic duct and sac is not totally understood, but it is believed that they help maintain the volume and ionic composition of endolymph necessary for transmitting hearing and balance nerve signals to the brain.

When a vestibular aqueduct is larger than normal, it is known as a large vestibular aqueduct (LVA) or by the term used here, enlarged vestibular aqueduct (EVA). Hearing loss or balance symptoms associated with an EVA can occur when the endolymphatic duct and sac expand to fill the larger space (see Figure). When EVA is associated with such symptoms, it is referred to as EVA syndrome (EVAS).
CAUSES OF AN ENLARGED AQUEDUCT

During fetal development, the vestibular aqueduct starts out as a wide tube. By the fifth week it narrows, and by midterm it approaches adult dimension and shape; however, the vestibular aqueduct continues to grow and change until a child is 3 to 4 years old. As yet incompletely understood genetic or environmental conditions cause EVA, which is often congenital (present at birth) or occurs during early childhood.

Hearing loss associated with EVAS can be syndromic deafness, a loss of hearing accompanied by physical signs and symptoms affecting other parts of the body. More commonly, it is nonsyndromic deafness, affecting only ear function (nonsyndromic deafness accounts for approximately 70% to 80% of all genetic hearing loss¹). Just as the cause of EVAS remains unclear, much of what is known about it—who the hearing loss pattern differs among patients, how many people actually have it, how it causes symptoms, how to effectively treat it, and what the prognosis might be—comes from isolated clinical observations and small studies, not from comprehensive scientific research.

Genetic testing often but not always reveals that EVA is associated with mutation of the SLC26A4 gene (also called the PDS gene) which also causes Pendred syndrome, a condition associated with syndromic hearing loss and thyroid disease. Pendred syndrome occurs in an estimated one-third of all cases of EVA2 and is an autosomal-recessive genetic disorder, meaning each parent must be a genetic carrier.
EVA can be associated with branchio-otorenal syndrome, which affects the anatomy of the ears, kidney, and neck. Additionally, it can be associated with other anatomical defects such as a Mondini malformation, an incomplete cochlear development that is also linked to a mutation of the PDS gene.

PREVALENCE

EVAS is considered to be rare, but as with many inner ear disorders, its true prevalence is difficult to assess because it is not always recognized during a medical evaluation. Estimates fall between as high as 5% to 15% in pediatric patients. More women than men are affected; for every two males with EVAS, there are three females who have the disorder. EVAS is associated with vestibular symptoms in a small percentage of people.

Hearing symptoms

It is hearing loss that usually brings EVAS to the attention of a physician. Such loss can be sensorineural, conductive, or both. Sensorineural hearing loss (SNHL) is deafness usually related to the cochlea, but sometimes to the vestibulocochlear nerve or the brain’s central auditory system. Conductive hearing loss involves reduced movements of the middle ear bones (malleus, incus, and stapes) which conduct external sound to the inner ear.

Some people are born with the hearing loss. However, in most cases of EVAS, a child will hear normally in the first years of life and then notice hearing loss later in childhood, or less commonly in adolescence or early adulthood. Generally, this occurs after a minor or major head impact, upper respiratory infection, or air pressure trauma, such as occurs during the rapid depressurization of an airplane. Even active play, especially jumping, can jar the head enough to result in hearing loss if an EVA is present. The loss can be progressive, fluctuating, stable, or sudden, and can involve tinnitus, a ringing in the ears. However, generally the hearing loss occurs in a series of steps. With each minor event, hearing drops one or more levels, a downward progression often culminating in profound hearing loss.

VESTIBULAR SYMPTOMS

Vestibular symptoms sometimes related to EVAS include episodic spinning vertigo, mild unsteadiness, trouble watching revolving objects, a feeling of vague instability, rocking sensations, jumping vision, decreased visual acuity in the presence of loud sounds, instability when leaning forward, vomiting, nausea, and drunken gait. A young child may also grab his or her head and walk in circles. The symptoms of vestibular disorders are notoriously difficult for children and adults to describe; for children, the task is even more challenging. Unless well trained in recognizing vestibular disorders, a physician may not ask the questions necessary to discover them. However, these signs and symptoms can also be seen in other types of vestibular disorders and are not unique to the diagnosis of EVAS.

Traditionally, physicians have devoted more attention and study to the effect of EVAS on hearing, but an increasing clinical awareness of the impacts of vestibular dysfunction on childhood development is starting to change that. These impacts manifest in outward signs such as reflex delays, visual-spatial problems, motion sensitivity, abnormal movement patterns, clumsiness, difficulty moving in the dark, and lingering anxiety. Many of childhood’s common milestones—climbing stairs, riding bicycles—may be delayed, or simply too difficult for a child with EVAS to
manage without treatment. It is also important to note that these symptoms can have other causes besides inner ear disorders, which is why a thorough medical evaluation by a pediatrician, otolaryngologist/neurotologist, and neurologist is needed.

HOW DOES EVAS CAUSE SYMPTOMS?

The SNHL and balance symptoms associated with EVAS may occur because the enlarged endolymphatic duct and sac are unable to maintain their normal functions. These include maintaining the endolymph volume and ionic composition (concentrations of sodium, potassium, calcium, and chloride) necessary for transmitting hearing and balance nerve signals to the brain. This disrupts inner ear homeostasis, which is ionic equilibrium among the compartments of the inner ear that contain either endolymph or perilymph, fluids that have specific and different concentrations of ions. If related to head trauma, EVAS may cause symptoms when the sudden fluctuation in cerebrospinal fluid (CSF) pressure on impact forces highly concentrated (hyperosmolar) proteins into the cochlear duct which connects the CSF space to the endolymph space inside the cochlea. This is called hyperosmolar reflux.

Conductive hearing loss with EVAS may occur because the increased endolymphatic pressure reduces the ability of the stapes to move the oval window, which is the membrane separating the middle ear from the fluid-filled inner ear. Because of this dysfunction, sound waves conducted through the middle ear can’t be transferred to the cochlea in the inner ear.

CLINICAL EVALUATION

The variable signs of EVAS indicate that diagnosis requires special care and attention to a person’s symptoms and medical history, especially those of children. In addition to a complete medical history and physical examination, the diagnostic process for uncovering EVAS usually involves hearing and balance testing and radiologic assessment. Thyroid, renal, and cardiac function are also usually analyzed, and genetic screening is sometimes also performed.

Audiologic testing often reveals low frequency conductive hearing loss, high frequency SNHL, or both. Vestibular tests may be useful even if a person with EVAS is not experiencing active vestibular symptoms such as vertigo. When electronystagmography (ENG) is used to measure eye movements and vestibular responses to thermal (caloric) or rotational stimulation, it may reveal loss of vestibular function in one or both ears and nystagmus, the abnormal eye movements commonly associated with vestibular dysfunction.

For radiologic assessment, fast spin-echo magnetic resonance imaging (MRI) is generally considered to be the most appropriate test because it permits precise imaging and measurement of the endolymphatic duct and sac soft tissues. High resolution computerized tomography (CT) scans of the temporal bone are also often used to confirm the bony enlargement and other bony abnormalities that are often associated with EVAS. No standard radiographic criteria exist yet to define what size constitutes an enlargement. Some studies suggest that a vestibular aqueduct is enlarged if it is more than 1.5 millimeters (mm) in diameter, while others define an EVA as being more than 4.0 mm in size.

TREATMENT
Historically, medical and surgical treatments have not reversed the progression of hearing or vestibular losses from an EVA. Currently, the cornerstone of management is prevention, primarily by protecting the head from traumas that will worsen the progression of hearing loss and vestibular symptoms. People with EVAS are cautioned to avoid contact sports and must wear a helmet while bicycling and performing other activities that elevate risk of head injury. Parents are often challenged to find a middle ground between allowing their children to enjoy the typical physical activities of childhood and preventing future losses to hearing and function. Each person must negotiate his or her own decisions about risk management.

With significant hearing loss, a hearing aid may be used, but often unsuccessfully. For some people, cochlear implantation has significantly improved hearing. An audiologist can help determine whether options such as communication training in sign language or speech could be helpful. In some people with related vestibular symptoms, treatment may include vestibular rehabilitation therapy; however, as is also common for patients experiencing an active phase of Ménière’s disease, EVAS may not respond well to vestibular rehabilitation.

Predicting what will ultimately happen in any one case of EVAS is difficult because the condition follows no typical course. No relationship exists between how large the aqueduct is and the amount of hearing loss a person may sustain. Some cases progress to profound deafness, some include vestibular losses or difficulties, and other cases lead to neither. It’s important to note that for some people with EVAS, loss of hearing begins in childhood, but vestibular symptoms are delayed until adulthood. Accordingly, people with EVAS should seek medical evaluation if they develop unexplained dizziness, vertigo, or other signs of vestibular dysfunction.

Additional Resources
CT images of EVA are available at the Cleveland Hearing and Balance Center website.
VEDA also produces these helpful documents:

- Pediatric Vestibular Disorders: Recognition, Evaluation, and Treatment
- Vestibular Rehabilitation: An Effective, Evidence-Based Treatment

References
Vestibular Migraine (a.k.a. Migraine Associated Vertigo or MAV)

MIGRAINE IS ONE OF THE MOST DEBILITATING CHRONIC DISORDERS IN THE UNITED STATES.

It is almost as prevalent as hypertension (high blood pressure) and is more common than asthma and diabetes mellitus. More importantly, migraine strikes people during what are expected to be their most productive years: between ages 20 and 40 for most women, with a slightly higher age range for men.

Despite better diagnostic capabilities and efforts to improve public awareness and education, it is estimated that approximately 50% of migraineurs go undiagnosed or mismanaged to this day. Many self-treat, or are treated inappropriately for sinus or other non-migrainous types of headache. Often described as “sick headache,” migraine is typically characterized by unilateral onset of head pain, severe progressive intensity of pain, throbbing or pounding, and interference with the person’s routine activities. Accompanying symptoms of photophobia (sensitivity to light) or phonosensitivity (intolerance to noise), as well as nausea and/or vomiting, are common, and often leads to the inability to perform daily tasks.

Following is a video filmed by Dr. P. Ashley Wackym of Portland, Oregon’s Ear & Skull Base Center, showing a patient who has suffered from vestibular migraine. When Dr. Wackym first
started seeing this girl she had to be confined to a wheelchair due to her severe symptoms. View this and more videos by Dr. Wackym on his You Tube Channel.

MIGRAINE AND VESTIBULAR DYSFUNCTION

Approximately 40% of migraine patients have some accompanying vestibular syndrome involving disruption in their balance and/or dizziness at one time or another. This may be prior to, during, after, or totally independent of their migraine event. Some interesting parallels exist between migraine and non-migrainous vestibular dysfunction. Many of the food and environmental triggers for migraineurs (see box on page 2) are the same as those for patients with non-migrainous vestibular dysfunction. Hormonal fluctuations, foods, and weather changes (barometric-pressure variations) often exacerbate both conditions. Finally, diet modifications and certain medications used in migraine management may ameliorate or prevent the vestibular component of the migraine.2,3 Interestingly enough, some of the analgesic medications for the pain do not resolve the dizziness and medications for the dizziness often do not resolve the painful headache.

The clinical presentation of vestibular symptoms that often correlate with migraine3 includes—but is not limited to—dizziness; motion intolerance with respect to head, eyes, and/or body; spontaneous vertigo attacks (often accompanied by nausea and vomiting); diminished eye focus with photosensitivity; sound sensitivity and tinnitus; balance loss and ataxia; cervicalgia (neck pain) with associated muscle spasms in the upper cervical spine musculature; confusion with altered cognition; spatial disorientation; and anxiety/panic.4 While migraine is often associated with benign recurrent vertigo of adults or paroxysmal vertigo of childhood,5,6,7 some migraine patients also present with true benign paroxysmal positional vertigo (BPPV) even after the migraine headache event has ceased. This is thought to be caused by a combination of vascular events along with an alteration of neural activity associated with the migraine event.8,9 It is believed that these changes more commonly affect the utricle and/or the superior portion of the vestibular nerve and anterior vestibular artery, rather than the saccule and the inferior portion of the vestibular nerve and posterior vestibular artery.10,11 This may explain why results within the normal range are often obtained with vestibular-evoked myogenic potentials (VEMP) testing of migraine patients in the absence of true BPPV. Similarly normal findings have been reported in cases of migraine in the apparent absence of inferior vestibular neuritis, leading to the belief that if inflammation is in fact present as a result of the migraine, and is a cause for utricular BPPV, the local inflammation of the peripheral blood vessels and/or cranial nerve branches is more prevalent in those supplying the utricle rather than the saccule. However, VEMP also now can be helpful in differentiating the clinical presentation of migraine vs Meniere’s syndrome or BPPV. Usually following a migraine event, the VEMP intensity measures are commonly hyperresponsive, whereas with Meniere’s exacerbation the affected ear intensity response is hyporesponsive, and with BPPV the affected ear latency response is typically prolonged.

TRIGGERS FOR MIGRAINE

Note: Some of the triggers below may also apply to other types of vestibular dysfunction.
Food triggers

- Aged or ripened cheeses (examples: Cheddar, Gruyère, Emmenthaler, Stilton, Brie, Gouda, Romano, Parmesan, feta, bleu, Camembert)
- Foods containing large amounts of monosodium glutamate (MSG). Asian foods often have large amounts of MSG.
- Smoked, cured, or processed meats such as bacon, sausage, ham, salami, pepperoni, pickled herring, bologna, chicken livers, and hot dogs
- Food prepared with meat tenderizer, soy sauce, vinegar (except white vinegar), or yeast extract; and food that has been fermented, pickled, or marinated
- Pea pods and pods of broad beans such as lima and navy beans
- Onions, olives, pickles
- Alcohol (especially red wine, port, sherry, Scotch, gin, and bourbon)
- Sour cream, yogurt, buttermilk
- Hot fresh bread, raised coffee cake, doughnuts
- Excessive aspartame (artificial sweetener)
- Chocolate, cocoa, carob
- Nuts, peanut butter
- Certain fruits, including figs, avocados, raisins, red plums, passion fruit, papaya, banana, and citrus fruit
- Excessive tea, coffee, cola

Other triggers

- Hormonal fluctuations
- Barometric-pressure variations
- Sleep disturbance
- Stress
- Medications

*Parts of this listing are adapted from Ronald J. Tusa, MD, PhD, “Diagnosis and Management of Neuro-otologic Disorders Due to Migraine,” chap. 12 in Vestibular Rehabilitation, ed. Susan J. Herdman, PhD, PT (Philadelphia: F.A. Davis Co., 1994).*
RECOGNITION OF MIGRAINE SYNDROMES

Most people associate migraine with severe head pain and a period of incapacitation. However, a large portion of people with migraine often have no accompanying pain, their predominant symptom instead being vertigo (a spinning sensation) or dizziness/ disequilibrium (balance loss), mental confusion, disorientation, dysarthria, visual distortion or altered visual clarity, or extremity paresis. This presentation may result in a visit to the emergency room and extensive laboratory, imaging, and other diagnostic evaluations—often with normal results, which lead to increased confusion and anxiety on the part of the patient. In addition, anti-emetic (anti-vomiting) medications are often given, which may have sedative side effects associated with increased postural instability and increased fall risks.

Clinicians are faced with the task of attempting to apply objective clinical testing methods to determine the etiology (cause) of a patient’s symptoms so as to optimize treatment. Often, a combination of etiologies exists, which can complicate or confuse the diagnostic process.

Physicians should be using the International Headache Society’s International Classification of Headache Disorders (2nd edition) in order to better diagnose patients with primary headache disorders. These criteria, used by neurologists and other headache specialists, are readily available in almost every library, either online or in print.

Migraine headaches (with or without aura), tension-type headache, cluster headache, paroxysmal hemicrania, and chronic daily headache constitute the vast majority of primary headache disorders. Variants of migraine, such as post traumatic headache from concussive injury, exertional migraine and benign orgasmic headaches, are becoming more frequently recognized. These variant presentations may also develop vestibular syndromes that are often more persistent and debilitating than the original headache.

MECHANISMS OF MIGRAINE

The emergence of new technologies, such as functional/dynamic imaging studies, has shown promise in documenting the evolution of the migraine processes. As a result, a better understanding of the vascular and neural processes of migraine has been developed.

The consensus is that the types of headache outlined above—especially migraine/vascular types—are related to a mixed pathophysiology, with cerebral spreading depression of Leão (a spontaneous spreading of an electrical charge along the cortex) followed by activation of pain receptors located in the brainstem, not far from the vestibular apparatus. The release of neurotransmitters then leads to the dilation of blood vessels near the scalp and other structures outside of the brain substance.

Migraine is also thought to be an inherited disorder giving rise to a “vulnerability” to an abnormal discharge of neurons (different from that seen in epilepsy) that preferentially affects brainstem regions and is triggered by a chemical event.
The vascular theory has been long accepted (and is perhaps better understood), which may make it difficult for some practitioners to accept the neural components and associated vestibular manifestations.

The exact mechanisms of migraine are still not completely understood. But since migraine pathophysiology has been shown to be not solely vascular, and is now thought to be a combination of altered vascular and neural processes, migraine-related vestibulopathy is easier to accept and to treat.12

EVALUATION AND TESTING

Migraine and its variants must be addressed in the clinical setting by a combination of medical management and comprehensive testing and rehabilitation techniques that offer the most complete and lasting benefit to the patient.

Traditionally, patients with recurrent vertigo associated with migraine are seen in consultation by neurologists. Otolaryngologists and internists are now becoming more familiar with this condition, but there remains a huge gap between those who care for migraine patients (with or without associated vertigo) and those who have remained “old school”—that is, not recognizing the peripheral and central vestibular components of migraine.

Patients with migraine associated vertigo (MAV) are often seen by audiologists and vestibular rehabilitation therapists for evaluation and treatment. These paramedical specialists are frequently needed to help the primary care doctor make a diagnosis of MAV.

After an initial, thorough subjective history is obtained, including a recitation of ongoing symptoms and disruption of activities of daily living, a battery of tests is typically performed, to determine a plan of care for optimized therapy. There are a large number of methods available for testing patients with MAV, and an optimal testing protocol is yet to be determined for this population. Some combination of computerized audiological and vestibular-function tests is typically employed, including positional testing with video-oculography; oculomotor and VOR (vestibulo-ocular reflex) assessments with gaze stability and/or dynamic visual acuity testing; horizontal canal testing with vENG (video electronystagmography), with caloric or rotational chair testing (preferred); audiogram and ABR (auditory brainstem response test); functional balance and gait assessments with CDP (computerized dynamic posturography); and VEMP.

In our clinic, a review of results obtained from such tests with MAV patients reveals a combination of findings that are attributable to both central processes and peripheral vestibular functions (see box on page 6).

An important component of the evaluation is reliable documentation of the degree of limitation of daily functional capacities. A number of questionnaires and inventories have been employed for this purpose, including the Jacobsen Dizziness Inventory, Dynamic Gait Index, Activities-Specific Balance Confidence Scale, Timed Up and Go test, and others.7,13

TREATMENT
The methodology believed to have the highest efficacy in the management of migraine dizziness is a combination of medications, vestibular rehabilitation, and lifestyle modifications that include limitation of the risk factors associated with migraine (those related to diet, sleep, stress, exercise, and environmental factors).

**MEDICATION**

Medications may be prescribed to prevent migraines or to stop a migraine that has already started. Drugs used to prevent frequent migraine attacks include beta-blockers, tricyclic antidepressants, calcium channel blockers, and certain anticonvulsant medications (Depakote and Topamax). Over the last several years, venlafaxine (Effexor XR) has become one of the favored preventative drug treatments for patients with migraine related vertigo. Drugs commonly used to stop migraine are aspirin, ibuprofen, isometheptene mucate, and the triptans, such as Imitrex and Relpax. Some of these medications work by blocking the action of serotonin (a neurotransmitter that causes large blood vessels to contract) or prostaglandins (a family of chemicals stimulated by estrogen that cause blood vessels to expand and contract). Generally the differentiation of whether to use a daily preventive vs an abortive type (taken to stop the already started migraine event) is the frequency and severity of the events. This is best determined by the patient’s discussion of options with the treating Neurologist.

**VESTIBULAR REHABILITATION**

The benefits of vestibular rehabilitation are well documented to reduce symptoms and restore function for vestibular-related disorders. With MAV, it is often helpful for the patient to have started the prescribed medications prior to beginning the vestibular rehabilitation course. This may allow for better tolerance to the exercise regimen without exacerbating the symptoms. The intensity of the rehabilitation course in gradually increased to the patient’s abilities, yet still at a low enough level so as to not initiate another migraine event.

For patients who have alterations in oculomotor functions and VOR deficits giving rise to visual perceptual dysfunction, a concentrated rehabilitation program consisting of VOR and gaze-stability exercises that emphasize visual acuity is effective. Various eye tracking devices are commercially available which allow the examiner to monitor not only the ability of the patient to visually track objects, but also allow the “method” of eye tracking employed by the patient to be evaluated. Spatial awareness may be altered, and exercises emphasizing proprioception and visual perception are helpful. Isolating visual fields incrementally during visual tracking exercises may be helpful in stabilizing alterations in positional sense. Vestibulo-visual interaction exercises also improve eye tracking abilities. It has become evident that velocity specific exercises are most effective. The velocity of the exercises needs to be matched to the measured velocity deficits on test results. Performing visual retraining exercises at random speeds rather than at specific velocities may be less effective. In cases where BPPV exists, performing canalith repositioning maneuvers is effective, and followed with home habituation exercises.

Postural instability and gait alterations respond to balance and gait-training tasks and exercises, employing both static and dynamic type balance exercises. Dual tasking and exercises that combine hand-eye coordination, balance maintenance, and gaze stability are effective as well, and can be combined with general conditioning exercises to the
extent tolerated by the patient’s general health. Performing exercises on various surface textures and variable stabilities also is recommended.

In patients with cervicalgia and cervical muscle spasms that limit range of motion, treatment may also include modalities and manual mobilization and stretching of the upper cervical segments, in order to diminish the muscle spasms and guarding and restore normal mobility to the neck. As an adjunct to therapy, greater occipital nerve block (GON) injections are often helpful in reducing symptoms and restoring motion. Some treating MD’s now use Botox for these injections for more lasting effect.

**LIFESTYLE MODIFICATIONS**

A consistent effort by the patient to adhere to necessary lifestyle modifications (including avoiding the migraine triggers mentioned above), medication usage as prescribed, and specific tasks and exercises performed independently at home are critical to the success of the overall rehabilitation program. Such adherence is essential for effective reduction of the symptoms and limitations of function caused by migraine associated vertigo (MAV).

**VESTIBULAR TEST RESULTS COMMONLY OBSERVED IN MIGRAINE-RELATED DIZZINESS PATIENTS**

During video-oculography, a prevalent feature is poor gaze stability with ocular “drift,” often accompanied by spontaneous up or downbeating directional nystagmus, which does not suppress with fixation-suppression testing added. Unilateral or bilateral gaze induced lateral nystagmus is commonly observed. There may also be a reduced ability to cancel or inhibit the vestibulo-ocular reflex (VOR) function, used for attaining simultaneous head and eye tracking maneuvers. These results may be due to the fact that the cerebellum, which is responsible for coordinating gaze-fixation functions, is thought to be involved in the vascular and neural changes associated with migraine.

Testing of other cerebellar functions (involving coordinated movements of the extremities) may give normal results, with no postural instability or ataxia/apraxia evident, but postural instability is often evident as well. Smooth pursuit tests often give abnormal results (although these must be distinguished from expected age-related changes). Thus, it may be that only those neural processes of the cerebellum associated with coordinated eye motions are affected in migraine, and not the neural connections involving postural stability.

Computerized dynamic posturography (CDP) may give positive results for postural instability, especially when used in combination with head motions for dual tasking and otolithic system involvement. Alterations in balance strategies are commonly measured, and need to be addressed with the specific balance exercises in accord with test measures.

Saccadic eye-motion testing is usually normal, but a rebound nystagmus may be present with hyperresponsive neural findings and presence of overshoot phenomenon. Directional gaze testing is usually abnormal, as is the Halmagyi head thrust test. HIT (head impulse test) may be helpful in documenting the objective findings of VOR
and gaze stability deficit. With Hallpike-Dix positional testing (unless true BPPV presents), no rotational component nystagmus is usually evident. However in acute migraine event, bilateral torsional nystagmus may present with positional testing and gaze added.

With passive VOR assessment via autorotation methods, or with mechanical rotational chair, an abnormal gain value with accompanying phase shift is usually evident. The visual-vestibular interaction can be markedly abnormal and may provoke symptoms of increased dizziness, often with accompanying nausea. Optokinetic after-nystagmus (OKAN) is commonly symmetrically prolonged. Subjective Visual Vertical assessment often is abnormal with accompanying spatial disorientation altered postural positional sense.

Active autorotation testing, which may be limited by cervicalgia and cervical muscle spasms with limited range of motion (often the patient moves “en bloc” to avoid eliciting dizziness), gives sporadic results. Gaze stability testing and dynamic visual acuity testing—after cervicalgia is resolved with appropriate treatments—are typically abnormal. Vestibular-evoked myogenic potentials (VEMP) testing has proven quite useful in determining differential diagnoses. Regularly, hyperactive VEMP responses are found in patients with MAV.

Audiometric testing in cases of migraine associated vertigo (MAV) typically reveals no changes in function other than occasional hyperacusis or noise sensitivity, which usually is temporary and resolves shortly after the migraine event ends. Tinnitus (most commonly associated with labyrinthitis rather than migraine), if present at all, is temporary. In cases of prolonged problematic tinnitus, tinnitus retraining therapy (TRT) may be helpful. Tinnitus masking devices are also commercially available.

**SUMMARY**

Migraine associated vertigo (MAV) afflicts a large percent of the population and continues to be a challenge to healthcare professionals. Technologies for measurement continue to expand and new medications continue to be manufactured for this affliction. Effective management of MAV necessitates a comprehensive effort and active participation of the patient, the treating physician, and the rehabilitation professionals. Proper identification, objective diagnostic measurements, and optimized treatment approaches net the best results.

**References**

Ménière’s Disease

HOW DID MÉNIÈRE’S DISEASE GET ITS NAME?

In 1861 the French physician Prosper Ménière theorized that attacks of vertigo, ringing in the ear (tinnitus) and hearing loss came from the inner ear rather than from the brain, as was generally believed at the time. Once this idea was accepted, the name of Dr. Prosper Ménière began its long association with this inner ear disease and with inner ear balance disorders in general.

WHAT IS MÉNIÈRE’S DISEASE?

Ménière’s disease is a chronic, incurable vestibular (inner ear) disorder defined in 1995 by the Committee on Hearing and Equilibrium of the American Academy of Otolaryngology—Head and Neck Surgery as “the idiopathic syndrome of endolymphatic hydrops.”¹ In plain language, this means that Ménière’s disease, a form of endolymphatic hydrops, produces a recurring set of symptoms as a result of abnormally large amounts of a fluid called endolymph collecting in the inner ear.

Ménière’s disease can develop at any age, but it is more likely to happen to adults between 40 and 60 years of age. The exact number of people with Ménière’s disease is difficult to measure accurately because no official reporting system exists. Numbers used by researchers differ from one report to the next and from one country to the next. The National Institutes of Health estimates that about 615,000 people in the U.S. have Ménière’s disease and that 45,500 new cases re-diagnosed each year.²

CAUSES
The exact cause and reason why Ménière’s disease starts is not yet known. Many theories have been proposed over the years. They include: circulation problems, viral infection, allergies, an autoimmune reaction, migraine, and the possibility of a genetic connection.

Experts aren’t sure what generates the symptoms of an acute attack of Ménière’s disease. The leading theory is that they result from increased pressure of an abnormally large amount of endolymph in the inner ear and/or from the presence of potassium in an area of the inner ear where it doesn’t belong. These conditions may be due to breaks in the membrane separating endolymph from the other inner ear fluid, perilymph. Some people with Ménière’s disease find that certain events and situations, sometimes called triggers, can set off attacks. These triggers include stress, overwork, fatigue, emotional distress, additional illnesses, pressure changes, certain foods, and too much salt in the diet.

**PROGRESSION OF SYMPTOMS**

Common symptoms of a Ménière’s disease attack do not reflect the entire picture of the disorder, because symptoms vary before, during, between, and after attacks, and also during the late-stage of Ménière’s disease.

Ménière’s disease may start with fluctuating hearing loss, eventually progressing to attacks of vertigo and dizziness. Oncoming attacks are often preceded by an “aura,” or the specific set of warning symptoms, listed below. Paying attention to these warning symptoms can allow a person to move to a safe or more comfortable situation before an attack.

- balance disturbance
- dizziness, lightheadedness
- headache, increased ear pressure
- hearing loss or tinnitus increase
- sound sensitivity
- vague feeling of uneasiness

During an attack of early-stage Ménière’s disease, symptoms include:

- spontaneous, violent vertigo
- fluctuating hearing loss
- ear fullness (aural fullness) and/or tinnitus

In addition to the above main symptoms, attacks can also include:

- anxiety, fear
• diarrhea
• blurry vision or eye jerking
• nausea and vomiting
• cold sweat, palpitations or rapid pulse
• trembling

  Following the attack, a period of extreme fatigue or exhaustion often occurs, prompting the need for hours of sleep.

  The periods between attacks are symptom free for some people and symptomatic for others. Many symptoms have been reported after and between attacks:

• anger, anxiety, fear, worry
• appetite change
• clumsiness
• concentration difficulty, distractibility, tendency to grope for words
• diarrhea
• fatigue, malaise, sleepiness
• headache, heavy head sensation
• lightheadedness (faintness)
• loss of self-confidence and self-reliance
• nausea, queasiness, motion sickness
• neck ache or stiff neck
• palpitations or rapid pulse, cold sweat
• sound distortion and sensitivity
• unsteadiness (sudden falls, staggering or stumbling, difficulty turning or walking in poorly lit areas, tendency to look down or to grope for stable handholds)
• vision difficulties (problems with blurring, bouncing, depth perception, glare intensification, focusing, watching movement; difficulty looking through lenses such as binoculars or cameras)
• vomiting
Late-stage Ménière’s disease refers to a set of symptoms rather than a point in time. Hearing loss is more significant and is less likely to fluctuate. Tinnitus and/or aural fullness may be stronger and more constant. Attacks of vertigo may be replaced by more constant struggles with vision and balance, including difficulty walking in the dark and occasional sudden loss of balance. Sometimes, drop attacks of vestibular origin (Tumarkin’s otolithic crisis) occur in this stage of Ménière’s disease and are characterized by sudden brief loss of posture without loss of consciousness. Some of these late-stage symptoms can become more problematic in conditions of low lighting, or with fatigue, or when a person is exposed to visually stimulating situations.

**DURATION AND FREQUENCY OF ATTACKS**

Attacks can last from 20 minutes to 24 hours. They can occur with the frequency of many attacks each week; or they can be separated by weeks, months, and even years. The unpredictable nature of this disease makes managing it challenging. It also complicates the ability of scientists and physicians to study it.

**IS THERE A CURE?**

To “cure” a disease means to eliminate the root cause of the disease and reverse the damage it has inflicted (on the inner ear, in this case). No treatment currently exists to cure Ménière’s disease. However, medical treatments exist that can help manage it.

**TREATMENT**

Existing treatments fall into two categories. Some treatments aim at reducing the severity of an attack while it is occurring; some treatments attempt to reduce the severity and number of attacks in the long term. Experts feel these medical treatments provide some degree of improvement in 60–80% of the treated people. Gentamicin is >80% effective at control of vertigo. The most conservative long-term treatment for Ménière’s disease in the U.S. involves adhering to a reduced-sodium diet and using medication that helps control water retention (diuretics or “water pills”). The goal of this treatment is to reduce inner-ear fluid pressure. Some physicians, more commonly outside of the U.S., also weigh the potential efficacy of using beta-histine HCl (Serc) as a vestibular suppressant for Ménière’s disease. Medications can be used during an attack to reduce the vertigo, nausea/vomiting or both. Some drugs used for this include diazepam (Valium), lorazepam (Ativan), promethazine (Phenergan), dimenhydrinate (Dramamine Original Formula), and meclizine hydrochloride (Antivert, Dramamine Less Drowsy Formula). Vestibular rehabilitation therapy (VRT) is sometimes used to help with the imbalance that can plague people between attacks. Its goal is to help retrain the ability of the body and brain to process balance information. When successful, this can help a person regain confidence in the ability to move about.

When conservative treatments don’t work: For the 20–40% of people who do not respond to medication or diet, a physician may recommend a treatment that involves more physical risk. One such method, a intratympanic gentamicin, destroys vestibular tissue with injections into the ear of the aminoglycoside antibiotic (gentamicin). Recently, intratympanic steroid injections have been used with less risk of hearing loss and persistent imbalance.
Another less conservative treatment method involves surgery. Two categories of surgery are available. The goal of the first type is to relieve the pressure on the inner ear. Surgery to reduce pressure is not as widely used now as it was in the past due to questions about its long-term effectiveness.

The goal of the second type of surgery is to block the movement of information from the affected ear to the brain. The process involves either destroying the inner ear so that the ear does not generate balance information to send to the brain, or destroying the vestibular nerve so that balance information is not transmitted to the brain. In either instance, physical therapy is useful to help the brain compensate from the loss of inner ear function due to surgery.

**PROGNOSIS**

It is difficult to predict how Ménière’s disease will affect a person’s future. Symptoms can disappear one day and never return. Or they might become so severe that they are disabling.

**COPING**

Coping with Ménière’s disease is challenging because attacks are unpredictable, it is incurable, some of the symptoms are not obvious to others, and most people know virtually nothing about the disorder. Many people with Ménière’s disease are thrust into the role of educator—they must teach themselves, their family, friends, coworkers, and sometimes even health care professionals about the disorder and how it impacts them. Key features of communicating with family and friends include informing them about what might happen with the onset of an acute attack and how they can help. If a low-sodium diet is effective, family and friends should be informed about how important it is for them to support adherence to the diet regimen. Changes in lifelong eating patterns can be easier with the assistance of others.

Managing an acute attack involves preparation. This includes consulting with a physician about any appropriate drugs that can be taken when an acute attack occurs, and deciding ahead of time when it is appropriate to go to a hospital. During an attack, it is helpful to lie down in a safe place with a firm surface, and avoid any head movement. Sometimes keeping the eyes open and fixed on a stationary object about 18 inches away is helpful. In order to control dehydration, a doctor should be called if fluid intake is not possible over time due to persistent vomiting.

After an acute attack subsides, it is not uncommon to want to sleep for several hours. Resting in bed for a short time is appropriate, if the person is exhausted. But it is also important for the person to get up and move around as soon as possible so that the brain readjusts to the changed balance signals. Precautions need to be taken in this process to accommodate any new balance sensations.

Successfully coping with symptoms involves understanding the disease. Talking with health care providers, communicating with other people who are experiencing the same disease, and reading books and articles about the topic are all helpful methods of learning more about Ménière’s disease.

*References*
Otosclerosis

WHAT IS OTOSCLEROSIS?

Otosclerosis is the abnormal growth of bone of the MIDDLE ear. This bone prevents structures within the ear from working properly and causes hearing loss. For some people with otosclerosis, the hearing loss may become severe.

How do we hear? Hearing is a series of events in which the ear converts sound waves into electrical signals and causes nerve impulses to be sent to the brain where they are interpreted as sound. The ear has three main parts: the outer, middle, and inner ear. Sound waves enter through the outer ear and reach the middle ear, where they cause the eardrum to vibrate. The vibrations are transmitted through three tiny bones in the middle ear called the ossicles. These three bones are named the malleus, incus, and stapes (and are also known as the hammer, anvil, and stirrup). The eardrum and ossicles carry the vibrations to the inner ear. The stirrup transmits the vibrations through the oval window and into the fluid that fills the inner ear.

The vibrations move through fluid in the snail-shaped hearing part of the inner ear (cochlea) that contains the hair cells. The fluid in the cochlea moves the top of the hair cells, which initiates the changes that lead to the production of the nerve impulses. These nerve impulses are carried to the brain, where they are interpreted as sound. Different sounds stimulate different parts of the inner ear, allowing the brain to distinguish among various sounds, for example, different vowel and consonant sounds.

HOW DOES OTOSCLEROSIS CAUSE HEARING IMPAIRMENT?

Otosclerosis can cause different types of hearing loss, depending on which structure within the ear is affected. Otosclerosis usually affects the last bone in the chain, the stapes, which rests in the entrance to the inner ear (the oval window). The abnormal bone fixates the stapes in the oval window and interferes with sound passing waves to the inner ear.
Otosclerosis usually causes a conductive hearing loss, a hearing loss caused by a problem in the outer or middle ear. Less frequently, otosclerosis may cause a sensorineural hearing loss (damaged sensory cells and/or nerve fibers of the inner ear), as well as a conductive hearing loss.

**WHAT CAUSES OTOSCLEROSIS?**

The cause of otosclerosis is not fully understood, although research has shown that otosclerosis tends to run in families and may be hereditary, or passed down from parent to child. People who have a family history of otosclerosis are more likely to develop the disorder. On average, a person who has one parent with otosclerosis has a 25 percent chance of developing the disorder. If both parents have otosclerosis, the risk goes up to 50 percent. Research shows that white, middle-aged women are most at risk.

Some research suggests a relationship between otosclerosis and the hormonal changes associated with pregnancy. While the exact cause remains unknown, there is some evidence associating viral infections (such as measles) and otosclerosis.

**WHAT ARE THE SYMPTOMS OF OTOSCLEROSIS?**

Hearing loss is the most frequent symptom of otosclerosis. The loss may appear very gradually. Many people with otosclerosis first notice that they cannot hear low-pitched sounds or they can no longer hear a whisper.

In addition to hearing loss, some people with otosclerosis may experience dizziness, balance problems, or tinnitus. Tinnitus is a sensation of ringing, roaring, buzzing, or hissing in the ears or head that accompanies many forms of hearing loss.

**HOW IS OTOSCLEROSIS DIAGNOSED?**

An examination by an otolaryngologist or otologist is needed to rule out other diseases or health problems that may cause these same symptoms. An audiologist is a hearing health care professional who is trained to identify, measure, and rehabilitate hearing impairment and related disorders. An audiologist uses a variety of tests and procedures to assess hearing and balance function. The audiologist may produce an audiogram (a graph that shows a person's hearing sensitivity) and a tympanogram (a graph that shows how well the middle ear functions to conduct sound). Discuss these results with your audiologist/otologist.

**HOW IS OTOSCLEROSIS TREATED?**

In many cases surgery is an option for treatment of otosclerosis. In an operation called a stapedectomy, a surgeon (otolaryngologist or otologist) bypasses the diseased bone with a prosthetic device that allows sound waves to be passed to the inner ear. It is important to discuss the risks and possible complications of this procedure, as well as the benefits, with the surgeon. In rare cases, surgery can worsen the hearing loss.
If the hearing loss is mild, surgery may not be an option. Also, on occasion, some hearing loss persists after surgery. A properly fitted hearing aid may help some people with otosclerosis in situations that include persistent hearing loss. A hearing aid is designed to compensate for a hearing loss by amplifying sound. An audiologist can discuss the various types of hearing aids available and make a recommendation based on the specific needs of an individual.

**WHAT RESEARCH IS BEING DONE ON OTOSCLEROSIS?**

Scientists are conducting research to improve understanding of otosclerosis. Genetic studies continue in order to identify the gene or genes that may lead to this disorder. Other researchers are studying the effectiveness of lasers currently used in surgery, of amplification devices, and of various stapes prostheses. Improved diagnostic techniques are also being examined and developed.

**Ototoxicity**

**WHAT IS OTOTOXICITY?**

Oto = ear; toxicity = poisoning

Ototoxicity is, quite simply, ear poisoning (oto = ear, toxicity = poisoning), which results from exposure to drugs or chemicals that damage the inner ear or the vestibulo-cochlear nerve (the nerve sending balance and hearing information from the inner ear to the brain). Because the inner ear is involved in both hearing and balance, ototoxicity can result in disturbances of either or both of these senses. The parts of the brain that receive hearing and balance information from the inner ear can also be affected by poison, but this is not technically considered ototoxicity and won’t be covered in this information sheet. (Poisoning of the brain is classified as neurotoxicity)

The occurrence and degree of inner ear poisoning depends upon the drug involved as well as other factors such as heredity. Ototoxicity can be temporary or permanent. The effect of certain drugs is often temporary, while other drugs typically produce permanent changes to the ear. Some drugs can cause either temporary or permanent problems. It is important to note here that the broad majority of people who experience ototoxicity have a temporary or reversible form that does not result in a major or long-term disruption in their lives.

With cochleotoxicity, hearing loss or the start or worsening of tinnitus (ringing in the ears) can occur through damage to the cochlea (the hearing apparatus) or the cochlear branch of the vestibulo-cochlear nerve. Vestibular ototoxicity or vestibulotoxicity are terms used to describe ototoxicity that affects the balance organs or the vestibular branch of the vestibulo-cochlear nerve.

It is important to note that no drug is known to cause Ménière’s disease, benign paroxysmal positional vertigo, or any other vestibular disorder causing fluctuating function.
HOW COMMON IS OTOTOXICITY?

No one knows how many people suffer from ototoxicity each year or the percentage of vestibular disorders caused by ototoxicity. What is known is that when permanent and extensive ototoxicity occurs, the effects can take a terrible toll on a person’s ability to function.

WHAT SUBSTANCES CAN CAUSE OTOTOXICITY?

Scientific studies are required to confirm whether a drug is ototoxic. Unfortunately, such research often involves years of study. When assessing the safety of a drug prior to releasing it on the market, the U.S. Food and Drug Administration does not require testing of inner ear function or examination of the inner ear structures. This is one reason it is almost impossible to say with confidence how many and which drugs cause ototoxicity and how many or which people are affected by it.

Problems with a particular drug are usually only discovered after enough people have suffered the consequences and when physicians or other health care professionals can see a probable connection between the symptoms or problems and a drug. This was the case with aspirin and quinine centuries ago, with the antibiotic streptomycin in the 1940s, and more recently with some anti-cancer drugs. Since then, scientific studies have shown that these drugs cause ototoxicity in animals and people. Other, newer drugs have been implicated as ototoxic as well, but solid scientific proof is often lacking.

Many chemicals have ototoxic potential, including over-the-counter drugs, prescription medications, and environmental chemicals. The information below includes substances thought to cause ototoxicity. The discussion is incomplete because of the limited research thus far. *Note: if you are taking drugs on the advice of your physician, DO NOT STOP TAKING THEM just because you see them listed here!* Speak with your doctor about your concerns to determine the best choice in your own unique situation.

**Aspirin and quinine** Aspirin (acetylsalicylic acid, ASA) and quinine are well known to cause temporary ototoxicity resulting in tinnitus. They may also reduce hearing, particularly when given at high doses. Quinine products can also temporarily reduce balance ability. Once aspirin or quinine is stopped, the ototoxicity generally disappears. Some quinine products include:

- chloroquine
- quinidine
- quinine (including Q-vel)
- tonic water

**Loop diuretics** are a specific family of “water pills” that is known to occasionally cause temporary ototoxicity. These drugs cause ringing in the ears or decreased hearing that reverses when the drug is stopped.

An increased probability of ototoxicity is thought to occur with loop diuretics when they are administered during the same time period that an aminoglycoside antibiotic (see next section) is given. The loop diuretics include:
• bumetanide (Bumex)
• ethacrynic acid (Edecrin)
• furosemide (Lasix)
• torsemide (Demadex)

*Note:* Hydrochlorothiazide (HCTZ) and Maxide—diuretics commonly prescribed to people with Ménière’s disease or other forms of endolymphatic hydrops—are not loop diuretics.

**Aminoglycoside antibiotics** All members of the aminoglycoside antibiotic family are well known for their potential to cause permanent ototoxicity if they enter the inner ear. Some of these drugs are more likely to cause hearing loss; others are more likely to cause vestibular loss. Others can cause either problem.

A higher risk for aminoglycoside-antibiotic induced ototoxicity occurs when a person receives concurrent ototoxic drugs (such as a loop diuretic or another antibiotic—vancomycin), has insufficient kidney function or is receiving a drug that causes insufficient kidney function, or has a genetic vulnerability.

The risk of ototoxicity also increases with an increasing amount of the drug that enters the blood stream, the longer the drug is in the body, and the duration of time the drug is taken.

Aminoglycoside antibiotics can enter the inner ear through the blood system or via diffusion from the middle ear into the inner ear. They enter the blood stream in largest amounts when given intravenously (by IV) and in the least amounts by pill. Inhaled drugs also enter the blood stream; an example of this is the use of inhaled tobramycin for long-term treatment of cystic fibrosis.

*Can ear drops containing aminoglycosides be problematic?* If they find their way into the middle ear in large enough quantities, such ear drops can diffuse into the inner ear and cause damage. Physicians do not agree about how often and under what circumstances this occurs. Many papers in medical journals address this argument.

Members of the aminoglycoside family include:

• amikacin
• netilmicin
• dihydrostreptomycin
• ribostamycin
• gentamicin
• streptomycin
• kanamycin
• tobramycin
Anti-cancer drugs work by killing cancer cells. Unfortunately some can also damage or kill cells elsewhere in the body, including the ears. **Cisplatin** is well known to cause massive and permanent hearing loss. **Carboplatin** is also known to be ototoxic.

**Environmental chemicals** have long been implicated in ototoxicity. Little research has been done to substantiate their precise effect on ears, but most are associated with hearing disturbances that may be permanent. In addition, mercury has also been linked to permanent balance problems. These include:

- butyl nitrite
- mercury
- carbon disulfide
- styrene
- carbon monoxide
- tin
- hexane
- toluene
- lead
- trichloroethylene
- manganese
- xylene

**WHAT DAMAGE OCCURS?**

Two areas can be damaged or destroyed through ototoxicity: the hair cells within the inner ear, and the vestibulocochlear nerve that links the inner ear to the brain. When damage occurs, any degree and combination of hearing loss and balance disruption are possible depending upon the part(s) affected.

Hair cells are located in both the cochlea and the vestibular areas of the inner ear. They are composed of a cell body with a hair-like attachment. When these “hairs” are normally bent with sound vibrations or movement, they send electrical signals to the brain about hearing or balance function. In ototoxicity, these hairs can be damaged to the point that they no longer stand up, thus reducing the auditory and/or balance signals sent to the brain.

**WHAT ARE THE SYMPTOMS?**
Cochleotoxicity symptoms range from mild tinnitus to total hearing loss, depending upon each person and the form and level of exposure to the ototoxin. They can include one-sided or two-sided hearing loss and constant or fluctuating tinnitus.

Vestibulotoxicity symptoms range from mild imbalance to total incapacitation. Symptoms of a vestibular or balance function loss depend upon the degree of damage, if the damage occurred rapidly or slowly, if it’s one-sided or two-sided, and how long ago the damage occurred. A slow one-sided loss might not produce any symptoms, while a rapid loss could produce enough vertigo, vomiting, and nystagmus (eye jerking), to keep a person in bed for days. Most of the time, the symptoms slowly pass, allowing a person to return to normal activities.

A two-sided loss in vestibulotoxicity typically causes headache, a feeling of ear fullness, imbalance to the point of being unable to walk, and a bouncing and blurring of vision (oscillopsia) rather than intense vertigo, vomiting, and nystagmus. It also tends to produce inability to tolerate head movement, a wide-based gait (walking with the legs farther apart than usual), difficulty walking in the dark, unsteadiness or the sensation of unsteadiness, lightheadedness, and significant fatigue. If the damage is severe, symptoms such as oscillopsia and problems with walking in the dark or with the eyes closed will not diminish with time.

**HOW IS OTOTOXICITY DIAGNOSED?**

The diagnosis is based upon the patient’s history, symptoms, and test results. There is no specific test for ototoxicity; this makes a positive history for ototoxin exposure crucial to the diagnosis. Some of the tests that may be used to determine how much hearing or balance function have been lost involve the vestibular autorotation test (VAT), vestibulo-ocular reflex testing equipment (VORTEQ), electronystagmography (ENG), computerized dynamic posturography (CDP), rotary chair (SHAT), head-shaking, electrocochleography (EcoG), auditory brainstem response (ABR), otoacoustic emissions, pure tone audiometry, speech discrimination, and most other tests often used to identify and quantify inner ear problems.

**WHAT IS THE TREATMENT?**

At present there are no treatments that can reverse the damage. Currently available treatments focus on reducing the effects of the damage and rehabilitating function. Specifically, individuals with hearing loss may be helped with hearing aids; those with profound bilateral (two-sided) hearing loss have been shown to benefit from cochlear implants. In fact, many early recipients of cochlear implants were victims of ototoxicity. When a loss of balance function has occurred, physical therapy can help the brain become accustomed to the altered balance signals coming from the inner ear. Physical therapy can also assist an individual in developing other ways to maintain balance such as emphasizing the use of vision and proprioception—the sensation felt by the soles of the feet, the ankles, knees, and hips—and structuring a program of general physical conditioning and exercises designed to strengthen and tone muscles.

**LONG-TERM GOALS**
The major long-term goals include continuing with conditioning activities to improve balance function, protecting the other systems involved with maintaining balance, and preventing further ototoxic damage.

Protection of other components of balance—vision and proprioception—is essential. Good vision is crucial in the face of a severe vestibular loss. Yearly ophthalmological examinations that include a glaucoma check should become routine. Use of ultraviolet (UV) eye protection in the sun and eye protection in the wind (such as goggles or sunglasses) should be considered.

Protecting proprioception involves taking precautions such as avoiding walking barefooted on any surface that could injure or damage the soles (such as on a macadam road surface), not wearing clothing that restricts circulation to the legs and feet (such as a tight girdle), and taking off excess body weight that can cause knee and hip difficulties.

Avoidance of ototoxic substances is also very important because individuals who have suffered from ototoxicity have a higher likelihood of experiencing it again, if exposed. A medic alert tag might be helpful for warning health care professionals about the need to avoid prescribing ototoxic medications unless needed to save your life. Such tags might also serve to flag an existing reduction in balance and/or hearing function.

**PREVENTION**

Limit using drugs to those that are absolutely needed and follow the instructions carefully for those medications that are prescribed for you. If possible, avoid taking multiple types of ototoxic drugs (aspirin, quinine, loop diuretics, and aminoglycosides). When using airborne chemicals that are potentially ototoxic, good ventilation should be used. Open the windows, turn on a fan, and refrain from using the chemical for any longer than necessary. Stay well hydrated.

**A LOOK AT THE FUTURE**

Ongoing related research addresses prevention and treatment. Chemicals are being evaluated for their ability to prevent ototoxicity and that might be prescribed in tandem with ototoxic drugs in the future. Investigators are also studying methods of hair-cell and nerve-cell regeneration. In the distant future, it may be possible to stimulate the ear into growing replacement hair cells and to repair damaged nerve fibers.

**ENDNOTE**

Most of the drugs listed in this document appear because strong evidence exists to show that they cause or probably cause ototoxicity. This evidence includes at least one of the following criteria:

- Large numbers of isolated reports about particular drugs or chemicals
- Experiments showing that animals develop ototoxicity when given the drug
Multiple post-mortem studies that demonstrate changes in the ear that are linked to ototoxins in people who took certain drugs and who subsequently developed symptoms of ototoxicity. (Such ear damage can only be observed after death, when the ears can be examined fully.) An example of this type of research is Zheng et al, 2001.

Scientific reports about groups of people tested before (if possible), during, and after their use of a drug, some of whom were found to develop ototoxicity while taking the drug. An example of this type of research is Black et al, 2001.

References
1. The entire October 1993 issue of The Otolaryngologic Clinics of North America is devoted to ototoxicity, as is the entire November 1999 issue of Annals of the New York Academy of Sciences.


**Pediatric Vestibular Disorders**

**VESTIBULAR DISORDERS AFFECT CHILDREN AS WELL AS ADULTS**

The vestibular system is important for the development of normal movement reactions, motion tolerance, and motor control for postural alignment, balance, and vision. A vestibular system that is damaged by disease or injury in childhood can have a major impact on a child’s development. Despite advances in testing and documentation of vestibular deficits in children, vestibular problems continue to be an overlooked entity in children. Many children are not receiving treatment that could significantly improve function and address developmental delays caused by vestibular disorders.

**STAGES OF DEVELOPMENT AND THE VESTIBULAR SYSTEM**

The vestibular organs provide sensory information about motion, and spatial orientation. The organs in each ear include the utricle, saccule, and three semicircular canals. The utricle and saccule detect gravity (vertical orientation) and linear movement. The semicircular canals detect rotational head movements and are located at right angles to each other. When these organs on both sides of the head are functioning properly, they send symmetrical signals to the brain that are integrated with other sensory and motors systems by age six.

If vestibular dysfunction occurs early in development, it slows the development of equilibrium and protective reflexes and motor-control tasks such as sitting unsupported, standing, and walking. In addition, an impaired vestibulo-ocular reflex (VOR) from vestibular dysfunction can have far-reaching impacts on a child’s ability to keep pace with schoolwork. The VOR is responsible for maintaining clear vision during rapid head movements. Stable vision is important for learning to read and write and for developing fine and gross motor control. If left untreated, a vestibular disorder can have adverse consequences for a range of functions as the child grows to adulthood.
SIGNS AND SYMPTOMS

Vestibular disorders are not as easily recognized in children as they are in adults, in part because children often cannot describe their symptoms well and may be unable to understand the concepts of vertigo and imbalance. Identification of pediatric vestibular dysfunction requires coordinating descriptions offered by the child, symptom reports from parents, and clinical observations by professionals.

SYMPTOMS AND SIGNS THAT MAY INDICATE VESTIBULAR DYSFUNCTION INCLUDE:

- Dizziness and visual acuity problems, especially with head movements such as when turning to look at something
- Poor spatial relationships, sometimes revealed by skipping words or letters while reading or by having a disorganized writing style.
- Nystagmus (involuntary, alternating, rapid, and slow eye movements)
- Difficulty navigating in the dark
- Hearing loss or tinnitus (ringing in the ears)
- Motion sickness or sensitivity (avoids or craves movement)
- Nausea
- Abnormal movement patterns, unsteady gait, clumsiness (including decreased eye-hand and eye-foot coordination), or poor posture— including a tendency to fall, lean, or tilt over
- Ear pressure
- Headaches with associated nausea and/or dizziness
- Developmental and reflex delays that are sometimes revealed by slower achievement of milestones such as riding a bicycle, swimming, hopping, and stair climbing involving alternating left-right leg movements

For each child, the specific set of signs and symptoms will differ based on whether the damage is peripheral (involving the organs in the inner ear), central (involving the brain and brain stem), or both. The signs and symptoms will also depend on whether the damage is unilateral (on one side) or bilateral (affecting both ears), and whether the disease or injury has caused a complete loss of function, reduced function (hypofunction), or increased sensitivity (hyperfunction).

CAUSES OF VESTIBULAR DYSFUNCTION

The interconnection of the vestibular system with many other body systems can result in vestibular dysfunction secondary to a range of medical conditions and histories. Histories sometimes associated with peripheral or central system dysfunction include:
- Chronic ear infections or otitis media\textsuperscript{10,11}
- Congenital sensorineural hearing loss\textsuperscript{1,12,13,14}
- Cytomegalovirus and other viral infections such as in Ramsay Hunt syndrome (an infection of the facial and cochleovestibular nerves caused by the herpes zoster virus, the same virus that is associated with chicken pox)
- Malformations from acquired or genetic conditions such as branchio-oto-renal syndrome, Mondini dysplasia, and Waardenburg syndrome
- Other genetic disorders such as Usher syndrome-type I (with severe profound sensorineural hearing loss and balance problems and deteriorating vision by age 10) or type III (with balance and vision problems appearing later in life)
- Anoxia (reduced oxygen at birth) or stroke
- Meningitis (inflammation of the membranes covering the brain and spinal cord, sometimes also affecting membranes in the inner ear)
- Neurological disorders or conditions such as cerebral palsy, hydrocephalus, a posterior brain tumor, or Wallenberg syndrome (caused by a stroke from blockage in the vertebral or posterior inferior cerebellar artery of the brain stem)
- Maternal drug or alcohol abuse during pregnancy\textsuperscript{15,16}
- Immune-deficiency disorders\textsuperscript{17,18}
- Metabolic disorders such as diabetes
- Vascular insufficiencies
- Head-neck trauma from car accidents or sports injuries\textsuperscript{19}

**SPECIFIC VESTIBULAR DISORDERS THAT CAN OCCUR IN CHILDHOOD INCLUDE:**

- Childhood paroxysmal vertigo (CPV), also known as benign paroxysmal vertigo (BPV), is the most common pediatric vestibular disorder associated with dizziness, and is sometimes referred to as migraine equivalent. It is a central vestibular disorder typically seen in children aged 2–12. It is characterized by true spinning vertigo, nystagmus, nausea, and vomiting. A child with CPV is often sensitive to motion, light, and sound, but is typically asymptomatic after sleeping. Children tend to grow out of CPV; however, sometimes CPV progresses into migraine-associated vertigo in adulthood. This disorder is also referred to with a variety of other names and acronyms, but it is not the same as benign paroxysmal positional vertigo (BPPV).
• **Vestibular neuritis** causes dizziness, and **labyrinthitis** causes both dizziness and hearing symptoms. Both result from severe ear infections and can cause acute symptoms of vertigo and nausea that usually, but not always, subside within 4–6 weeks. These disorders are attributed to a viral infection of the trigeminal ganglion, a collection of nerve cells located just behind the ear, (vestibular neuritis) or bacterial or viral infection of the inner ear (labyrinthitis).

• **Ototoxicity** (ear poisoning) is caused by medications that destroy the hair cells of the inner ear, which transmit balance signals from the inner ear to the brain. Drugs that can cause significant bilateral damage are intravenously administered aminoglycoside antibiotics (such as gentamicin) and certain types of chemotherapies, which in children can result in severe imbalance, falls, and visual-motor problems, including oscillopsia (bouncing vision). Many children who have experienced ototoxicity have difficulties at school, working at a computer, or learning to drive an automobile.

• **Ménière’s disease** is less common in children. It is a peripheral disorder involving an imbalance of an inner ear fluid called endolymph. Symptoms include fluctuating hearing loss, tinnitus, ear pressure or fullness sensations, and episodes of vertigo and nausea.

• Some less common vestibular disorders in children include these peripheral disorders: **perilymph fistula** (a tear in the oval or round window membranes of the inner ear), and **enlarged vestibular aqueduct syndrome** (an abnormally large tube connecting the inner ear’s endolymph to the endolymphatic sac). **Benign paroxysmal positional vertigo** (BPPV), a condition caused by dislodged otoconia in a semicircular canal that abnormally stimulate movement-sensing nerve cells, is sometimes seen in children. However, it has a much lower incidence in children than in adults. It is also sometimes observed as a surgical complication of cochlear implantation.

**EVALUATION**

Diagnosing and evaluating a vestibular disorder appropriately involves collaboration among specialists. Medical evaluation is essential and will begin with a comprehensive history taking and physical exam by a physician. To help rule out non-vestibular causes of symptoms, this physician may order imaging tests such as a MRI or CT scan. A referral to a specialist (an otolaryngologist, ophthalmologist, otologist, or neurotologist) may lead to diagnostic tests that measure hearing, eye movement, and peripheral vestibular function (performed by an audiologist) and an assessment of balance and functional impairments (evaluated by physical and occupational therapists with advanced training in balance disorders).
TESTS OF VESTIBULAR AND BALANCE FUNCTION

Vestibular function testing is designed to determine if peripheral unilateral or bilateral damage has occurred and whether there is a resulting absence of function, hypofunction, or hyperfunction. In most cases, vestibular function tests can be performed on children aged one year or older by specialists whose training in pediatric care equips them to modify the equipment and techniques as needed, provide assurances that minimize potential apprehension, and ensure cooperation.

Electronystagmography (ENG) or videonystagmography (VNG) is a group of eye-movement tests used to look for signs of vestibular dysfunction or neurological problems. ENG/VNG is used to measure the ability of the semicircular canals to stabilize vision with different stimuli such as with head rotation or movement in different planes, and with warm or cold water circulating in one ear canal (a caloric test). ENG/VNG can evaluate whether signals originating from one side are consistent (symmetrical) with signals from the other side or if responses are absent, hyperactive, or hypoactive. In addition, ENG/VNG can be used with off-vertical axis rotation to test the function of the utricle.

Vestibular evoked myogenic potential (VEMP) testing evaluates whether the saccule and the inferior vestibular nerve are intact and functioning normally. When sound is transmitted through earphones or via bone conduction with a specialized vibrator placed behind the ear, electrodes placed on the neck muscles record the response of the muscle to the vestibular stimuli.

Balance and posture evaluations address whether sensory information from the vestibular system coordinates and integrates with vision and somatosensory information from the muscles and joints. Such tests require the child to balance under various vision conditions (e.g., with eyes open and closed or with a moving visual field) and varied standing surfaces (e.g., on a firm, soft, or moving surface). For diagnostic purposes, balance testing may be performed with sophisticated equipment such as computerized dynamic posturography (CDP). For treatment evaluation purposes, CDP or simpler equipment such as a pillow of dense foam may be used. In addition, Standardized Developmental Motor Scales and other assessments employ various eye-hand and eye-foot coordination tasks, balance activities, fine motor tasks, and gaze stability measurements to evaluate the child’s development, reflexes, and ability to use the vestibular system for balance, coordination, and visual-motor control.

TREATMENT

Evaluation and assessment results help determine the most effective treatment plan. For example, dietary changes are critical in children with childhood paroxysmal vertigo.

Depending on the diagnosis, recovery can be complete or nearly complete with unilateral peripheral vestibular dysfunction. With bilateral and central problems, some adaptation is needed to achieve near normal function.

Children with vestibular disorders often respond well to a specialized form of therapy called vestibular rehabilitation therapy (VRT). If surgery is needed to stabilize or correct a condition, VRT can also help with post-surgery
recovery. With unilateral peripheral or central dysfunction, VRT may focus on habituation and training in using remaining vestibular function; however, with bilateral dysfunction, VRT must focus on training substitution (i.e., heavier reliance on sensory information from the visual and somatosensory systems).

VRT is an exercise program tailored to address eye-movement control, dynamic visual acuity, balance, developmental reflexes, and body-movement functions. VRT exercises should be based on a child’s age, interests, level of comprehension, and test results. For example, appropriate balance or eye-movement exercises for a child aged 4 may involve block designs or balancing on a therapy ball. For a child aged 7, the same goal may be accomplished with a visual maze or balance obstacle course.

VRT can be effective for reducing or eliminating vertigo, improving visual-motor control, improving balance and coordination, improving visual acuity, and promoting normal development. Children typically respond more quickly to VRT than adults, because of their greater plasticity—the ability of their neurological systems to more quickly compensate for and adapt to vestibular deficits. In addition, children tend to be less fearful of movement than adults, so they participate well in the balance and movement aspects of therapy, if the therapist keeps it fun and interesting.

COPING

The effectiveness of a child’s VRT treatment program depends in part upon the cooperation, patience, and understanding of parents and caretakers in supporting compliance and progress. Parents should be provided with a specific home-exercise program to reinforce the VRT movements and activities performed at the clinic. Teachers and occupational and physical therapists can also help integrate vestibular training into activities at school. They should work closely with the parents to identify the child’s tolerances for vestibular therapy and to develop and modify the individualized therapy program accordingly. Calming activities should be included as part of the program in order to help the child avoid becoming overly stimulated, sick, frightened, or stressed. A well-structured VRT program can make a significant difference in a child’s function, learning abilities, development, balance, and self-confidence.

References
Perilymph Fistula

WHAT IS A PERILYMPH FISTULA?

A perilymph fistula (PLF) is an abnormal connection (a tear or defect) in one or both of the small, thin membranes (the oval window and the round window) that separate the air filled middle ear and the fluid filled perilymphatic space of the inner ear. This small opening allows perilymph (fluid) to leak into the middle ear.

Changes in air pressure that occur in the middle ear (for example, when your ears “pop” in an airplane) normally do not affect your inner ear. However, when a fistula is present, changes in middle ear pressure will directly affect the inner ear, stimulating the balance and/or hearing structures within and causing PLF symptoms.

The perilymphatic space of the inner ear is connected to the cerebrospinal fluid (CSF) that surrounds the brain. Perilymphatic fluid, which is high in sodium (Na+), is similar in composition to CSF. When an abnormal connection between the membranes between the middle and inner ear exists, perilymph in the inner ear escapes, driven by the hydrostatic pressure of the CSF, and is replaced by CSF. This can also result in lower than normal levels of CSF fluid around the brain and spinal cord, which may result in symptoms such as mild headache.

Patients with PLF often feel frustrated and depressed because, while they don’t feel well, they look fine to others. PLF patients specifically and vestibular patients in general often have a challenging time explaining to friends and family what they are going through. Sometimes it is enough to ask your support network for patience and understanding while you explore diagnosis and treatment options and learn to cope with the symptoms brought on by persistent dizziness.
HISTORY
The small amount of fluid leaking from the middle ear to the inner ear is not detectable by the patient and is not generally visible to the surgeon who sets out to patch the leak. 40 years ago, when PLF first became an item of concern, the presumption was that there was a tear in the round window membrane or the ligamentous attachment of the footplate of the stapes to the edge of the oval window. A novel idea was put forth by Dr. Robert Kohut, based on post-mortem examination of temporal bones in patients who had suffered sudden hearing loss. His pioneering work indicated that the leak sites could be microfissures in the area just in front of the oval window or in the floor of the round window niche.

SYMPTOMS
The symptoms of a PLF most commonly include ear fullness, fluctuating or “sensitive” hearing, dizziness without true vertigo (spinning), and motion intolerance. Vertigo or sudden hearing loss can occur from a PLF. Most people with fistulas find that their symptoms get worse with changes in altitude (fast elevators, airplanes, and travel over mountain passes) or increased CSF pressure resulting from heavy lifting, bending over, and coughing or sneezing.

CAUSES
Head trauma is the most common cause of fistulas, usually involving a direct blow to the head or in some cases a "whiplash" injury. Other common causes include ear trauma, objects perforating the eardrum, or “ear block” on descent of an airplane or SCUBA diving. Fistulas may also develop after rapid increases in intracranial pressure, such as may occur with weightlifting or childbirth.

Fistulas are infrequently present from birth. A long-running controversy has surrounded the idea of a “spontaneous PLF.” Instead, what may occur is that a patient has a causative event but does not see an ear specialist right away. The passage of time blurs the memory of such an event so that the PLF might seem to have been spontaneous. Rarely, PLF’s occur in both ears, and only after a severe head injury.

Following is a video filmed by Dr. P. Ashley Wackym of Portland, Oregon's Ear & Skull Base Center, showing a patient who has suffered from a perilymph fistula. View this and more videos by Dr. Wackym on his YouTube channel.

DIAGNOSIS
There is no positive way to diagnosis a PLF. For many years it was thought that it could be confirmed by performing a tympanotomy (surgical exploration of the middle ear) and directly viewing the area of the suspected fistula to detect a fluid leak. However, since the leak would be only a few microliters of clear fluid, visual detection has been found to be virtually impossible. Larger amounts of fluid leakage may indicate a CSF leak due to a congenital defect in the inner ear.
A physician can arrive at a presumptive diagnosis through a thorough probing for events close in time to the onset of symptoms, along with a variety of tests. These tests can include hearing tests (audiogram, ECOG), balance tests (VNG, VEMP) and some form of a “fistula test.”

Historically, a platform pressure test developed by Dr. F. Owen Black was seen to be the most reliable test to determine if a PLF was present. However, this equipment is no longer in production, and only a small number still exist today.

In the end, a physician has to present the possibility of a PLF to the patient based on history, test results, and the lack of spontaneous resolution of symptoms. Together the physician and patient (or guardian) must decide whether to undertake an operation to patch the oval and round window areas. Immediately following surgery there is a period of bed rest, followed by a period of restricted activity. Four to six weeks later a reassessment of the patient’s symptoms is done to determine if the patching successfully corrected the PLF.

**TREATMENT**

When a traumatic event results in sudden onset of hearing loss or dizziness, the patient is advised to severely restrict physical activity for 7-14 days. If the symptoms do not improve or they plateau, testing is ordered. If the tests are compatible with the diagnosis of PLF, a surgical intervention may be considered. Persons with diagnosed fistulas who are awaiting surgery should avoid lifting, straining and bending over as these activities can cause a worsening of the symptoms.

A PLF repair involves an operation, often under general anesthesia, working through the ear canal. The eardrum is lifted up and minute soft tissue grafts are placed around the base of the stapes (stirrup) and in the round window niche. The operation usually takes about 45-60 minutes to complete. There is very little, if any, pain. Some patients are kept overnight to restrict activity. Once discharged the patient is advised to spend three days at home with limited activity. After three days the patient may return to sedentary work activities. The patient is advised to avoid lifting more than 10 lbs. for one month and avoid sporting activities. After one month there are additional restrictions suggested on activities such as contact sports, diving, weight lifting, and roller coasters. All of these activities have resulted in recurrent PLF’s after an initial successful repair.

**Secondary Endolymphatic Hydrops (SEH)**

**WHAT IS ENDOLYMPHATIC HYDROPS.**

Although its underlying cause and natural history are unknown, it is believed to result from abnormalities in the quantity, composition, and/or pressure of the endolymph (the fluid within the endolymphatic sac, a compartment of the inner ear).
CAUSES

Endolymphatic hydrops may be either primary or secondary. Primary idiopathic endolymphatic hydrops (known as Ménière’s disease) occurs for no known reason. Secondary endolymphatic hydrops appears to occur in response to an event or underlying condition. For example, it can follow head trauma or ear surgery, and it can occur with other inner ear disorders, allergies, or systemic disorders (such as diabetes or autoimmune disorders).

In a normal inner ear, the endolymph is maintained at a constant volume and with specific concentrations of sodium, potassium, chloride, and other electrolytes. This fluid bathes the sensory cells of the inner ear and allows them to function normally. In an inner ear affected by hydrops, these fluid-system controls are believed to be lost or damaged. This may cause the volume and concentration of the endolymph to fluctuate in response to changes in the body’s circulatory fluids and electrolytes.

SYMPTOMS

Symptoms typical of hydrops include pressure or fullness in the ears (aural fullness), tinnitus (ringing or other noise in the ears), hearing loss, dizziness, and imbalance.

DIAGNOSIS AND TESTING

There is no vestibular or auditory test that is diagnostic of endolymphatic hydrops. Diagnosis is clinical—based on the physician’s observations and on the patient’s history, symptoms, and symptom pattern. The clinical diagnosis may be strengthened by the results of certain tests. For example, certain abnormalities in electrocchleography (which tests the response of the eighth cranial nerve to clicks or tones presented to the ear) or audiometry (which tests hearing function) may support a hydrops diagnosis.

TREATMENT GOALS

Ménière’s disease (primary idiopathic endolymphatic hydrops) is discussed in detail in a separate publication of the Vestibular Disorders Association. In brief, Ménière’s disease is characterized by sudden, violent attacks or episodes of vertigo, tinnitus, hearing loss, and aural fullness. The attacks typically occur at intervals of weeks to months, with symptom-free periods between attacks. Over several years, there is partial destruction of hearing and sometimes of balance function. Ménière’s disease affects both ears in up to half of those who experience it.

Treatment is intended to improve symptoms, manage acute attacks, deal with the damage to hearing and balance, and maintain quality of life.

The treatment of secondary endolymphatic hydrops (SEH) is somewhat different. Since SEH is secondary to (that is, results from) an underlying disorder, the symptoms tend to be present more continuously, rather than occur in spontaneous attacks. However, they are often less violent, and SEH may cause less damage to hearing and balance than does Ménière’s disease.
Treatment of SEH has five goals: to stabilize the body's fluid and electrolyte levels; to identify and treat the underlying condition that is driving the SEH; to improve daily symptoms; to manage persistent symptoms and changes; and to maintain quality of life.

The five goals of treating secondary endolymphatic hydrops:

1. Stabilize the body’s fluid and electrolyte levels.
2. Identify and treat the underlying conditions.
3. Improve daily symptoms.
4. Manage persistent symptoms and changes.
5. Maintain quality of life.

GOAL 1: STABILIZING THE BODY’S FLUID AND ELECTROLYTE LEVELS

Fluctuations in body fluids and electrolyte levels may affect the amount and composition of the endolymph, leading to hydrops symptoms. Stabilizing the fluid and electrolyte levels may help reduce or relieve the symptoms. Modifications in diet may be necessary in order to achieve such stabilization.

A hydrops diet regimen (HDR) often makes many people with SEH feel significantly better without any other treatment. The HDR is the cornerstone of stabilizing overall fluid levels. The most important aspect of this regimen is constancy. Eating a balanced diet in moderate amounts at regular intervals—with meals and snacks of a consistent size, eaten at about the same time every day, without skipping meals or alternating tiny snacks with huge meals—helps the body’s fluid and electrolyte levels remain stable.

A second key element of the HDR is minimizing the use of solutes (salts and sugars) in the diet. Solutes require the body to use large amounts of fluid for dilution and digestion. This causes large fluctuations in body fluids and consequently in the endolymph, which can trigger hydrops symptoms. An important starting point of this diet is to avoid adding salt or sugar to food and to avoid eating prepared foods (those that come out of a can or other container).

Adequate fluid intake is another mainstay of the HDR. In order to function at its best, the body needs lots of water—six to eight glasses spaced evenly throughout the day. It is also important to anticipate and replace the additional fluid lost through perspiration during exercise, fever, or hot weather. Other fluids—for example, low-sugar sodas, herbal teas, and low-sugar fruit and vegetable juices—may also make up part of the daily allotment. Caffeine (found in coffee, tea, some herbal teas, colas, chocolate, and some medications) and alcohol have strong diuretic properties and may need to be restricted, because they can cause the body to lose more fluid than it has taken in.

Physicians may prescribe diuretics as part of treatment—not to cause fluid loss, but rather to “push” the kidneys to excrete a constant amount of urine throughout the day, thus helping to minimize large swings in the body’s fluid.
content. With diuretic use, drinking lots of water is important, in order to avoid dehydration. Certain diuretics require the use of a potassium supplement to replace potassium lost through the urine.

**GOAL 2: IDENTIFYING AND TREATING THE UNDERLYING CONDITION**

This goal is complex and will likely involve both the otologist (ear specialist) and the primary care provider (internist, family practice physician, nurse practitioner, etc.). Once an underlying condition is identified and treated, SEH symptoms tend to improve over time. Hydrops associated with head trauma or ear surgery usually improves over the course of one to two years following the causative event.

**GOAL 3: IMPROVING DAILY SYMPTOMS**

With the use of the HDR and possibly also a diuretic, balance symptoms may improve dramatically. Other medications may be used to help with persistent dizziness, nausea, or vomiting.

Other strategies to reduce the symptoms of SEH include:

- Maintaining normal weight, or losing any excess weight. When a person is overweight, the vestibular system must struggle to deal with a larger-than-normal and displaced center of gravity.
- Avoiding aspirin in high doses, which can cause temporary tinnitus.
- Avoiding ibuprofen and other NSAIDS (nonsteroidal anti-inflammatory drugs), which can have a direct effect on fluid balance and may increase symptoms in some SEH patients.
- Stopping smoking. Smoking constricts the single, tiny artery that feeds the ear, thus depriving it of oxygen and hemoglobin.
- Maintaining general health by getting adequate exercise and sleep.

Vestibular rehabilitation, a type of specialized physical therapy for vestibular patients, can improve tolerance for activity, overall energy level, and symptoms of dizziness and imbalance. The cognitive symptoms that often accompany vestibular disorders—for instance, trouble with concentration, short-term memory, reading, or prioritizing tasks—may diminish as the hydrops is brought under control.

**GOAL 4: MANAGING PERSISTENT SYMPTOMS AND CHANGES**

If dizziness and vertigo become intractable, more aggressive measures may be considered. For example, the hair-cell structures of the inner ear may be selectively destroyed with the careful use of ototoxic (ear-poisoning) medication.
In rare cases, surgery may be recommended. Endolymphatic decompression procedures aim at relieving fluid pressure in the inner ear. Another type of surgery is a labyrinthectomy, which destroys the membranous structures of the inner ear that detect gravity and motion changes. Semi-circular canal plugging and neurectomy (cutting the vestibular nerve between the ear and the brain) create mechanical changes that prevent abnormal inner ear signals from reaching the brain, thus reducing symptoms. These procedures do not cure the underlying disorder and are not without risk, but they may improve symptoms in some cases.

SEH does not usually result in significant hearing loss. If it does occur, modern hearing aids and other assistive devices may be useful. In addition, tinnitus-masking devices can be used to help deal with annoying tinnitus.

Very often, people with inner ear disorders attempt to avoid aggravating their symptoms by restricting their activity and becoming reclusive. This is counterproductive. In fact, remaining as active and busy as possible (within safe limits) helps the brain adjust to changes in inner ear function and helps control symptoms. The physician may recommend a course of vestibular rehabilitation therapy to help the brain compensate for changes in balance function.

**GOAL 5: MAINTAINING QUALITY OF LIFE**

As with any chronic disorder, maintaining a healthy outlook and as normal a routine as possible is essential. Creating a safe physical environment in the home is also important, as well as taking into consideration whether one should undertake potentially hazardous activities such as driving a car or climbing ladders.

As an “invisible” disability, an inner ear disorder can be frustrating to manage. A person may feel miserable, yet “look normal” to friends and family. Educating others about the illness can help them better understand the difficulties and consequences of having a vestibular disorder. Counseling or participating in a support group for people with inner ear disorders may help to deal with the confusion or secondary depression that often accompanies these conditions. In any case, the physician and the physician’s staff remain the patient’s primary resource in understanding and dealing with SEH.
Spontaneous????

SEEMS LIKE MAGIC BUT IT IS JUST THE EDUCTOR
Superior Semicircular Canal Dehiscence (SSCD)

WHAT IS SUPERIOR SEMICIRCULAR CANAL DEHISCENCE?

Vestibular and auditory symptoms and signs can result from a dehiscence (opening) in the bone overlying the superior semicircular canal of the inner ear. This clinical syndrome—superior semicircular canal dehiscence syndrome (SSCD)—was first described by Minor and colleagues in 1998. Patients with SSCD can experience vertigo and oscillopsia (the apparent motion of objects that are known to be stationary) evoked by loud noises and/or by maneuvers that change middle-ear or intracranial pressure (such as coughing, sneezing, or straining). Auditory manifestations of the syndrome include autophony (increased resonance of one’s own voice), hypersensitivity to bone-conducted sounds, and an apparent conductive hearing loss revealed on audiometry. Some patients have exclusively vestibular symptoms and signs; some have both auditory and vestibular manifestations; and still other patients have exclusively auditory complaints.

Following is a video filmed by Dr. P. Ashley Wackym of Portland, Oregon's Ear & Skull Base Center, showing a patient who has suffered from superior semicircular canal dehiscence. View this and more videos by Dr. Wackym on his YouTube channel.

CAUSES

With a dehiscence in the bone that is supposed to cover the superior semicircular canal, the fluid in the membranous superior semicircular canal (which is located within the lumen—tubular cavity—of the bony canal) can be displaced by sound and pressure stimuli. There are normally only two points of increased compliance (yielding to pressure) in the inner ear: the oval window, through which sound energy is transmitted into the inner ear via the stapes bone; and the round window, through which sound energy is dissipated from the inner ear after traveling around the cochlea. SSCD creates a third mobile window into the inner ear. The signs and symptoms in this syndrome are due to the physiological consequences of this third window.
The mean age at the time of diagnosis is around 45 years. Unilateral SSCD occurs relatively equally in the right and left ears. About one-third of patients have evidence of bilateral SSCD at the time of diagnosis. In patients with bilateral dehiscence, there is typically one ear from which the symptoms and signs are greater. In patients with unilateral dehiscence, the bone overlying the contralateral superior canal (in the opposite ear) is often abnormally thin.

These findings support the notion that SSCD is due to a developmental abnormality. Temporal-bone histopathological studies suggest that 1–2% of the population have abnormally thin bone overlying the superior canal (Carey et al. 2000). Disruption of this thin layer (as may perhaps occur with trauma or over time due to the pressure of the overlying temporal lobe of the brain) leads to the onset of symptoms and signs.

**DIAGNOSIS**

**Vestibular symptoms and signs:** The vestibular symptoms in SSCD can be debilitating and often provoke patients to seek medical attention. Patients may note that loud noises cause them to see things moving or that they experience a similar sensation when they cough, sneeze, or strain to lift something heavy. They may perceive that objects are moving in time with their pulse (pulsatile oscillopsia). Some individuals can bring on the sensation of motion—and cause their eyes to move—by pressing on their tragus (the area of skin and cartilage located just outside the ear canal). Patients may experience a feeling of constant disequilibrium and imbalance.

The signs of vestibular abnormalities in SSCD relate directly to the effect of the dehiscence in creating a third mobile window into the inner ear. One of the most important functions of the vestibular system is to keep the eyes focused on objects of interest during head movements. A principle underlying the organization of these vestibulo-ocular reflexes is that the eyes move in the plane of the semicircular canal that is being activated.
Analysis of the eye movements evoked by sound and pressure stimuli in patients with SSCD led Minor and colleagues (1998) to the identification of this syndrome. These evoked eye movements often align with the plane of the superior canal. Furthermore, the direction of the eye movements provides support for the theory of a third-mobile-window mechanism. Stimuli that result in inward motion of the stapes footplate (such as loud sounds, applying pressure to the external ear canal, or blowing pressure through the nose while pinching the nostrils) produce an excitation of the superior canal. Evoked eye movements in these situations are typically vertical-torsional, with the eyes moving up and the superior pole of each eye moving away from the SSCD ear. Stimuli that result in outward motion of the stapes footplate (such as negative pressure applied to the external ear canal) or that increase intracranial pressure (such as taking a deep breath and bearing down, or compressing the jugular vein with pressure on the neck) typically result in eye movements that are in the same plane but opposite in direction. The eyes move down, and the superior pole of each eye moves toward the SSCD ear.

**Auditory symptoms and signs:** The auditory symptoms and signs in SSCD may mimic those in other ear disorders and may at times seem bizarre. Some patients have a conductive hearing loss for low-frequency sounds that can resemble the pattern in otosclerosis. These diagnostic entities can be distinguished by acoustic reflex testing: patients with otosclerosis lose the acoustic reflex response in the affected ear early in the course of the disorder, whereas the response remains intact in SSCD. This distinction is important because patients who have conductive hearing loss resulting from SSCD will not benefit from stapedectomy surgery. Patients with SSCD may also complain of symptoms such as hearing their eye movements, hearing their own voice too loudly in the affected ear (autophony), or having a distorted sensation of sound in the affected ear during activities such as running. These auditory symptoms and signs are also manifestations of the third mobile window created by the dehiscence. Bone-conducted sounds are amplified by the effects of the dehiscence, whereas the energy from air-conducted sounds is partially shunted away from the cochlea and through the dehiscence.

**CT imaging:** High-resolution CT scans of the temporal bones are very useful in making the diagnosis of SSCD. These scans demonstrate the opening in the bone that should cover the superior canal. Care must be exercised, however, because such scans may miss a thin layer of intact bone overlying the canal. Applying specific parameters for the CT imaging can improve the specificity of the scans (Belden et al. 2003), but false positives can still occur, even with the highest-resolution scans. Individuals who are suspected of having SSCD are strongly urged to have their CT scans performed at a center experienced in the diagnosis of SSCD. It must be emphasized that even with these scans, the diagnosis depends upon characteristic clinical findings and other physiologic tests.

**Vestibular evoked myogenic potentials (VEMP):** In SSCD, loud tones evoke a short-latency relaxation potential in the ipsilateral sternocleidomastoid muscle. Patients with SSCD typically have a lower-than-normal threshold for the VEMP response, and the amplitude of the VEMP waveform in an SSCD ear is greater for comparable stimulus intensities than in an ear without dehiscence. The VEMP test plays an important role in the evaluation of patients with suspected SSCD (Zhou et al. 2007).

**TREATMENT**

Many patients with SSCD are able to tolerate their symptoms and reduce the more severe effects by avoiding the stimuli that make the symptoms worse, such as loud noises. For other patients, the symptoms are much more
debilitating. Pulsatile oscillopsia, chronic disequilibrium, and autophony are some of the symptoms for which avoidance of stimuli is unlikely to be helpful.

Surgical correction: For patients whose well-being is severely affected by SSCD, surgical repair of the dehiscence can be very beneficial. The middle cranial fossa approach has been used most commonly. Plugging of the canal with fascia (fibrous tissue), using small bone chips to secure the fascia in place, has been shown to be more effective than canal resurfacing in achieving long-term control of symptoms. The main risk of this procedure is hearing loss in the affected ear, although this risk is low in patients who have not undergone prior SSCD surgery or prior stapedectomy. The procedure is very effective in relieving both the vestibular symptoms and the autophony associated with SSCD. Plugging of the superior canal typically results in decreased function in this canal alone, while preserving function in the other semicircular canals (Carey et al. 2007). The reduction of function in the superior canal has minimal negative functional consequences for the patient. In patients with bilateral SSCD, surgery on the more severely affected ear may be sufficient to control their symptoms.

CONCLUSIONS

Dehiscence of bone overlying the superior semicircular canal can cause a constellation of vestibular and auditory symptoms and signs. These abnormalities can be understood in terms of the effect of the dehiscence in creating a third mobile window into the inner ear. The diagnosis is made based upon characteristic symptoms, specific findings on clinical examination, CT imaging, and findings on VEMP testing. The diagnosis should never be made exclusively on the basis of CT findings.

For many patients, an understanding of the cause of the symptoms and avoidance of provocative stimuli such as loud noises may be sufficient. For those patients who are debilitated by their symptoms, surgical plugging of the superior canal can be very beneficial in alleviating or substantially reducing the symptoms and signs.

References
Tinnitus

WHAT IS THAT RINGING IN MY EARS?

Tinnitus is abnormal noise perceived in one or both ears or in the head. Tinnitus (pronounced either “TIN-uh-tus” or “tin-NY-tus”) may be intermittent, or it might appear as a constant or continuous sound. It can be experienced as a ringing, hissing, whistling, buzzing, or clicking sound and can vary in pitch from a low roar to a high squeal.

Tinnitus is very common. Most studies indicate the prevalence in adults as falling within the range of 10% to 15%, with a greater prevalence at higher ages, through the sixth or seventh decade of life. Gender distinctions are not consistently reported across studies, but tinnitus prevalence is significantly higher in pregnant than non-pregnant women.

The most common form of tinnitus is subjective tinnitus, which is noise that other people cannot hear. Objective tinnitus can be heard by an examiner positioned close to the ear. This is a rare form of tinnitus, occurring in less than 1% of cases.

Chronic tinnitus can be annoying, intrusive, and in some cases devastating to a person’s life. Up to 25% of those with chronic tinnitus find it severe enough to seek treatment. It can interfere with a person’s ability to hear, work, and perform daily activities. One study showed that 33% of persons being treated for tinnitus reported that it disrupted their sleep, with a greater degree of disruption directly related to the perceived loudness or severity of the tinnitus.

CAUSES AND RELATED FACTORS

Most tinnitus is associated with damage to the auditory (hearing) system, although it can also be associated with other events or factors: jaw, head, or neck injury; exposure to certain drugs; nerve damage; or vascular (blood-flow) problems. With severe tinnitus in adults, coexisting factors may include hearing loss, dizziness, head injury, sinus and middle-ear infections, or mastoiditis (infection of the spaces within the mastoid bone). Significant factors associated with mild tinnitus may include meningitis (inflammation of the membranous covering of the brain and spinal cord), dizziness, migraine, hearing loss, or age.

Forty percent of tinnitus patients have decreased sound tolerance, identified as the sum of hyperacusis (perception of over-amplification of environmental sounds) and misophonia/phonophobia (dislike/fear of environmental sounds). While most cases of tinnitus are associated with some form of hearing impairment, up to 18% of cases do not involve reports of abnormal hearing.

EAR DISORDERS

Hearing loss from exposure to loud noise: Acute hearing depends on the microscopic endings of the hearing nerve in the inner ear. Exposure to loud noise can injure these nerve endings and result in hearing loss. Hearing damage from noise exposure is considered to be the leading cause of tinnitus.
Presbycusis: Tinnitus can also be related to the general impairment of the hearing nerve that occurs with aging, known as presbycusis. Age-related degeneration of the inner ear occurs in 30% of persons age 65–74, and in 50% of persons 75 years or older.10

Middle-ear problems: Tinnitus is reported in 65% of persons who have preoperative otosclerosis (stiffening of the middle-ear bones),11 with the tinnitus sound typically occurring as a high-pitched tone or white noise rather than as a low tone.12 Otitis media (middle-ear infection) can be accompanied by tinnitus, which usually disappears when the infection is treated. If repeated infections cause a cholesteatoma (benign mass of skin cells in the middle ear behind the eardrum), hearing loss, tinnitus, and other symptoms can result.13 Objective tinnitus has been associated with myoclonus (contraction or twitching) of the small muscles in the middle ear.14,15 Conductive hearing loss resulting from an accumulation of earwax in the ear canal can sometimes cause tinnitus.

Vestibular disorders: Hearing impairment and related tinnitus often accompany dysfunction of the balance organs (vestibular system). Some vestibular disorders associated with tinnitus include Ménière’s disease and secondary endolymphatic hydrops (resulting from abnormal amounts of a fluid called endolymph collecting in the inner ear) and perilymph fistula (a tear or defect in one or both of the thin membranes between the middle and inner ear).

VESTIBULO-COCHLEAR NERVE DAMAGE AND CENTRAL AUDITORY SYSTEM CHANGES

The vestibulo-cochlear nerve, or eighth cranial nerve, carries signals from the inner ear to the brain. Tinnitus can result from damage to this nerve. Such damage can be caused by an acoustic neuroma, also known as a vestibular schwannoma (benign tumor on the vestibular portion of the nerve), vestibular neuritis (viral infection of the nerve), or microvascular compression syndrome (irritation of the nerve by a blood vessel).

The perception of chronic tinnitus has also been associated with hyperactivity in the central auditory system, especially in the auditory cortex.16 In such cases, the tinnitus is thought to be triggered by damage to the cochlea (the peripheral hearing structure) or the vestibulo-cochlear nerve.

HEAD AND NECK TRAUMA

Compared with tinnitus from other causes, tinnitus due to head or neck trauma tends to be perceived as louder and more severe. It is accompanied by more frequent headaches, greater difficulties with concentration and memory, and a greater likelihood of depression.17

Somatic tinnitus is the term used when the tinnitus is associated with head, neck, or dental injury—such as misalignment of the jaw or temporomandibular joint (TMJ)—and occurs in the absence of hearing loss.

Characteristics of somatic tinnitus include intermittency, large fluctuations in loudness, and variation in the perceived location and pattern of its occurrence throughout the day.18

MEDICATIONS

Many drugs can cause or increase tinnitus. These include certain non-steroidal anti-inflammatory drugs (NSAIDs, such as Motrin, Advil, and Aleve), certain antibiotics (such as gentamicin and vancomycin), loop diuretics (such as Lasix), aspirin and other salicylates, quinine-containing drugs, and chemotherapy medications (such as carboplatin and cisplatin). Depending on the medication dosage, the tinnitus can be temporary or permanent.3

VASCULAR SOURCES
**Pulsatile tinnitus** is a rhythmic pulsing sound that sometimes occurs in time with the heartbeat. This is typically a result of noise from blood vessels close to the inner ear. Pulsatile tinnitus is usually not serious. However, sometimes it is associated with serious conditions such as high or low blood pressure, hardening of the arteries (arteriosclerosis), anemia, vascular tumor, or aneurysm.

**OTHER POSSIBLE CAUSES**

Other conditions have been linked to tinnitus: high stress levels, the onset of a sinus infection or cold, autoimmune disorders (such as rheumatoid arthritis or lupus), hormonal changes, diabetes, fibromyalgia, Lyme disease, allergies, depletion of cerebrospinal fluid, vitamin deficiency, and exposure to lead. In addition, excessive amounts of alcohol or caffeine exacerbate tinnitus in some people.

**DIAGNOSIS**

Examination by a primary care physician will help rule out certain sources of tinnitus, such as blood pressure or medication problems. This doctor can also, if necessary, provide a referral to an ear, nose, and throat specialist (an otolaryngologist, otologist, or neurotologist), who will examine the ears and hearing, in consultation with an audiologist. Their evaluations might involve extensive testing that can include an audiogram (to measure hearing), a tympanogram (to measure the stiffness of the eardrum and help detect the presence of fluid in the middle ear), otoacoustic emissions testing (to provide information about how the hair cells of the cochlea are working), an auditory brainstem response test (to measure how hearing signals travel from the ear to the brain and then within parts of the brain), electrocochleography (to measure how sound signals move from the ear along the beginning of the hearing nerve), vestibular-evoked myogenic potentials (to test the functioning of the saccule and/or inferior vestibular nerve), blood tests, and magnetic resonance imaging (MRI). Neuropsychological testing is also sometimes included to screen for the presence of anxiety, depression, or obsessions—which are understandable and not uncommon effects when tinnitus has disrupted a person’s life.

**TREATMENT**

If a specific cause of the tinnitus is identified, treatment may be available to relieve it. For example, if TMJ dysfunction is the cause, a dentist may be able to relieve symptoms by realigning the jaw or adjusting the bite with dental work. If an infection is the cause, successful treatment of the infection may reduce or eliminate the tinnitus.

Many cases of tinnitus have no identifiable cause, however, and thus are more difficult to treat. Although a person’s tolerance of tinnitus tends to increase with time, severe cases can be disturbing for many years. In such chronic cases, a variety of treatment approaches are available, including medication, dietary adjustments, counseling, and devices that help mask the sound or desensitize a person to it. Not every treatment works for every person.

**MASKING DEVICES**

A masking device emits sound that obscures, though does not eliminate, the tinnitus noise. The usefulness of maskers is based on the observation that tinnitus is usually more bothersome in quiet surroundings and that a
competing sound at a constant low level, such as a ticking clock, whirring fan, ocean surf, radio static, or white noise produced by a commercially available masker, may disguise or reduce the sound of tinnitus, thus making it less noticeable. Some tinnitus sufferers report that they sleep better when they use a masker. In some users, maskers produce residual inhibition—tinnitus suppression that lasts for a short while after the masker has been turned off. Hearing aids are sometimes used as maskers. If hearing loss is involved, properly fitted hearing aids can improve hearing and may reduce tinnitus temporarily. However, tinnitus can actually worsen if the hearing aid is set at an excessively loud level.

Cochlear implants, used for persons who are profoundly deaf or severely hard-of-hearing, have been shown to suppress tinnitus in up to 92% of patients. This is likely a result of masking due to newly perceived ambient sounds or from electrical stimulation of the auditory nerve. Other devices under development may eventually prove effective in relieving tinnitus. For example, the recently introduced acoustics-based Neuromonics device involves working with an audiologist who matches the frequency spectrum of the perceived tinnitus sound to music that overlaps this spectrum. This technique aims to stimulate a wide range of auditory pathways, the limbic system (a network of structures in the brain involved in memory and emotions), and the autonomic nervous system such that a person is desensitized to the tinnitus. Assessing the true effectiveness of this device will require further scientific study, although observations from an initial stage of clinical trials indicate that the device can reduce the severity of symptoms and improve quality of life.

**TINNITUS RETRAINING THERAPY**

Tinnitus retraining therapy (TRT) is designed to help a person retrain the brain to avoid thinking about the tinnitus. It employs a combination of counseling and a non-masking sound that decreases the contrast between the sound of the tinnitus and the surrounding environment. The goal is not to eliminate the perception of the tinnitus sound itself, but to retrain a person’s conditioned negative response (annoyance, fear) to it. In one comparison of the effectiveness of tinnitus masking and TRT as treatments, masking was found to provide the greatest benefit in the short term (three to six months), while TRT provided the greatest improvement with continued treatment over time (12–18 months).

**PSYCHOLOGICAL TREATMENTS**

Chronic tinnitus can disrupt concentration, sleep patterns, and participation in social activities, leading to depression and anxiety. In addition, tinnitus tends to be more persistent and distressful if a person obsesses about it. Consulting with a psychologist or psychiatrist can be useful when the emotional reaction to the perception of tinnitus becomes as troublesome as the tinnitus itself and when help is needed in identifying and altering negative behaviors and thought patterns.

**MEDICATION**

No drug is available to cure tinnitus; however, some drugs have been shown to be effective in treating its psychological effects. These include anti-anxiety medications in the benzodiazepine family, such as clonazepam (Klonopin) or lorazepam (Ativan); antidepressants in the tricyclic family, such as amitriptyline (Elavil) and
nortriptyline (Aventyl, Nortrilen, Pameler); and some selective serotonin reuptake inhibitors (SSRIs), such as fluoxetine (Prozac).  

Other drugs have been anecdotally associated with relief of tinnitus. These include certain heart medications, anesthetics, antihistamines, statins, vitamin or mineral supplements, vasodilators, anticonvulsants, and various homeopathic or herbal preparations. Scientific evidence is lacking to support the effectiveness of many of these remedies. Some appear to be placebos, while some are possibly mildly or temporarily effective but with potential side effects that are serious.

Examples of recent research studies on some of these anecdotal treatments follow, although this list is not exhaustive:

- In assessing the effectiveness of atorvastatin (Lipitor) in the treatment of tinnitus, scientists observed a trend toward relief of symptoms; however, this trend was not statistically significant when compared with results produced by administration of a placebo.

- The relationship between low blood zinc levels and subjective tinnitus was inspected in a small placebo-controlled study. Administration of oral zinc medication produced results that prompted the researchers to note that additional tests were needed to investigate whether duration of treatment might be a significant factor.

- Immediate suppression of subjective tinnitus has been observed in patients administered intravenous lidocaine, although such relief has been shown to be very short term. The effect of such tinnitus treatment is thought to occur in the central auditory pathway rather than in the cochlea.

- Scientists demonstrated that the anticonvulsant gabapentin (Neurontin) is no more effective than placebo in treatment of tinnitus.

- When scientists reported their finding that Ginkgo biloba extracts and placebo treatments produce very similar results, they also noted that use of the extract could lead to adverse side effects, especially if used unsupervised and with other medications.

Some alternative approaches may eventually yield helpful options in tinnitus treatment. However, most scientists agree that additional well-constructed research is needed before any anecdotally associated preparation can be applied as a proven and effective treatment option.

**SURGERY**

Treating tinnitus with surgery is generally limited to being a possible secondary outcome of surgery that is used in cases when the source of the tinnitus is identified (such as acoustic neuroma, perilymph fistula, or otosclerosis) and surgical intervention is required to treat that condition.
OTHER PROPOSED TREATMENTS

Stress-reduction techniques are often advocated for improving general health, as they can help control muscle groups and improve circulation throughout the body. Such relaxation training, the use of biofeedback to augment relaxation exercises, and hypnosis have been suggested as treatments for tinnitus. Limited research is available on the effectiveness of these methods.

Acupuncture, electrical stimulation, application of magnets, electromagnetic stimulation, and ultrasound have been found to be placebo treatments for tinnitus or to have limited scientific support for their effectiveness. Recent and ongoing research studies have attempted to assess whether transcranial magnetic stimulation could be an effective tinnitus treatment. This application is based on the thought that tinnitus is associated with an irregular activation of the temporoparietal cortex (a part of the brain), and thus that disturbing this irregular activation could result in transient reduction of tinnitus.

PREVENTION

Precautionary measures to help lessen the severity of tinnitus or help a person cope with tinnitus are related to some of the causes and treatments listed above. Avoiding exposure to loud sounds (especially work-related noise) and getting prompt treatment for ear infections have been identified as the two most important interventions for reducing the risk of tinnitus. Wearing ear protection against loud noise at work or at home and avoiding listening to music at high volume can both help reduce risk.

Other important factors are exercising daily, getting adequate rest, and having blood pressure monitored and controlled, if needed. Additional precautionary measures include limiting salt intake, avoiding stimulants such as caffeine and nicotine, and avoiding ototoxic drugs known to increase tinnitus (some of which are listed above under “Causes and Related Factors”).

SUMMARY

Tinnitus is a common condition that can disrupt a person’s life. Our understanding of the mechanisms of tinnitus is incomplete, and many unknown factors remain. These limitations contribute to the lack of medical consensus about tinnitus management, stimulate continued research efforts, and motivate anecdotal and commercially based speculation about potential but unproven treatments. Prior to receiving any treatment for tinnitus or head noise, it is important for a person to have a thorough examination that includes an evaluation by a physician. Understanding the tinnitus and its possible causes is an essential part of its treatment.

References


Vestibular Hyperacusis

ARE YOU SENSITIVE TO CERTAIN SOUNDS?

Hyperacusis is the perception of an unusual auditory sensitivity to some environmental noises or tones. The particular symptoms of cochlear hyperacusis and vestibular hyperacusis can help physicians and audiologists distinguish between the two disorders. The effects of hyperacusis can range from a mild sense of unease to a complete loss of balance or upright posture with severe ear pain. In serious cases, it can cause seizure-like activity in the brain.

Hyperacusis can be associated with auto-immune disorders, traumatic brain injury, metabolic disorders, and other conditions. It has not been sufficiently studied in the adult population and is often ascribed to psychological conditions rather than being recognized as a physiologic symptom of cochlear or vestibular damage.

The hearing and balance systems of the inner ear are interconnected. Both systems are filled with fluid whose movement stimulates tiny sensory cells. Sounds are detected as energy vibrations; the human cochlea can hear best the frequencies associated with speech. The balance system uses lower-frequency sensations to help maintain posture in relation to gravity.

Hyperacusis is an abnormal condition in which the complex electrical signals generated by sound vibrations are misinterpreted, confused, or exaggerated. The signals coming in are identical to those that present to a normal ear, but the reaction in the abnormal system is markedly different: for example, the sounds in a quiet library may seem like a loud parade to a person with hyperacusis.

COCHLEAR VS. VESTIBULAR HYPERACUSIS
With cochlear hyperacusis, subjects feel ear pain, discomfort, annoyance, and irritation when certain sounds are heard, including those that are very soft or high-pitched. Most people react by covering their ears or leaving the room. Severe emotional reactions may also occur; crying or panic reactions are not uncommon.

In vestibular hyperacusis, exposure to sound can result in falling or a loss of balance or postural control. Such disturbances have been called by various names, including Tullio’s syndrome and audiogenic seizure disorder. Some of the same reactions as with cochlear hyperacusis can also occur, along with sudden severe vertigo or nausea. In some cases, vestibular hyperacusis can affect the autonomic system and cause problems such as loss of consciousness, mental confusion, nausea, or extreme fatigue.

In both cochlear and vestibular hyperacusis, headache is common. In addition, many subjects with hyperacusis feel distinct cognitive changes during these exposures and will describe themselves as being “out of myself” or disassociated from reality, unable to take in other stimuli, having an immediate feeling of something being wrong or a sensation of being unwell, or experiencing severe confusion.

WHAT CAUSES HYPERACUSIS?

The physiologic conditions underlying these symptoms cannot be identified with certainty because of difficulties involved with studying the very small inner ear structures without damaging them.

A suspected cause of cochlear hyperacusis involves a loss of the regulatory function provided by the system that conducts impulses along the auditory neural pathways. In hyperacusis, the mechanism that regulates amplification erroneously magnifies the incoming sounds and noises instead of reducing them. For example, the sound of a passing car is interpreted as comparable to the roar of a jet engine!

Other possible explanations of cochlear hyperacusis involve brain-chemistry dysfunction or head trauma that damages the chain of tiny bones in the middle ear that amplify sound and help transmit vibrations to the inner ear fluid. Changes in the transmission of electrical signals along complex neural pathways are also highly possible in cases of head injury.

In vestibular hyperacusis, we suspect that the main pathology results from damage to the nerve cells in the balance system. These cells may suffer damage from trauma such as head injury, metabolic disruptions due to chemical ingestions (e.g., medications or anesthesia), or circulatory changes due to heart disease or artery blockages. In addition, autoimmune disease, which can be triggered by many different causes, can harm the balance organ. Head trauma in a motor vehicle accident can set off an autoimmune reaction in the inner ear that can destroy the nerve cells, often weeks or months after the initial injury.

In one clinic, several serious cases were evaluated where simple soft auditory stimulations of less than 30 decibels (comparable to a mid-pitch musical note played at a very soft level) elicited loss of consciousness and seizures. All of these patients had suffered head and/or neck injuries in motor vehicle accidents that affected the brain stem and higher areas of the central nervous system. None of these patients had significant hearing loss or previous balance
problems. One person loses balance and consciousness frequently and must use earplugs and earmuffs all of the time to avoid injury from falling.

**TESTING AND TREATMENT INNOVATIONS**

Special audiologic tests can reveal the presence and severity of cochlear hyperacusis. Simple tests such as the Loudness Discomfort Level test (promoted for use in hyperacusis assessment by Drs. Pawel Jastreboff and Jonathan Hazell) and balance screening using an audiometer and observation take only a few moments and can yield significant information.

Cochlear hyperacusis can be treated with acoustic therapies such as tinnitus retraining therapy (TRT). The Jastreboff TRT method is the treatment of choice and can result in recovery of normal or near-normal dynamic ranges of sound tolerance.

Vestibular hyperacusis, however, continues to go untreated or unrecognized in many cases. When vestibular hyperacusis is recognized, the treatment protocols vary widely, depending on the level of expertise and interest of the treating physician. Treatment with a low-salt diet combined with anti-nausea drugs still dominates medical approaches, although there are some pioneers—such as John Epley, MD (Portland Otologic Clinic, Portland, Oregon)—who have had promising results introducing anti-inflammatory medicines directly into the cochlear/vestibular system using catheters.

For individuals who complain of loss of balance with exposure to sound, thorough diagnostic testing should be completed in otology, neurology, and audiology offices. Innovative testing protocols could be devised to provoke or produce the response in a clinical setting. In the clinic, presenting a tone at 500 Hz and gradually increasing the loudness can often induce vestibular hyperacusis.

It is important that clinicians present tests tailored to the individual patient’s situation. For example, if someone complains of falling when large vehicles pass by, identifying the specific problem area may require changing a test to include lower-frequency tones at very low volume levels, or narrow-band noise, or even white noise. A portable audiometer might be used in conjunction with a computerized dynamic posturography test so that various sounds can be presented to induce a balance response. Another possibility is to utilize electroencephalography (EEG) with an audiometer to present sound stimulation, so that shifts in brain-wave patterns in response to sound can be observed. We used this strategy in our clinic recently to produce clear evidence of brain-wave anomalies, providing proof to a patient that the source of her troubling symptoms was organic. Her constant falling and loss of consciousness were based on a physiologic condition, not a psychological one. These results provided a sense of relief to the patient, whose previous EEG results, without sound stimulation, had been normal.

Adapting clinical assessment tools with the use of various stimuli and then making careful observations may allow medical providers to identify patients with vestibular hyperacusis and to devise better therapeutic strategies.
"If you’re interested in ‘balancing’ work and pleasure, stop trying to balance them. Instead, make your work more pleasurable.” Donald Trump

I never imagined I would quite Donald Trump, but then again, the Donald is having a good time doing what he does best and making millions doing it. He has it right here—that feeling pleasure is key.

Balance—some imagined ideal state of perfect equilibrium—is just not what it is about. Many aspire to it, some say it is impossible, and most don’t even care.

I committed to a blog challenge 3 days ago and I missed yesterday. I was committed from 6:30 am until an unforeseen adventure in the evening. There was no way….or rather…..I chose fun over the writing of the blog.

Some would say my adventure was worth not getting all the work done I had planned. Others would say I got off track.

I say it was all an experiment that turned out just great. And I did not write the blog I had promised I would.
What do we do in these instances? We have an intention, and then, life takes over. Let me share about yesterday.

I began the day at 5:30 with coffee and meditation. Off to a 6:30 am Toastmaster’s meeting. Back to walk the dog and make a green smoothie, then off to a networking meeting.

Lunch is a “working” lunch with my mastermind buddy, then to my place to help a friend of a friend open a file on my Mac that she cannot on her PC.

Off I go to Lake Oswego to get haircut and color, picking up messages on the way, since there had been no time before then. My calendar says “Hair—4-5.”

I finally check my voicemail messages and hear, “You had a 9:30 am appointment, and it is 9:45, Did you forget? Please call.”

Holy shit!!! Something was screwed up. It is 4/5, and I thought the appointment was from 4-5. Ugh! I realize the mix-up. I call, but he has left the shop. I had waited 3 weeks
for that appointment, and I am going away next week. He says he can get me in 12 days from today.

Big disappointment!!!! I feel sad. I lose my balance big time.

I stop and think about the fact that I have a choice. I wanted the pleasure of a haircut and color. Any woman over a certain age can relate to that pleasure of being transformed to feel more beautiful.

I pull over and park my car. I ask myself what I want to feel. I want to feel happy, pleasure. It is Friday afternoon, and I worked hard all week. I have so much to do, but right now, I want to feel light and happy.

I walk into a clothing store and ask if the woman knows of a good salon in the area. Indeed she does, has been going there for 11 years, and hands me their card. I drive up the road and turn into a hidden spot where the salon is still open. There are no openings at the moment, but the gal puts me on the phone with Danny, who is available tomorrow morning. I thank my lucky stars.

Now what? I had no plans since I had thought I would be having my hair done. I have so much to do.

I stop and ask myself what I want to feel. There it is again. I want to feel pleasure. There is a world class wine shop nearby, and I decide to go in. There is the weekly Friday evening wine tasting, and the place is lively and filled with smiling people drinking really good wine, eating snacks, and chatting loudly.

I ask a single woman my age if she has been here before. She invites me to sit down and says she is waiting for a blind date. We hit it off and she asks if I will stay and check him out. Of course I will. We are sisters.

I will make the long story of the evening a shorter version. It was one of the most delightful evenings of my life. The blind date was a gem of a man, and he brought his well-worn Tarot Deck and we had a blast! We were 3 peas in a pod, as they say.

We all three head to Terry’s house nearby, where we eat delicious Thai food and talk until the wee hours, as our souls were so overjoyed to have found one another.

I felt such pleasure, and when I went to sleep last night, I realized that this pleasure, this joy in being alive and giving myself over to the joy of the moment is what I want to feel when I work in my business. When I allow these kind of experiences into my life, I can
tap into the joy and pleasure that is my basic nature. Then, when I have to do the hard stuff in my business, I can bring a sense of pleasure more easily into the experience.

I am exercising the muscle of pleasure, rather than the muscle of avoidance of pain. I see the value in stepping back from the undending “to do list” that every entrepreneur has, and giving myself over to spontaneity, which always increases my creativity. Almost all of the choices we make in our daily life are motivated by one of two powerful forces. The more powerful of these is the desire to avoid pain, and the second is the desire for pleasure.

Avoidance of pain often leads to things not getting done, but going for pleasure is a far more energized state of being. You can, for instance, see getting a certain challenging task done as bringing you pleasure, thus shifting the Pain-Pleasure paradigm to support you fulfilling an obligation.

The real balancing act is the dance between the pain and pleasure of life. It is a dynamic state, like the various standing balancing poses in yogo, where thousands of muscle fibers are firing to keep you steady in tree pose. You are anything but static—more like a willow than an oak.

The master Abraham says, “When you get hold of an idea, play it out for the pleasure in it. If you are doing it for any other reason, then you are not connecting to your Source Energy.”

I know that I want to be connected to Source Energy in my business and in my life. If it means going for pleasure, so be it. I am in. I believe it is the key to balance.

And now I will post this blog, since I let myself have an experience that is worth sharing.

What a pleasure!