

causes a broader spectrum of fatigue and depression than a migraine might. Current studies have found that individuals with chronic tension-type headache report a lower quality of life.

TREATMENT. The first step in treatment should be a written record of the frequency and severity of attacks and of the medication currently consumed for them; often there is more medication used than the patient had been aware of. Overuse of analgesics and nonsteroidal antiinflammatory drugs (NSAIDs) may increase the frequency of tension-type headaches (see later). Estrogenic hormones, including oral contraception or hormonal replacement therapy, may worsen tension-type headaches.

Analgesics and NSAIDs are the most typical medications used for control of tension-type headaches, and often the individual will self-select the dosage. Most often, the doses taken are not adequate to control the pain but are taken more often than is effective, leading to rebound as a trigger for headache. Ibuprofen appears to be the fastest acting and is superior to aspirin for relief of headache pain; it has a lower risk of gastrointestinal bleeding compared to other NSAIDs. Combination therapy using analgesics can enhance the effect and allow lower dosages and lower risk of side effects. Caffeine has long been used as an analgesic adjuvant, often combined with aspirin or ibuprofen. It has been common to include codeine and butalbital, but dependency on the drug may develop. When migraine headache and tension-type headache are occurring at the same time, use of triptans shows effectiveness for both headache types. As the role of serotonin becomes better understood in tension-type headaches, medication directed at modulation of serotonin will become more common.

Tricyclic antidepressants such as amitriptyline are useful in chronic tension-type headache. If depression is comorbid, tricyclics are generally regarded as more effective than selective serotonin reuptake inhibitors. Stress management strategies are effective, as are cognitive-behavioral therapies. A combination been shown to be better than either strategy alone.⁵²

PROGNOSIS. Over time there is an increased risk of the episodic tension-type headache developing into a chronic tension-type headache.

Migraine

Overview and Definition

The definition of the World Federation of Neurology contains the following language regarding migraine: "a familial disorder characterized by recurrent attacks of headache widely variable in intensity, frequency and duration. Attacks are commonly unilateral and are usually associated with anorexia, nausea and vomiting. In some cases they are preceded by neurologic and mood disturbances."¹⁰⁴

Migraine syndrome is difficult to define due to the fact that the attacks vary widely between individuals. Within one individual, each attack may have different characteristics. The prodrome or aura, the nature of the attack, and the type of pain may represent separate pathologic pro-

cesses that happen at the same time or within the cycle of the headache. The trigger that causes one type of migraine may be different from the one that causes another type. Most authors agree that stressful periods are linked to the onset of the headache, although the manifestations may be quite different.

In the United States, migraine headaches often go undiagnosed or are misdiagnosed as tension-type or sinus headaches. There also seems to be two types of migraine sufferers, those who seek medical intervention and those who self-medicate without medical consultation. The ability to determine the appropriate link between pathologic processes and intervention remains a medical challenge. As a result, many migraine sufferers may not receive effective treatment.¹⁶

Incidence

Migraine headaches are the second most common type of primary headache. An estimated 28 million people in the United States (about 12% of the population) will experience migraine headaches at some point.⁷⁸ An estimated 6% of men and up to 18% of women have migraine headaches. In 90% of migraineurs, the first attack generally develops before the age of 40 years.⁵³ Clinical and epidemiologic investigations indicate that adult women are at greater risk for the development of migraine than are adult men. In women the frequency of headaches is highest during their reproductive years, when estrogen levels are higher, and decreases to some extent after menopause. Frequency in adult men does not appreciably change between ages 20 and 65 years.^{54,77}

About 45% of cases of migraine emerge during childhood or adolescence. Migraine headaches are estimated to appear earlier in males than in females, and in both genders migraine with aura is more likely to develop at an earlier age than migraine without aura.^{17,77} Young boys have a frequency and intensity slightly higher than young girls.

Childhood headaches include complaints of bitemporal pain and a shorter duration (less than 2 hours). Headaches appear more frequently in children than in adults. Episodic vertigo, abdominal pain, and autonomic responses are more common during an attack in a child than is headache. Cyclic vomiting syndrome (CVS) appears to be a common precursor to migraine in children.

Etiologic and Risk Factors

It has been determined that there is not a "migraine personality" but rather that those individuals who are susceptible to migraine may also be at risk for other alterations of the nervous system. Anxiety disorders, especially panic attacks and depression, are characterized by disturbances in the same neurochemical systems, and therefore the syndromes may overlap or could even be considered one syndrome with a subset of conditions. There may also be a central serotonin demodulation that is seen within the context of psychologic disorders. Chronic pain related to migraine may also lead to changes in behavior.^{22,26,64}

Comorbidity with possible association to other diseases has been studied. A relationship between migraine

and stroke has been found in many studies, but the mechanism is not clear, and there seem to be different risk factors with age and type of migraine attack. In young women 20 to 45 years, there is an increased incidence of stroke in the migraine population. Smoking or using oral contraceptives increases this risk.⁶⁶ Common elements such as abnormal platelet aggregation, a well-known component of migraine and a possible stroke risk factor, may explain the correlation. Migraine is common in the young population; stroke is not. There appears to be a higher incidence of stroke in between migraine attacks in individuals with aura. The activation of brainstem structures as the mechanism for migraine may predispose individuals to posterior fossa stroke, the most common type found in young stroke victims with migraine. Posterior fossa strokes are also more common in the general younger population. In migraine-induced stroke, the symptoms of aura last longer than 1 hour, and neuroimaging confirms infarction.⁶⁵

Despite an apparent reported connection with heart disease and mitral valve prolapse, the link does not appear to be strong. Hypertension may be one of the factors that changes episodic migraine into a chronic daily headache. Allergy, Meniere's disease, lupus, and epilepsy have all been reported often as comorbidities, and the changes in migraine status as a result of interventions directed toward these conditions may help to describe the link.⁶⁸

For many migraineurs, certain foods must be avoided. The foods most likely to be linked to migraine are milk, eggs, corn, and wheat.⁷⁶ Environmental factors such as food additives and colorings could also be a trigger for migraine. Coffee, alcohol (especially red wine), and sugar can set off an episode in vulnerable individuals.

Often the headaches are associated with hormonal changes of menstruation and use of oral contraceptives. Pure menstrual migraines appear to be more frequently migraine without aura. It appears that these migraines may be related to the abrupt decrease in estradiol that occurs immediately before menstruation after several days of exposure to high levels of estrogens. It also appears that there may be an intrinsic estrogen receptor sensitivity that differs among women. This may therefore create an individual response to the hormone level. Attempts to find differences in ovarian hormone levels between women with menstrual migraine and controls have not yielded consistent results.⁶¹

When the headaches follow a pattern related to hormone changes, pregnancy brings periods of both exacerbation and relief. In the first trimester of pregnancy, the number and severity of migraines rise. The headaches abate during the second trimester and then increase again in the third trimester. Migraine can be triggered during delivery and may be more prevalent in the weeks and months following childbirth.¹⁵

There is a positive family history of migraine in about 60% of cases, which suggests a hereditary factor. However, the patterns of migraine inheritance are complex, both in the mode of inheritance of migraine and in the role of genetic factors in the pathogenesis. Studies show an increased risk of migraine with aura among first-degree relatives. Twin studies show that migraine with aura is

caused by a combination of genetic and environmental factors.^{63, 97}

Multiple genes probably contribute to the genetic susceptibility for migraine, with different genes associated with the different types of migraine. The failure to identify a simple genetic mechanism does not rule out genetic determinants of migraine, but it suggests either that not all migraine syndromes are genetically equivalent or that environmental factors also play a significant role.

In familial hemiplegic migraine, mutations of genes have been found for at least two separate chromosomes, and there is overlap with the gene mutations found in a benign form of epilepsy that is only briefly active in children. These mutations may make the brain more susceptible to prolonged cortical spreading depression via excessive glutamate release or decreased removal of glutamate and potassium from the synaptic cleft. It is believed that there is another gene that may be responsible that has yet to be identified.³³

Study of mutations in the motor nerve terminals that contain P-type calcium channels in mice may shed some light on the mutations that would contribute to changes in acetylcholine release identified with a variety of movement disorders and may also identify part of the mechanism of the pain of migraine. Susceptibility to migraine during periods of recovery from prolonged stress suggests that the mechanism might involve a switch in autonomic balance from sympathetic to parasympathetic dominance.²²

Pathogenesis

The components of migraine are complex, and it appears that the mechanisms vary among individuals and among attacks in the same individual. The more that is known about migraine, the more levels of dysfunction are identified.⁷¹ The mechanism that causes pain may be different from the mechanism that causes the neurologic symptoms. The trigger for the migraine may give the clue to the nature of the symptoms. Fig. 37-2 illustrates the possible mechanisms related to migraine headache.³⁶

Pain is the most common complaint in migraine and may relate to several causal factors. The trigeminal complex, including cranial nerve V, is a key component in the distribution of the pain within the head and neck associated with migraine. Surrounding the large cerebral vessels, pial vessels, large venous sinuses, and dura mater is a plexus of largely unmyelinated fibers that arise from the ophthalmic division of the trigeminal ganglion and the upper cervical dorsal roots. The dural nerves that innervate the cranial vessels consist largely of small-diameter myelinated and unmyelinated fibers that almost are nociceptive in function, so that stimulation of the cranial vessels causes pain. The meninges that cover the brain parenchyma, and the arteries of the cortex are richly supplied by nerve fibers from the trigeminal ganglion and the upper cervical dorsal roots. These fibers contain substance P and calcitonin gene-related peptide that can be released when the trigeminal ganglion is stimulated, resulting in nociception, or pain. Sensitization of the mechanoreceptors in these structures enhances the responses to mechanical stimuli.⁶⁷ Sensitization of the meningeal primary afferents creates a painful response to

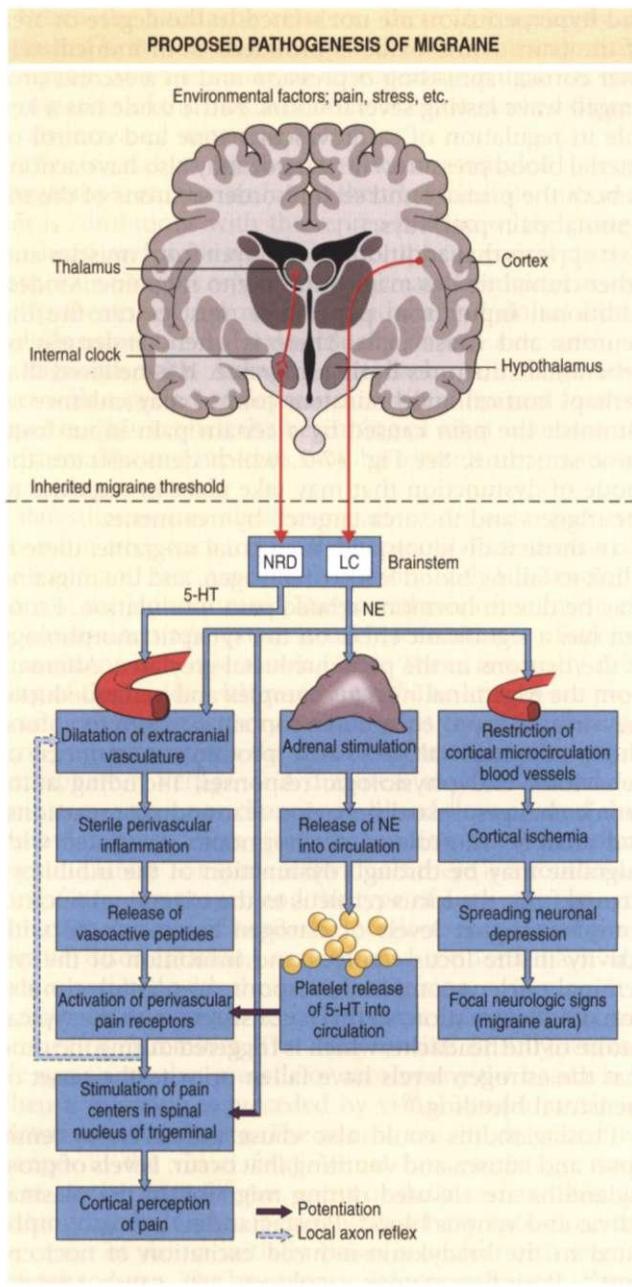


Figure 37-2

Mechanisms that are related to the pathogenesis of migraine headache based on current proposals. (From Weinstein JM: Headache and facial pain. In Yanoff M, Duker JS, Augsburger JJ, et al, eds: *Ophthalmology*, ed 2, St Louis, 2004, Mosby.)

traction and pressure. This is thought to be responsible for the diffuse pain often noted in one area of the head and the throbbing pain that seems to be associated with the heartbeat.¹⁵ Routine physical activities such as coughing, sneezing, bending over, or performing the Valsalva maneuver can cause pain through this mechanism. Sterile neuroinflammation is another possible result of the peptide release; however, there does not appear to be the level of tissue damage found in other inflammatory processes. As in other inflammatory disorders, inflammation in migraine is neither the original nor the leading event,

but one of the mechanisms involved in its pathogenesis, helping to explain findings such as sensitization.

The central modulation of pain is related to a complex neurochemical and neurophysiologic system in the brainstem. The central processes of the trigeminal afferents play a role in migraine pain. Long-lasting central sensitization may prolong the effects of a pain stimulus related to the trigeminal vascular system. Common to the complaints of migraine is the phenomenon of allydema, or pain responses to common activities such as hair combing, shaving, or wearing earrings or a necklace. This is the result of sensitization of the neurons of the medullary dorsal horn that receive sensory input from the dura and skin. This abnormal input is sent to the thalamus and then relayed to the cortex.

Results obtained by positron emission tomographic (PET) scan during a spontaneous migraine attack suggest that the pathologic activity in brainstem nuclei regulating antinociception and vascular control may be related to the pathogenesis of migraine. The brainstem contains networks that modulate pain transmission. In the medulla are so-called off-cells that inhibit pain and on-cells that facilitate pain. It is believed that these cells modulate the activity of the trigeminal nucleus ceruleus and neurons of the dorsal horn. Increased on-cell firing or decreased off-cells firing could be responsible for the increased sensitivity to both painful and nonpainful stimuli. Both pain and stress can turn on the system and may account for some of the association between pain and stress. Pain tolerance decreases in individuals who develops headache. Normally, stimulation of sensory fibers excites γ -aminobutyric acid (GABA) receptors, and the inhibition reduces the excitability of pain neurons in the dorsal horn of the spine in order to modulate pain response to stimulation. This mechanism may be aberrant in migraine sufferers, and so there is less inhibition. It has been found that tenderness in the neck, not muscle contraction, correlates with headache development.⁷³

Neurovascular transmitters and receptors play a role in headache through their influence on the regulation of tone in cranial blood vessels. Acetylcholine and vasoactive intestinal polypeptide in the cranial arteries cause relaxation of the vessels. Vascular changes occur before and during the migraine headache. In the headache with aura, cerebral blood flow is reduced by about 20% before the headache, resulting in cerebral hypoxia. This cerebral hypoxia is thought to be responsible for the neurologic defects that characterize the aura. Blood flow increases during the headache, and there appears to be some relationship to the arteries, to referred pain patterns of the veins, and to the site of pain. Dilatation of the middle cerebral artery and the superficial temporal artery on the pain side during migraine has been demonstrated.

Endothelial cells found in the vessel wall synthesize and release substances that produce vasodilation and vasoconstriction of the cerebral vessels. Cerebral blood vessels are very responsive to chemical stimuli such as changes in carbon dioxide tension and oxygen level. Bradykinin, adenosine, and histamine are released not only by neurons but also by endothelium, platelets, and mast cells. These substances have been proposed as local regulators of cerebral blood flow.¹⁸

The neurotransmitter 5-hydroxytryptamine (5-HT), known as serotonin, appears to be a part of the pathogenesis of migraine. Chronically low availability of serotonin is found in individuals with migraine, so a hypersensitivity to the presence of serotonin develops. Platelets contain virtually all the serotonin present in the blood and release serotonin during aggregation. Platelets play an important role in the acute inflammatory response. Many persons with migraine show hyperaggregability of platelets when free from headache. Aggregated platelets release catecholamines and serotonin that may cause the initial stage of vasoconstriction. Platelet aggregation is increased during the prodromal stage of migraine. This could be due to an increase in epinephrine, thrombin, or arachidonic acid in the circulation in response to anxiety. There appears to be a decrease in aggregation during the actual headache. Reported changes in plasma serotonin may be due to changes in platelet function. Serotonin also excites pain nerve endings and increases the pain response. The serotonergic neurons also have a close association with cortical astroglial cells that help to maintain homeostasis in the brain.⁷¹

The brainstem is considered to be implicated in the changes in cerebral blood flow by stimulation of the locus ceruleus or the nuclei raphe. The nuclei raphe appear to increase blood flow in the brain. Although there is no direct anatomic connection between raphe neurons and the meningeal and cerebral blood vessels, these vessels can respond to changes in central serotonin neurotransmission. Serotonergic neurons located in brainstem nuclei raphe are involved in the regulation of multiple other physiologic functions, including stress, pain, appetite, mood, and sleep. The neurons change their firing rate during the sleep-wake cycle. This may explain why sleep is often the best antidote to a migraine.

Cortical spreading depression is believed to be part of the neuronal abnormality leading to migraine. It is a mechanism that starts with a small excitatory response that begins to spread through the brain and then causes a suppression of electroencephalographic (EEG) activity that moves through the cortex. Cortical spreading depression involves gross disturbances of the brain's extracellular environment. The potassium level increases, the extracellular glutamate level increases considerably, and the extracellular calcium level decreases. Calcium ion channels control and maintain electrical potential across cell membranes and generation of electrical signals (see Chapter 28). Abnormal ion channel function is believed to be the mechanism in the rare form of familial hemiplegic migraine.³⁷

Cortical spreading depression may be responsible for the reflex activation of sensory pathways that induce inflammation within the meninges as described earlier. Cortical spreading depression is accompanied by local vascular responsiveness. Spread of first aura symptoms and spread of cortical depression proceed at the same speed. This may explain the initial hypoperfusion that is related to the aura of headache. Cortical hyperemia may be responsible for the flashing jagged light that sometimes occurs just before the pain begins. Once the headache is underway, it appears that the vascularity changes

and hyperperfusion are not related to the degree or area of the pain. Nitric oxide is produced both immediately after cortical spreading depression and in a second prolonged wave lasting several hours. Nitric oxide has a key role in regulation of cardiovascular tone and control of arterial blood pressure. Nitric oxide may also have actions at both the primary and second-order neurons of the trigeminal pain pathways.

It appears that additional pain input from muscles and other cranial tissues may contribute to migraine. Modest additional input from pericranial muscles can fire the neurons and cause pain. There is often tenderness of pericranial structures during migraine. It is believed that perhaps cortical and brainstem control may enhance or diminish the pain caused by a certain pain input from these structures. See Fig. 37-2, which demonstrates the mode of dysfunction that may take place in response to the triggers and the area targeted by treatments.

In those individuals with menstrual migraine, there is a link to falling blood levels of estrogen, and the migraine may be due to hormone-related pain modulation. Estrogen has a significant effect on the synaptic morphology of the neurons in the periaqueductal grey area. Afferents from the trigeminal nucleus complex and periaqueductal grey areas that project to them contain estrogen receptors. The pariaqueductal grey area produces a number of behavioral and physiologic responses, including autonomic changes, sexual behavior, fear and rage reactions, and anxiety. The role of sex hormones associated with migraine may be through dysfunction of the inhibitory circuits from the locus ceruleus to the trigeminal nucleus complex. Higher levels of estrogen are correlated with activity in the locus ceruleus and inhibition of the trigeminal nucleus complex in response to painful stimulation of inflammation, so this is consistent with the typical nature of the headache, which is triggered during the time that the estrogen levels have fallen prior to the onset of menstrual bleeding.⁵⁰

Prostaglandins could also cause the severe systemic upset and nausea and vomiting that occur. Levels of prostaglandins are elevated during migraine in the plasma, saliva, and venous blood. Prostaglandins are also implicated in the bradykinin-induced excitation of nociceptors.³¹ Proinflammatory cytokines can cause sensory hypersensitivity and are expressed in the central nervous system by neurons.

Migraine associated with food containing tyramine may occur in persons who have a deficiency of tyramine-o-sulfatase. The excess tyramine could be responsible for the release of catecholamines and could initiate the vasoconstrictive stages of migraine.

Clinical Manifestations

Migraine Without Aura. Migraine headaches may be dull or throbbing. They usually build up gradually and may last for 4 to 72 hours. Headache is aggravated by routine physical activity, and there is an association with nausea. Photophobia, or increased sensitivity to light, is common, and the migraineur will often seek a dark, quiet place. There are often various combinations of fatigue, difficulty in concentrating, neck stiffness, blurred vision, yawning, and pallor.

When the headache resolves, there is commonly a feeling of heaviness and aching in the head, the scalp may be tender, and there may be considerable fatigue. At the termination of the headache there is often marked diuresis, or increased urination.^{17,30}

The trigeminal brainstem nuclear complex has a somatotopic representation of the trigeminal dermatome that is continuous with the representation of the posterior head and neck region in the upper cervical dorsal horn. Pain patterns representing increased neuronal activity in the trigeminal nucleus caudalis and dorsal horn include upper cervical pain, usually on one side only.

Migraine with Aura. The headache will frequently start with a period of depression, irritability, and loss of appetite. This is often the beginning of what is known as the aura. These symptoms may be a result of spreading depression.

Paresthesias can also be a part of an aura and are second in frequency to visual symptoms. Paresthesias of the hand and face are most common, with less involvement of the leg and trunk. The paresthesia often involves the tongue, something that is rarely seen in cases of stroke or transient ischemic attack (TIA). There is usually tingling of the hands with numbness.³⁰ The attacks are predictable using the sensory homunculus. Speech difficulty during an aura reflects the involvement of the dominant hemisphere.³⁰ Vertigo and dizziness may be related to brainstem activity or changes in blood flow around the vestibular mechanism (see the section on Vestibular Migraine later). For some individuals there is a combination of aura symptoms, and the type of aura may change with each episode of headache. Neglect, spatial and geographic disorientation, anxiety, and amnesia have all been associated with the aura of migraine.

Autonomic changes can also result in paresthesia of the face or extremities. The aura consists of fully reversible symptoms that precede or accompany the headache. When a migraine is preceded by visual symptoms, it is known as a visual aura. The aura is usually described as changes in the visual field. Visual images change, and there can be loss of focus, spots of darkness, and zigzag flashing lights. It often begins with a hazy spot close to the center of vision; initially vision is unclear or it seems difficult to focus. The hazy spot will form into a star shape that further develops into a shape known as a fortification because it is semicircular with angles like those of fortifications. This scintillating vision consists of luminous, bright, flickering colors of the spectrum, much like a prism catching light. It can be combined with a scotoma, or an area of vision that appears to be obstructed, or missing. The visual image fades as the headache begins. The headache is intense, throbbing, and usually contralateral to the visual field changes.¹⁷ Fig. 37-3 shows the image as it would appear during a migraine.

Typical Aura Without Headache. In some cases, the aura symptoms are not followed by headache. When recurrent aura symptoms occur in the absence of headaches, the distinction from mimicking conditions becomes more important. This is especially important when the symptoms are very short or very long, or they begin after the age of 40. Aura without headache is seen primarily in men and in advancing age.³⁰

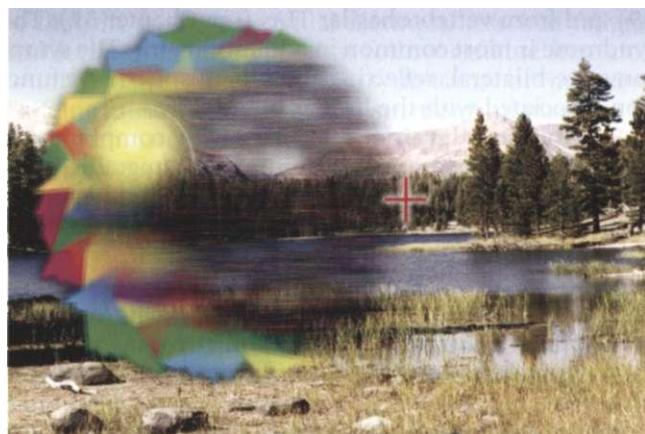


Figure 37-3

Scintillating scotoma in migraine with aura. The leading edge of the scotoma is "positive" (i.e., it consists of bright flickering imagery that obscures or replaces the normal visual field), whereas the trailing edge of the scotoma often is "negative" (i.e., it displays a relatively dark area that fully or partially obscures the visual surround). The illustration depicts a typical fortification scotoma with sharply angulated borders; many other variants of the migraine scotoma may occur. (From Weinstein JM: Headache and facial pain. In Yanoff M, Duker JS, Augsburger JJ, et al, eds: *Ophthalmology*, ed 2, St Louis, 2004, Mosby.)

Sporadic and Familial Hemiplegic Migraine. In sporadic and familial hemiplegic migraine, the presentation is migraine with aura, including motor weakness. Paresis can be associated with impaired coordination. The person experiences symptoms of numbness and weakness preceding the onset of the headache. The numbness in the face and arm may spread to involve one side of the body. Basilar symptoms may be present with dysphasia or aphasia causing difficulty with speech. Comprehension is preserved. This type of migraine may alternate with headaches without hemiplegic symptoms. Pain may be either contralateral or ipsilateral to symptoms, and on rare occasions there is loss of consciousness. Although the symptoms appear to resemble a TIA or mild stroke, migrainous infarction is rare. Age of onset for the hemiparetic migraine is 10 to 15 years, and there is usually a history within the family. Familial hemiplegic migraine appears to be related to genes on chromosome 19 or 1.²⁵ The migraine is considered sporadic if there is no first- or second-degree relative with hemiplegic migraine. There is high variability of the attacks, symptoms, and the disease course among individuals with familial hemiplegic migraine.²⁸

Basilar-Type Migraine. Basilar migraine can occur throughout life in both sexes. Basilar migraines have a symptom profile that suggests posterior fossa involvement localizing to the vascular territory of the basilar artery—the brainstem, cerebellum, and occipital lobes. The prodromal symptoms reflect brainstem dysfunction. There are complaints of altered level of consciousness, dysarthria, tinnitus, ataxia, diplopia, symptoms in both the temporal and nasal fields of both eyes, and peripheral dysesthesias, followed by occipital headache.¹ Given how often vertigo, fluctuating hearing, and tinnitus occur together, basilar migraine may be difficult to distinguish from peripheral disorders such as hydrops (see Chapter

39) and from vertebrobasilar TIAs (see Chapter 32). The syndrome is most common in adolescent girls. The symptoms are bilateral, reflecting the bihemispheric dysfunction associated with the basilar blood flow.⁷

Vestibular Migraine. Dizziness with complaints of true vertigo is extremely common in migraine, and migraineurs often have a lifelong or childhood history of motion sensitivity, with symptoms provoked by amusement park rides, riding in the back seat of a car, or reading in the car. Most migraine sufferers will report some motion sensitivity during a severe headache and will often prefer to lie still in a dark, quiet room, although these symptoms alone are not sufficient to be considered vestibular migraine.⁶⁴

When dizziness is the primary complaint or is the predominant component of the aura, then it may be considered as vestibular migraine. Vertigo occurring as an aura may arise from the same transient inhibition of neuronal function responsible for the visual aura. Episodic vertigo from vestibular migraine can be thought of as a subset of basilar migraine. Spells usually last approximately an hour but can last several hours or days in patients who have severe symptoms.¹² Nausea, vomiting, hypersensitivity to motion, and postural instability are cardinal signs. Some persons complain of vertigo, a swimming sensation, or imbalance without a complaint of headache. Dizziness does not always accompany the headache but may intermittently coincide with the headache. Changes are rarely seen on electronystagmography (ENG), indicating a peripheral deficit that is transient. The vestibulocochlear symptoms occur most often as a part of the headache. Auditory symptoms also occur in migraine. Phonophobia is the most common, with complaints of tinnitus more frequent than in the general population. Hearing loss alone is rarely associated with migraine.⁴⁸ Box 37-2 defines both definite and probable vestibular migraine.⁷⁵

The mechanisms are still not clearly understood, but it may be that neuropeptides released during the migraineous event could increase the firing of the peripheral vestibular apparatus, or the vestibular nuclei.⁷⁵ This may result in a transient increase in the firing on one side of the system, causing the subjective sensation of moving. Changes in cerebral blood flow during the migraine may contribute to the dizziness. The inner ear is supplied by branches of the anterior inferior cerebellar artery arising from the basilar artery, and the anterior inferior cerebellar artery also supplies the anterolateral pons, middle cerebellar peduncle, and flocculus. Prolonged vasoconstriction of vertebral and basilar arteries may be responsible, or the cause could be the direct mechanical pressure of the dilated vessel during the vasodilating period of the headache. The dizziness can be associated with the position of the head, indicating an association with benign paroxysmal positional vertigo (see Chapter 39).

Acute attacks can be treated with various analgesics. Preventive treatment is most frequently accomplished with amitriptyline, β -blockers, calcium channel blockers such as verapamil, and acetazolamide. Migraine may be associated with many vestibular symptoms.⁵¹

Ophthalmoplegic Migraine. Ophthalmoplegic migraine results in pain around the eye and paralysis in

Box 37-2

DEFINITE AND PROBABLE VESTIBULAR MIGRAINES

Definite Vestibular Migraine

- Episodic vestibular symptoms of at least moderate severity (vertigo, self-motion); vestibular symptoms are "moderate" if they interfere with but do not prohibit daily activities and "severe" if patients cannot continue daily activities
- Current or previous history of migraine according to the 2004 criteria of the International Headache Society (IHS)
- Migraine symptoms during two or more attacks of vertigo, including migraine headache, photophobia, phonophobia, visual or other auras
- Other possible causes ruled out

Probable Vestibular Migraine

- Episodic vestibular symptoms of at least moderate severity
- Current or previous history of migraine according to the 2004 criteria of the IHS
- Migraine symptoms during vestibular symptoms as stated above
- Precipitants of vertigo in more than 50% of attacks consistent with migraine: food triggers, sleep irregularities, hormonal change
- Relief of symptoms in response to migraine medications in more than 50% of attacks
- Other causes ruled out

the distribution of the third, fourth, or sixth cranial nerve and can produce diplopia, or double vision. One of the features of ophthalmoplegic migraine is that the headache always precedes the oculomotor deficit by several hours or days. The paralysis can progress from being transient to lasting several days, and in some persons it becomes permanent. People with ophthalmoplegic migraine typically give a history of many years' duration of migraine without oculomotor involvement before the ophthalmoplegia develops.^{7,101}

Retinal Migraine. Retinal migraine is repeated attacks of monocular visual disturbance, including scintillations, scotomata, or blindness, associated with migraine headache. Visual changes are strictly unilateral. There may be neuronal spreading depression or involvement of the posterior ciliary vasculature. TIA must be ruled out, as emboli from the carotid artery can cause similar symptoms.⁵

Status Migrainosus. The headache of status migrainosus (SM) lasts more than 72 hours, and a usual trigger will set it off. SM typically resembles the individual's usual severe migraine, but there may be a spread of pain to other areas as the headache persists, and the spread of allodynia occurs. The premenstrual part of the female cycle is a time of particular risk for SM, and changes in hormone status, pregnancy, miscarriage, or change in birth control pills can be factors. Upper respiratory or urinary tract infections can also be the trigger for SM. Overuse of analgesics and rebound withdrawal-type headache, the headache that comes when not taking medication, can trigger SM. Ruling out other possible causes is critical.

Prolonged vomiting to the point of dehydration is common. Severe pain and fatigue limit activity, and hospital admission may be appropriate. Ergots and antiemetics are effective to some degree. Corticosteroids may be helpful, and opioids are sometimes used. Comorbid depression is frequently seen.¹⁰

Migraine Aura Status. Migraine aura status is a rare subtype with persistent aura without infarction. One or more symptoms that the individual usually experiences as a part of his or her typical migraine aura persist for more than 1 week. Visual symptoms are most common, but it can be any symptom. Most typical treatment is ineffective, and the problem often must run its course. Valproate has been helpful in a small number of patients.

Chronic Migraine. Most cases of chronic migraine start as migraine without aura. The criterion for chronic migraine is headache for 15 or more days a month for more than 3 months. Medication overuse is the most common cause of chronic migraine, with rebound or withdrawal of medications as a trigger. Women predominantly report chronic migraine, and they report a change in symptoms over time, with decreasing photophobia, and nausea and a headache that resembles a mixture of tension-type headache and migraine.⁸⁷

MEDICAL MANAGEMENT

DIAGNOSIS. In most cases the diagnosis of migraine can be established by the history alone. The neurologic examination is normal. Diagnostic procedures are unnecessary and may lead to confusion. An EEG may show focal slowing if taken during an attack of migraine and may create the impression of a space-occupying lesion or infarction. Magnetic resonance imaging (MRI) scans show diffuse white matter changes in the frontal subcortical and deep white matter at the level of the basal ganglia. This may be due to platelet aggregation with microemboli, abnormal cerebrovascular regulation, and repeated attacks of hypoperfusion during the aura.

TREATMENT. It is clear that the causes of migraine are multifactorial, and therefore the most successful treatment is directed at the system that is most likely implicated. Avoidance of precipitating factors becomes a natural component of controlling frequency. Rest in a quiet, dark room is often necessary during the attack. Pharmacologic treatment can be abortive or preventative by reducing frequency and severity. Because the migraine type may vary in one individual in different attacks, prevention is sometimes difficult. Timing of medication related to migraine type may be critical. There may be a change to the blood-brain barrier during migraine that allows inhibitory substances to pass into the brain, which changes pain perception; however, the gastric stasis that can be part of the aura can limit or slow the absorption of medication.³⁸

Vasoconstricting drugs such as Cafergot, a combination of ergotamine tartrate and caffeine, are often helpful. Caffeine has several properties that may be of benefit in the treatment of migraine. Concurrent oral administration of caffeine with aspirin increases the peak plasma concentration of aspirin. It has mood-altering properties,

so that the increased mental alertness, lessened fatigue, and feeling of well-being that follow caffeine ingestion counterbalance some of the symptoms of migraine. Caffeine causes cerebral vasoconstriction and may also be antiinflammatory.⁸⁸ Caffeine use is a major risk factor for development of chronic daily headache and can cause increases nervousness, sleeplessness, and anxiety.

NSAIDs are useful analgesics for migraine-type pain. They also play a role in the inhibition of prostaglandin synthesis in brain neurons. NSAIDs are particularly effective when inflammation has caused sensitization of pain receptors to mechanical or chemical stimuli. They appear to prolong the catecholamine and serotonin turnover in brain neurons and block the release of serotonin. Antinflammatories inhibit platelet formation that becomes abnormal during a headache phase. Aspirin and other NSAIDs are quickly absorbed. Contraindications for use are ulcers and bleeding disorders.¹⁸ Increasing the dosage of these medications does not significantly increase their effectiveness, and there is a dose-dependent increase in the side effects, so use of aspirin and other NSAIDs at a low dose should be tried before going on to higher doses. There is no evidence that trying another type of NSAID after a failure of one type is helpful.

Ergot alkaloids provide vasoconstriction and are more active on large arteries. There is a temporary increase in blood pressure. These drugs may decrease activity in the central trigeminal neurons. Ergot alkaloid can be used to abort a headache. There is some concern about prescribing a drug that induces vasospasm in individuals with aura or those with hemiplegic migraine.⁹⁵ A short-term increase in blood pressure may limit use in some patients.

Triptans are effective in controlling the symptoms associated with migraine. The drug action is at the level of the serotonin receptor agonist. Typically, the sensation of throbbing is diminished with use of triptans. There is evidence suggesting that triptans may also activate descending pain-modulating pathways that affect the trigeminal system. The most common triptan, sumatriptan, is a rapidly effective agent for aborting attacks when given subcutaneously by an autoinjection device. In less severe cases, sumatriptan is given orally. Other triptans now in use are zolmitriptan, rizatriptan, naratriptan, eletriptan, and frovatriptan. Headache recurrence is a problem with all triptans, and they do not appear to be effective during aura. Triptans primarily cause constriction in the cranial arteries with an effect in the coronary arteries as well. These drugs therefore should be used with caution in individuals with coronary artery disease.⁸⁴ The nausea and vomiting induced by migraine can be controlled by drugs such as metoclopramide, which works as an antiemetic, counteracting gastric stasis by increasing intestinal motility and increasing the absorption of other migraine drugs.

Prophylactic treatment may be necessary if migraine occurs more than three times per month. The mode of action may involve both an effect on extracerebral vasculature and the stabilization of serotonergic neurotransmission. The β -blocking drugs affect the central catecholaminergic system, relating to the central mechanism of the headache. In this class of drugs are

Box 37-3**TREATMENT OF MIGRAINE****Acute**

- Rest in dark, quiet room
- Nonsteroidal antiinflammatory drugs
- Ergotamine and caffeine
- Narcotics
- Antiemetics
- Ergot alkaloids
- Triptans

Prophylactic

- β -Blockers
- Calcium channel antagonists
- Carbamazepine
- Verapamil
- Tricyclic antidepressants
- Antiserotonin drugs
- Nonsteroidal antiinflammatory drugs
- Neuroleptic drugs
- Behavioral treatment

propranolol, metoprolol, timolol, nadolol, and atenolol.¹⁷ Side effects are fatigue, gastrointestinal symptoms, and dizziness. Vivid dreams, nightmares, insomnia, depression, and memory disturbances will cause termination of use.⁹⁴

Calcium channel antagonist drugs such as verapamil may reduce the frequency of attacks after an interval of several weeks, but the severity and duration of attacks are not influenced.¹⁻⁹⁶ Flunarizine can be used for migraine prophylaxis if β -blockers are ineffective or contraindicated. Box 37-3 summarizes the acute and prophylactic treatment of migraine.

With the current understanding of the mechanism of migraine, new treatments are being developed. Drugs directed at inhibiting spreading depression, reducing glutamate levels, and controlling nitric oxide biosynthesis show promise. Antiemetics combined with other drugs are being studied, and the use of neuroleptics with an effect on serotonergic, adrenergic, and cholinergic neurotransmitter systems are in clinical trials.⁹

Antiepileptic drugs such as gabapentin, valproate, and topiramate can be effective for migraine prophylaxis with an effect on levels and action of GABA in the central nervous system and influence on the activity of sodium and calcium channels. Gabapentin is effective in both episodic and chronic migraine. Valproate blocks neurogenic inflammation. Topiramate inhibits the activation of trigeminocervical neurons. Paresthesias, cognitive effects, and dizziness are side effects that may limit use. Valproate should not be used during pregnancy, due to possible neural tube defects.^{89,91}

Botulinum toxin (Botox) has been used for migraine prophylaxis but has not been well studied. Central effects may be in the hippocampal neurons and astrocytes, leading to a decreased release of neuropeptides and possible blockade of glutamate.

Herbal therapies have also been used for many years with reported beneficial outcomes. Feverfew (*Tanacetum*

parthenium) appears to decrease headache intensity and nonheadache symptoms of nausea, vomiting, photophobia, and phonophobia. Feverfew is rich in sesquiterpenelactones, which may be a nonspecific serotonin, bradykinin, prostaglandin, and acetylcholine antagonist. Long-term use has not been carefully evaluated.

Studies indicate that magnesium given as trimagnesium dicitrate appears to have some effect on the intensity and duration of menstrual migraine, and shows promise in use in children. Riboflavin may improve oxygen metabolism at the level of the mitochondria. It has been effective in reducing attack frequency, intensity and duration, but there are few studies to support this. Coenzyme Q10, also working at the level of the mitochondria, has been proven to decrease the frequency of migraine.

Acupuncture has been used with limited results, and the effects are temporary. Its effect is most likely related to changes in stress levels and control of pain responses. Hypnosis has also been used for a long time; however, it has been difficult to determine its effects in current studies. Behavioral treatment has targeted coping with pain and relaxation and examining unrealistic beliefs and their role in creating stress. It includes distraction, imagery, assertiveness training, and problem solving.²⁶

PROGNOSIS. Men have a 48% persistence rate, whereas women have a 79% rate. Menopause and the resultant decrease in estrogen usually result in a decrease in the frequency and severity of migraine. However, there have been reports of migraine attacks beginning after the onset of menopause.

There has been some question about the relationship of migraine to increased chance of stroke. Migrainous cerebral infarction is associated with an exceptionally severe migraine with changes seen on computed tomographic (CT) scan.¹¹

SPECIAL IMPLICATIONS FOR THE THERAPIST**37-1****Migraine****PREFERRED PRACTICE PATTERN**

5D: Impaired Motor Function and Sensory Integrity Associated with Nonprogressive Disorders of the Central Nervous System—Acquired in Adolescence or Adulthood

Biofeedback is the best known and most widely used nonpharmacologic procedure for the treatment of migraine. Biofeedback allows the client to make intrinsic changes in both the autonomic and somatic nervous systems. The use of biofeedback is not effective for all migraineurs, probably because of the variety of causes of migraine. The mechanism is believed to be a change in the levels of neurotransmitters produced when a person is able to control the stress response. Both thermal biofeedback (the control of temperature in the digits) and EMG feedback (the control of muscle activity) have been used by therapists in the treatment of migraine.

There appears to be a direct relationship between cervical spine dysfunction and a sympathetic nervous

system initiation of vascular spasm. Some migraineurs have relief of symptoms when the cervical spine is treated with manipulation or other physical modalities. Although manipulation has been used for the control of migraine headache, there is an increased risk of stroke from chiropractic manipulation of the neck.

Treatment of the soft tissues of the neck and occipital structures often will bring some relief to the client with migraine. This may be due to the general relaxation of the skeletal musculature, which reduces the stress responses. More research into the sympathetic changes associated with these techniques may bring the therapist further into the treatment of migraine. A better understanding of the relationship of the upper cervical musculature to the pain pattern of migraine may also lead to techniques that will help to provide relief of the pain.

Consistent and aggressive exercise may also create relief of migraine headache. Although current reports have been anecdotal, further investigation may lead to an exercise protocol as a part of the overall management of migraine.

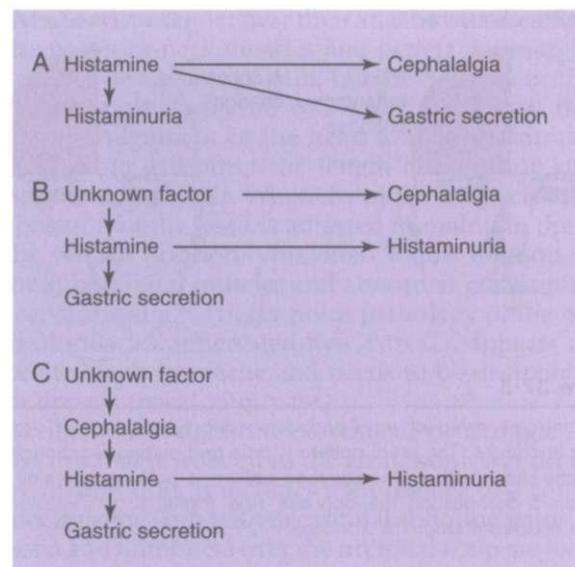


Figure 37-4

Possible relationship between histamine headache and gastric secretion in cluster headache. **A**, Histamine as the causative agent. **B**, Histamine level as an independent phenomenon. **C**, Increased histamine as a secondary product. (From Sjaastad O: *Cluster headache syndrome*, London, 1992, Saunders, p 150.)

Cluster Headache

Overview and Definition

Cluster headaches (CHs) are rare but are the most painful of the primary headaches. There are two types of CH. The episodic type is the most common, constituting 80% of all cases. Episodic CH is defined by periods of susceptibility to headache, called cluster periods, alternating with periods of remission. The headache can occur daily or several times per day for a period of several weeks, known as the cluster period. The headache will go away for several months and recur as another series or duster.^{69,90}

Chronic cluster headache is a term used when the headaches have gone on for at least 12 months and remissions last less than 14 days. Chronic CH evolves from episodic CH.⁴⁹

Incidence

CHs affect approximately 1% to 4% of the population. CHs occur mostly in men between the ages of 20 and 50 years and predominantly between ages 27 and 30 years, approximately 10 years later than the typical onset of migraine. Black males appear to have a higher incidence than males of other racial backgrounds. The ratio favoring men over women is getting smaller over time.¹⁰²

There is a correlation between CH and prior head trauma. There is question as to whether this may be related to venous vasculitis, since there is evidence of inflammation.¹⁰²

Studies of genetic risk are beginning to show some prevalence of CH in certain families, with a positive history in more than 7% of individuals with CH. The possibility of gene mutation is strong with a high concordance found in monozygotic twin studies.⁸³

Pathogenesis

Like migraine, CH appears to have multifaceted components and dysfunction at many levels. There is vasodilation of the ophthalmic and internal carotid arteries, ipsilateral to the pain. The vascular changes may be a result of stimulation of the trigeminal complex, rather than a casual factor. Stimulation of the trigeminal ganglion causes a cerebral vasodilator response. There is an increase in the level of calcitonin gene-related peptide and substance P. The cerebrospinal fluid shows changes consistent with both parasympathetic and sympathetic and central involvement in CH.

Platelet levels of histamine and serotonin increase during the episode of headache and decrease in the absence of headache. Fig. 37-4 shows the relationship among histamine, gastric secretions, and pain. Blood pressure typically increases; heart rate decreases during attacks of CH. The CH period may be characterized by disturbances in neuroendocrine substances based on the circadian rhythm, which may explain its cyclic nature.^{23,46}

The carotid body may play a major role in the pathogenesis of CHs by its receptor mechanism associated with blood pressure and heart rate. Throughout the course of the cluster period, it may be that the chemoreceptor activity is affected by central control of sympathetic and parasympathetic pathways.³⁹

Hypothalamic regulation of the endocrine system may explain the cyclical nature of the headaches. There is decreased plasma testosterone during the CH period and alterations of production of hormones and melatonin. Melatonin levels are generally low during the day and increase during the hours of darkness and sleep. The nature of the hypothalamic disturbance is not clear.

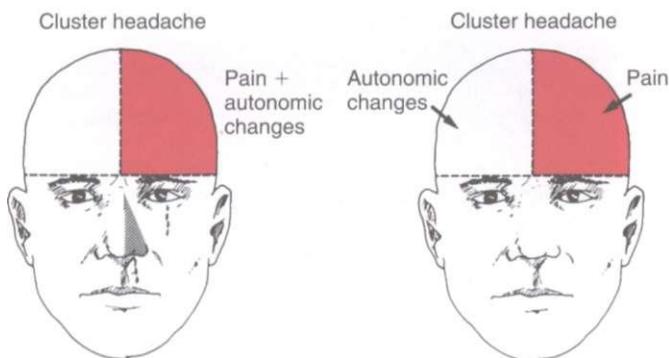


Figure 37-5

The interrelationship of pain and autonomic symptoms and signs in cluster headache. The usual pattern is pain and autonomic changes on the same side (left). A less common pattern is pain on one side and autonomic changes on the opposite side (right). (From Sjaastad O: *Cluster headache syndrome*, London, 1992, Saunders, p 49.)

Clinical Manifestations

The onset of CH is sudden, with excruciating pain. In the majority of CH cases, the headache remains on one side of the head in all recurrences throughout life. The headache is usually localized to one eye and the frontotemporal region (see Fig. 37-1). The pain is boring and nonthrobbing. Autonomic symptoms, which are often on the opposite side from the pain, include photophobia, tearing, and nasal congestion. Autonomic changes may be bilateral. Occasionally Horner's syndrome (constricted pupil, droopy eyelid) or forehead sweating will appear on the uninvolved side of the face.⁶ Fig. 37-5 shows the interrelationship of pain and autonomic symptoms in CH. Persons will occasionally complain of blurred vision on the painful side. Persons with CH generally prefer to assume an erect rather than a reclining posture during attacks.²⁹

Vasodilators such as nitroglycerin will induce cluster attacks. Often, but not always, alcohol induces an acute cluster attack while the person is in an active cluster period.

Attacks are commonly induced on awakening from an afternoon nap or from sleep during the night, most commonly 90 minutes after falling asleep. Melatonin levels are low during the day and increase during the hours of darkness and sleep; in individuals with CH, the 24-hour production of melatonin is reduced, and the acrophase (the time from midnight to the moment of peak hormone level) is moved forward. Low serotonin level may be responsible for the low melatonin production. Sleep apnea and hypoxemia are associated with CHs.²⁹

CHs follow a circadian rhythm and occur often during the same time of year. They occur most during the summer or winter solstice, appearing shortly after either the longest or shortest day of the year. Even the setting of the clock for daylight saving time will affect the headache cycle, often bringing an end to an episode.¹⁰²

Persons with CH have some similar physical characteristics, such as a hypermasculine appearance that includes increased and asymmetric facial wrinkles and

thick orange peel skin, giving them the appearance of people with alcoholism. Men with CH are on average 3 inches taller than age-matched controls. The typical client with CH smokes cigarettes and drinks alcohol. Personality inventories show a relationship to anxiety, compulsivity, and hypochondria.¹⁹

MEDICAL MANAGEMENT

DIAGNOSIS. Diagnosis is based on the symptoms and history. Paroxysmal hemicrania, trigeminal neuralgia, and temporal arteritis may have similar symptoms, but they are not episodic. Diagnostic criteria are strict unilaterality, severe intensity, orbital localization, and short duration. Differential diagnosis includes migraine, trigeminal neuralgia, chronic paroxysmal hemicrania, pericarotid syndrome, sinusitis, and glaucoma.¹³

TREATMENT. Education should be provided regarding precipitating factors, including alcohol, abrupt changes in sleeping patterns related to travel, and work shift changes. Lack of sleep or afternoon naps may precipitate a headache. Bursts of anger or prolonged anticipatory anxiety can provoke a headache during the cluster period. Altitude hypoxia can increase incidence at levels above 5000 feet, including on airplane flights. Avoiding these situations may reduce the frequency of CH during a cluster period. Upright activities, including walking, appear to be of some relief. Few people choose to lie down during an attack.¹⁹

Treatment is similar to that for migraine. Sumatriptan and ergotamine-containing drugs are fast acting and effective for controlling acute symptoms. For prevention of headache during cluster periods, verapamil can be combined with prednisone. Methysergide is effective, but side effects include fibrotic complications, and it appears to lose its effectiveness with repeated use for some. Valproic acid is effective but needs to be carefully monitored for liver, pancreas, and blood count changes. Lithium carbonate leads to long-term remission, but has a narrow therapeutic window and side effects that include tremor and renal and thyroid dysfunction.⁷⁰ Combinations of these drugs are often used rather than one drug in maximal dosages.

Melatonin therapies can be helpful, with remission coming within 3 to 5 days. Oxygen inhalation gives symptomatic relief of headache but does not change the cluster period.

Surgical procedures, including radiosurgeries directed toward the sensory trigeminal nerve, have been the most successful. Hypothalamic implants have been used in individuals with intractable chronic CH. The results are encouraging, with notable pain reduction. All deep-brain electrode implantation procedures carry a small risk of mortality due to intracerebral hemorrhage. Future studies are directed toward other areas of the pain matrix that may be suitable targets for neuromodulation in patients with CH who do not respond to hypothalamic modulation.⁵²

PROGNOSIS. In the natural course of CH, around 10% of those affected will transition from episodic CH to chronic CH, and that same number may change from chronic CH

to episodic CH. One third will suffer for around 20 years and then experience a complete remission. In another one third the attacks will be mild and no longer require medical intervention. The final one third continue to have attacks in the same pattern and to the same degree. CHs do not begin in old age.

SPECIAL IMPLICATIONS FOR THE THERAPIST 37-2

Cluster Headache

PREFERRED PRACTICE PATTERN

5D: Impaired Motor Function and Sensory Integrity Associated with Nonprogressive Disorders of the Central Nervous System—Acquired in Adolescence or Adulthood

It is important for the therapist to be able to recognize the symptoms of CH and to differentiate it from tension headache. Tension headache often is improved when drinking alcohol; CH may be triggered by alcohol. Relaxation and stress reduction may be of benefit if the client has a history of episodes triggered by stress response.

Use of thermal and EMG biofeedback may be of benefit (see the section on Migraine). Aggressive exercise appears to improve symptoms in CH as it does in migraine.

the head and are tender near their attachment sites. Spasm of the posterior neck muscles and pericranial muscles is believed to be a source of pain, but the evidence is mixed. Abnormal posture of the trunk and neck may trigger headache. Alignment of the head in relationship to the cervical spine will affect the length and tension of the muscles and ligaments. When the upper back is rounded, the position of the head is adjusted to maintain the eyes in the vertical position. This often results in shortening of the suboccipital muscles and abnormal positioning of the cervical spine.¹² Trigger point pathology of the upper cervical muscles, innervated by C1 to C3, appears to be associated with headache and needs to be distinguished from upper cervical joint pain.

Occipital neuralgia can be related, or a single-point complaint that is isolated to the greater or lesser occipital nerve distribution. It includes the presence of a constant deeper burning pain with occasional shooting pains. Paresthesia and numbness over the occipital scalp are usually present. Occipital neuralgia is believed to arise from trauma or entrapment of the occipital nerve as it goes through the suboccipital muscles, or it may be related to the C2 spinal root.

Temporomandibular joint (TMJ) dysfunction should be ruled out as a cause of tension headache. Damage to alar ligaments can cause headache and can be ruled out with use of functional CT.

The C2-C3 apophyseal joint is suspected of producing cervicogenic headache related to its biomechanical vulnerability as a transitional joint between the C1 and C2 vertebra, which rotate the head, and the C3 to C7 vertebra, which flex and extend the neck. It bears both vectors of stress and the result is a mechanical irritation of C3; this joint and the third occipital nerve appear most vulnerable to trauma from acceleration-deceleration or whiplash injuries. If that is the case, blockades of C3 can relieve headache pain.¹

It is difficult to diagnosis cervicogenic headache, because there is overlap in the symptoms, including unilateral, occipital pain radiating to the upper cervical area. Head movement can increase migraine pain related to allodynia but may not be the primary trigger.²¹ There is no consensus on what constitutes an adequate physical examination of the neck or the techniques for performing the examination. It is not even clear what findings are significant.²² Passive neck positioning in extension and rotation toward the side of pain or digital pressure to the involved facet regions or occipital nerve are typically reported as painful.⁴⁰ Muscular trigger points are usually found in the suboccipital, cervical, and shoulder musculature and may refer to the head. These findings are in the absence of neurologic findings of cervical radiculopathy, although the patient might report scalp paresthesia or dysesthesia.^{100,103}

MEDICAL MANAGEMENT

The failure to demonstrate conclusively a specific disease or dysfunction of the neck is an impediment to specific treatment for the individual with possible cervicogenic headache.⁸²

Cervical epidural steroid injections may be indicated for multilevel disk or spine degeneration. Nerve blocks,

SECONDARY HEADACHES

Cervicogenic Headaches

In cervicogenic headache, pain is localized to the neck and occipital regions and may project to the forehead, orbital region, temples, vertex, or ears. Pain is precipitated or aggravated by special neck movements or sustained neck posture. There can be resistance to or limitation of passive neck movements, and texture, tone, or pain response to active and passive stretching and contraction. Radiologic examination reveals at least one of the following: movement abnormalities in flexion/extension, abnormal posture, fractures, congenital abnormalities, bone tumors, rheumatoid arthritis, or other distinct pathology (but not spondylosis or osteochondrosis, because these are conditions that are common incidental findings in asymptomatic individuals). The criteria do not insist that the lesion be located within the neural territory of C1 to C4 but that it be in the neck.⁸¹ The mechanism, which may be related to disk disease in the lower neck, may involve compensatory increase in movement, especially in the facet joints in the upper cervical spine, thus causing pain to travel along the C1 to C4 nerve to the interface of the trigeminal complex. It is believed that nociceptive impulses and other sensory impulses can be involved in sensitization of the brainstem and thus be involved in the spread of pain into the head as described earlier.

Disorders of the cervical spine and atlantooccipital junction may create localized pain and referred pain to the head. Muscles innervated by C1 to C3 all attach to

trigger point injections, or radiofrequency thermal neuromodulation may block the cascade of sensitization to the central mechanisms. Surgical interventions such as neurotomy, dorsal rhizotomy, and microvascular decompression of nerve roots to perform blocks or reduce pressure on a nerve often also provide only temporary relief, with the possibility of longer intensification of pain. Nerve stimulators have been used successfully in migraine and may be of some benefit in cervicogenic headache.⁷

Tricyclic antidepressants have been used at a lower dosage than that required for the treatment of patients with depression. Simple analgesics may be used to manage pain but have long-term side effects that should be considered.

Muscle relaxants may be of benefit as they relate to the central nervous system.⁴²

Studies using botulinum toxin have been mixed, and this agent may be more effective if the "follow the pain" strategy is used, injecting the muscles that are reported as tender.⁴¹

Physical and manual therapies are important modalities for treatment of cervicogenic headache. Long-term prevention and control of headaches appears greatest in individuals who are involved in ongoing exercise and physical conditioning programs.^{8,47}

SPECIAL IMPLICATIONS FOR THE THERAPIST 37-3

Cervicogenic Headaches

PREFERRED PRACTICE PATTERN

5D: Impaired Motor Function and Sensory Integrity Associated with Nonprogressive Disorders of the Central Nervous System—Acquired in Adolescence or Adulthood

The chronic headache that arises from the atlantooccipital and upper cervical joints is a unique syndrome and needs to be differentiated from disorders at other vertebral levels.^{57,58} Although this type of headache is responsive to therapy oriented at treating the soft tissue restrictions, the method of examination, assessment, and treatment needs to be specific to the neck and occiput.¹²

If the headache is related to TMJ dysfunction, this should be addressed by the appropriate therapist. Intervention can include relaxation, body awareness, physical therapy modalities, transcutaneous electrical nerve stimulation (TENS), and soft tissue techniques.

Posttraumatic Headache Syndromes

Overview, Definition, and Risk Factors

Posttraumatic headache syndrome is not a single pathology but a group of traumatically induced disorders with overlapping symptoms. The headache is a manifestation of the brain dysfunction and can have contribution from the musculoskeletal injuries. The headache can last longer after the trauma than the musculoskeletal problems.⁵⁹ When the headache develops within 7 days of the trauma and resolves within 3 months, it is considered acute. The

headache can begin weeks or months after the trauma and can persist for years. This headache is considered chronic. The risk of developing chronic posttraumatic headache is greater for mild or moderate head injury than for severe head injury.⁴³

Pathogenesis

The mechanisms of posttraumatic headache mirror those of primary headaches. Both central and peripheral sensitization may play a role in posttraumatic headache. Neurochemical changes similar to those found in migraine include excessive release of excitatory amino acids, especially glutamate; an increase in extracellular potassium, intracellular sodium, or calcium; and an abnormal accumulation of serotonin. There is a decline in magnesium levels, abnormalities in nitric oxide formation, and changes in catecholamines and endogenous opioids. Chronic local inflammation can release substance P, bradykinin, and other pain-producing peptides. The nerve endings responding to these neurochemicals can trigger spasm and pain.³²

Diffuse axonal injury resulting from the shear forces to the brain and brainstem remain difficult to image. With newer MRI techniques, PET, and single-photon emission computed tomography (SPECT), the parenchymal delayed cerebral atrophy and axonal damage related to membrane injury may be identified. The ability to detect metabolic brain changes using magnetic resonance spectroscopy will help to determine the level of brain damage sustained. Xenon inhalation techniques provide additional insight into brain perfusion abnormalities.

The connection between the cervical nerves (C1 to C3) and the cervical portion of the trigeminal nucleus forming the trigeminocervical nucleus may play a role in a similar manner as in both tension-type headache and migraine. Pain from the upper cervical nerves is then referred to the ophthalmic division of the trigeminal nerve, and the pain is felt in the orbital, frontal, and temporal regions. Although there is no direct head trauma with some whiplash injury, studies show that these accidents can cause cerebral hemorrhages on the surface of the brain and upper cervical spinal cord.

Clinical Manifestations

Posttraumatic tension-type headaches are generally mild to moderate with dull, aching, pressing, tightening feelings that are usually not pulsating. They are often bilateral and bandlike. These headaches do not involve nausea or vomiting and are not aggravated by bright lights.

Posttraumatic migrainelike headaches can become incapacitating when combined with other sequelae of brain injury. They are described as pulsating or throbbing. They trigger nausea and vomiting, and they are triggered by bright lights, noise, and odor. Increased activity can make the headache worse, and often the challenge of daily tasks can be overwhelming.

Posttraumatic dysautonomic cephalgias is a result of neck injury and is similar to migraine. The pain is unilateral and throbbing, and there are pupillary dilation and excessive sweating on the side of the headache during the headache phase. Miosis can occur between headaches.⁹³

Posttraumatic clusterlike headaches appear as CHs do. Trauma may trigger a cluster period during what had been a remission.

Cervicogenic headache related to cervical acceleration/deceleration syndrome is associated with an injury to the neck without a direct strike to the head. Usually there is both shear damage to the brain and musculoskeletal damage to the soft tissues in the neck, jaw, and scalp. There are complaints of neck pain, headache, visual disturbances, dizziness, weakness, paresthesias, and concentration and memory disturbances as well as psychologic symptoms. Complaints of memory and concentration disturbances after whiplash injury is common.

Dizziness and vertigo are often reported along with headache. When the dizziness is triggered by activity, the headache can be worse. In an effort to control the sensation of dizziness that comes with head position changes or when there is excessive visual stimulation, the individual will co-contract the muscles of the neck to limit stimulation of the vestibular system resulting in a cervicogenic headache. See Chapter 38 for further description of the mechanism of cervicogenic dizziness. Box 37-4 lists the typical complaints associated with posttraumatic headaches.

Headache status before trauma does not make one more or less likely to develop headache after trauma. But the incidence of headache after trauma is high regardless of prior experience with headache. Mechanical impact factors such as an abnormal position of the head (i.e., rotation) increase the risk of posttraumatic headache.

MEDICAL MANAGEMENT

TREATMENT. The treatment of posttraumatic headaches proceeds in the manner described earlier for the type of headache that is manifested. Early and intensive treatment may be the main factors in stopping headache and preventing a chronic condition. Recognizing this type of headache is critical in the course of treatment of an individual with brain injury related to a blow to the head or other causes of whiplash. These headaches are often not recognized by the medical practitioners who provide the follow-up care. Individuals with persistent complaints of headache are often assigned the label of malingerer or symptom magnifier.

SPECIAL IMPLICATIONS FOR THE THERAPIST 37-4

Posttraumatic Headache Syndromes

PREFERRED PRACTICE PATTERN

5D: Impaired Motor Function and Sensory Integrity Associated with Nonprogressive Disorders of the Central Nervous System—Acquired in Adolescence or Adulthood

Persons with posttraumatic complications are seen by therapists in many different settings. With moderate to severe head injury, the headache is a part of so many other issues that are critical to the return of function that it may be dismissed. The individual may also have difficulty in describing the headache secondary to cognitive changes. Therapists working in rehabilitation and long-term care are often confronted with vague

Box 37-4

TYPICAL COMPLAINTS ASSOCIATED WITH POSTTRAUMATIC HEADACHE SYNDROME

Head and Face Pain

- Cervicogenic or tension-type headaches
- Cranial myofascial injury
- Secondary to neck injury (cervicogenic)
 - Myofascial injury
 - Intervertebral disks
 - Cervical spondylosis
 - C2-C3 facet joint (third occipital headache)
- Temporomandibular joint injury
- Occipital neuralgia
- Migraine with and without aura
- Cluster headache
- Dysautonomic cephalgia
- Supraorbital and infraorbital neuralgia
- Hemorrhagic cortical contusions
- Subdural or epidural hematomas
- Low cerebrospinal fluid pressure syndrome

Brainstem Symptoms

- Dizziness
- Vertigo
- Tinnitus
- Hearing loss
- Blurred vision
- Diplopia
- Convergence insufficiency
- Light and noise sensitivity
- Diminished taste and smell

Psychologic and Somatic Complaints

- Irritability
- Anxiety
- Depression
- Personality change
- Posttraumatic stress disorder
- Fatigue
- Sleep disturbance
- Nausea/vomiting
- Decreased appetite

Cognitive Impairment

- Memory dysfunction
- Impaired concentration and attention
- Slowing of reaction time
- Slowing of information-processing speed

complaints of pain and dizziness that may limit activity. With better understanding of the mechanisms of pain associated with these injuries, the therapist may be better able to address the specific issues.

Headache is a common complaint reported to the therapist treating soft tissue dysfunction associated with trauma. Often there are complaints of excessive fatigue, eye strain, inability to concentrate, depression, and dizziness in these same patients. The nature of the headache should be evaluated to help determine the appropriate intervention. Working with a medical practitioner to get the appropriate medication can be a part of the therapist's intervention.

Continued.

Trigger point treatment of the pericranial and cervical tissue has been used to help control the pain. As the understanding of the relationship of the pericranial structures and central modulation of pain emerges, combining treatment of soft tissue and use of CNS-mediating drugs may bring more relief of these disabling symptoms.

Medication Overuse Headaches

Frequent intake of medications used for headache can lead to chronic headache. It appears that all drugs used for the treatment of headache may lead to chronic headache. Triptan overuse is common, and the time of onset of chronicity is shorter compared to ergots and analgesics. Women appear to be more prone to overuse than men. Migraineurs are most susceptible to headache related to excessive medication. If a migraineur is taking an analgesic for arthritis, for example, there can be an increase in migraine; this is not seen as significantly with CHs, and an individual taking medication for arthritis who has no history of headache does not begin to have headaches because of the medication.¹⁰⁵ Sensitization of receptors and changes in receptor thresholds may explain the phenomenon. Circumstances in which there is more consequence to the headache, as before an important date or during a period of work that should not be interrupted, and the instruction to take the medication as early as possible at the onset of a headache may lead to excessive use of medications.⁸⁰

Overuse of analgesic medications resulting in analgesic-rebound phenomenon is common when the headache type has not been identified appropriately.¹⁰⁶ Intervention by therapists to relieve the mechanical contribution to cervical pain is critical, in addition to pharmacologic treatment.¹⁰⁷

Headaches in the Elderly

Primary headaches continue into the later decades of life, with a decrease noted in migraine, cluster headache, and hemicrania. New-onset headaches in the elderly that are related to another disorder are more common than in the young. Increased use of nitrates for control of angina can cause headache. Chronic daily headache often persists into old age. Hypertension and atherosclerotic and hemorrhagic cerebrovascular disease can produce new-onset headache in the elderly.²⁸ Diseases of the aged such as chronic obstructive lung disease can cause dull, throbbing headaches. Anemia, the hypercalcemia related to malignancy, and chronic renal failure can all be a cause of headache. There is a dull headache that can be a part of the pathology of Parkinson's disease and can clear with the use of antiparkinson medication. The incidence of intracranial disease increases with age, with tumors and subdural hematomas creating space-occupying lesions (see Chapters 30 and 32). Cervical spondylosis and TMJ related to loss of teeth increase with age and may be a cause of headache pain. Depression, which is often comorbid with headache, may be underreported in the

Box 37-5

SYMPTOMS RELATED TO TEMPORAL ARTERITIS

- Presence of pain or tenderness around the temporal arteries
- Scalp tenderness
- Pain with chewing (i.e., jaw or tongue claudication)
- Diplopia, which is generally thought to result from extraocular muscle ischemia rather than from cranial neuropathy
- Transient visual loss as a result of optic nerve or retinal ischemia

aged, and when the headache is treated with antidepressants, both conditions are improved.

Temporal Arteritis

Headache is the most common symptom in temporal arteritis, and this diagnosis should be considered in all adults older than 50 years who have headache or facial pain. Symptoms of vasculitis and vascular insufficiency are common relating to extracranial carotid circulation. Box 37-5 gives the typical complaints related to temporal arteritis.

Temporal arteritis may be related to widespread rheumatologic involvement such as polymyalgia rheumatica. These symptoms may be rather nonspecific and include malaise and easy fatigability, weight loss, anorexia, and unexplained fevers, as well as proximal myalgias.

The headache of temporal arteritis classically is located over a branch of the superficial temporal artery and is described as a dull ache that persists throughout the day. It may be accompanied by tenderness of the artery and overlying scalp. If severely affected, the artery may be nonpulsatile.

Sedimentation rate should be measured to rule out temporal arteritis in individuals older than 60 years who have headache or facial pain, unless the pain obviously results from another cause. Visual loss may result and can be avoided with early diagnosis and aggressive treatment. Findings from a temporal artery biopsy can confirm the diagnosis. Unless otherwise contraindicated, corticosteroid therapy should be instituted, pending results of the biopsy.

Sinus Headaches

The diagnosis of pain that results from acute sinus inflammation is rarely difficult. Often a prior history of sinus inflammation or respiratory allergies is elicited. In general, the pain is of low to moderate intensity and is present on a daily basis. The pain usually is localized to the frontal or maxillary area, and there is tenderness to tapping over the affected sinus. The pain is often worsened by bending forward and may be triggered by blowing the nose or sneezing. Symptoms of nasal stuffiness are usually present, and mucopurulent drainage from the nostrils may be seen. If the nasal passages are blocked, use of a nasal decongestant can be useful diagnostically and often results in discharge. In doubtful cases, a simple plain film

of the sinuses or an opinion from an otolaryngologist should be obtained.

Postdural Puncture Headaches

Another disabling headache can occur after lumbar puncture or with epidural anesthesia. If a myelogram is performed, it can trigger this type of headache. The headache is due to leakage of cerebrospinal fluid and changes in the pressure within the skull. The headache is usually self-limiting but can last 3 to 5 days. An individual with this type of headache must lie still, because movement, including Valsalva maneuver, increases pain. Hydration helps, in combination with analgesics and antiemetics. Intravenous caffeine sodium benzoate constricts cerebral

blood vessels. An epidural blood patch may be necessary. Blood taken from the peripheral vein and put into the epidural space clots and forms a patch. There is increased risk of pain, infection, and spinal cord compression. An epidural blood patch cannot be used in persons taking anticoagulants. Aspirin is not used as an analgesic due to its anticoagulant properties.¹¹

References

To enhance this text and add value for the reader, all references are included on the companion Evolve site that accompanies this textbook. The reader can view the reference source and access it on-line whenever possible. There are a total of 107 cited references and other general references for this chapter.

CHAPTER 38

Vestibular Disorders

KENDA S. FULLER

VESTIBULAR DISORDERS

Definition and Overview

The organs and nervous system circuitry make up the vestibular system, which detects both the forces created by gravity and the forces generated as we move. The ability to maintain clear vision during head motion and to determine head position or speed and direction of movement is provided by the vestibular system, which generates appropriate postural reflexes to maintain balance.³⁴ Until the system fails, the activity of the vestibular system is noticed only in particular circumstances when it is stimulated beyond normal, such as during a long spin or an amusement ride. Vestibular disorders, with the exception of aggressive form of neoplasm, are not life threatening but can cause significant disability.

Because the vestibular system provides information about orientation in space, disorders of the vestibular system can cause a devastating abnormal sense of movement, visual instability, and loss of balance. The vestibular system works as part of the sensory triad for postural stability and must be integrated with somatosensory and visual input to determine appropriate postural strategies. The disorders of the vestibular input cannot always be assumed to be an actual loss of function, or even hypo-function, as many individuals complaining of dizziness and imbalance have normal tests of vestibular function but do not make appropriate use of the information within the environmental context.

Incidence and Etiologic and Risk Factors

The vestibular system can be involved in many conditions either directly or indirectly; therefore it is difficult to determine the incidence within the general population. Even the most common causes of lesions vary. Box 38-1 gives common causes of vestibular disorders. Although dizziness is common in all age groups, the frequency of dizziness increases with age. Aging has a significant effect on vestibular function but may not be the primary cause of imbalance. Hair cell loss occurs with aging, particularly in the ampulla. It has been estimated that neuronal loss in the vestibular nuclei occurs at a rate of approximately 3% per decade from the age of 40 years.³⁵ Nearly 10% of people older than 45 years visit their physicians with this complaint. Most of this group

is older than 75 years.¹⁶ Changes in the vestibular system happen during the normal aging process, but the problems reported to physicians are most likely related to a pathologic process. Dizziness is reported more frequently by women than by men.²¹

A dysfunction can affect the system at any level (Box 38-2). Disorders of the labyrinth, or inner ear, can affect the sensory end organs, resulting in abnormal input into other levels of the vestibular system. Damage to the peripheral vestibular nerve can be from a neurologic pathology, mechanical deformation from a nonneurologic pathologic condition, or trauma to the structures surrounding the nerve. A central deficit can be the result of damage to the area in the brainstem or cortex that processes vestibular information. Often the signs and symptoms help to indicate the location of the lesion or level of dysfunction.

At its most peripheral level, the end organ of the vestibular system is primarily a mechanical system designed to identify movement of the head. It consists of a fluid-filled labyrinth and is bordered laterally by the middle ear and medially by the petrous portion of the temporal bone. The pathologic condition at this level is related to the structures involved. Figs. 38-1 and 38-2 show the mechanism of the vestibular system and the relationship of the vestibular system to the nearby structures of the brain and bony structures.

The semicircular canals are ring-shaped, fluid-filled structures oriented in three dimensions that provide sensory input about head velocity, or angular acceleration of the head. This is accomplished by movement of the fluid (endolymph) in the direction opposite to the head movement. Fig. 38-3 represents the relationship of the movement of the head and the resulting effect on the direction of the endolymph movement. The ampulla is deflected away from the direction of head movement by the movement of the endolymph. Figs. 38-4 and 38-5 show the relationship between the movement of the fluid within the canal, the orientation of the canal to the head, and the ampullar deflection. The speed and direction of the deflection of the hair cells of the ampulla determine the rate of firing of the vestibular nerve. The difference between the rate of firing of each vestibular nerve is interpreted by the brain as to the amount of angular acceleration of the head.⁴² Disorders at this level are most often

Box 38-1**COMMON CAUSES OF VESTIBULAR DISORDER**

- Benign positional vertigo
- Menière's syndrome
- Viral infection
- Bacterial infection
- Perilymph fistula
- Vascular disease
- Neoplasia
- TBI
- Metabolic disorders
- Toxicity
- Allergies
- Mal de débarquement
- Immune disease

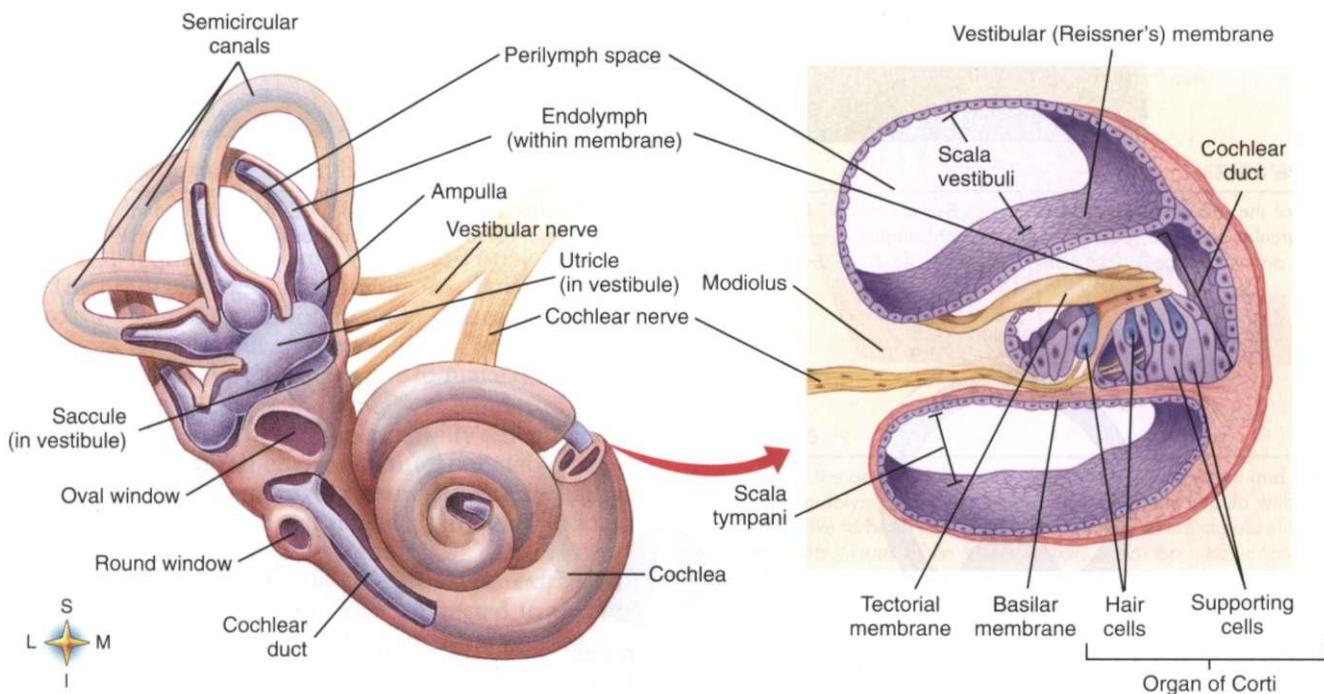
Box 38-2**POTENTIAL LOCATION OF LESIONS THAT MAY AFFECT THE VESTIBULAR SYSTEM**

- Vestibular end organ and vestibular nerve terminals
- Vestibular ganglia and nerve within the internal auditory canal
- Cerebellopontine angle
- Brainstem and cerebellum
- Vestibular projections to the cerebral cortex

related to fluid pressures, changes in the contents of the fluid, and inflammatory or infectious agents that affect the homeostasis of the system. Blows to the head or acceleration-related injuries can cause direct damage to the labyrinthine system. Ischemia in the surrounding vasculature can cause disrupted function.

Both ends of each fluid-filled semicircular duct open into the otolith, which contains the utricle and saccule. A portion of the floor of the utricle and the saccule is thickened and contains hair cells covered with a gelatinous membrane known as the otolithic membrane. Calcium carbonate crystals, or otoconia, adhere to this membrane. Fig. 38-6 shows how the otoconia sit above the hair cells. The weight of the otoconia produces a shear force on the hair cells with acceleration of the head. Information about linear acceleration, the tilt of the head, or the static position of the head with respect to gravity from the movement of the hair cells is transmitted to the vestibular nerve. Changes in the position of the hair cell respond to changes in head position in relation to gravity. Fig. 38-7 shows the relationship of the otolith to head positions changes. When this system is not working properly, there is loss of the ability to orient to gravity, and the person complains of bouncing or feels as if he or she were on a ship. It is difficult to orient to the environment when a person is not able to orient to gravity.^{5,23}

The hair cells, or nerve endings in both the semicircular canal and the otolith, work together to determine head position and rate of movement. There is a tonic resting firing rate of the hair cells that is carried through the vestibular nerve. The vestibular nerve transmits these

**Figure 38-1**

Components of the vestibular system and cochlea with distribution of neural connections. (From Thibodeau GA: Anatomy and physiology, ed 6, St Louis, 2006, Mosby.)

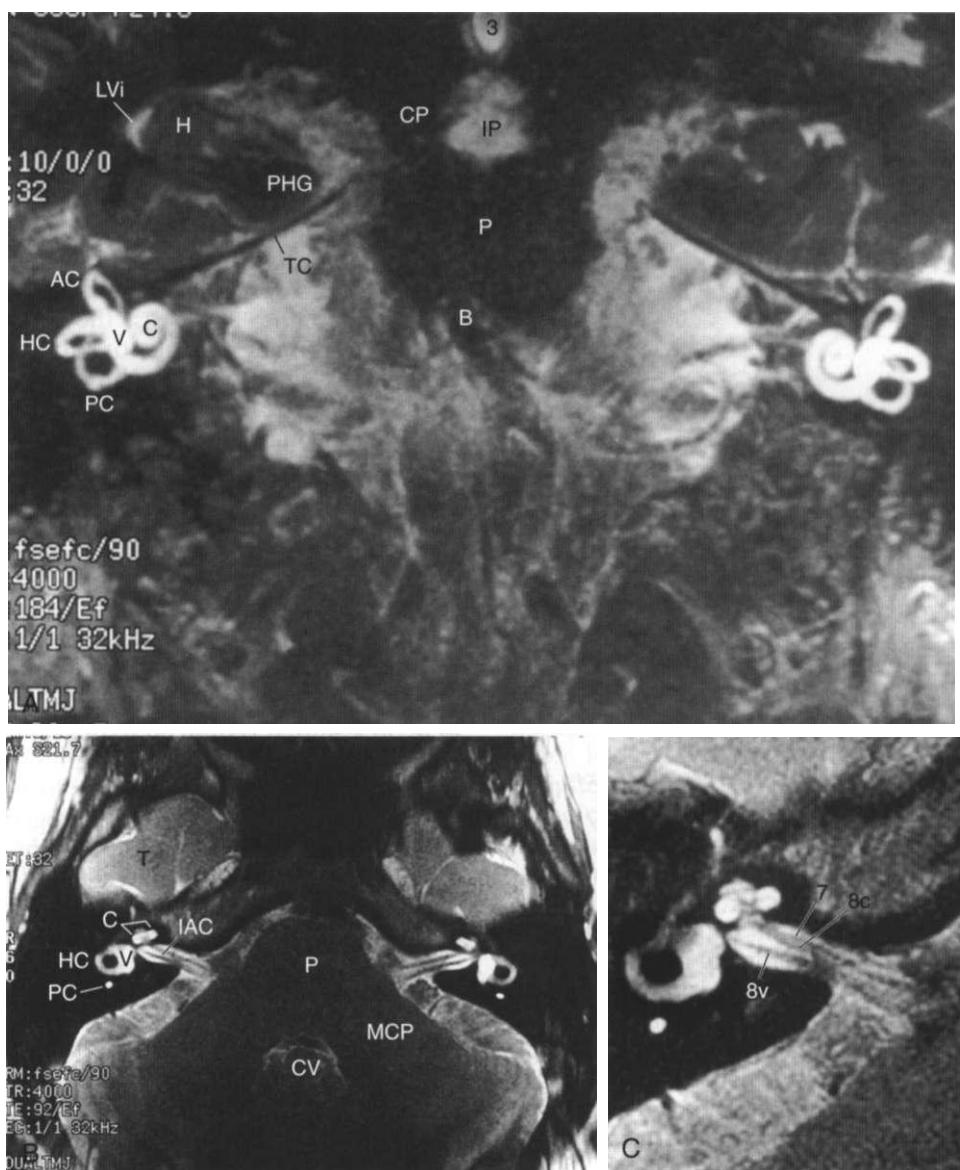


Figure 38-2

MRIs of the labyrinth. **A**, Coronal view. **B**, Axial view. **C**, Enlarged view. AC, Anterior semicircular canal; HC, horizontal canal; PC, posterior semicircular canal; C, cochlea; IAC, internal auditory canal; P, pons; 8v, eighth cranial nerve (vestibulocochlear nerve). (From Nolte J: *The human brain: an introduction to its functional anatomy*, ed 5, St Louis, 2002, Mosby.)

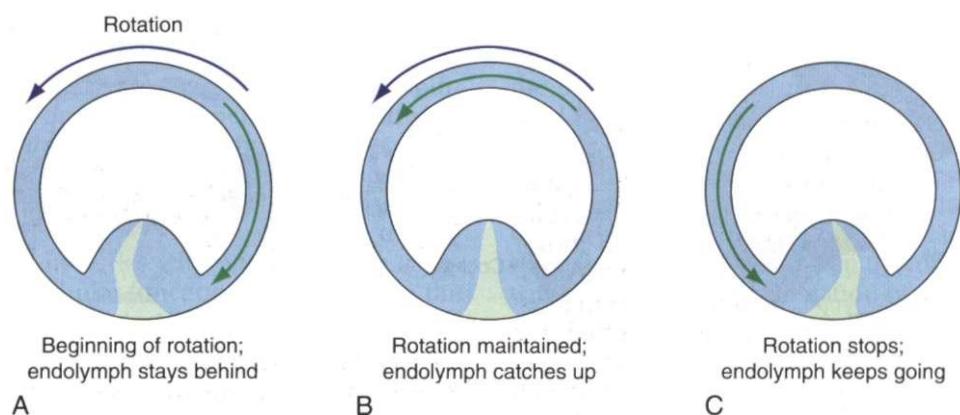
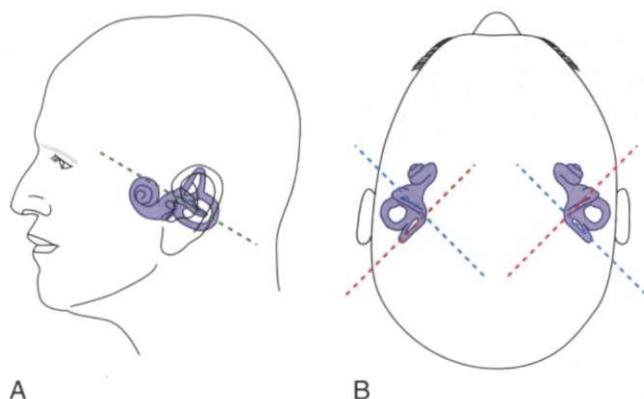


Figure 38-3

Relative movement of semicircular canals. **A**, Beginning of rotation. **B**, During rotation. **C**, End of rotation. (From Nolte J: *The human brain: an introduction to its functional anatomy*, ed 5, St Louis, 2002, Mosby.)

**Figure 38-4**

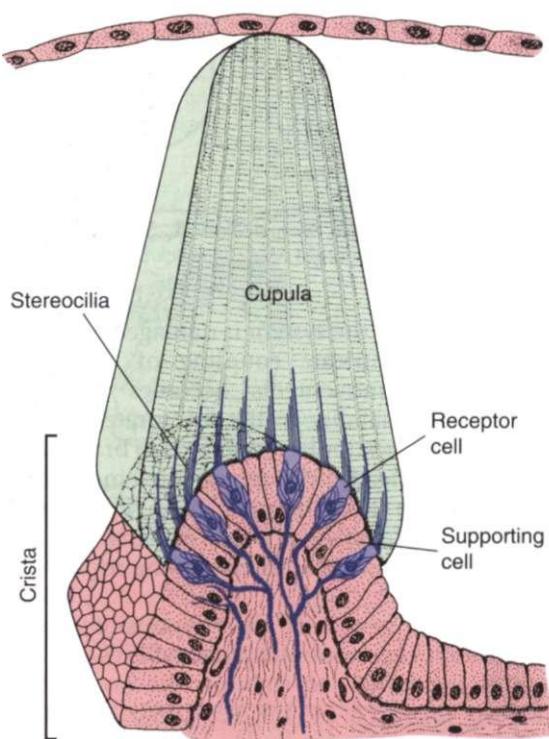
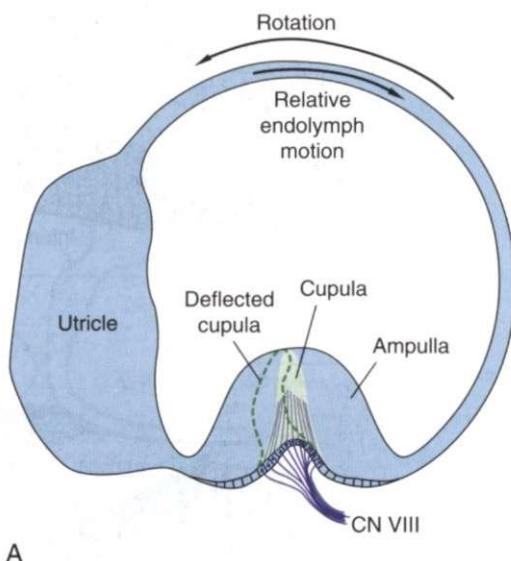
Orientation of the labyrinth. **A**, Side view. **B**, From the top. Labyrinth is enlarged relative to the head in this drawing for clarity. [From Nolte J: *The human brain: an introduction to its functional anatomy*, ed 5, St Louis, 2002, Mosby.]

afferent signals from the labyrinths, extending through the internal auditory canal and entering the brainstem at the pontomedullary junction. Somatosensory, visual, tactile, and auditory information is also processed in the vestibular nuclei.^{13,27} The brain uses the combined input in determining orientation within the environmental context.

When there is disruption of the vestibular nerve on one side causing loss of tonic firing, it results in abnormal information relayed to the brain about the position or movement of the head. The brain, as it compares the two sides, then interprets the abnormal input as movement when the head actually is at rest. This is the phenomenon of vertigo, or the illusion of turning or spinning. The individual will sense that the head is turning or the room is rotating.

The brain, however, has a remarkable ability to be able to adapt this system, and if the lesion is stable or non-fluctuating, the brain will learn to interpret movement based on the information that is available from the intact vestibular nerve on the opposite side. This adaptation depends on intact function of the central nervous system (CNS) and requires time and available visual information. The brain can then compare the vestibular information to the visual information in order to recalibrate. This adaptation takes about 2 weeks in an intact nervous system.^{5,10,17} When the CNS is unable to make the adaptation, or it is not adequate for the task at hand, the resulting sensation is one of spinning, especially when the eyes are closed or the visual environment is not stable.³¹

The vestibular system drives eye movement through its connections in the vestibular nucleus through the oculomotor nuclei to the extraocular muscles. This is known as the vestibulo-ocular reflex (VOR). The extraocular muscles are arranged in pairs and connected to the vestibular system so that a pair of canals is connected to a pair of extraocular muscles. This allows conjugate movement of the eyes as a result of head movement. Through the complex connections of the vestibular and oculomotor nuclei, information regarding direction and speed of head movement is directed to the eye muscles so that

**Figure 38-5**

Two views of the ampulla. **A**, View of the semicircular canal and movement of the endolymph and resulting deflection of the cupula within the ampulla relative to head motion. **B**, Enlargement of the cupula showing its components. [From Nolte J: *The human brain: an introduction to its functional anatomy*, ed 5, St Louis, 2002, Mosby.]

they react and move in the opposite direction as the head is moving. This keeps the visual environment in focus during rapid head movements. Fig. 38-8 shows the connection between stimulation of the semicircular canals and eye movement.

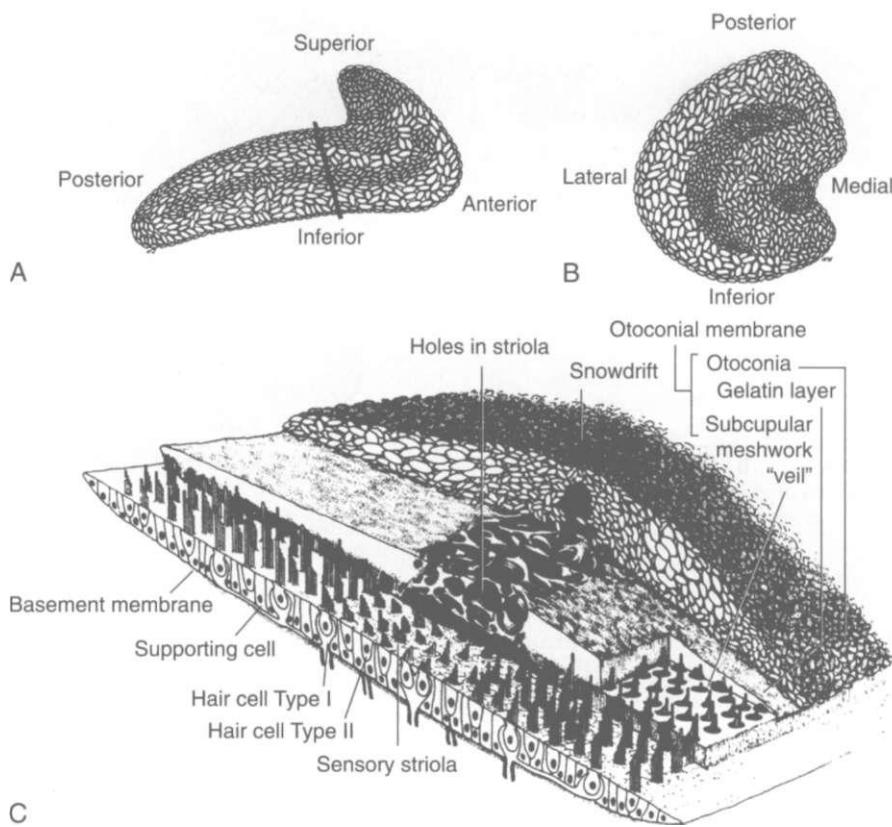


Figure 38-6

Arrangement of otoliths in the two maculae. **A**, Saccule. **B**, Utricle. **C**, Composition of the saccular otoconial membrane in a section taken at the level shown in **A**. (From Paparella MM, Shumrick DA, eds: *Textbook of otolaryngology*, vol 1, Philadelphia, 1980, WB Saunders.)

When one side of the system is damaged or stimulated abnormally, the intricate coupling of the vestibular system and eye movements becomes disturbed.^{17,33} Because of the imbalance in the firing rate between the two sides of the vestibular system, the brain perceives movement of the head, although no movement may have occurred. Through the VOR mechanism, eye movements are triggered as if the head were moving. As the eyes move, the sensation created is one of the room spinning around.

The vestibular system is also responsible for motor output through the vestibulospinal reflexes (VSRs). The purpose of the VSR is to stabilize the body using the information provided by the vestibular system. Three pathways connect the vestibular nuclei to the anterior horn cells of the spinal cord. The lateral vestibulospinal tract, with its connection to the lateral vestibular nucleus, receives the majority of its input from the otoliths and the cerebellum. It is responsible for postural activity in the lower extremities in response to head position changes that occur with respect to gravity.³² The medial vestibulospinal tract from the medial, superior, and descending vestibular nuclei gets its input from the semicircular canals and triggers postural responses with regard to angular head motion. The medial vestibulospinal tract descends only through the cervical spinal cord. The reticulospinal tract gets its input from all the vestibular nuclei in addition to other systems concerned with maintaining balance. These reflexes together provide automatic control

of the activity of the postural muscles in the trunk and limb. Because these tracts are the output for the vestibular system, damage to any part of the system can result in abnormal postural responses to movement.⁴³

The vestibular nuclei are connected to each other in a complex manner across the brainstem. The descending nucleus is a relay between all the other nuclei and also to the cerebellum. The cerebellum receives output from the vestibular nucleus. The vestibular reflexes become uncalibrated and ineffective when the cerebellum is dysfunctional (see Chapter 28).

The brainstem can be affected in many ways. Pathologic processes can be direct, as in mechanical damage of the neurons in brain injury, bleeding disorders, and neoplasia. Hypoxic damage can occur in stroke or other conditions that cause decreased profusion of oxygen into the brainstem. Degenerative diseases such as multiple sclerosis (MS) and Alzheimer's disease can cause abnormal function in the brainstem and its connections.

The vertebral-basilar artery supplies blood to the components of the vestibular system. The posterior and inferior cerebellar arteries feed the CNS. Because there is redundant blood supply via the circle of Willis, ischemia in this area is rare. The anterior inferior cerebellar arteries supply the peripheral mechanism via the labyrinthine, common cochlear, and anterior vestibular arteries.¹³ Disorders that affect the small vessels can cause direct damage to the peripheral vestibular system, or the ischemia can cause direct damage to the vestibular nuclei. Fig. 38-9

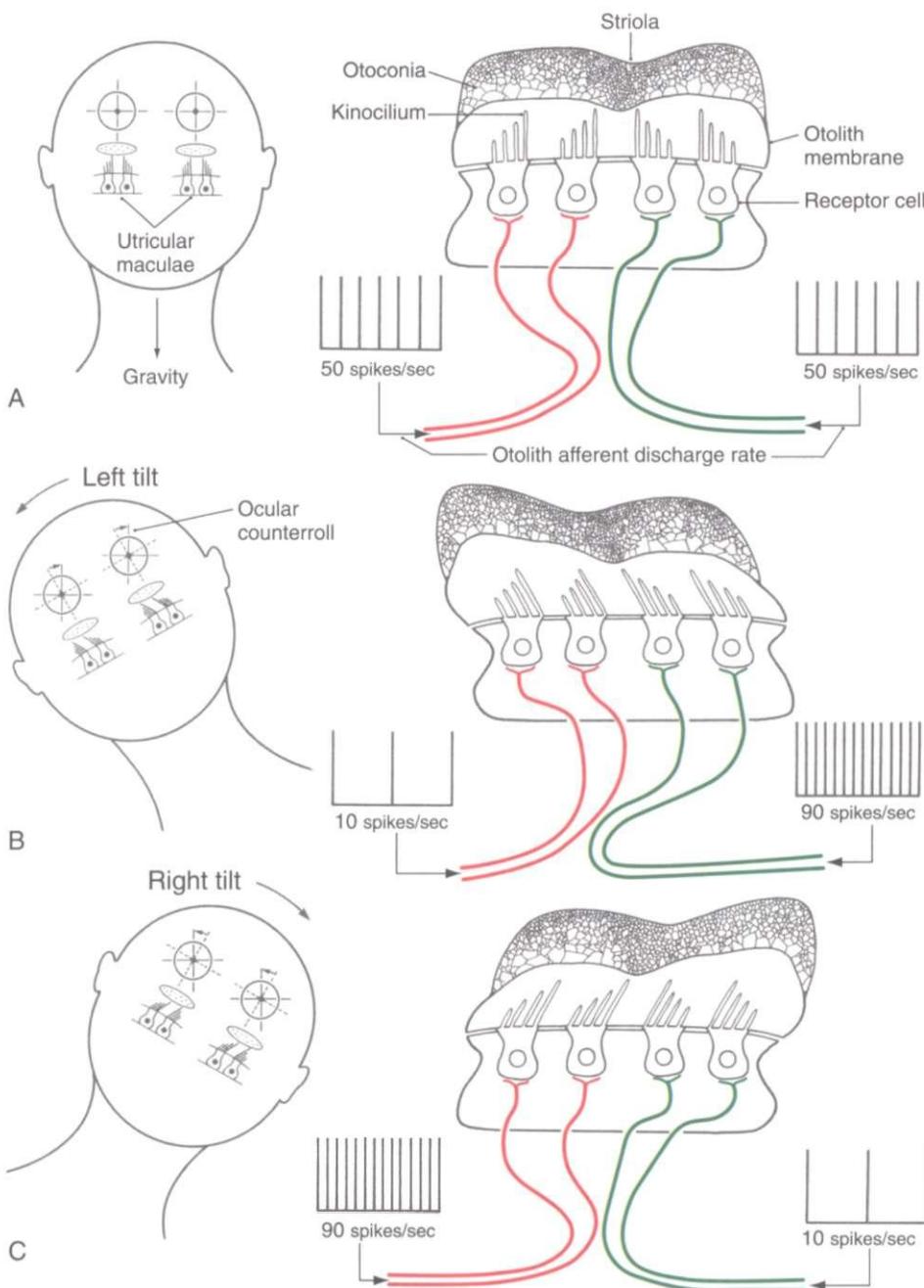


Figure 38-7

Patterns of excitation and inhibition for the left utricle and saccule when the head is tilted with the right ear 30 degrees down (**A**), upright (**B**), and tilted with the left ear 30 degrees down (**C**). The utricle is seen from above and the saccule from the left side. The background color represents baseline activity and black and white represent depolarization and hyperpolarization, respectively. (From Haines DE: *Fundamental neuroscience for basic and clinical applications*, ed 3, 2006, Churchill Livingstone.)

demonstrates the pathways associated with the end organ, vestibular nuclei, fibers of the cerebellum, and the extraocular muscles. This pathway involves inhibition in the process of modulating output to extraocular muscles.

MEDICAL MANAGEMENT

DIAGNOSIS

History. Dizziness is one of the most common clinical complaints when the vestibular system is involved in some way. Dizziness is a broad term for the sensation

that results from the disruption of information from the vestibular, visual, and somatosensory systems. The description of dizziness may reflect the nature of the disorder. The history of the dizziness should give an indication of the cause of the symptoms and lead toward diagnosis. The symptoms as reported can reflect an abnormality in the vestibular system or may indicate some other general medical cause. Often the history is complex, and much insight is gained by listening carefully and allowing the client to provide his or her own thoughts

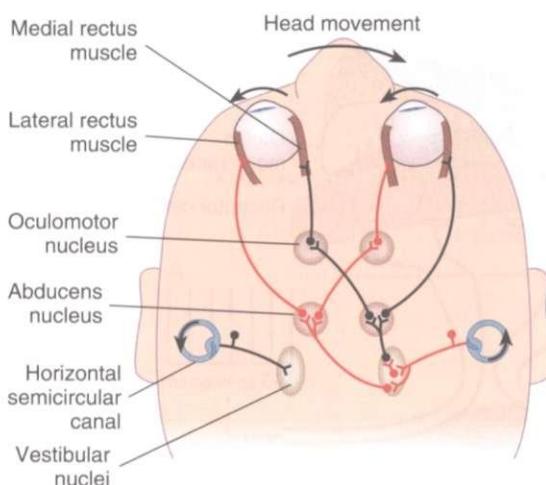


Figure 38-8

Neural connections in the direct pathway for the VOR from excitation of the left horizontal canal (left HC). As seen from above, a counter-clockwise head rotation (head) produces relative endolymph flow in the left HC that is clockwise and toward the utricle. The cupular deflection excites the hair cells in the left HC ampulla, and the firing rate in the afferents increases (inset). Excitatory interneurons in the vestibular nuclei (vest. N.) connect to motor neurons for the medial rectus muscle in the ipsilateral third nucleus (III) and lateral rectus muscle in the contralateral sixth nucleus (VI). Firing rates for these motor neurons increase (bar graphs). The respective muscles contract and pull the eyes clockwise—opposite the head—during the slow phases of nystagmus. Inhibitory interneurons in the vestibular nuclei connect to motor neurons for the ipsilateral lateral rectus and contralateral medial rectus. Their firing rates decrease (bar graphs), and these antagonist muscles relax to augment the eye movement. (From Lundy-Ekman L: *Neuroscience: fundamentals for rehabilitation*, ed 3, Philadelphia, 2007, Saunders.)

on the probable cause. See Box 38-1 for a list of common vestibular causes of dizziness.

Static disturbances come about because of the tonic imbalance of vestibular tone when the head is still. Determining sensation when the head is stable, with the individual sitting unsupported, can give initial cues. A sensation of rotation may represent maladaptation of the input, reflecting a unilateral disorder. Floating, swimming, or leaning may represent abnormal otolith response.

When the complaint is of feeling lightheaded, several conditions should be considered. Presyncope is related to blood flow and may be related to orthostatic hypotension. Anxiety or panic can trigger sensation of lightheadedness, but usually there are other abnormal sensations such as paresthesia or shortness of breath. (See section on somatoform disorders later in this chapter.) When the complaint of lightheadedness is accompanied by a lack of feeling grounded, the integration between the vestibular and somatosensory system may be disrupted.

Head motion-provoked disturbances reflect abnormalities in the ability of the system to identify movement accurately, with abnormal gain of the system. This can be tested through responses to head rotation. The time it takes the brain to resettle after this motion can also reflect lack of CNS recalibration.

Nausea is a frequent complaint associated with dizziness and reflects the stimulation of the medullary centers.

Box 38-3

THREE TYPES OF PHYSIOLOGIC NYSTAGMUS

- Vestibular induced, or spontaneous
- Visually induced, or optokinetic
- Cerebellar, or end-gaze induced

Direct damage to the medulla causes the most severe nausea, as in stroke, but it can also be reflected in traumatic brain injury. Sudden-onset conditions often create a high level of nausea that decreases as the system recalibrates. Canal stimulation can cause nausea; horizontal canal stimulation provokes higher levels of nausea than does the posterior or anterior canal. Headache pain can have a relationship to dizziness and reflects a concomitant disorder of the CNS. Migraine headache and traumatic brain injury may trigger dizziness as a part of the abnormal processing of input through the brainstem.

Measurement of the interaction of the visual and vestibular systems can be accomplished by a variety of tests, with each test providing information that can be compared with the results of other tests to help determine the diagnosis.

The visual-ocular system is measured when looking for a disorder of the vestibular system and, when abnormal, can provide some clues as to where the damage to the system is located. If the motor component of the VOR is damaged, visual and vestibular controlled eye movements are abnormal. If the sensory component is damaged, visually controlled eye movements are usually normal, but vestibular-dependent eye movements are abnormal. Nystagmus is involuntary, rhythmic oscillation of the eye. Nystagmus can be a part of another brain dysfunction in addition to vestibular dysfunction. The nystagmus related to vestibular disorders usually has a particular direction, intensity, and shape. The characteristics can often offer clues to the pathologic condition. Pathologic nystagmus can be spontaneous, gaze evoked, or positional. Box 38-3 describes three types of physiologic nystagmus. Examination for pathologic nystagmus must include observation of the effects of fixation, eye position in the orbit, and head position.⁶²

Disorientation is often described as feeling out of sync with the environment and is usually associated with other deficits, such as short-term memory loss, lack of concentration, and irritability, reflecting the intricate integration with the limbic, hippocampal, and reticular systems through the brainstem.⁶³

Disequilibrium is reported as feeling unsteady or clumsy or as if one were swaying, and a history of falls should be obtained. Fear of falling is also of importance, and the efforts made to avoid a fall can shed light on the degree of relative vestibular spinal responses.

The side effects associated with use of some medications, anemia, hypoperfusion of the brain from postural hypotension, cardiac arrhythmia, endocrine disorders, and hypoglycemia may mimic a vestibular disorder. If the cause is a medical condition or use of medications, the symptoms should decrease when the appropriate condition is treated or when the medication is stopped.³⁰

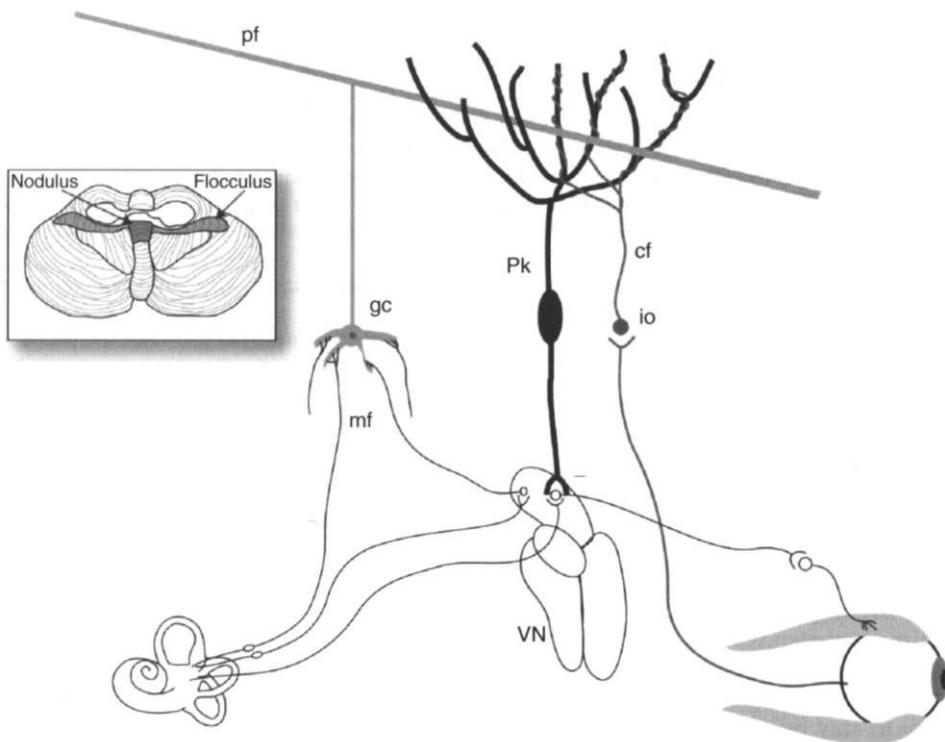


Figure 38-9

Circuitry of the cerebellum involved in modifying the VOR. Inputs from primary vestibular afferents and secondary neurons in the vestibular nuclei (VN) form mossy fiber (mf) inputs to cerebellar granule cells (gc). Parallel fibers (Pf) that originate from granule cells synapse weakly with Purkinje cells (Pk) and cause a high tonic inhibitory output of simple spikes from the Purkinje cells onto secondary vestibular neurons controlling the VOR. Climbing fiber (cf) input from the inferior olive (io) carries sensorimotor error information such as retinal slip. Climbing fibers make extensive and strong synapses onto Purkinje cells. Climbing fiber activity leads to complex spikes in the Purkinje cells, which can alter the efficacy of the parallel fibers' synapses onto the Purkinje cells, a form of learning. (From Cummings CW, Haughey BH, Thomas R, et al: *Cummings otolaryngology: head and neck surgery*, ed 4, St Louis, 2004, Mosby.)

Elderly patients tend to show reductions in the gain of the VOR and smooth pursuit and increases in saccade latencies. Such age-related alterations might interfere with appropriate responses to fast head movements. Initially, reduction of the inhibitory system mediated through the cerebellum can compensate for decreased sensitivity. Over time, the combination of reduced peripheral sensitivity and central inhibition decreases the range in which the system can respond.⁵¹ Most falls in the elderly population result from an accidental slip or trip, often in association with an unsteady gait. Fewer than 10% of falls in the elderly population are a result of an acute attack of vertigo or dizziness. It appears that over time there is selective nonuse of vestibular cues during balance activities. Age-related increases in sway have been found for conditions that involved sway referencing of visual or somatosensory cues.

Video Nystagmography or Electronystagmography. Capturing eye movements related to vestibular dysfunction can be done through use of video goggles (video nystagmography [VNG]) or by surrounding the orbit with electrodes (electronystagmography [ENG]). Most testing now is done using VNG. The angular velocity, amplitude, and frequency of nystagmus can be quantified. The ability to move the eyes quickly from one target to another intended target can be tested for accuracy and is called *saccadic eye movement*. When the target is missed on the first move-

ment, there is a catch-up saccade used to move the eye directly to the target. Catch-up saccades are normal in some cases, such as for jumps made more than 20 degrees, because there is a consistent undershooting. Overshooting rarely occurs in normal individuals. Slow saccadic eye movements can be related to lesions in the central pathways that control their movement. This may be an abnormal motor response of the VOR, or it can be a part of many other degenerative or static lesions that can affect the same part of the system, such as MS or parkinsonian disorders. Saccadic eye movements can be induced by using dots or lights at different amplitudes and asking the client to move the focus quickly from target to target. Accuracy can be recorded using ENG to demonstrate the undershooting and overshooting resulting from disorders affecting control of eye movement.

The ability to follow a target through a trajectory, or smooth pursuit, is another ocular skill that reflects an intact visual-ocular system and vestibulo-ocular output. An acute vestibular lesion will cause an impairment of smooth pursuit, but accuracy should naturally return. Abnormalities of the smooth pursuit system can reflect disorders throughout the CNS. Cerebellar lesions are often a cause of abnormal smooth pursuit. Table 38-1 describes different functions of eye movements.

Smooth pursuit, or tracking of the target, is observed by recording the eye movement as the client attempts to

Table 38-1 Different Functions of Eye Movements

Class of Eye Movement	Main Function
Vestibular	Holds images steady on the retina during brief head rotations
Visual fixation	Holds the image of a stationary object on the fovea
Optokinetic	Holds images steady on the retina during sustained head rotation
Smooth pursuit	Holds the image of a small moving target on the fovea or holds the image of a small near target on the retina during linear self-motion; with optokinetic responses, aids gaze stabilization during sustained head rotation
Nystagmus quick phases	Reset the eyes during prolonged rotation and direct gaze toward an oncoming visual scene
Saccades	Bring objects of interest onto the fovea
Vergence	Moves the eyes in opposite directions so that images are placed or held simultaneously on both foveae

follow targets at varying velocities. Use of a pendulum or computer-generated movement of lighted targets allows a sinusoidal tracking that can be measured by electrooculographic recording. Two measures are usually applied to the recordings. Pursuit gain compares the velocity of the eye to the velocity of the target. Pursuit phase lag refers to the difference in time between waveforms of the target and the eye movement. A person with normal tracking will predict the target motion and make the precise eye movements necessary to stay on target.⁷⁰

Optokinetic nystagmus is the eye movement elicited by tracking a field instead of a target. The purpose of optokinetic nystagmus is to stabilize visually an entire moving visual field. Optokinetic nystagmus is recorded by ENG when a striped drum surrounding the client is moved at a constant velocity at a slow speed of 30 degrees and a fast speed of 60 degrees. Abnormalities of slow components parallel those of smooth pursuits and are related to disorders of the cortex, brainstem, diencephalon, or cerebellum. Abnormalities of the fast components reflect damage, as do abnormal saccades. ENG is used to measure eye movements caused by activation of the VOR and records spontaneous eye movements caused by alteration of the vestibular discharge rate.

The limitations of smooth pursuit and optokinetic nystagmus illustrate the important concept that reflexive sensorimotor systems have optimal operating ranges. Smooth visual pursuit functions are best for low-frequency and slow head movements. Autonomic gravity receptors function best for static and very-low-frequency conditions. These and other reflexes overlap with the vestibular system for part of its operating range, but nonvestibular systems largely break down during quick head movements. The VOR is essential for gaze stabilization during high-frequency, high-velocity, and high-acceleration head movements.

The direction of the nystagmus reflects possible origin. Purely vertical or torsional spontaneous nystagmus suggests a central origin. Nystagmus caused by a central lesion can be in any direction (vertical, oblique, horizontal, rotational), may change direction as gaze direction changes, and is not suppressed by fixation of gaze.⁴⁷

Bithermal Caloric Test. Manipulation of endolymphatic flow in the semicircular canals by creating a temperature gradient is known as *bithermal caloric stimulation*. When warm air is infused in the external auditory meatus, the skin of the horizontal canal is heated, resulting in a

temperature change that is transmitted to the horizontal semicircular canal. The endolymph closest to the canal wall is heated, causing it to become relatively less dense than the surrounding endolymph. The fluid movement that results from the heating of the endolymph deflects the hair cells in the ampulla, simulating head movement; the result is a nystagmus with the fast component moving toward the canal that was stimulated. Cold air or water has created movement in the opposite direction. The mnemonic COWS (cold opposite, warm same) describes the movement of nystagmus related to the temperature of the stimulus used. The caloric examination allows the clinician to evaluate each horizontal semicircular canal separately.⁴¹ VNG recordings during this stimulation can indicate abnormalities in different locations of the vestibular system and brain. Paresis can indicate damage anywhere from the end organ to the entry of the nerve root in the brainstem. A central disorder that affects the nerve root, such as MS or brainstem strokes, can cause paresis recording on caloric testing. Lesions of the cerebellum evoke heightened caloric responses and a suppression fixation deficit. Abnormalities in the characteristics of the nystagmus, such as vertical or oblique responses, are associated with CNS disorders.⁴⁷ Fig. 38-10 describes three ways to invoke nystagmus.

Rotational Chair Testing. Rotational testing of the horizontal semicircular canal is provided by use of a motorized rotational chair. Angular acceleration can be controlled and responses to angular acceleration measured. Persons who suddenly lose vestibular function on one side have asymmetric responses to rotational stimuli. Rotational stimuli are ideally suited for testing persons with bilateral peripheral vestibular lesions because both labyrinths are stimulated simultaneously and the degree of function is accurately quantified. As with lesions of the peripheral vestibular structures, lesions of the central VOR patterns can lead to changes in the gain of rotational-induced nystagmus. Cerebellar dysfunction will result in abnormal amplitudes and arrhythmia.

Subjective Visual Vertical or Horizontal. The ability to recognize vertical and horizontal in a darkened room in relation to gravity indicates intact functioning of the utricle. This is known as *subjective visual vertical* or *subjective visual horizontal*. There is an ocular torsion that occurs after unilateral lesions of the vestibular system that is related to dysfunction of the otoliths. It is believed that this ocular rotation results in the inability, in the acute

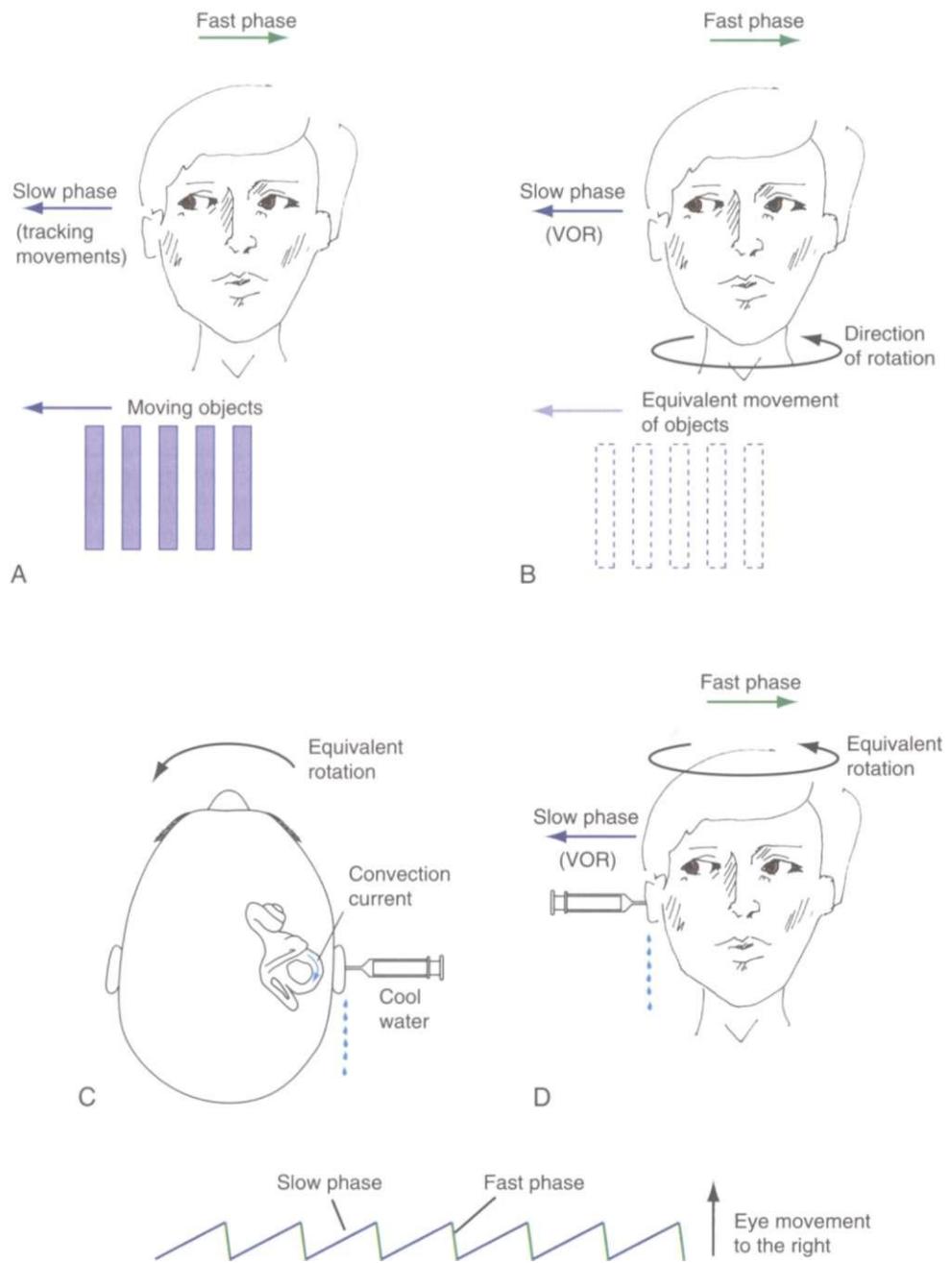


Figure 38-10

Three different ways to cause nystagmus. **A**, Movement of a series of objects to the individual's right causes slow tracking eye movements to the right followed by rapid movements to the left. **B**, Rotation to the left is equivalent to the movement of objects to the right as perceived by the retina. **C**, Cool water or air placed near the horizontal canal via the external ear canal causes the same movement of the endolymph as head rotation in **B**. **D**, Electrical recording of horizontal nystagmus with its fast phase to the left. (From Nolte J: *The human brain: an introduction to its functional anatomy*, ed 5, St Louis, 2002, Mosby.)

phase of vestibular damage, to accurately determine vertical or horizontal visually. This is tested in a darkened room using a light bar that is controlled by the client's relation to vertical or horizontal. Clients are asked to determine when the bar is in either a horizontal or vertical position in relationship to gravity as they sit in a chair. The offset from the true position is often up to 15 degrees. With a compensated lesion, the accuracy improves to 4 degrees but does not ever appear to return to normal. This test will not detect bilateral utricular lesions.^{34,67}

Vestibular-Evoked Myogenic Potentials. Vestibular-evoked myogenic potentials (VEMPs) may be another way to test otolith function in the clinic. The VEMP is generated by synchronous discharges of groups of muscle cells innervated by a single motor unit, or myogenic potential; this response is found in the sternocleidomastoid as part of the vestibulocolic reflex. It appears that the VEMPs arise from stimulation of the saccule, and the saccule is very sensitive to sound. Click-sensitive neurons in the vestibular nerve are the same neurons that respond

to tilts. VEMP testing involves generating a standard sound set and looking for inhibition of the sternocleidomastoid recorded via electromyography (EMG). When one side of the system is dysfunctional there will be a corresponding decrease in EMG activity in one of the sternocleidomastoid muscles. It appears at this time that the VEMP test can provide information in addition to the tests of the semicircular canals to isolate dysfunction in the otolith.

Posturography. Testing of VSRs is performed by posturography, which evaluates somatosensory and visual influences on posture and equilibrium. The clinical applicability of computerized dynamic posturography (CDP) assessment varies with the chief complaints of the individual. CDP has the most clinical value in persons with symptoms of unsteadiness, disorientation, and vertigo in whom the history and physical examination do not suggest an obvious localized cause.⁴⁴ In the *motor control test* the individual's postural responses are recorded during displacement of the support surface using EMG recordings that reflect the activation of the segmental, spinal, and long-loop response pathways. Prolonged motor control test latencies are evidence for abnormality in any one or a combination of components making up the long-loop automatic system and therefore are strong indications for nonvestibular, spinal cord, brainstem, and subcortical involvement.^{14,54,55} The *sensory organization test*, the second component of the CDP, defines six different sensory conditions in which an individual's postural sway is measured. These six conditions vary the amount and accuracy of the sensory information (somatosensation, vision, vestibular input) available to maintain balance.⁵³ Vestibular dysfunction patterns are seen in virtually all persons with bilateral peripheral vestibular deficits. These persons are able to maintain balance when the vision or somatosensation is available, but they free fall repeatedly when conditions require dependence primarily on vestibular input.⁷ Similar vestibular dysfunction patterns are seen in persons with peripheral vestibular lesions and CNS lesions affecting central pathways of the vestibular system. Persons who compensate after a unilateral lesion will have a normal sensory organization test result 2 to 4 weeks after the initial insult.²⁸ An abnormal preference for vision, either alone or in combination with vestibular dysfunction, is most frequently observed in persons with posttraumatic vertigo and unsteadiness. Fig. 38-11 shows sensory conditions during the sensory organization test.

Measurement of movement strategies (ankle, hip, stepping) used to maintain balance is the third component of the CDP. Inappropriate use of ankle movements during large-amplitude sway might be an abnormal adaptation used to minimize head movements and associated stimulation of the vestibular system.⁵³

Electrococleography. Electrococleography is used to record elevation of endolymphatic fluid pressure. It is the recording of acoustically evoked electrical potentials arising from the cochlea and the eighth cranial nerve. An electrode is placed on the tympanic membrane or on the wall of the external ear canal near the tympanic membrane.

		VISUAL CONDITION		
		Fixed	Eyes closed	Sway-referenced
SUPPORT CONDITION	Fixed	1	2	3
	Sway-referenced	4	5	6

Figure 38-11

Diagram of the sensory conditions related to the sensory organization test, a component of posturography. (Courtesy NeuroCom International, Clackamas, OR.)

Imaging. When a central cause is suspected based on clinical findings and vestibular testing, magnetic resonance imaging (MRI) with contrast is indicated. Central brainstem lesions, such as stroke, trauma, or MS, can be identified. Neoplasms, including schwannomas, meningiomas, and metastases, can also be identified. Vascular loop or sling can be identified on MRI. Inflammatory lesions can be identified with MRI, including labyrinthitis and inflammatory lesions of the eighth cranial nerve.

TREATMENT. Treatment of vestibular disorders reflects the spectrum of etiologies. A multidisciplinary approach is critical in the management of symptoms to determine the role of medications, surgical interventions, and therapy. The need for lifestyle changes should be carefully evaluated related to the effect of other interventions.

When symptoms are due to a peripheral vestibular lesion, functional recovery will begin within 2 days to 4 weeks through the adaptive mechanism of the brain.⁷⁰ If the symptoms are severe, sedatives may be given, ideally for the first 24 hours only. Vestibular rehabilitation should begin within the first 3 days if possible. Treatment of recurrent dizziness depends on the nature of the underlying disorder. The goal is to eliminate the frequency and duration of the abnormal sensation of motion and the symptoms of nausea, vomiting, and anxiety.⁴

If the central adaptation process is inadequate, vestibular suppression can be helpful but must be used judiciously. Benzodiazepines, antihistamines, or anticholinergic agents are used frequently. Serotonin 5-HT receptor antagonists have been reported to be of some use.

Surgical intervention is considered when the symptoms are unrelenting and the underlying condition is determined but is unresponsive to other medical measures. Local application of gentamicin to selectively destroy the end organ is often used rather than ablative surgery. Surgery is indicated with neoplasia (see Chapter 30) perilymph fistula, persistent benign paroxysmal positional vertigo, hydrops, and vascular loop. These conditions are covered later in this chapter.⁴⁶

PROGNOSIS. When there is a unilateral lesion in the peripheral vestibular system and the CNS is intact, recovery of function is possible. The recovery related to the static imbalance, the difference in the tonic firing rate within the vestibular nuclei, is spontaneous. The spontaneous nystagmus at rest resolves, and there is no longer a sensation of movement at rest. The recovery of dynamic disturbances, reflecting the relationship of the coupling of the two sides of the system during movement, requires visual experience. The visual experience allows the brain to recognize the error of the system through the retinal slip, causing oscillopsia that occurs immediately after the lesion. It is this sensory mismatch provided by the visual system that allows the CNS to recognize the need for adaptation and drives the readjustment of motor reflexes.⁴⁴ There is a limitation to this recovery, and the system is able to recover function related to slow to moderate movements only. The VOR should return to near normal within 2 to 6 weeks for slower movements, so that motion of the head during typical activities no longer disrupts vision or causes nausea. However, there will continue to be an abnormal reaction to rapid head turns. Recovery is minimized when the response of the individual experiencing the symptoms is to avoid the provoking motions and the CNS is never given the opportunity to adapt to the asymmetric firing patterns of the peripheral vestibular system.

Complete bilateral loss is relatively rare; more often there is decreased function in both sides, often to a different degree on each side. Rehabilitation can provide adaptation to whatever degree possible and compensation for the remaining loss of function.⁴⁵ In the instance of complete bilateral dysfunction, substitution of the other intact sensory inputs for balance is required. The use of visual and somatosensory inputs to substitute for loss of vestibular function is possible when the environment provides adequate cues, as in well-lighted environments and firm, level surfaces. Substitution of small saccades in the direction opposite to head rotation can augment inadequate gain. Smooth pursuit accuracy can improve, and predictive strategies can improve gaze stability. More accurate use of spatial localization is reflected in the ability to imagine the location of stationary targets.⁷⁰

The VSRs are slower to return, and the individual may continue to experience instability when turning quickly or walking on uneven surfaces or in the dark for weeks or months after the insult.

Recovery rate in a central vestibular system disorder causing dizziness or disequilibrium depends on the nature of the lesion and the concomitant neurologic dysfunction. If vertigo is part of a progressive disease, the prognosis is less optimistic than if it is part of a transient disorder, such as a minor vascular incident. A disease such as MS can cause episodes of symptoms and progressive dysfunction with poor adaptation and compensation because of the other damage within the CNS.⁸ In a disorder of the central as well as the peripheral vestibular system, as in head injury or multisensory disorders, the recovery is significantly reduced.⁵⁸ Recognizing and treating the individual components will successfully enhance the recovery.

Benign Paroxysmal Positional Vertigo

Episodic, intense vertigo related to head position is most often a benign disorder called benign paroxysmal positional vertigo (BPPV), also known simply as benign positional vertigo (BPV). It is considered a benign condition, since it is not the sign of a disease process but a mechanical disorder of the labyrinths.

Incidence and Risk Factors. BPPV is the most common cause of vertigo seen by otolaryngologists, representing 20% to 40% of patients with peripheral vestibular disease. The incidence is difficult to estimate given the benign, typically self-limited course of the disease. It is thought to vary from 10 to 64 cases per 100,000 persons. Affected women outnumber men by a ratio of 1.6:1. Involvement in more than one canal is found in 20% of cases. Spontaneous remissions are common, but the disorder can recur in up to 40% of the cases. The condition may trouble the individual intermittently for years, but in this condition a close examination of potential causes will often identify an underlying disorder, and recurrences decline when the disorder is managed. Increased fluid pressure within the labyrinth may dislodge otoconia (see discussion of endolymphatic hydrops below). Migraine-induced ischemia may be responsible for the release of otoconia. Head trauma or infectious or inflammatory disorders may precede the onset by months or even years. Adverse life events are reported often prior to the onset of BPPV. Aging, perhaps because of the increase of dehydration, can increase incidence.¹⁸

Clinical Manifestations. Typically, a person with BPPV will complain of brief episodes of vertigo precipitated by rapid head movement in a specific direction. The vertigo follows getting into bed and lying down. Often the first report is that of waking up suddenly in the night with the room spinning. Bending, looking up to take an object off a shelf, tilting the head back to shave, getting a haircut, or turning the head rapidly while backing up a car can trigger symptoms. These single bouts of vertigo are frequently clustered in time and separated by remissions lasting months or more. Lightheadedness, worsened by head movement, is common and balance problems develop over time when the crystals are not removed from the canals. Symptoms occur suddenly and typically last 20 seconds, but not more than a minute. The subjective impression of attack duration reported is frequently longer than the actual period of dizziness.^{25,36} Box 38-4 gives typical complaints related to BPPV.

Pathogenesis. The otoconia in the otolith can become loose, clump together, and form densities known as canaliths. Canaliths move through the fluid and become problematic when they float into the semicircular canals. There is potential for them to go into any of the three canals, but most often they will move into the posterior canal because of the relationship of the posterior canal to the otolith. The posterior canal is placed in the vertical position when the body is supine and the head is extended beyond neutral and rotated 45 degrees to the same side. The canaliths drift out of the otolith through the opening to the posterior canal and begin moving through the endolymph in the semicircular canal, causing drag on the endolymph and creating cupular deflection, or move-

Box 38-4**COMMON CHARACTERISTICS OF BENIGN PAROXYSMAL POSITIONAL VERTIGO**

- Episodic sensation of intense vertigo with head position changes
- Sensation stops after 20 to 30 seconds in static position
- Nausea with vertigo with reports of spinning inside the head
- Autonomic changes such as sweating, feeling like passing out
- Sensation of movement of the environment and blurred vision
- Reports of disequilibrium during typical activities
- Waking up dizzy at night after rolling over in bed
- Symptoms during head movement or bending forward during typical activities

Box 38-5**CANALITHIASIS OF BENIGN PAROXYSMAL POSITIONAL VERTIGO**

- The canalithiasis mechanism explains the latency of nystagmus as a result of the time needed for motion of the material within the posterior canal to be initiated by gravity.
- The nystagmus duration is correlated with the length of time required for the dense material to reach the lowest part of the canal.
- The vertical (upbeating) and torsional (superior poles of the eyes beating toward the lower-most ear) components of the nystagmus are consistent with eye movements evoked by stimulation of the posterior canal nerve in experimental animals.
- The reversal of nystagmus when the patient returns to the sitting upright position is due to retrograde movement of material in the lumen of the posterior canal back toward the ampulla, with resulting ampullipetal deflection of the cupula.
- The fatigability of the nystagmus evoked by repeated Dix-Hallpike positional testing is explained by dispersion of material within the canal.

merit of the ampulla. This movement of the cupula is interpreted in the brain as movement of the head. There is an intense sensation of rotation. As the canaliths reach the bottom of the semicircular canal, they slow down and stop moving, stimulation of the vestibular nerve ceases, and the sensation of movement stops. This typically takes about 15 to 60 seconds. Canalithiasis of the posterior semicircular is the most frequent cause of BPPV. Box 38-5 describes the phenomenon of canalithiasis.

A less common variant of BPPV is cupulolithiasis. It is believed that the otoconia can become adherent to the ampulla. In this instance there is an inappropriate deflection of the hair cells with movement, causing vertigo and nausea. The symptoms will begin immediately with movement of the head in the provoking position and will persist as long as the head is held in the position. The symptoms may decrease slightly because of adaptation of the CNS and therefore may mimic the fatigue of canalithiasis.³⁸

The horizontal or semicircular canal has been identified as the affected structure in about 17% of cases.

Cupulolithiasis, either occurring independently or in combination with canalithiasis, is more likely to be involved in the etiology of lateral canal BPPV than is the case for posterior canal BPPV.

MEDICAL MANAGEMENT

DIAGNOSIS. The diagnosis of BPPV is made based on a suggestive history and report of current clinical manifestations as described above. The classic eye movements are found when performing the Dix-Hallpike maneuver. The maneuver is repeated with the head in the opposite direction.

Dix-Hallpike Maneuver. Diagnostic criteria for BPPV include whirling vertigo induced by a specific head position or movement. There is a burst of positioning nystagmus that is rotary with latency of symptoms on placing the head in a provoking position; the nystagmus is *fati-gable*, that is, it diminishes with repeated positioning and lasts less than 60 seconds.

The Dix-Hallpike maneuver, or passive movement of the head from the upright position to one with the head hanging extended and rotated to 45 degrees, is the standard test performed to establish the diagnosis of BPPV. When the head is in this position, the clinician observes the eyes for evidence of nystagmus. It may take at least 20 seconds for the nystagmus to occur, and the nystagmus should fatigue within 60 seconds. The nystagmus occurs when the affected ear is placed down. Fig. 38-12 shows the testing positions. The pattern of response is torsional nystagmus that is identified by rotation of the superior poles (the center, vertical component of the iris) toward the involved ear. This is accompanied by an upbeat nature to the nystagmus. There is a latency of onset of nystagmus. Duration of nystagmus is less than 1 minute. Vertiginous symptoms are reported as a sensation that the room is spinning, or the individual is spinning or falling out of control. Symptoms often recur, and the nystagmus beats in the opposite direction on return of the head to the upright position. There are also reports of BPPV affecting the anterior semicircular canal, but they are found infrequently. The nystagmus expected in cases of anterior canal BPV would be torsional, such that the superior poles would beat to the right but would have a downward nature.¹⁸ Although a positive test is pathognomonic for BPV, a negative test indicates only that BPV is not active at that movement.

To test for BPPV in the horizontal canal, the head is first brought to the supine position resting on the examining table or is slightly flexed. The head is then turned rapidly to the right so that the patient's right ear rests on the table. The nystagmus with lateral canal BPV is horizontal. It often begins with a shorter latency, increases in magnitude while maintaining the test position, and is less susceptible to fatigue. Dizziness is likely to occur with head rotation in both directions, but the direction that causes the most intense horizontal nystagmus or is the most symptomatic is considered the side involved. If a brief period of dizziness is experienced during the Dix-Hallpike maneuver as the head is approaching the full supine position, and there is no evidence of posterior or anterior canal BPPV, there is a possibility of horizontal canal involvement, which should be further investigated.

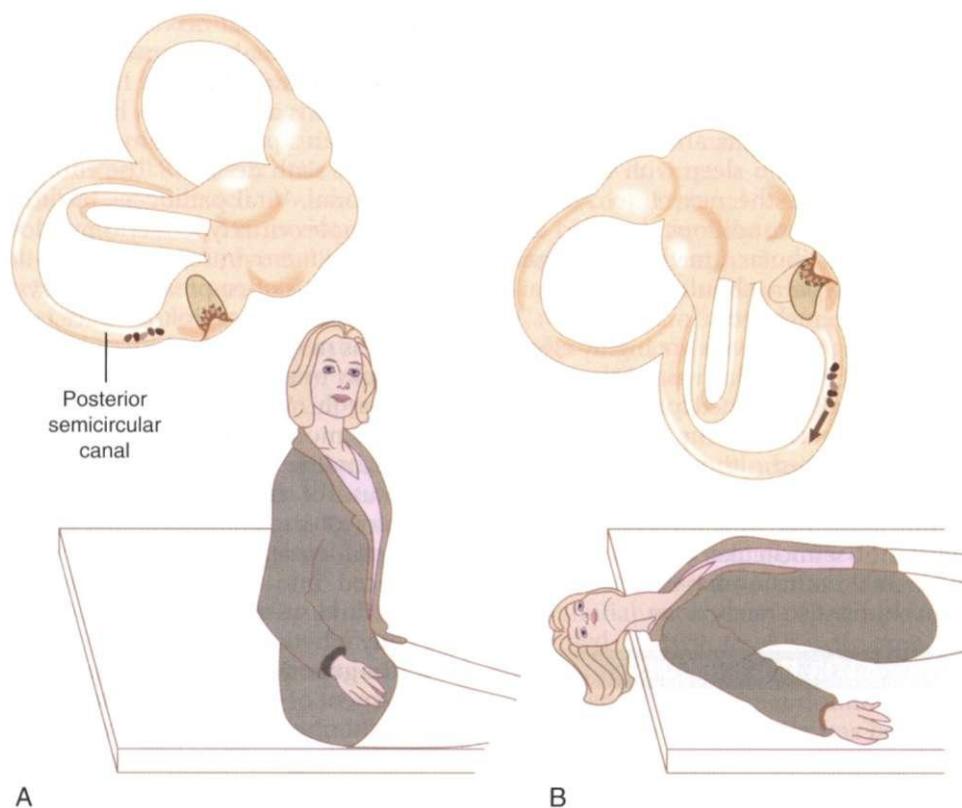


Figure 38-12

The Dix-Hallpike maneuver. **A**, Starting position with head rotated toward the side to be tested. **B**, Lowering the patient's head backward and to the side allows debris in the posterior canal to fall to its lowest position, activating the canal and causing eye movements and vertigo. (From Lundy-Ekman L: *Neuroscience: fundamentals for rehabilitation*, ed 3, Philadelphia, 2007, Saunders.)

Horizontal canalithiasis can also occur, producing symptoms provoked by rolling in the supine position. Horizontal nystagmus may occur in either direction but will be greatest when rolling the head to the involved side.^{38,57}

Another form of positional dizziness that must be ruled out in making the diagnosis of BPPV is central positional vertigo. Nystagmus that is sustained and not suppressed by visual fixation reflects a central lesion. These findings can be indicative of ischemia of the pontomedullary brainstem or another part of the central vestibular pathway. Cerebellar involvement will cause dizziness in positions of supine, head rotated left with extension, and head rotated right with extension. Pure downbeating nystagmus in either the sitting or supine position may indicate an infratentorial disorder. This can be related to vascular insults such as Arnold-Chiari malformation, stroke, or subdural hematoma. MS lesions are common in this area and are associated with the progressive forms. Unilateral lesions, perilymphatic fistula, superior semicircular canal dehiscence, or middle ear problems can cause positional dizziness and are discussed later in this chapter. Intermittent dizziness that is made worse with position changes could also be part of the aura of migraine.

It should also be noted that it is typical to experience dizziness with the head pitched backward, as it puts the otoliths at an angle that is not a part of daily activity.

Orthostatic hypotension can also create a brief episode of dizziness when going from supine to standing.

TREATMENT. Several approaches have been developed to treat persons with BPPV, including maneuvers or positions that attempt to dislodge the provoking substances and move them through the canal system back to the otolith, known as canalith repositioning procedures or the liberatory maneuver. Individuals can also be instructed in exercises (e.g., Brandt's) to try to habituate to the stimulus. The use of gravity to move canalith debris out of the affected semicircular canal and into the vestibule is a safe and reliable method. Even patients who have been symptomatic for years can be cured. The posterior and superior canal maneuver begins with placement of the head into the Dix-Hallpike position that evokes vertigo. Lateral or horizontal canal BPPV responds to positional procedures that involve head rolling with the horizontal canal in the plane and allow gravity to pull the otoconia through the horizontal canal. The maneuvers are repeated until no nystagmus is elicited. In some cases this may take several repositioning sessions. In the case of bilateral BPPV, the canals cannot be treated on the same day, as the maneuvers move the crystals in the opposite direction. Oscillation to the mastoid process can be used in conjunction with the repositioning maneuvers but should be avoided in patients who have had retinal detachment or who might be susceptible to such a detachment because

of high myopia. A low dose of meclizine or a benzodiazepine can be given 1 hour earlier if the patient is unusually anxious or susceptible to nausea and vomiting with vestibular stimulation. After undergoing one of the canalith repositioning maneuvers, patients are instructed to avoid bending over and are told to sleep with the head elevated at least 30 degrees for the next 1 to 2 days. Because BPPV can be related to conditions that also are responsible for abnormal vestibular function, exercises stimulating the vestibular system should follow treatment of BPPV once it has been cleared.²⁸

The canalith-repositioning maneuvers are highly effective in the treatment of BPPV in the vast majority of patients. Chairs that are specially designed for precise positioning or repositioning are in use in some centers. Surgical procedures have been described, although their current use is rare. These procedures include singular neurectomy, which is rarely used due to the potential for sensorineural hearing loss, and posterior semicircular canal occlusion. Posterior semicircular canal occlusion has resulted in the abolition of symptoms but is also rarely done.

SPECIAL IMPLICATIONS FOR THE THERAPIST 38-1

BPPV

PREFERRED PRACTICE PATTERNS

- 5A: Primary Prevention/Risk Reduction for Loss of Balance and Falling
- 5D: Impaired Motor Function and Sensory Integrity Associated with Nonprogressive Disorders of the Central Nervous System—Acquired in Adolescence or Adulthood
- 5E: Impaired Motor Function and Sensory Integrity Associated with Progressive Disorders of the Central Nervous System
- 5F: Impaired Peripheral Nerve Integrity and Muscle Performance Associated with Peripheral Nerve Injury

Canalith repositioning is effective as performed by the therapist. When the individual has had a period of dizziness, and especially when the BPPV has recurred or has been persistent over years, there is a high incidence of maladaptation of sensory integration for balance, resulting in somatosensory and visual dependence for balance. There is imbalance when the brain is using primarily the vestibular system for balance. Although the individual no longer complains of dizziness, there may be imbalance in low light or when there are compliant surface conditions. Visual motion hypersensitivity is common and responds well to intervention. Evaluation for BPPV should include the balance system, and intervention for balance disorder should follow clearing of BPPV.

Infection

Acute unilateral vestibulopathy, or vestibular neuritis, is the second most common cause of vertigo. Viral infection is common and usually affects the vestibular nerve unilaterally. (See section above on unilateral lesions.) Vestibular neuritis can be a partial unilateral vestibular lesion, and this partial lesion can affect the superior division

of the vestibular nerve, which includes the afferents from the horizontal and anterior semicircular canals.²⁴

Incidence and Etiology. Incidence is 3.5 in 100,000. Viral infections are common between the ages of 30 and 60 years, with a peak for women in the fourth decade and men in the sixth decade.²³ The etiology of the disease is multifactorial. Viral pathogens include mumps, rubella, herpes simplex virus type 1, cytomegalovirus, and Epstein-Barr virus. Enteroviruses are among the other rare viral causes. Onset is often preceded by a systemic viral illness, such as an upper respiratory tract infection or gastritis. The illness may precede the vestibular dysfunction by up to 2 weeks but can happen within the course of a bout of cold or flu. When illness such as measles, mumps, or infectious mononucleosis is the source, hearing loss may accompany the vestibular symptoms.

The use of antibiotics in general has decreased the incidence of bacterial infections affecting the vestibular system. However, infections still do arise and can be introduced into the vestibular apparatus through the various fluid systems involved or through breakdown of the bony labyrinth.

Pathogenesis. Histopathologic studies have suggested involvement of the superior vestibular nerve and vestibular ganglion, often with little or no involvement of the actual end organ. This is true of vestibular neuritis, in which there is no cochlear involvement. However, many viruses do damage throughout the labyrinth and cochlea. Typically, in vestibular neuritis, the end organ is filled with lymphocytes. Intracytoplasmic particles have been found in the vestibular ganglia. These particles are thought to be dormant forms of a virus that may produce infection, with resultant inner ear disease.³

Persons with bacterial meningitis develop labyrinthitis when bacteria enter the perilymphatic space from the cerebrospinal fluid by way of the cochlear aqueduct or the internal auditory canal. Some bacterial infections result in biochemical irritation of the membranes through a toxic reaction. Both congenital and acquired syphilitic infections produce labyrinthitis as a latent manifestation.

Clinical Manifestations. Unilateral vestibular neuritis causes sudden onset of rotatory vertigo, spontaneous horizontal nystagmus, nausea, and vomiting. Immediately after the onset of unilateral vestibular hypofunction there is intense disequilibrium. There are profound disturbances of position and motion perception. There is a false sense of angular motion (i.e., rotation). With the eyes closed, there is an illusion of spinning, or sense of motion of the body turning on its long axis toward the involved side. When the eyes are open, the illusion is of spinning of the environment in the opposite direction. There is a tonic ocular tilt reaction consisting of head tilt, conjugate eye torsion, skew deviation, and lateropulsion, seen as an abnormal weight shift toward the side of the lesion. This is due to the otolithic hypofunction on the side with the lesion.^{53,54} This sensation of movement at rest should resolve within 2 days.

MEDICAL MANAGEMENT

DIAGNOSIS. The VOR becomes abnormal with unilateral hypofunction. There is loss of gaze stabilization with

head movement. Movement of the head then causes blurring of vision, resulting in dizziness and loss of ability to use visual cues to maintain balance. EMG testing shows a unilateral hypofunction.

A spontaneous mixed torsional and horizontal nystagmus is often present immediately after unilateral hypofunction. The slow phases are directed toward the side of the lesion, and the quick phases are to the intact side. As the eyes are observed, it is the quick phases that are apparent, and therefore the nystagmus appears to be moving away from the side with the lesion. The nystagmus is suppressed by gaze fixation, or looking at a static object.⁵⁴ Peripheral vestibular nystagmus is usually mixed torsional and horizontal in a fixed direction opposite the side of the lesion.

In severe unilateral vestibular hypofunction, passive head shaking for about 20 seconds will elicit a quick burst of nystagmus directed toward the intact end organ. The nystagmus can be suppressed with visual fixation.

TREATMENT. The main groups of drugs used for symptoms of acute vertigo include antihistamines, anticholinergic agents, antidopaminergic agents, steroids, and antivirals such as acyclovir.

Polyneuritis of the seventh and eighth cranial nerves, known as Ramsay Hunt syndrome, can be the result of herpes zoster, causing perivascular, perineural, and intraneuronal infiltration. Other viruses are suspected but have yet to be positively identified. The cortical vestibular projection fibers can be affected by herpes zoster encephalitis, which has a predilection for the temporal lobe.

Recovery of function and resolution of dizziness is accomplished through a program to facilitate the vestibular system. Recalibration, or adaptation of the system, comes by facilitating the integration of somatosensory input and recovery of normal postural responses. See Special Implications for the Therapist: Vestibular Dysfunction in this chapter.

PROGNOSIS. The symptoms slowly resolve over 6 weeks to 3 months, but there can be persistent complaints of imbalance, motion intolerance, and headache related to decrease in the natural motion of the head. Over time, decreased functional use of the vestibular mechanism develops and is related to excess caution on uneven surfaces, dizziness that is more easily provoked, an overall decrease in activity level, or avoidance of activity that may cause dizziness.

Endolymphatic Hydrops and Meniere's Syndrome

Definition and Overview. Endolymphatic hydrops is a disorder relating to the membranous inner ear as a consequence of the overaccumulation of endolymph compromising the perilymphatic space. There is lack of absorption of endolymph in the endolymphatic duct and sac and fluid backs up into the system. Meniere's syndrome is the most common form of endolymphatic hydrops and is characterized by episodic vertigo; fluctuating, sensorineural hearing loss; sensation of fullness in the ears; and tinnitus. Tinnitus is an abnormal sound in the ear usually described as a ringing, buzzing, clicking, or crackling sound. It is often associated with other

abnormal sensations, such as fullness of the ear. Vertiginous attacks are the most debilitating symptom, with intervals of hours to days. Acute attacks can be superimposed on a gradual deterioration in sensorineural hearing in the involved ear, typically in the low frequencies initially. Over time, a reduction in responsiveness of the involved peripheral vestibular system can occur.⁵ Fig. 38-13 shows the fluid mechanism of the inner ear, including the endolymphatic sac.

Incidence and Etiologic and Risk Factors. Female Caucasians are most prone to the disorder. However, diagnostic criteria have varied across epidemiologic studies. These vary from 157 per 100,000 persons in England to 46 per 100,000 in Sweden and 7.5 per 100,000 in France. The peak incidence is in the 40- to 60-year-old age group, with a nearly equal female to male ratio (1.3:1). Estimates of symptoms arising in the opposite ear vary from 2% to 50%. Whether the variability in prevalence rates is caused by differences in environment, genetics, or diagnostic criteria is unclear.⁶³

Familial occurrence of Meniere's syndrome has been reported in 10% to 20% of cases. Genetic inheritance plays a role. The mode of transmission appears variable; however, an autosomal-dominant mode of inheritance with increased penetrance has been documented.

The incidence of Meniere's syndrome is greater in individuals with certain genetically acquired major histocompatibility complexes. Specifically, human leukocyte antigens B8/DR3 and Cw7 have been associated with Meniere's syndrome. The etiology for disease in these individuals may be autoimmune.

The cause of endolymphatic hydrops is multifactorial and may be related to fibrosis, atrophy of the sac, obstruction of the endolymphatic duct, infection, or the vascularity in the region of the inner ear. It can also be caused by otosyphilis, or involvement of the inner ear in collagen vascular diseases. Immune responses are likely within the complex, including the endolymphatic sac, related to allergic reactions and histamine. There may be a predisposing viral infection that may cause the inner ear to be more susceptible to changes in thyroid, sodium, or hormone dysfunction. The deficit may also be related to overproduction of endolymph by the stria vasularis.¹⁹ Posttraumatic endolymphatic hydrops can be observed following a blow to the head, a fall, or flexion or extension injury sustained in an automobile accident.

Pathogenesis. Although the pathophysiologic mechanism involves a disruption of homeostasis of inner ear fluid, the mechanism and the pathophysiology of the symptoms are uncertain. Endolymphatic hydrops may be an epiphénoménon rather than directly responsible for the symptoms. Deficits are related to the volume and pressure changes within closed fluid systems. The increase in the volume of endolymph causes the membranous labyrinth to progressively dilate until the wall makes contact with the stapes footplate and the cochlear duct fills the entire scala vestibuli, causing both vestibular and cochlear dysfunction. Fig. 38-14 shows the cross view of the canal and the changes that can occur with increased pressure. Distension of the otoliths can put pressure on the ampulla, creating the sensation of spinning that is characteristic of acute unilateral dysfunction. The mem-

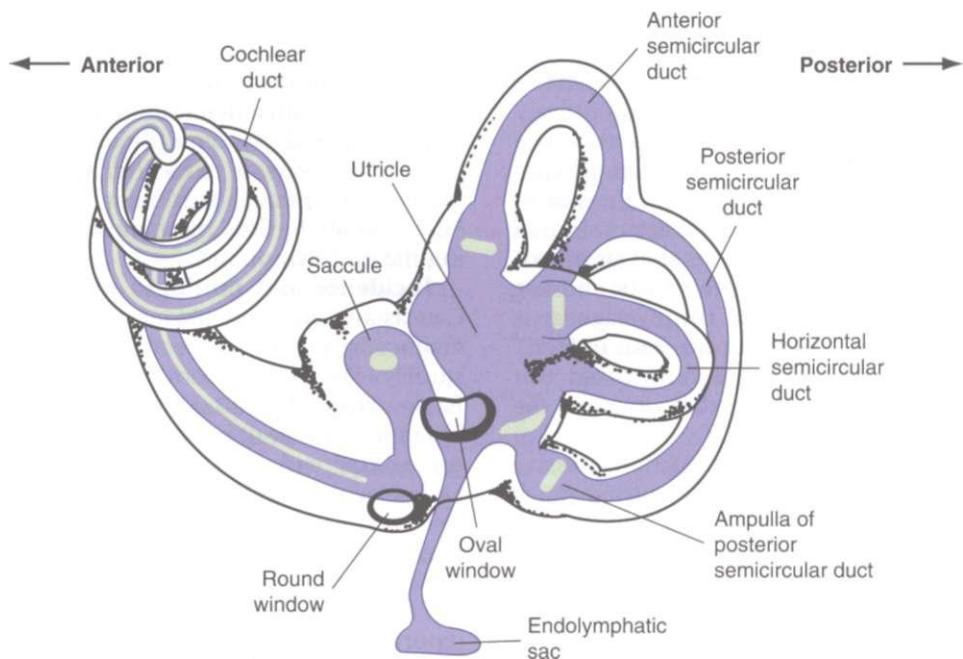


Figure 38-13

Membranous labyrinth as seen through an outline of the bony labyrinth. The endolymphatic sac provides the absorption of endolymph. This is the probable site of dysfunction in hydrops, causing a backup of fluid within the membranous labyrinth. (From Nolte J: *The human brain: an introduction to its functional anatomy*, ed 5, St Louis, 2002, Mosby.)



Figure 38-14

Cross-section of human cochlea demonstrating endolymphatic hydrops in a patient with Menière's syndrome. Note the distension of Reissner's membrane into the scala tympani in the apical turn of the cochlea (arrowhead). (From Cummings CW, Haughey BH, Thomas R, et al: *Cummings otolaryngology: head and neck surgery*, ed 4, St Louis, 2004, Mosby.)

brane that separates the endolymph from the perilymph may rupture, and there is a leakage of potassium into the endolymph that may result in palsy of the vestibular nerve fibers.

Pathologic studies of the human sac in the hydrops patient have recorded ischemia and fibrosis around the endolymphatic sac. Alterations in the size of the endo-

lymphatic duct and sac along with reductions in the lining of these structures have also been noted in both the diseased and nondiseased inner ears. This supports the theory that an abnormal endolymph drainage system may predispose individuals to the future development of Meniere's syndrome. Alterations in calcium and chloride metabolism may alter the osmotic gradients in the endolymph, causing accumulation of endolymph as well as the loss of endocochlear potentials necessary for hair cell function. These ionic disturbances may be caused by ischemia resulting from changes in local vasculature. A common vascular mechanism for migraine headaches and Meniere's disease has also been proposed. A hydropic state may leave those persons sensitive to stresses to the inner ear.⁶³

Clinical Manifestations. The typical attack of hydrops related to Meniere's syndrome is experienced as an initial sensation of fullness of the ear, a reduction in hearing, and tinnitus. This is usually followed by a rotational vertigo, postural imbalance, nystagmus, and nausea. The vertigo may last from 30 minutes to 24 hours. The symptoms abate over time, and the individual regains the ability to maintain balance. However, there may still be some sense of disequilibrium. Hearing slowly returns, but over time there may be a permanent loss of hearing. Tinnitus is common in hydrops and is commonly described as a low-pitched roaring or similar to a seashell noise. Box 38-6 gives some of the characteristics consistent with the diagnosis of Meniere's syndrome.

MEDICAL MANAGEMENT

DIAGNOSIS. The complaints of vertigo, hearing loss, and tinnitus do not automatically confirm a diagnosis of

Box 38-6**SOME CHARACTERISTICS CONSISTENT FOR THE DIAGNOSIS OF MENIERE'S SYNDROME**

The diagnosis of Meniere's syndrome is primarily based on the medical history. According to established guidelines, a definite diagnosis requires the following:

- Two or more definitive episodes of spontaneous rotational vertigo lasting at least 20 minutes
- Low-frequency sensorineural hearing loss documented by audiometry
- Tinnitus or aural fullness in the affected ear
- Exclusion of other causes for the symptoms

autoimmune and inflammatory injury in the etiology of hydrops.¹⁸

Intratympanic gentamicin is used in the treatment of chronic, unrelenting unilateral hydrops. Gentamicin typically results in a reduction of vestibular function in the treated ear. Surgical treatment to restore normal endolymph volume includes endolymphatic decompression procedures. Other procedures designed to ablate vestibular function in the affected ear without damage to the cochlea include vestibular neurectomy, intratympanic gentamicin, cryosurgery, ultrasound, and cochlear dialysis.¹⁸

PROGNOSIS. The natural history of endolymphatic hydrops and Meniere's syndrome is highly variable. Clusters of attacks may be separated by periods of long remission. Balance function between attacks can be normal, although a sense of disequilibrium often persists later in the course of the disorder. Meniere's syndrome initially affects only one ear. The attacks increase in frequency in the first years and then decrease. The frequency of bilateral disease ranges from 2% to 78%, with an average incidence of 45%. If bilateral involvement has not occurred within 5 years of onset of disease in the first ear, then there is less likelihood of developing bilateral involvement.

The hearing loss in Meniere's syndrome is a fluctuating, low-frequency sensorineural loss early in the clinical course. Eventually, the loss becomes irreversible, often progressing in severity with involvement of higher frequencies and loss of speech discrimination.

An estimated 2% to 6% of patients with Meniere's syndrome of long duration can experience "drop attacks" known as otolithic crisis of Tumarkin, characterized by being abruptly thrown to the ground without loss of consciousness and with little or no vertigo.²⁹

SPECIAL IMPLICATIONS FOR THE THERAPIST 38-2***Endolymphatic Hydrops*****PREFERRED PRACTICE PATTERNS**

5A: Primary Prevention /Risk Reduction for Loss of Balance and Falling

5D: Impaired Motor Function and Sensory Integrity Associated with Nonprogressive Disorders of the Central Nervous System—Acquired in Adolescence or Adulthood

5E: Impaired Motor Function and Sensory Integrity Associated with Progressive Disorders of the Central Nervous System

5F: Impaired Peripheral Nerve Integrity and Muscle Performance Associated with Peripheral Nerve Injury

Endolymphatic hydrops leads to great insecurity. If an acute attack occurs in social situations, it understandably can lead to social phobic reactions, even if no particular psychopathologic predisposition is present. Comorbidity with somatoform dizziness attacks may develop. Whether trigger stress that in turn leads to psychologic symptoms or stress factors are involved primarily in the pathogenesis cannot be

Continued.

endolymphatic hydrops or Meniere's syndrome. The definitive vertiginous attack is sudden in onset with nausea and vomiting, lasts 20 minutes, but abates by 24 hours. Typically, any movement during an attack aggravates the vertigo. The presence of neurologic signs or symptoms such as syncope, visual aura, and motor weakness suggest another diagnosis. Disorders that can present with similar symptoms include migraine, acoustic neuroma, perilymphatic fistula, dehiscence of the superior semicircular canal, labyrinthitis, autoimmune inner ear disorder, and MS.

An audiogram, or test of hearing, typically demonstrates low-frequency hearing loss on one side. Perceived hearing loss can be difficult to verify without audiometry, especially during an exacerbation of tinnitus or aural pressure. Improvement in audiograms can reflect better control of fluid, and decreases in hearing are suggestive of progressive disease.⁶³

Electrocochleography may provide objective evidence of the presence of endolymphatic hydrops in the presence of large summing potentials. However, the diagnostic utility of the test is limited by the variability of the ratio both in individuals with hydrops and in normal individuals. The results should be used in the face of other subjective history.

TREATMENT. Diuretics can control vertigo and stabilize hearing in more than half of the individuals with reported hydrops. Restricting salt, caffeine, alcohol, and nicotine is routinely suggested. The goal of salt restriction and diuretics is to reduce endolymph volume by fluid removal and reduced production.

Histamine is used in the belief that hydrops are a result of striaal ischemia or immune modulated disorders. Other agents used include the papaverine analog eupavarine, nicotinic acid, adenosine triphosphate, and dipyridamole. Antivertiginous medications, antiemetics, sedatives, antidepressants, and psychiatric treatment have been reported to be beneficial in reducing the severity of the vertigo and vegetative symptoms.

Corticosteroid infusion of the middle ear via a trans-tympanic route is under investigation as hearing and vestibular conservation therapy for endolymphatic hydrops. The presumed site of action is in the inner ear. The rationale for the treatment is based on the apparent role of

answered at this time. The anticipation anxiety that is typical for phobic disorders arises from the unpredictable pattern of endolymphatic hydrops, the feeling of being overcome by the attacks, being completely at their mercy, and having to stop all activity until the attacks subside. The concern over not finding a place where they can stay while experiencing an acute attack, of receiving negative reactions from those around them, and of being in a situation where they are embarrassed and helpless can cause them to withdraw from social life. A severe anxiety disorder can develop, so that in addition to the endolymphatic hydrops, symptoms of somatoform dizziness arise that then often are attributed wrongly to the endolymphatic hydrops. When invasive treatments such as gentamycin injections or sac decompression are performed, it increases the complexity of the disorder. Severe cases of Meniere's syndrome ultimately can lead to reactive depressive disorders. Antidepressant medications may be helpful. The physical therapy may be working in conjunction with behavioral therapy to improve a person's ability to cope with the disorder. This might include active reduction of stress factors, dealing with phobic anxieties, and learning relaxation techniques.

Perilymph Fistula

Definition and Overview. Perilymph fistula, an abnormal communication of the inner and middle ear spaces, can cause vertigo. Fistulas commonly occur at the round and oval windows of the middle ear. Attempts to identify the prevalence and characterize auditory and vestibular symptoms have been inconclusive. Some studies report vestibular symptoms as the major presenting complaint, whereas others indicate hearing loss equal to or more common than balance-related symptoms. It is believed that most fistulas result from congenital malformations or prior ear surgery. Damage can also result from pressure applied via the external ear, via the eustachian tube, or by an increase in the pressure of the cerebrospinal fluid.⁹

Clinical Manifestations. Characteristics of perilymph fistula include easing of symptoms at rest and increases with activity, including the Valsalva maneuver. Barotrauma, violent exercise, heavy lifting, or even sneezing may cause a fistula. Other mechanisms include head trauma, explosive blast, or barotrauma. Sensorineural hearing losses vary from an isolated high-frequency loss to a low-frequency or flat one. Speech discrimination test results are not characteristic. Both the pure-tone threshold and speech discrimination scores have been noted to fluctuate. Isolated mild conductive losses have been noted. Vestibular symptoms are also variable and include episodic incapacitating vertigo, equivalent to a Meniere's attack, positional vertigo, motion intolerance, or occasional disequilibrium. Disequilibrium after increases in CSF pressure (e.g., nose blowing, lifting), called *Hennebert's sign*, has been noted, as has vertigo after exposure to loud noises, which is known as Tullio's phenomenon.

MEDICAL MANAGEMENT

DIAGNOSIS. The inability to reliably predict the presence of a fistula before surgical exploration, as well as the lack of standard criteria for recognizing a fistula intraoperatively, have resulted in confusion and even doubt as to the existence of symptomatic fistulas. Because fistulas have been identified intraoperatively and their repair has resulted in symptomatic and objective improvement, this diagnosis must be kept in mind in the evaluation of the vertiginous patient.

Audiologic tests considered to be helpful in the diagnosis include electrocochleography. This demonstrates a larger summating potential due to endolymph/perilymph disequilibrium. However, the test is not sensitive or specific for perilymph fistula.

Results of vestibular testing are nondiagnostic. The most consistent abnormality seen is a unilateral reduced caloric response in the affected ear.

A fistula test is done by introducing positive pressure into the suspected ear, either by rapid pressure on the tragus, compression of the external canal, or use of a pneumatic otoscope, while observing the eyes. A positive fistula sign consists of conjugate contralateral slow deviation of the eyes followed by three to four beats of nystagmus. Vertigo is usually elicited at the same time. Measuring body sway during pressure on the tympanic eardrum can help make the diagnosis.

TREATMENT. Recommendations with suspected inner ear fistula include head elevation during bed rest, laxatives to reduce the risk of increased intracranial pressure, and monitoring of both hearing and vestibular function. In those instances in which hearing loss worsens or vestibular symptoms persist, surgical exploration is warranted.

Intraoperative identification of a fistula, regardless of criteria used, is reported in about 50% of individuals explored. At the time of surgery, the oval and round windows are patched with tissue, such as blood clot, fat, fascia, or absorbable gelatin sponge.¹⁸

PROGNOSIS. The outcome of surgical repair is variable. An appropriate surgical candidate is probably the most significant factor in outcome. Reduction in vestibular-related complaints has been reported in more than 50% of surgeries. Hearing is improved about 25% of the time.

SPECIAL IMPLICATIONS FOR THE THERAPIST 38-3

Perilymphatic Fistula

PREFERRED PRACTICE PATTERNS

5A: Primary Prevention/Risk Reduction for Loss of Balance and Falling

5D: Impaired Motor Function and Sensory Integrity Associated with Nonprogressive Disorders of the Central Nervous System—Acquired in Adolescence or Adulthood

5E: Impaired Motor Function and Sensory Integrity Associated with Progressive Disorders of the Central Nervous System

5F: Impaired Peripheral Nerve Integrity and Muscle Performance Associated with Peripheral Nerve Injury

The therapist can be instrumental in helping to determine the possibility of perilymph fistula. An individual who is compliant with prescribed exercise but lacks progress over time and reports or demonstrates fluctuating symptoms and status of balance may be suspect for possible fistula. It is important to maintain close contact with the physician, as these individuals become very frustrated and may not follow through with medical attention. Vestibular therapy should be continued after surgery as good recovery is expected. Often the physician is uncertain whether the patient's complaints are because of stable vestibular disease with inadequate central compensation or by unstable labyrinthine function. In this setting, a trial of vestibular rehabilitation is appropriate and assists in the diagnosis by clarifying this important distinction. Failure to improve with vestibular rehabilitation lends further credibility to the diagnostic impression that the lesion is unstable or progressive. It is then suitable to proceed with appropriate surgical management if the symptoms are severe enough to warrant the procedure and they emanate from the end organ.

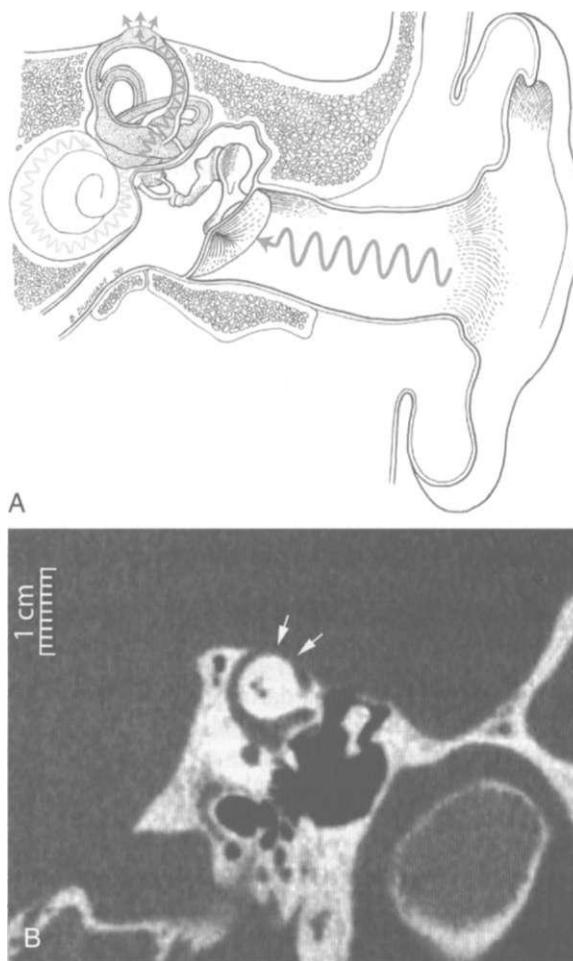


Figure 38-15

A, In superior semicircular canal dehiscence syndrome, sound waves can excite the superior canal because the "third mobile window" created by the dehiscence allows some sound pressure to be dissipated along a route through the superior canal in addition to the conventional route through the cochlea. **B,** CT scan demonstrating dehiscence (arrows) of the superior canal. (From Cummings CW, Haughey BH, Thomas R, et al: *Cummings otolaryngology: head and neck surgery*, ed 4, St Louis, 2004, Mosby.)

the drug. It appears that there is a possible genetic vulnerability, which is under study. Streptomycin and gentamicin specifically target the vestibular end organ. Other members of this group of drugs include kanamycin, tobramycin, amikacin, netilmicin, and sisomicin. Otoxicity is usually seen in individuals who are given multiple doses over time or one large dose, usually aimed at managing a threatening infection. Damage to the hair cells in the inner ear can result in complete loss of vestibular function within 2 to 4 weeks after these drugs are given.³²

Etiologic and Risk Factors. Approximately 3% of an orally administered aminoglycoside is absorbed from the gastrointestinal tract. They are normally injected for severe systemic infections. Penetration of the blood-brain barrier is generally poor, so that aminoglycosides are injected intrathecally to treat meningitis. Aminoglycosides are excreted primarily by the kidney by glomerular filtration, and therefore high concentrations of drug in

Superior Semicircular Canal Dehiscence Syndrome

Definition and Overview. Superior semicircular canal dehiscence syndrome is a syndrome of vertigo and oscillopsia induced by loud noises or by stimuli that change middle ear or intracranial pressure in patients with a dehiscence of bone overlying the superior semicircular canal. Tullio's phenomenon (eye movements induced by loud noises) or Hennebert's sign (eye movements induced by pressure in the external auditory canal) develop and often there is chronic disequilibrium.

Pathogenesis and Clinical Manifestations. The dehiscence creates a "third mobile window" into the inner ear, thereby allowing the superior canal to respond to sound and pressure stimuli. The evoked eye movements in this syndrome typically align with the affected superior canal. Loud sounds, positive pressure in the external auditory canal, and the Valsalva maneuver can cause characteristic eye movements. A larger length of dehiscence overlying the superior canal (5 mm or greater) can lead to dysfunction in the affected canal when evaluated by responses to rapid head movements in the plane of the superior canal. Visual fixation can suppress the evoked eye movements.²⁰ Fig. 38-15, A, shows the schematic representation of superior semicircular canal dehiscence; Fig. 38-15, B, shows the condition as seen on MRI.

Treatment. The window is repaired surgically, with good results.

Ototoxicity

Overview. The aminoglycoside antibiotics can be ototoxic, with auditory toxicity estimated at 20% and vestibular toxicity affecting 15% of the individuals receiving

the urine may be achieved. Impaired renal function reduces the rate of excretion. Therefore renal failure is a risk factor for ototoxicity, and dosing of aminoglycosides must be modified to compensate for delayed renal excretion. Measurement of peak and trough serum levels of aminoglycosides provides rough guidelines for therapeutic efficacy but is not an absolute guarantee for prevention of ototoxicity, particularly vestibular ototoxicity.¹⁸

Pathogenesis. The aminoglycoside reacts with inner ear tissues to form an active, ototoxic metabolite. The drug in its inactive form combines with iron to form an ototoxic complex. This complex reacts with oxygen to produce reactive oxygen species. These species can then react with various cell components—including the phospholipids in the cell membrane, proteins, and DNA—to disrupt the function, primarily in the outer hair cell. This process can then trigger programmed cell death, resulting in apoptosis. Histopathologic studies demonstrate that the cochlear and/or vestibular hair cells serve as primary targets for injury. Inner hair cells seem to be more resistant to injury than the outer hair cells. This could be a result of the higher concentration of the natural antioxidant glutathione in the inner hair cells. In some cases, spiral ganglion cells may be damaged directly by aminoglycosides without injury to outer hair cells. The stria vascularis may become thinner as a result of cell death. The damage is primarily bilateral, although there may be a difference in severity of loss between the two inner ear systems.

When there is bilateral dysfunction of the vestibular system the ability to determine head position in space can also be lost or decreased. Gaze stability with head movement is severely affected as the vestibular ocular reflex is disabled. With loss of the vestibular system function the individual must rely on somatosensory and visual cues for perception of stability and movement.

Clinical Manifestations and Treatment. One of the most debilitating early symptoms is oscillopsia. Oscillating vision is the illusion of environmental movement caused by excessive motion of images of stationary objects on the retina, known as retinal slip. Oscillopsia of vestibular origin is brought on or accentuated by head movement. It is due to insufficient VOR. When the vestibular system is unable to keep an image stationary by controlling the movement of the eyes, this slip occurs. Severe or complete bilateral loss of peripheral vestibular system function will result in inability to stabilize vision during head movement and produce oscillopsia.¹⁹

MEDICAL MANAGEMENT

PROGNOSIS. It is generally considered that the damage is permanent and that the recovery of function of the vestibular mechanism is limited. Initially there is severe disability related to sensation of dizziness, oscillopsia, and imbalance. Compensatory strategies using visual references and somatosensory input can improve mobility, taking approximately 6 weeks to become effective. There are some circumstances that will always be problematic such as walking in the dark, especially on uneven surfaces, or swimming. Night driving or driving in inclement weather should be avoided as vision is unavailable to provide stability.

SPECIAL IMPLICATIONS FOR THE THERAPIST 38-4

Ototoxicity

PREFERRED PRACTICE PATTERNS

5A: Primary Prevention/Risk Reduction for Loss of Balance and Falling

5D: Impaired Motor Function and Sensory Integrity Associated with Nonprogressive Disorders of the Central Nervous System—Acquired in Adolescence or Adulthood

5E: Impaired Motor Function and Sensory Integrity Associated with Progressive Disorders of the Central Nervous System

5F: Impaired Peripheral Nerve Integrity and Muscle Performance Associated with Peripheral Nerve Injury

When there is bilateral vestibular loss secondary to ototoxicity or by any other means that is considered permanent, the individual must be "uptrained" in the use of vision and somatosensation. They will essentially be "hanging on" with their eyes. Exercises for individuals with bilateral peripheral vestibular loss must be aimed at substitution of visual and somatosensory cues for the lost vestibular function.^{37,59} The ability to maintain stable gaze is critical. Gaze stability exercise incorporates the VOR, facilitating any possible remaining vestibular function. Modifications in eye movements can be used to improve gaze stability with head movement. No mechanism to improve gaze stability will fully compensate for the loss of the VOR, and clients will continue to have difficulty seeing during rapid head movements. Some persons can learn to close their eyes during a turn and then focus quickly on a stable object to regain stability. This technique increases stability because the disruptive visual input is temporarily eliminated.¹⁷ Strategies must be developed when the environment provides less than optimal sensory information, such as the use of an assistive device on uneven surfaces and the use of night lights to provide light at night.

Mal de Débarquement

Overview and Definition. Mal de débarquement (MDDS) is a syndrome that is named essentially for the symptoms related to "getting off the boat." It is usually triggered after a long time spent on a ship, such as during a cruise, or by an extended train ride. The complaints occasionally occur after international or extended air travel, especially if there is turbulence. The symptoms of dizziness and disequilibrium that usually subside within hours become persistent and can last for weeks, months, and even years. It is also reported as rocking vertigo, since the sensation is usually one of rocking back and forth and is experienced to a greater degree at rest than during movement.¹² The individual with MDDS would prefer to be moving in a car rather than standing still.

Pathogenesis, Clinical Manifestations, and Treatment. The basis for MDDS is uncertain but seems to be related to abnormal adaptation from one sensory context to another. During passive motion there may be a mis-

match between the vestibular inputs that encode head motion and other sensory and motor cues. Central adaptation may minimize the symptoms provoked by this mismatch, but when the movement ends, the brain must readapt to the stationary environment. It has been suggested that patients with MDDS undergo a physiologic adaptive process during passive motion, but for unknown reasons they do not readily adapt back to the stable environment. This impaired return to baseline results in a perception of motion when the patient is stationary.⁴⁹ Somatosensory input is no longer able to override vestibular input for stability. This sensation of constant motion at rest can be debilitating in that the person has difficulty managing the symptoms and tends to limit activity or overmedicate to dampen the sensation.^{43,56} Providing cues outside the vestibular system is helpful, and using somatosensory weighting or providing cues about position in space using an external level appear to help the system to recalibrate.

Autoimmune Ear Disease

Overview and Definition. Autoimmune ear disease (AIED) is rapidly progressive bilateral sensorineural hearing loss that responds to the administration of immunosuppressive therapy. The disease is typically characterized by symptoms of pressure and tinnitus in the ears with or without dizziness. Autoimmune disease can affect the inner ear with resultant vertigo, sensorineural hearing loss, aural fullness, and tinnitus.

Clinical Manifestations. Symptoms usually progress over weeks or months, and there is often a known systemic immune disease, such as rheumatoid arthritis. The otologic symptoms may occur as a direct assault by the immune system or as the deposition of antibody-antigen complex in the capillaries or basement membranes of inner ear structures. AIED mimics Meniere's disease with fluctuating hearing loss and vestibular dysfunction. Approximately 50% of patients testing positively for AIED have positive serum antibody tests. The Western blot is currently the most widely reported diagnostic tool used in the diagnosis of AIED with autoantibodies directed against the 68 kilodalton (kd) protein.¹¹

Treatment. Typical patients with AIED are managed medically with corticosteroids at the lowest dose that prevents fluctuating hearing. Most patients achieve mild recovery of sensorineural hearing loss on corticosteroids. In more severe cases there is report of medical management with methotrexate.³⁵

Neoplasia

Overview. Primary carcinoma can directly involve the end organ, the middle ear, or the mastoid. Glomus tumors are the most common tumor of the middle ear, arising from the chemoreceptor system of the ninth through twelfth cranial nerves and producing focal symptoms. See Chapter 30 for more information on the type of tumor presented here and information regarding treatment.

Schwann cell tumors arise from the nerve sheath of the vestibular nerve. The term *acoustic neuroma* is commonly used to describe this tumor, especially with regard to surgery. The tumor usually arises from the vestibular

component of the nerve rather than the cochlea. Schwannomas are usually small, firm, encapsulated tumors that grow very slowly. They form in the internal auditory canal or cerebellopontine angle and produce symptoms by compressing the nerve. Initial symptoms are usually related to hearing loss. Eventually mild symptoms of dizziness or balance disorders can develop as the tumor increases in size and the ability to adapt to the loss of function is lost. Fig. 38-16 shows the resulting findings on audiogram and imaged with MRI.

MRI with gadolinium contrast is used to diagnose these tumors. Surgical removal of schwannoma can be achieved by performing middle fossa craniotomy. The translabyrinthine approach is appropriate for tumors up to 3.0 cm. Radiosurgery (a single treatment of high-dose irradiation stereotactically administered) has been used successfully with fewer complications than surgery. There is a chance of progressive loss of hearing in the years following this procedure.

Meningiomas arise from the arachnoid layer in the area of the petrosal and sigmoid sinuses. The tumor is encapsulated and therefore does not invade the neural tissue. Displacement of the cranial nerves, brainstem, and cerebellum is common, causing complaints consistent with compressive damage.

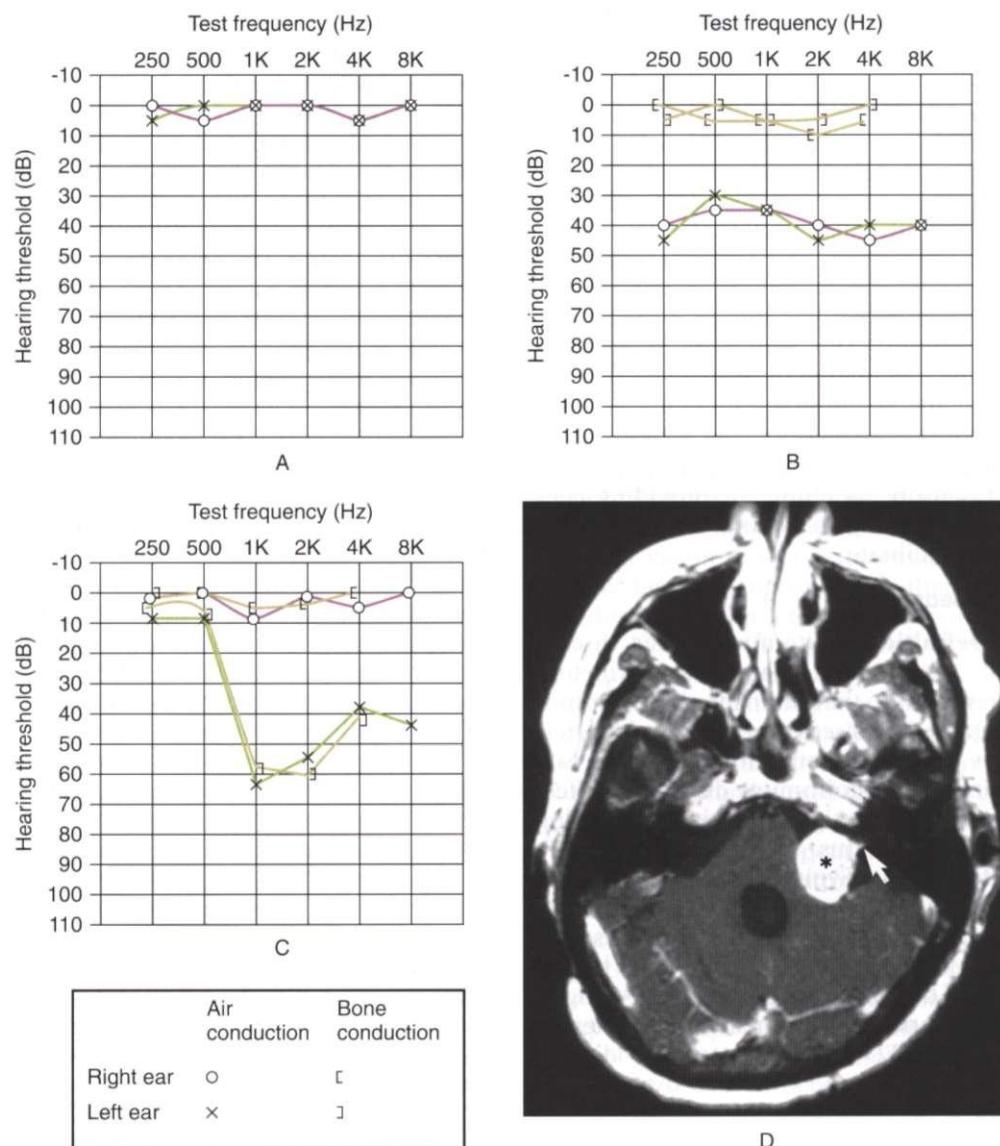
Gliomas arising from the brainstem grow slowly and progressively disrupt the brainstem centers, invading the vestibular and auditory systems in 50% of cases.¹⁶ Gliomas arising in the cerebellum are relatively silent until there is compression of the brainstem or obstruction of cerebrospinal fluid. Medulloblastomas, which occur primarily in children and adolescents, are rapidly growing tumors of the vermis and hemispheres of the cerebellum.

Metastatic neoplasms involve the vestibular and auditory functions primarily through involvement of the temporal bone. The internal auditory canal is a frequent site of metastatic tumor growth. Medical interventions to control growth can cause disruption of the neural tissue and trigger edema in the area of the vestibular system. This can cause acute symptoms; there is usually a good response to vestibular rehabilitation in that case.

Traumatic Brain Injury

Risk Factors. Complaints of dizziness and imbalance are common after traumatic brain injury (TBI) and may affect as many as 50% of the individuals who experience TBI. There may be direct injury to the vestibular mechanism as a result of temporal bone fracture resulting in tearing of the tympanic membrane or the membranous labyrinth. The vestibular and cochlear nerve can be damaged in the trauma, producing peripheral vestibular disorders. Labyrinthine concussion can trigger BPPV.⁵¹ Posttraumatic endolymphatic hydrops can cause intermittent dizziness but responds to management as described above. Perilymphatic fistula is more common after head injury and can be difficult to assess given the fluctuating nature of brain injury and difficulty with maintaining consistent performance of home programs.

Pathogenesis. Direct damage of the vestibular nuclei or cerebellar connections can cause persistent and disabling positional dizziness.^{3,48} Brainstem or midbrain

**Figure 38-16**

A, Normal audiogram. **B**, Abnormal audiogram in an individual with bilateral hearing loss. **C**, Abnormal audiogram in left ear with loss of both air and bone conduction. **D**, Vestibular schwannoma pressing against the left side of the brainstem and into the internal auditory canal. (A, B, and C redrawn from and D from Nolte J: *The human brain: an introduction to its functional anatomy*, ed 5, St Louis, 2002, Mosby.)

damage involving the vestibular nuclei can create inability to integrate somatosensation, visual, or vestibular input. In this case the individual will often choose to decrease the use of the vestibular system by limiting head motion and using predominantly visual or somatosensory cues for balance. The ability of the vestibular system to perform is further decreased by general lack of challenge. Then when the vestibular system is stimulated during daily activity, there is a sensation of dizziness and loss of postural stability. The individual with TBI may have more difficulty providing a clear history and may be plagued by other issues such as photophobia, hyperacusis, and visual motion sensitivity. Because of the damage to the central mechanisms that are necessary for the adaptation of vestibular impairment, recovery can take up to three times as long as would otherwise be expected. Flexion-extension injury is common and may limit the

ability to perform therapy involving head motion. There is often a cervicogenic component to the dizziness that can respond well to intervention. See Chapter 33 for further discussion of the brainstem disorders.

Psychologic Disorders and Somatoform Dizziness

Overview and Definition. Forty percent of dizzy individuals have psychologic disorders and, in general, individuals with psychiatric disorders report more disability with dizziness than do individuals without psychiatric involvement. Determining the correct etiology is critical in determining the treatment. Interest in this area is growing, and methods to more accurately identify the cause of the abnormalities are being developed. Connections between the locus coeruleus and lateral vestibular nucleus and serotonergic effects have implications for both processes. In a somatoform disorder the cause of the

Box 38-7**PHOBIC DIZZINESS**

Phobic disorders are characterized by the following:

- Dizziness and subjectively perceived disturbance of balance while patients are walking or standing but with normal balance test outcomes
- Fluctuating attacks or episodes of unsteadiness with the feeling of illusory body sway lasting from seconds to minutes
- Visually induced episodes with rapidly developing avoidance behavior
- Possible anxiety symptoms and distressing vegetative symptoms occurring during or after the dizziness sensations
- Possible compulsive personality traits, mild depressive symptoms, and instability of affect
- Symptoms often emerging after specific emotional distress, a severe illness, or an organic dizziness disorder

to half an hour or up to 2 hours and can be accompanied by vegetative symptoms, feelings of depersonalization, feelings of anxiety and panic, and hypochondriacal anxieties. In the case of panic disorder, recurrent dizziness attacks (i.e., several attacks over the course of 1 month) justify the diagnosis. In severe cases, the attacks may occur several times a day. Initially, the dizziness attacks cannot be related to any specific trigger situation. Nevertheless, the result is that patients frequently tend to avoid those places where they experienced their first dizziness attack. Not until later, during the course of psychotherapy, does it become clear what unconscious conflicts are underlying.

Nighttime dizziness attacks combined with sleep disorders also can occur. The patients subjectively believe that they have been awakened by the dizziness and afterwards often are unable to sleep for long periods. After the attacks, patients frequently feel exhausted. It is not uncommon for these patients to contact emergency medical services. Many patients develop hypochondriac anxieties and insist on repeated diagnostic measures. Episodes of hyperventilation also may be associated with panic disorder, yet patients subjectively may not perceive them as such. The hyperventilation leads to subjective feelings and sensations, such as dizziness, emptiness, lightheadedness, fear of fainting, and paresthesias, which in turn intensify the fear in a vicious circle because patients interpret these sensations as an expression of a severe physical illness.

In generalized anxiety disorder patients have dizziness attacks and permanent dizziness with fluctuating intensity. In addition to the symptoms, patients complain of constant inner tension and nervousness, a persistent generalized diffuse anxiety accompanied by psychomotor agitation and anxiety about the future. Establishing a diagnosis of somatoform dizziness as an expression of a panic disorder or a generalized anxiety disorder is simple after careful clinical examination. Anxiety disorders can be associated with depression. Often depressive disorders are not distinguished easily from each other.²²

Agoraphobia and social phobia are the two phobic disorders underlying somatoform dizziness most frequently. On closer exploration, it becomes clear that the dizziness frequently occurs in specific situations (e.g., agoraphobia when shopping in the supermarket, waiting in line, or driving a car). A classic phobia involves the fear of failing in such situations, becoming panicked, and falling into a state of helplessness and powerlessness, ultimately leading to embarrassment and shame. Avoidance behavior is not perceived subjectively as such; rather, it is a logical consequence of the dizziness. They often conceal their psychologic symptoms of anxiety and phobic avoidance and they are able to continue with their activities of daily life and work.

Somatoform dizziness as an expression of a depressive disorder usually takes the form of permanent diffuse dizziness. As with the anxiety disorders and phobic disorders, all further symptoms are interpreted as resulting from the dizziness. The symptoms can be manifold: depressive moods; rapid changes in mood; loss of drive and concentration; sleep disorders; or typical somatic symptoms, such as loss of appetite, loss of weight, and

dizziness symptoms cannot be traced to any objective organic pathologic findings and are experienced by patients as symptoms resulting from the dizziness and not the cause of it. Dizziness is usually the chief symptom and remains dominant after several years. There appears to be greater impairment in work and daily activities when the condition has a somatoform nature than for individuals who have dizziness with a known cause.

Anxiety and phobic disorders are the most frequent psychologic disorders underlying somatoform dizziness. Primary somatoform dizziness may develop without a prior fundamental organic disease, with individuals exhibiting dizziness sensations of varying quality and associated with impairment of daily activities as the chief symptom. Examination reveals typical symptom complexes for each of the underlying psychiatric disorders, such as anxiety, phobia, depression, or dissociation. Box 38-7 provides guidelines regarding phobic dizziness.

Secondary, reactive, or comorbid psychiatric disorders can emerge as a consequence of organic dizziness. Vestibular neuritis and BPPV are two common causes of dizziness that respond well to intervention. Often these individuals are given the standard intervention and acute symptoms subside, but recovery is illusive because of the underlying psychiatric disorder. Patients who have predisposed personality traits or previously latent anxiety disorders interpret the physical sensations catastrophically. The physical sensations function as stimuli triggering panic attacks. As a consequence, panic attacks can be triggered by other stimuli through conditioning, thereby persisting despite adequate central vestibular compensation.²²

Vestibular disorders have an influence on autonomic regulation. In somatoform disorder there is often a higher frequency of other autonomic symptoms such as heart palpitations, fainting spells, and chronic fatigue. As central neurologic links are found to exist between the vestibular and autonomic systems, it is hypothesized that dizziness and panic symptoms may have a common neuroanatomic basis. Vertiginous migraineurs have a higher frequency of comorbid anxiety disorders.

Clinical Manifestations. Dizziness attacks can be an expression of a panic attack lasting from a few minutes

libidinal disorders. In severe cases, social withdrawal, fear of the future, nihilistic thoughts and, rarely, suicidal indications may occur. Older patients often are misdiagnosed as suffering from vascular diseases or dementia. An organic lesion can act as a trigger and can make these persons feel completely unsure of themselves, leading in some instances to withdrawal from social life and isolation. Older people generally are poorly cared for in terms of psychotherapeutic and psychiatric therapy. In particular, older patients benefit most from appropriate treatment to improve their quality of life when it is hampered considerably by somatoform dizziness.

Somatoform dizziness can be an expression of a dissociative disorder (conversion disorder). In these cases, dizziness has the psychodynamic meaning of a conversion symptom. Careful clinical diagnostics commonly reveal a close temporal connection between past conflicts or stress situations and the onset of dizziness, a relationship of which patients initially are not aware. In contrast to patients who have anxiety or phobic or depressive disorders, these patients initially do not show such pronounced suffering and are less limited in their daily life and work activities. They do tend to demonstrate clear signs, however, of reaping secondary gains from the disorder, which primarily is connected with acute relief from unsolved, usually subconscious, internal conflicts. During the course of illness, however, secondary conditioning processes can start. Then the dizziness eventually occurs independently of the triggering conflict situation whenever low-level tension or mental irritation arises. In these cases, diagnosis can be difficult because the original conflict that triggered the dizziness is hidden. Therefore it is important to identify the exact time of onset of dizziness and the associated living circumstances at that time.

Psychogenic hearing disorders nearly always are dissociative disorders (conversion disorders). Initially, patients are not aware of the underlying conflicts. They willingly tell about their hearing disorder, which they often attribute to an earlier illness or previously occurring event, such as loud music and noises, an upper respiratory tract infection, or an ear infection. Frequently, it is only during thorough psychiatric-psychodynamic diagnostic procedures that conflicts come to light that are connected with the hearing disorder. Depending on the underlying conflict, long-term psychotherapy may be required in some cases.

Pathogenesis. The parabrachial nucleus holds particular importance as it is tied to the central core of the amygdala and the infralimbic cortex through reciprocal connections. A special significance is attributed to these brain structures during the development of fear and panic sensations and the conditioning of avoidance. States of hyperventilation can alter balance of the vestibular system, induce nystagmus, and provoke dizziness sensations in patients who have subclinical vestibular disorders.

Patients who suffer from a depersonalization or derealization syndrome often describe themselves as having diffuse dizziness or feeling dazed. Frequently, the depersonalization disorder is overlooked, but closer examination reveals that patients are not suffering from dizziness per se. The patients describe a strange feeling in their

head, feeling dazed, like being wrapped in cotton, feeling unreal or foreign, as though they are outside their body. They often barely can express these symptoms in words, as they find the symptoms highly disturbing and embarrassing. Isolated depersonalization syndromes are rare; a depersonalization disorder usually is accompanied by anxiety and depressive disorders. In the presence of a depressive disorder or an anxiety disorder, the existence of a depersonalization disorder often is underestimated.

MEDICAL MANAGEMENT

DIAGNOSIS. Somatoform dizziness usually is included late in the differential diagnosis or not at all, which is why the diagnosis is delayed in most cases—often after several months or years of the disorder—or is not made at all.

Individuals often consult first with an ear, nose, and throat physician, neurologist, or internist. To physicians who are less familiar with somatoform dizziness, these symptoms do not seem suspicious, and they usually are not recognized because the patients present with dizziness as the chief symptom. Initially it seems logical that, for example, a specific avoidance behavior or the reported vegetative symptoms result exclusively from organically caused dizziness.

This circumstance often increases the tendency of somatoform dizziness to become chronic, frequently leading to severe impairment of the quality of life, even early retirement, and high health care costs. Patients who have somatoform dizziness lasting more than 12 weeks and involving marked impairment should undergo interdisciplinary diagnostics routinely, including medical and ophthalmologic diagnostics, in addition to differentiated neurologic or neuro-otologic and psychosomatic diagnostics. Over long periods misdiagnoses are made or novel diagnoses invented, such as cervical dizziness or Meniere's disease.

When complex persistent dizziness syndromes are present, careful organic diagnostics always must be performed followed by a differentiated psychiatric-psychodynamic examination performed by an experienced clinician; this practice makes possible the diagnosis of somatoform dizziness in approximately two thirds of patients.

Attempts at symptomatic treatment such as long-term use of antivertiginous drugs are based on the assumption of residual signs and symptoms after a previous organic vertigo syndrome. These treatments typically lead to a short-term placebo effect, if they are successful at all, and contribute to perpetuating the dizziness symptoms by encouraging patients to believe their dizziness to be organic in origin. The consequence is that other symptoms, such as reactive depression and motivational disorders, crop up in addition to the primary underlying psychiatric disorder because all attempts at treatment are unsuccessful.

Clinical interactions have an important influence on the course of somatoform dizziness. When somatoform dizziness develops as a consequence of a vestibular disorder, explaining the psychosomatic connections in detail can be the first step in recovery. Patients may insist on new diagnostic measures because of hypochondriac

anxieties. If their health care providers do not give in to patients' insistence, instead reassuring them that their symptoms are taken seriously and repeatedly pointing out the necessity of psychosomatic diagnostics and therapy, they may help prevent the development of chronic symptoms.

TREATMENT. Treatment is determined by clinical presentation. Psychotherapy should begin immediately. Psychodynamic techniques and behavioral therapy are effective. The indication is determined by the clinical findings and the underlying conflict and stress situation. If the disorder is of short duration and pronounced only mildly, focused outpatient treatment can be successful. Depending on the underlying conflict, a long-term psychodynamic therapy should be the treatment of choice. If the illness is severe, pharmacotherapy to help manage symptoms may be necessary.

PROGNOSIS. Follow-up studies of the course of complex somatoform dizziness show an urgent need for early differentiated interdisciplinary diagnostics. The follow-up period in individual studies ranges from 4 months to 2 years. Depression, anxiety, and compulsion are influenced more heavily by how the dizziness is experienced subjectively than by the objective balance disorder. Subjective experiences depend on the underlying or associated psychologic disorder. The extent of the handicap and form of emotional distress interact with each other; depression and anxiety impede the ability to cope, thus exacerbating the dysfunction.

Tinnitus can be a trigger for depressive disorders. Sleep disorders, concentration difficulties, irritability, and a low threshold for stress can cause depression and lead to social withdrawal. The subjectively perceived intensity of the tinnitus often depends on the level of stress. Phobic reactions may develop, particularly when the tinnitus is accompanied by hyperacusis.

symptom is an expression of a psychologic disorder and must be understood as a somatoform symptom. The patients, however, perceive the symptoms subjectively as an expression of a physical illness. (2) Reactive or comorbid psychologic disorders present in underlying otoneurologic disorders may result in difficulties in coping with the illness. Individuals who are predisposed to anxiety and panic reactions can interpret a physical sensation, such as dizziness, or a physical symptom of illness as catastrophic or a sign of a severe and threatening illness. This can result in an escalated anxiety or panic reaction. This anxiety reaction leads to a further increase in the level of autonomic-nervous system arousal, leading to panic and, in some, fear of death. When this symptom complex is joined by episodes of hyperventilation with the corresponding physical consequences, such as paresthesia resulting from alkalosis, an increase in feeling of dizziness, and presyncope, this cycle can escalate further. Unexpected dizziness creates a feeling of helplessness and can trigger typical anticipation anxiety and phobic avoidance behavior. Avoidance behavior usually means avoiding body movements, thereby causing sensory integration to improve slower. The subjective symptoms persist, made worse by the anxious introspection, ultimately becoming chronic. Frequently, there is a change in the quality of dizziness from rotational vertigo to a more diffuse dizziness and lightheadedness. Although it can be interpreted as improvement or a normal progression of rehabilitation, it can cause further panic. Chapter 3 provides further information on these disorders and guidelines for the appropriate strategies for intervention.

SPECIAL IMPLICATIONS FOR THE THERAPIST 38-5

Somatoform Dizziness

PREFERRED PRACTICE PATTERNS

5A: Primary Prevention/Risk Reduction for Loss of Balance and Falling

5D: Impaired Motor Function and Sensory Integrity Associated with Nonprogressive Disorders of the Central Nervous System—Acquired in Adolescence or Adulthood

5E: Impaired Motor Function and Sensory Integrity Associated with Progressive Disorders of the Central Nervous System

5F: Impaired Peripheral Nerve Integrity and Muscle Performance Associated with Peripheral Nerve Injury

When discussing otoneurologic symptoms, it is important to distinguish between the following disorders: (1) disorders expressing themselves primarily through otoneurologic symptoms, yet caused by underlying psychologic disorders. The physical

Comorbid Disorders with Vestibular Consequences

Congenital Vestibular Loss. Events before birth can cause loss of vestibular function and are related to genetics or intrauterine infection, intoxication, or anoxia. Rubella and cytomegalovirus are responsible for most cases. Thalidomide, no longer commonly used, can cause loss of vestibular function.²⁵

Vascular Disease. Vertebrobasilar insufficiency is a common cause of vertigo in persons older than 50 years. This is often due to atherosclerosis of the vertebral and basilar arteries. Ischemia confined to the labyrinthine artery distribution results in infarction of the vestibular labyrinth and cochlea. Ischemia can also cause vertigo without hearing loss. A vascular loop compressing the eighth cranial nerve can cause vertigo, tinnitus, and hearing loss. Spontaneous hemorrhage into the inner ear, resulting in vertigo and hearing loss, mainly occurs in persons with underlying bleeding disorders. Leukemia is the most common disorder associated with labyrinthine hemorrhage.

Cerebellar infarction simulating vestibular neuritis is more common than previously thought. Early recognition of the pseudo-vestibular neuritis of vascular cause may allow specific management. Infarcts located dorsolateral to the fourth ventricle can cause interruption of

the vestibular nuclei-archicerebellar loop seems to be responsible for central paroxysmal positional vertigo.

Vestibular dysfunction is a common component with brainstem stroke and may respond to intervention. See Chapter 32 for a more complete description of these stroke syndromes.³

Vestibular Migraine. A type of migraine headache that results in symptoms of dizziness is associated with vascular changes in the area of the vestibular apparatus and is common in both children and adults. (See Chapter 37 for the full description of vestibular migraine.) Approximately 30% of the population with migraine report episodic vertigo, in some cases during the headache-free period.

Metabolic Disorders. When vascular and nerve changes associated with diabetes mellitus occur in the area of the peripheral or central vestibular system, symptoms may develop. Loss of proprioceptive input and visual degeneration will further exacerbate the sense of disequilibrium. Metabolic disorders affecting the resorption of bone, such as *otosclerosis* and *Paget's disease*, may develop to the point of causing degeneration of the labyrinths or nerves, resulting in vertigo and dizziness.

Allergies. Adverse reactions to foods and chemicals have been recognized as important etiologic agents in allergy. Otolaryngologic allergists address the clinical aspects of food sensitivity and dizziness after exposure to specific chemicals. There is clinical evidence of the relationship of dizziness with food allergies. The pathophysiology, however, remains unclear.^{47,56}

Aural Cholesteatomas. Aural cholesteatomas are cysts of the middle ear or mastoid arising from squamous epithelial lining, containing keratin. Cholesteatomas erode the temporal bone and may be congenital or acquired. The cholesteatoma can cause resorption of the adjacent bone by a process of pressure erosion. This process can cause damage to the labyrinths and result in vestibular dysfunction. Annual incidence in children is 3 per 100,000 and 12.6 per 100,000 in adults. The diagnosis of aural cholesteatoma is made on otoscopic examination, including endoscopic and microscopic evaluation or surgical exploration. Special imaging procedures, such as high-resolution CT scanning and MRI, may suggest the presence of cholesteatoma within the temporal bone and may be used to complement the clinical examination. High-resolution CT scanning is useful for operative planning and is recommended for all revision mastoid operations. The symptoms of cholesteatoma vary; some cholesteatomas are asymptomatic, whereas others become infected and rapidly cause bone destruction.

Malignant External Otitis. Malignant external otitis, an infection affecting older people with diabetes or immunosuppression, begins in the external auditory canal and spreads to the temporal bone, putting pressure on the facial nerve or the surrounding nerves.

The most common intracranial complication of otitic infections is *extradural abscess*, a collection of purulent fluid between the dura mater and bone of the middle or posterior fossa. Spread of the infection across the dura from the epidural space may result in thrombophlebitis of the lateral venous sinus, subdural abscess, meningitis, and brain abscess.

In some cases the damage is temporary, as in serous labyrinthitis, and vestibular and auditory function return to varying degrees. Permanent damage is possible when the infection causes damage to the structures of the labyrinth or the eighth cranial nerve. When the membranous labyrinth becomes permanently damaged, endolymphatic hydrops may result (see the section on Meniere's disease in this chapter) and the symptoms can become episodic.

Malignant external otitis in the mastoid bone is a major clinical problem. It often occurs in persons with a chronic illness, such as diabetes, or a malignancy who are receiving broad-spectrum antibiotics. The infection enters the middle and inner ear via the nasal sinuses. The infection can spread into the intracranial cavity and create thrombosis of the cerebral arteries. The eighth cranial nerve can be affected along with surrounding cranial nerves in the presence of basilar meningitis.⁵

SPECIAL IMPLICATIONS FOR THE THERAPIST 38-6

Vestibular Dysfunction

PREFERRED PRACTICE PATTERNS

5A: Primary Prevention/Risk Reduction for Loss of Balance and Falling

5D: Impaired Motor Function and Sensory Integrity Associated with Nonprogressive Disorders of the Central Nervous System—Acquired in Adolescence or Adulthood

5E: Impaired Motor Function and Sensory Integrity Associated with Progressive Disorders of the Central Nervous System

5F: Impaired Peripheral Nerve Integrity and Muscle Performance Associated with Peripheral Nerve Injury

Vestibular rehabilitation is used extensively for individuals with vestibular dysfunction with good evidence for effectiveness.^{37,49,58} The appropriate choice and progression of exercises are critical for the outcome.^{54,64}

Examination

Clinical evaluation by the therapist of the client with vestibular dysfunction integrates the history with the objective findings of sensory and movement disorders. The therapist is able to look at vestibuloocular function as well as vestibulospinal function. The history can guide the therapist regarding the mechanism of injury and location within the system. The nature of the symptoms and the precipitating, exacerbating, and relieving factors are reviewed.^{1,52,53}

The use of clinical syndromes can help make the connection between evaluation and intervention. Identification of sensation of motion at rest points to basic lack of adaptation of tonic firing rate, whereas motion-provoked dizziness occurs when the calibration between the two sides is inefficient during movement. Visual and somatosensory dependence develops early when the vestibular input is inadequate and can persist throughout the course of rehabilitation if not identified and treated. Visual motion sensitivity results

from the inability to filter out movement in the peripheral fields and is related to, but not exactly the same as, visual dependency. Positional dizziness should be fully evaluated. Head-righting response should be evaluated to determine if there is ability to use the otolith mechanism without override of the somatosensory system.

Testing in the clinic includes motor control of the visual-ocular component. VOR integrity is demonstrated by the client's ability to maintain visual focus during quick head turns. Interpretation of reflexive eye movements and postural changes to determine vestibular dysfunction is common. Other reflexive systems can compensate for the loss of vestibular reflexes and make it appear that there is no deficit, so these factors must be recognized when performing the tests. For example, a patient with well-compensated, long-standing bilateral loss of vestibular function may appear to have no problem keeping vision fixed on the examiner as the examiner rotates the patient's head slowly from side to side. In such individuals smooth pursuit, optokinetic nystagmus, and the cervico-ocular reflex make up for the vestibular deficit. This is an example of a head movement that *can* be sensed by the vestibular system, but that is not in the range of frequencies and accelerations sensed *exclusively* by the vestibular system. However, rapid head rotation can pull the eyes off target. The vestibular deficit can thus be unmasked by very dynamic head movements.^{26,66}

Evaluating the provoking positions and fatigability of dizziness may point toward a canal problem. A careful look at the CNS, through testing of function associated with cranial nerves, cerebellum, and brainstem and cortical connections, is critical to determine if there is a central cause. Often, both central and peripheral lesions can be identified. Sensory deficits should be determined. Control of balance in altered sensory conditions that isolate vision, somatosensory, and vestibular inputs is evaluated (see earlier discussion of the sensory organization test).

Musculoskeletal and neuromuscular evaluations are critical to determine compensatory movements based on deficits in these areas that may mimic vestibular dysfunction or detract from recovery. When observing upright balance, evaluation of strategy selection is helpful in identifying abnormal hip or stepping strategy, a common finding in clients with vestibular disorder.^{1,35}

Intervention

Recovery from the dynamic disturbance requires both visual input and movement of the body and head. There must be an error signal produced to let the brain know that adaptation is needed. For example, when the client is unable to maintain gaze stabilization and the image moves or slips on the retina, the nervous system will identify this error and try to adapt and modify the gain of the vestibular system. This is the process of adaptation that is facilitated by vestibular rehabilitation.^{26,29,31} Depending on the severity of the symptoms, these exercises may begin in a supine or sitting position in order to decrease the vestibulospinal challenge. The system is progressively challenged by adding activities incorporating movement of the eyes, head, and body, such as bouncing a ball off the wall. During the acute stage, the client may experience severe vertigo and may experience nausea and vomiting. Exercise will make these symptoms worse. The client should be encouraged to perform the established activities even if only for brief periods.

Adaptation of the vestibular system is context specific. Treatment should therefore address the multitude of environments that the person will encounter. Exercises should stress the integration of all systems involved in balance, synthesizing the visual and somatosensory cues with the vestibular cues.¹¹ To stimulate use of somatosensory inputs, environments are designed to place less emphasis on vision while providing reliable somatosensory inputs. To stimulate the use of visual inputs, environments are designed to place less emphasis on somatosensation while providing reliable visual cues. The vestibular system is naturally stimulated when visual and somatosensory cues are distorted.²⁷

When the balance disorder results in an inability to move the center of gravity through the ranges necessary to perform mobility tasks, the client will benefit from activities directed toward increasing the limits of stability.⁴¹ Center-of-pressure biofeedback provides input regarding control of center of gravity during weight shifts.⁵⁵

In clients with chronic complaints of dizziness, effective treatment is often challenging. For example, a therapist treating an individual with TBI who is complaining of dizziness must try to identify if there is a vestibular component. The intervention should provide the challenge necessary for adaptation of the vestibular system in addition to addressing the other important issues of TBI.^{1,51}

With mild to moderate TBI, often associated with auto accidents or falls, the dizziness persists or is reported to start weeks after the accident. Often the client has self-limited movement of the head and avoids certain changes of position. Symptoms may be easily provoked, and movement may trigger other associated brainstem dysfunction. The intervention in this instance may involve a gradual increase of activity. Practicing activities that increase reliance on existing sensory input can also improve the client's confidence in moving about when the environment is more complex.^{50,59,60}

As the system recovers, typical activity should provide adequate stimulus to maintain vestibular function. However, individuals will often avoid situations on the edge of their tolerance, especially if there is additional brain dysfunction. These individuals may show some decline in function over time if the system does not continue to be challenged as it is during the rehabilitation process.

One of the greatest challenges in working with a client with symptoms of dizziness and imbalance is making the correct determination of precipitating factors and understanding concomitant disorders of

Continued.

the brain and musculoskeletal system. Often health care providers stop short of the success that can be achieved because they do not fully understand the very complex mechanisms related to balance and dizziness.

Altered postural alignment is common with vestibular dysfunction. Sudden loss of vestibular function can result in both lateral flexion of the head and an abnormal shift of the center of gravity to the side of the lesion. Vestibular pathologic conditions result in an inability to accurately determine one's limits of stability, resulting in maintaining the center of gravity outside of or on the edge of the actual limits of stability, which may cause loss of balance.³² It is often noted that the person is unable (or unwilling because of fear of falling) to move the center of gravity far enough to perform a functional task such as descending stairs.

Lesions of the vestibular system can result in abnormal postural reactions to changes in head and body positions. When the individual relies on vestibular cues for the primary input for balance, the impaired system fails to provide the necessary input. Orientation to gravity or to support surface changes that include dorsiflexion or plantar flexion is inadequate to maintain upright position. This is seen in the inability to maintain balance on a compliant surface or on a surface that rotates at the ankle. It becomes difficult to walk in the dark, on soft surfaces, and up and down ramps or stairs.

Postural strategies, or postural control synergies, are triggered in preparation for movement and in response to changes in the environment. An ankle strategy is used when the client is standing on a firm, flat surface. The torque for the motion is controlled at the ankle. Vestibular dysfunction does not appear to affect this strategy since the input to drive the response comes primarily from the somatosensory system. However, initially there is difficulty performing this ankle strategy without the use of vision as a second sensory system with which the brain can compare.³³

A hip strategy is used when the client is standing on a narrow (beamlike) surface or when the surface is soft. A hip strategy is used on a flat surface when the

center of gravity moves beyond the base of support. In a hip strategy, the torque is controlled at the hip. Individuals with a vestibular loss have difficulty using the hip strategy. It becomes difficult to perform activities such as single-leg stance, heel-toe walk, and standing on a beam or a compliant surface. The individual with vestibular loss can generate a hip strategy but have difficulty maintaining control with perturbations of the surface and tends to use a stepping strategy to compensate for inability to maintain balance using the hip strategy.^{1,32,33} Stepping strategy is used to bring the center of gravity over the base of support when it moves too far to control with an ankle or hip strategy.

There is an abnormal perception of self-motion in relationship to movement in the environment. Therefore walking in a crowded environment, riding on an escalator, or walking in a grocery store may create increased symptoms of dizziness. Walking is easier in an empty shopping mall than in a crowded one, and walking with the crowd is less taxing than walking against the crowd.

It is important for the therapist to educate and support the individual during the natural course of the recovery. A program should be designed to facilitate the use of vestibular input for balance progressively so that the individual does not become overdependent on vision or somatosensation for balance. It appears that many individuals who have been considered compensated show less natural use of the vestibular system compared with individuals who have never had a vestibular problem. Current research is oriented toward more novel and effective ways to reestablish vestibular function.

References

To enhance this text and add value for the reader, all references are included on the companion Evolve site that accompanies this text book. The reader can view the reference source and access it online whenever possible. There are a total of 71 cited references and other general references for this chapter.

CHAPTER 39

The Peripheral Nervous System

MARCI A. SMITH

The peripheral nervous system (PNS) includes somatic motor and sensory components of cranial and spinal nerves arising from neurons whose cell bodies are located within the brainstem and spinal cord or lie in dorsal root ganglia. In addition, peripheral aspects of the autonomic nervous system (ANS) also contribute to axons found in peripheral nerves. Axons from the three components extend from the cell bodies to form peripheral nerves. Disorders of the PNS can be broadly divided into neuropathies, in which the pathology is confined to the nerve, and myopathies, in which the pathology occurs in muscle. Disorders of the PNS can be subdivided further according to site of anatomic involvement.⁴⁶

Signs and symptoms of PNS involvement relate to the motor and sensory systems, as well as the ANS. Motor involvement, termed *lower motor neuron* (LMN) involvement, occurs when any of the following sites is affected:

- Cell body of the alpha motor neuron (anterior horn cell) located within the spinal cord or brainstem
- Axons that arise from the anterior horn cell that form spinal and peripheral nerves and cranial nerves
- Motor endplate of the axon
- Muscle fibers innervated by the motor nerve axon

Sensory fibers of the PNS will show involvement if a lesion occurs in the dorsal root ganglion where the cell body is located or in the nerve root proximal to the ganglia, or distally in fibers of the peripheral nerve (Fig. 39-1). Similarly, when the ANS preganglionic or postganglionic motor fibers are involved, involuntary motor function of organs will be affected, and when sensory ANS fibers are affected, unconscious sensory functions (such as baroreceptors signaling arterial pressure, receptors within organs signaling irritants, distention, hypoxia, and so forth) will have transmission into the central nervous system (CNS) altered.

STRUCTURE

Nerves in the PNS are supported and covered by three connective tissue coverings that act like a tube surrounding the nerve. The inner-most covering is the *endoneurium*, which surrounds each individual axon. The middle layer, or *perineurium*, envelopes groups, or fascicles, of axons and is responsible for maintaining the blood-nerve

barrier. The outer-most layer, or *epineurium*, surrounds the entire nerve and provides cushioning for the entire nerve.¹³¹ The surface of an axon is formed by a phospholipid membrane called the *axolemma*. Lying between the axolemma and the endoneurium are Schwann cells (Fig. 39-2). Throughout life, axonal-Schwann cell molecular signaling occurs. In large diameter axons (greater than 1 μm), the Schwann cell receives a signal to wrap its membrane around the axon, thus creating myelin. In small diameter axons, the Schwann cell merely envelopes and supports nonmyelinated fibers. Myelin not only provides electrical insulation essential for rapid saltatory conduction of the axon potential but also affects axonal properties. The presence of myelin causes sodium channels to cluster at the nodes of Ranvier, thus reinforcing efficient saltatory conduction.⁹⁷ In the smallest axons, Schwann cells do not make myelin but do provide support for these unmyelinated fibers, whose action potentials are conducted by local circuit conduction (Table 39-1). Within a peripheral nerve, only about 25% of the fibers are myelinated.⁵²

Normal propagation of the action potential also requires sufficient energy, supplied by a vascular plexus interlaced between connective tissue layers. Each peripheral nerve receives an artery that penetrates the epineurium; this artery's branches extend into the perineurium as arterioles, and branches from the arterioles enter the endoneurium as capillaries. Vessels supplying peripheral nerves appear coiled when a limb is in a shortened position, but uncoiled after movement so that neural vascular supply is not impaired with a limb's normal excursion. This rich vascular supply makes peripheral nerves relatively resistant to ischemia.¹⁵⁶

PERIPHERAL NERVOUS SYSTEM CHANGES WITH AGING

Changes that occur in the PNS may be considered as one component of a continuum that relates to normal growth and development, or the changes may represent a combination of pathologic processes superimposed on the normal aging process. Because of the difficulty of studying human peripheral nerves *in vivo*, experimental animals have been used to assess the effects of aging.

Age does not affect the size or number of fascicles, but the perineurium and epineurium do thicken with age and the endoneurium often becomes fibrosed with increased collagen. Even with these changes, the cross-sectional area decreases slightly with age because there is a reduced number of unmyelinated and myelinated fibers. Ventral root fibers controlling motion are more affected than

dorsal root fibers controlling sensation. Blood vessels to nerves may become atherosclerotic with aging, and occlusion may contribute to loss of nerve fibers. The prevalence of peripheral neuropathies seen in older people has been attributed to this vascular pathology.

Decreases in protein production are hypothesized to cause myelin deterioration.¹⁶⁴ When individual myelinated fibers are examined, shorter internodes are seen, suggesting that a demyelinating-remyelinating process occurs with aging. This structural alteration in peripheral nerve myelination may be reflected in diminished appreciation of vibratory sense.

ANS dysfunction is more common in the elderly. This dysfunction may be related to the changes seen in the nervous system in the elderly. Cell bodies show chromatolysis, as well as an accumulation of lipofuscin, representing a diminished ability of the cell to rid itself of toxins. Loss of cell bodies has been observed in the sympathetic ganglia, along with a loss of unmyelinated fibers in peripheral nerves. Sympathetic control of dermal vasculature shows an age-related decline that leads to a diminished wound repair efficiency. In an aging animal model, transcutaneous nerve stimulation (TENS) improved the vascular response. Peripheral activity of sympathetic nerves were affected by the low frequency electrical stimulation.⁷⁷

When the motor endplate is examined, age-related changes have occurred, but these changes are seen as early as the third decade of life and are not reported in all muscles. When sensory receptors have been evaluated, density and morphology have been found to be altered in the elderly. Altered axonal myelination creates slowing of nerve conduction velocities (NCVs) in the elderly. In addition, the loss of fibers decreases the amplitude of the potential. Simultaneous with the decreased protein production is a decrease in intraaxonal transport by cytoskeletal elements in the peripheral nerve. Electromyographic (EMG) studies of elderly people without evidence of neurologic disorders of the PNS show loss of motor units, as well as signs of reinnervation. Morphologic changes observed in people over 60 years of age are manifested by decreased strength and sensory changes.¹⁰⁸

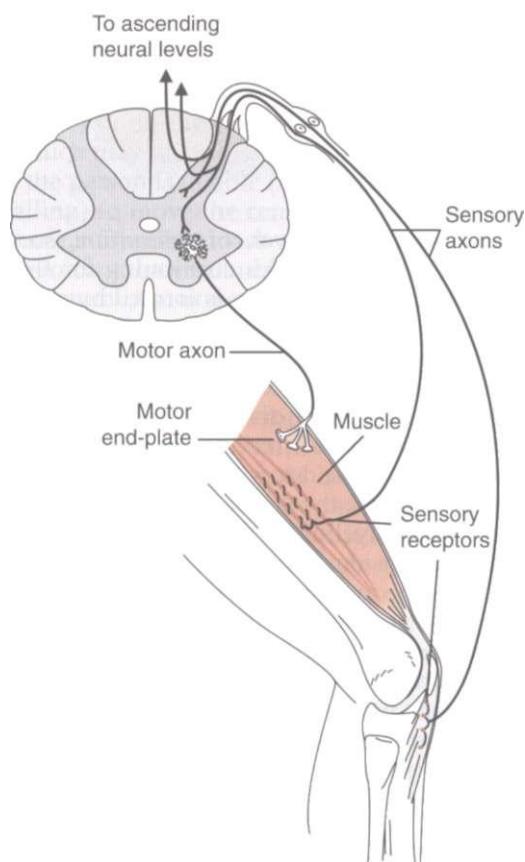


Figure 39-1

Potential sites of involvement in the peripheral nervous system. Motor: motor neuron cell body, axon, motor endplate, muscle fiber. Sensory: cell body in ganglion, axon, sensory receptor.

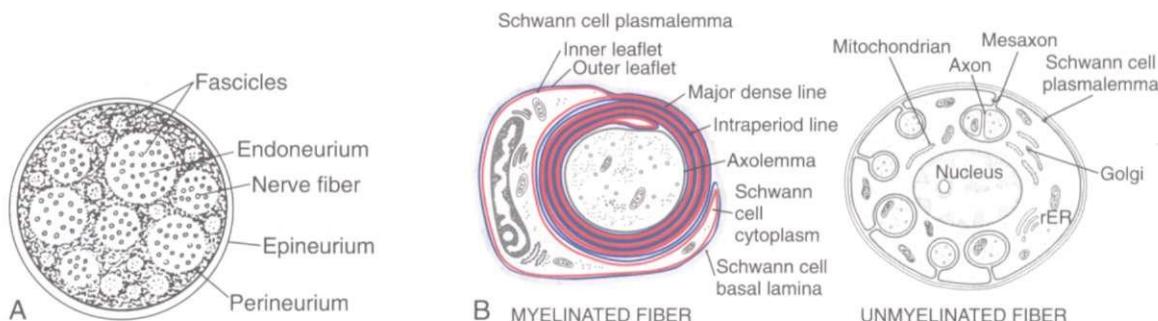


Figure 39-2

A, Cross-section of a peripheral nerve showing connective tissue coverings. Externally, the nerve is enveloped by the epineurium; internally, individual axons are surrounded by endoneurium. The perineurium surrounds groups of axons, termed fascicles. **B**, Although the majority of the fibers are unmyelinated, they are still associated with supporting Schwann cells. In myelinated fibers (left), each Schwann cell forms a myelin internode whose borders are formed by the nodes of Ranvier. Schwann cell sheath supporting unmyelinated fibers (right). (A, From Wildsmith JAW: Peripheral nerve and local anesthetic drugs, *Br J Anaesth* 58:692, 1986. Reproduced by permission. B, From Haines DE: *Fundamental neuroscience for basic and clinical applications*, ed 3, Edinburgh, 2006, Churchill Livingstone.)

Table 39-1 Relationship of Myelin Thickness, Conduction Velocity, and Sensory and Motor Fibers

Myelin	Conduction Velocity	Sensory Fibers	Motor Fibers
Very thick	Very fast	Proprioception (muscle spindle and Golgi tendon organ)	To skeletal muscle fibers (alpha)
Thick	Fast	Touch, pressure	To muscle spindle (gamma)
Thin	Slower	Touch, temperature	To ANS ganglia
None	Slow	Pain	From ANS ganglia to smooth muscle

ANS, Autonomic nervous system.

Healthy elderly, with no evidence of neurologic disease, may provide a clinical history suggestive of peripheral neuropathy. This includes numbness and tingling in the hands and feet along with mild, diffuse weakness—especially in the distal muscles of the hand. Sensory alterations may lead to poor balance and gait instability. On examination, sensory thresholds are increased.

The cause of an aging neuropathy can be attributed to a combination of factors. First, loss of both motor and sensory cell bodies; second, a dying-back condition, suggesting neurons can metabolically support a limited number of fibers or receptors, similar to that seen in other systemic neuropathies; and last, over the course of a lifetime, chronic compression of the peripheral nerves or repetitive trauma may have damaged the nerves. All these factors, combined with coexisting medical conditions, atherosclerosis, and nutritional deficiencies, may create this neuropathy of aging.

When the aging PNS is damaged, Wallerian degeneration is delayed and regeneration takes longer because secretion of trophic factors is slower than in younger individuals. Density of regenerating axons is less. In a partial nerve injury, collateral sprouting is reduced, further limiting recovery of function.¹⁶⁴

RESPONSE TO INJURY

Peripheral nerve damage occurs by any one of several causal conditions: heredity, trauma, infections, toxins, and metabolism.²⁶ When either motor or sensory nerves are affected, there is a limited response to injury, regardless of the cause. Either fibers demyelinate or fibers degenerate. Segmental demyelination occurs when nerves are subject to external compression or disease. Degeneration occurs in any peripheral nerve disorder that directly affects the axon, including physical injury (crush, stretch, or laceration), as well as disease.

Loss of myelin, typically in segments, leaves the axon intact but bare where the myelin is lost. This is called *segmental demyelination*. More severe involvement causes axonal degeneration, distal to the lesion (termed *anterograde*, or *Wallerian degeneration*^{147,27}), that begins immediately after involvement and is completed over a period of a few weeks. Neuropathic diseases that affect the axon or its cell body causing axonal degeneration typically affect the longest nerve fibers first (a length-dependent process), with signs and symptoms beginning distally and spreading proximally as the disease progresses. Because nerves in the legs are longer, the feet and lower legs are involved

long before the fingers and hands. Those conditions that affect only myelin cause segmental demyelination in both sensory and motor fibers. Thus disruption of the conduction of the action potential from proprioceptors and mechanoreceptors causes sensory changes. Those neuropathies that affect myelin cause demyelination of motor nerves to muscle and preganglionic fibers of the ANS create weakness, proprioceptive and tactile changes, and autonomic involvement by disrupting conduction of the action potential.

CLASSIFICATION OF NERVE INJURY

Traumatic injury to peripheral nerves from mechanical involvement secondary to compression, ischemia, and stretching can be classified using one of two systems based on the structural and functional changes that occur. Seddon¹⁴⁰ initially divided nerve injury into three categories: neurapraxia, axonotmesis, and neurotmesis (Fig. 39-3). Sunderland¹⁵³ divided this classification into five categories, based on axonal and connective tissue covering involvement.

Neurapraxia involves segmental demyelination, which slows or blocks conduction of the action potential at the point of demyelination in a myelinated nerve. Neurapraxias often occur after nerve compression that induces mild ischemia in nerve fibers. When segmental demyelination occurs because of disease, the response may be termed a *myelinopathy*. Conduction of the action potential is normal above and below the point of compression, and because the axon remains intact, muscle does not atrophy. *Axonotmesis* occurs when the axon has been damaged, but the connective tissue coverings that support and protect the nerve remain intact. Prolonged compression that produces an area of infarction and necrosis causes an axonotmesis. In the presence of disease, Wallerian degeneration creates an axonopathy, which is analogous to an axonotmesis. *Neurotmesis*, the most severe axonal loss, is the complete severance of the axon, as well as the disruption of its supporting connective tissue coverings (endoneurium, perineurium, and/or epineurium) at the site of injury. Neurotmesis is caused by gunshot or stab wounds or avulsion injuries that disrupt a section of the nerve or entire nerve. When axonal continuity is lost (either axonotmesis or neurotmesis), axons distal to the lesion degenerate (Wallerian degeneration). Because muscle fibers innervated by the axon depend on the nerve cell body as a source of nourishment or trophic control,

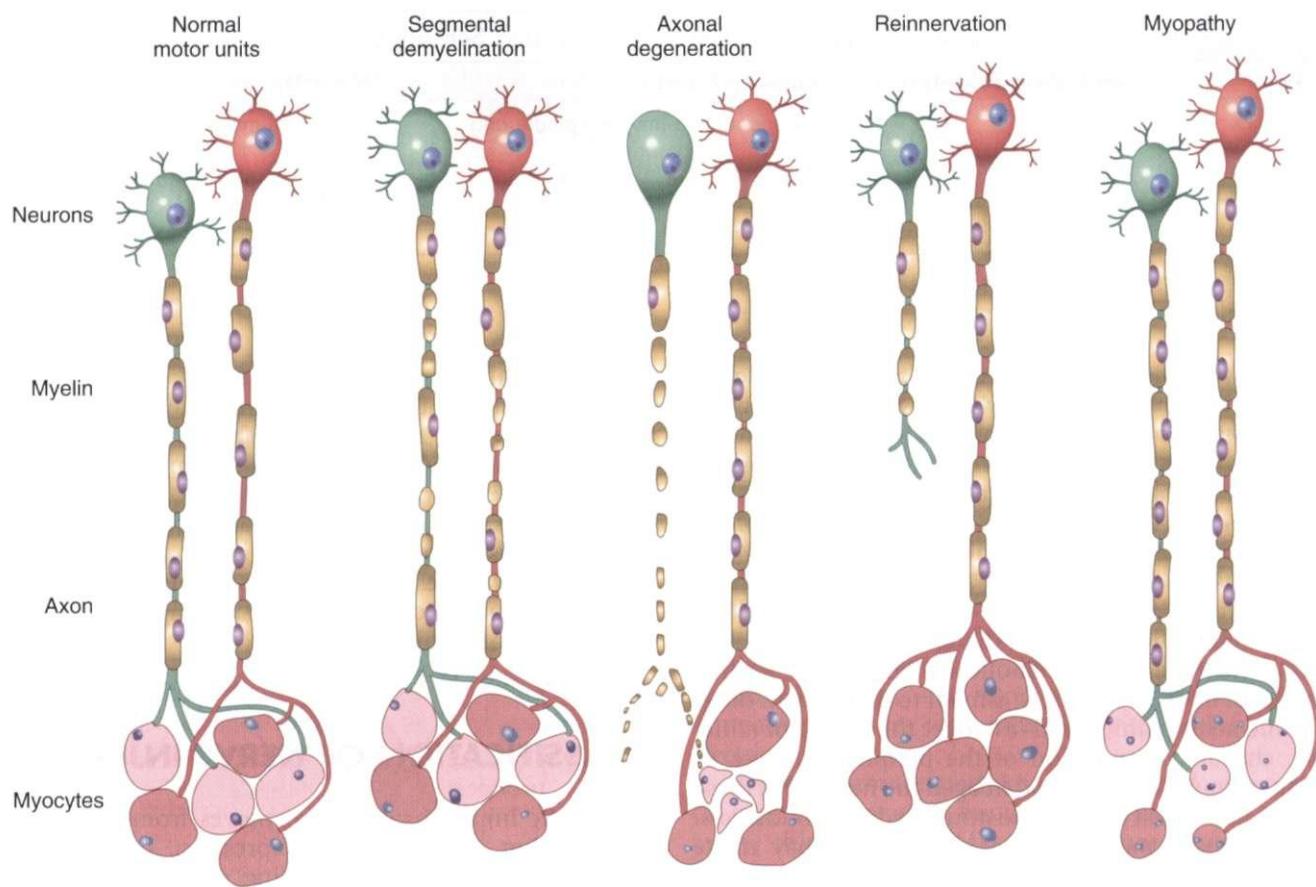


Figure 39-3

Types of nerve involvement and recovery that occur in peripheral nerves. **A**, Illustration of two adjacent normal myelinated nerves. **B**, Segmental demyelination. Several internodes of myelin have demyelinated, but the axon remain intact. The repair process for segmental demyelination occurs rapidly because Schwann cells divide and remyelinate the bare portion of the axon. Shorter internodal distance occurs with remyelination, thus nerve conduction velocity may not return to normal, even though muscle contracts normally. **C**, Illustration of axonal degeneration. The axon and myelin have degenerated, but the connective tissue covering remains intact in an axonotmesis. In neurotmesis, the connective tissue covering is disrupted at the lesion site. Signs of chromatolysis occur in the cell body after axonotmesis and neurotmesis. Note that muscle atrophies rapidly because it has lost the trophic influence from the nerve cell body. **D**, The repair process for axonotmesis and neurotmesis is more complex. Growth cones from the proximal axon must cross the lesion site and regrow down the connective tissue channels and reestablish a motor endplate or sensory connection before remyelination occurs. In partial nerve injuries, while the injury axon is regrowing, adjacent motor units sprout collateral fibers, leading to expansion of the size of this (red) motor unit. **E**, In myopathic conditions, scattered muscle fibers in adjacent motor units are small (degenerating or regenerating), while the neurons and axons are normal. (From Kumar V, Fausto N, Abbas A: Robbins and Cotran pathologic basis of disease, ed 7, Philadelphia, 2005, WB Saunders.)

when axons degenerate, muscle fibers rapidly atrophy (Table 39-2).

If segmental demyelination has occurred, molecular signaling to remaining Schwann cells causes them to begin dividing mitotically. Newborn Schwann cells move to envelope the denuded segment of nerve and once these cells are in place they will begin to form myelin (Fig. 39-3, B). The potential for regeneration after axonal/Wallerian degeneration is possible as long as the nerve cell body remains viable; new axons can sprout from the proximal end of damaged axons (Fig. 39-3, C). However, successful functional regeneration requires that the proximal and distal ends of the connective tissue tube are aligned. This occurs in an axonotmesis because the connective tissue coverings remain intact. In a neurotmesis, without surgical intervention, recovery is less likely

because the proximal end of the endoneurium is not approximated to the distal endoneurium. Without surgery, axonal sprouts often enter nearby soft tissue and form a neuroma, or axonal regrowth occurs down the incorrect endoneurial tube, rendering reinnervation non-functional.¹⁶⁴ Once the axon has established a distal contact either with muscle or sensory receptor, remyelination will begin. When partial axonal degeneration occurs, adjacent noninvolved axons will produce collateral sprouts that will innervate muscle fibers before the damaged axons have time to grow and reinnervate those muscle fibers. This results in an enlarged motor unit for the neuron that has collateral sprouts (Fig. 39-3, D). Numerous reports in the literature link various molecular factors to nerve regeneration and healing following repair.^{116,155}

Table 39-2 Relationship of Nerve and Muscle Responses to Disease and Trauma

Level of Severity	Response to Disease	Response to Trauma	Response of Muscle
Mild	Myelinopathy (segmental demyelination)	Neurapraxia (segmental demyelination)	Paresis/paralysis, no atrophy
Severe	Axonopathy (Wallerian degeneration)	Axonotmesis (wallerian degeneration)	Paresis/paralysis with atrophy
Severe	—	Neurotmesis (wallerian degeneration)	Paresis/paralysis with atrophy

Table 39-3 Causes of Peripheral Neuropathies and Myopathies and Their Effects

Cause	INVOLVEMENT*					
	Axonal Degeneration	Demyelination	Motor Endplate	Muscle	Motor	Sensory
Charcot-Marie-Tooth disease	X	X			X	X
Mechanical compression/entrapment						
Neurapraxia		X			X	X
Axonotmesis	X				X	X
Neurotmesis	X				X	X
Postpolio syndrome	X				X	
Diabetic mellitus	X	X			X	X
Alcohol	X	X			X	X
Guillain-Barré syndrome	X	X			X	X
Toxins						
Lead	X				X	X
Organophosphate	X	X			X	X
Myasthenia gravis			X		X	
Botulism			X		X	
Muscular dystrophy					X	X
Inflammatory myopathy					X	X
Steroid-induced myopathy					X	X
Overuse myopathy					X	X
Aging	X	X	X	X	X	X
AIDS/HIV (including associated vasculitis)	X	X			X	X
Vitamin B12 deficiency	X	X				X
Chronic renal failure	X				X	X

AIDS, Acquired immunodeficiency syndrome; HIV, human immunodeficiency virus.

*X indicates the most common types of involvement for each cause.

CLASSIFICATION OF NEUROPATHY

Neuropathies include a wide variety of causes and can be classified in many ways, including the rate of onset, type and size of nerve fibers involved, distribution pattern, or pathology (Table 39-3). For example, when a single peripheral nerve is affected the result is a *mononeuropathy*, which is commonly a result of trauma. The term *polyneuropathy* indicates involvement of several peripheral nerves. A *radiculoneuropathy* indicates involvement of the nerve root as it emerges from the spinal cord, and *polyradiculitis* indicates involvement of several nerve roots and occurs when infections create an inflammatory response.

In addition to involvement of the peripheral nerve, the motor endplate or muscle itself may be involved in a peripheral disorder. Involvement of muscle, termed *myopathy*, follows a different clinical pattern than nerve. When muscle is involved, the disorder typically is reflected

by proximal weakness, wasting, and hypotonia without sensory impairments (see Chapter 23).¹⁷³

SIGNS AND SYMPTOMS OF PERIPHERAL DYSFUNCTION

The presence of signs and symptoms aid in the localization of the level or levels of involvement. Loss of sensory function will follow a peripheral nerve distribution if that is the anatomic region involved, or it will follow a dermatomal pattern when the spinal nerve or dorsal root ganglia (cell body) has been affected (Fig. 39-4).

Similarly, when a peripheral nerve has motor involvement, paresis or paralysis will occur in muscles innervated by that nerve distal to the lesion. When spinal motor nerves are involved, weakness occurs in all the muscles receiving axons from that spinal level (a myoto-

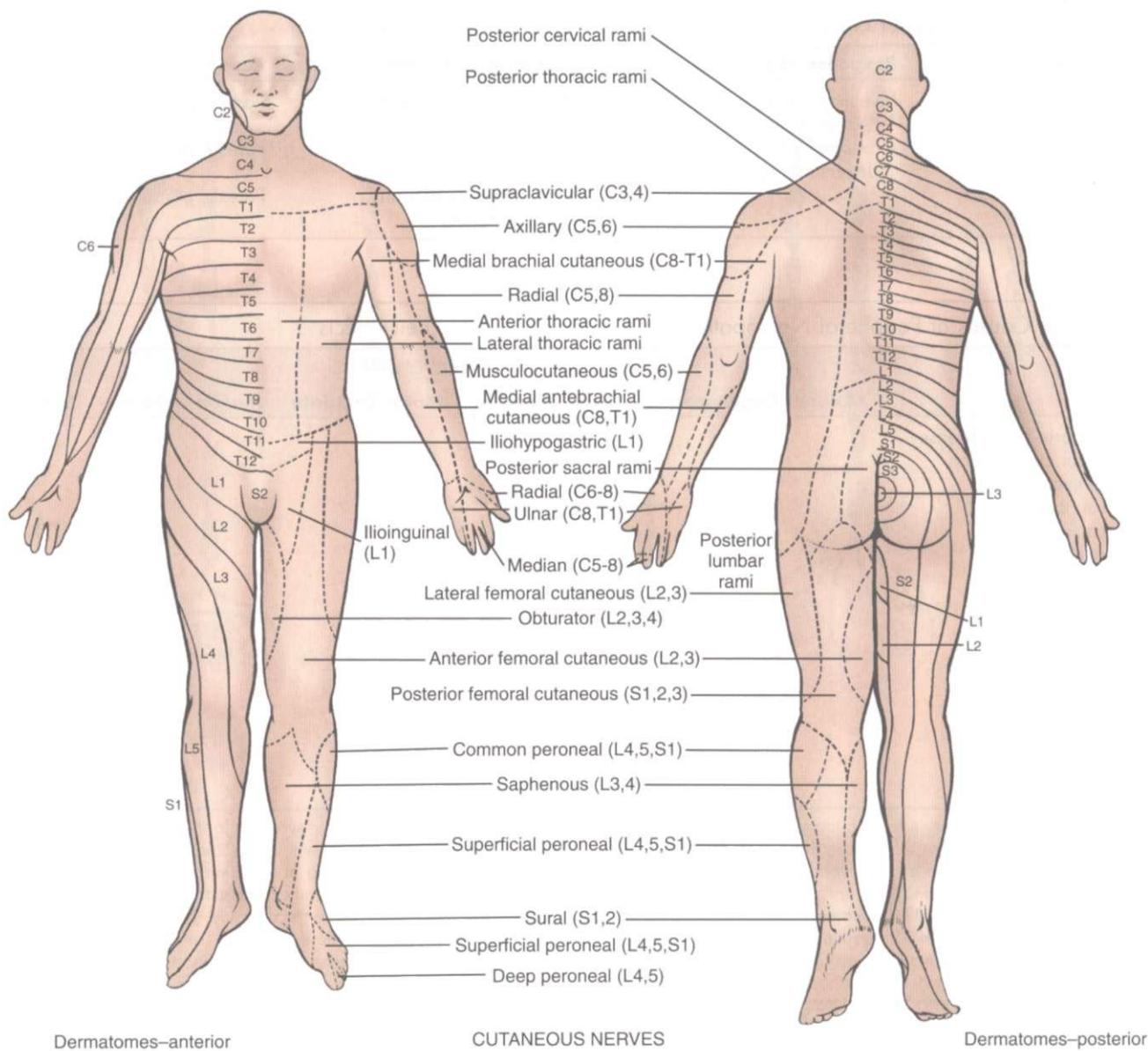


Figure 39-4

Dermatomal (right side of body, anterior and posterior) and peripheral sensory nerve (left side of body, anterior and posterior) patterns. (From Auerbach PS: *Wilderness medicine*, ed 5, St. Louis, Mosby, 2007.)

mal pattern). Individuals with only peripheral nerve involvement will have no signs or symptoms of CNS dysfunction.

Although differences occur in symptom evolution and in progression and severity of a neuropathy, a classic pattern of involvement would occur as follows. Involvement of sensory fibers is reflected by distal sensory deficits with the longest nerves in the body involved first. The first noticeable features of neuropathies are often sensory and consist of tingling, prickling, burning, or bandlike *dysesthesia*s and *paresthesias* in the feet. When more than one nerve is involved, the sensory loss follows a glove-and-stockin distribution that is attributed to the dying-back of the longest fibers in all nerves from distal to proximal (Fig. 39-5).

The most common symptoms of motor nerve involvement include distal weakness and abnormalities of tone (hypotonicity or flaccidity). When clients are asked to walk on their heels or toes, weakness of dorsiflexors or plantarflexors, respectively, becomes apparent. Deep tendon reflexes (DTRs) are diminished or absent, and distal-most DTRs will be affected first. In the presence of axonal degeneration, rapid atrophy occurs, along with electrophysiologic changes (Tables 39-4 and 39-5). Prolonged paralysis gives rise to secondary complications like contracture formation and edema.

In addition to weakness and hypotonia, a diagnosis of any one of the muscle diseases may be associated with muscle tenderness or cramping. Classically, the motor involvement in a myopathy is opposite to that of a neu-