REVIEW ARTICLE

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Tinnitus, Hyperacusis, Otalgia, and Hearing Loss

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ABSTRACT

PURPOSE OF REVIEW: **This article reviews the causes of tinnitus, hyperacusis, and otalgia, as well as hearing loss relevant for clinicians in the field of neurology.**

RECENT FINDINGS: **Important causes of unilateral and bilateral tinnitus are discussed, including those that are treatable or caused by serious structural or vascular causes. Concepts of hyperacusis and misophonia are covered, along with various types of neurologic disorders that can lead to pain in the ear. Hearing loss is common but not always purely otologic.**

SUMMARY: **Tinnitus and hearing loss are common symptoms that are sometimes related to a primary neurologic disorder. This review, tailored to neurologists who care for patients who may be referred to or encountered in neurology practice, provides information on hearing disorders, how to recognize when a neurologic process may be involved, and when to refer to otolaryngology or other specialists.**

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Drs Fife and Tourkevich report no disclosures.

UNLABELED USE OF PRODUCTS/INVESTIGATIONAL USE DISCLOSURE:
Drs Fife and Tourkevich discuss the unlabeled/investigational use of carbamazepine, gabapentin, oxcarbazepine, or pregabalin for the treatment of certain types of pulsatile pulse-asynchronous tinnitus; baclofen, carbamazepine, clonazepam, onabotulinumtoxinA, or oxcarbazepine for the treatment of palatal or middle ear myoclonus; and betahistine for the treatment of Ménière disease.

INTRODUCTION

Hearing loss and tinnitus are exceptionally common symptoms encountered in nearly all areas of medicine and may affect patients of all ages. Hearing loss severe enough to impair daily communication is seen in more than half of individuals aged 60 to 70 years and affects 80% of those 85 years and older.[1,2] Hearing loss as a measure of years of disability is the fourth leading cause of disability globally.[1] Similarly, clinically significant chronic tinnitus affects about 10% to 15% of people, and the incidence, like that of hearing loss, increases with age. Tinnitus is frequently also accompanied by hearing loss.[3] Hyperacusis, an abnormally low tolerance for ordinary sounds, is far less common but may be related to tinnitus or may occur as an isolated condition that can affect both adults and children.[4]

Neurologists may be asked to consult on sudden hearing loss or acute tinnitus, especially when it is either unilateral or occurs in association with neurologic symptoms. Neurologists should be especially aware of neurologic conditions that may include tinnitus, hyperacusis, and hearing loss. It is valuable to have some familiarity with hearing and other auditory symptoms and to be able to recognize when to refer to the otolaryngologist. This article begins by focusing on tinnitus, then on hyperacusis and a brief section on unexplained unilateral otalgia, followed by a discussion of hearing loss. The final two sections include a brief overview of audiometry and current hearing augmentation.

TINNITUS

Tinnitus is the perception of sound such as ringing, crickets, buzzing, or hissing that occurs in the absence of an external sound stimulus. The word comes from the Latin *tinnire* meaning *to ring*. Tinnitus is considered a symptom rather than a discrete disease entity, although, in many cases, the specific cause remains undetermined. Many people (12% to 30% of the general population) experience tinnitus and are aware of it but are not particularly bothered by it. About 0.5% to 3% of those with tinnitus find it intrusive and very distressing. The prevalence of tinnitus increases with age. Risk factors include hearing loss, increased age, male sex, noise exposure, family history of hearing loss or tinnitus, temporomandibular joint (TMJ) syndrome, eustachian tube dysfunction, ceruminous impaction of the external ear, hypertension, diabetes, and exposure to ototoxic drugs such as aminoglycosides, salicylates, loop diuretics, and quinine. TABLE 9-1 outlines some conditions associated with tinnitus.

Most tinnitus is subjective, meaning no source of the perceived sound can be identified and it is evident only to the individual. Objective tinnitus, which is much less common, is generated by vascular or other anatomic mechanisms that

TABLE 9-1 Conditions and Medications Associated With Tinnitus

Inner ear

- Bilateral sensorineural hearing loss
- Loud noise exposure (bilateral)
- Ménière disease (unilateral)
- Labyrinthitis (unilateral)
- Sudden sensorineural hearing loss (unilateral)
- Labyrinthine barotrauma (unilateral or bilateral)
- Autoimmune inner ear disease (bilateral)
- Superior canal dehiscence syndrome (unilateral or bilateral)[a]
- Pregnancy (bilateral)
- Hyperviscosity states (often bilateral)
- Sudden brief unilateral tapering tinnitus (unilateral)

Middle ear or external ear

- Otosclerosis (unilateral or bilateral)
- Ceruminous impaction (unilateral or bilateral)
- Cholesteatoma (unilateral)
- Eustachian tube dysfunction (unilateral or bilateral)[a]
- Stapedius or tensor tympani spasms (unilateral)

Nonotologic causes

- Stress, generalized anxiety (bilateral)
- Temporomandibular joint syndrome (unilateral or bilateral)

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occur in or near the ear and with proper methods may be audible to an external observer.

Pathophysiology of Tinnitus

The precise mechanism of chronic subjective tinnitus is still not well understood. In most cases, dysfunction affecting the peripheral auditory system with associated loss of cochlear hair cell function underlies tinnitus. The loss of the usual cochlear input that results from the lesion causes a frequency-specific increase in output to the central auditory pathways. The reduction of sound that results from reduced hearing causes the plastic changes in the brain to replace the missing auditory stimulation. In people with persisting tinnitus, impairment of the limbic system and central auditory processes seems to lead to reduced ability to suppress phantom sounds.[5] By at least one theory, tinnitus may be akin to denervation hypersensitivity or phantom limb pain, which occurs in the somatosensory system. At least the denervation hypersensitivity may lead to the initial tinnitus, whereas long-term persistence may be related more to the limbic system and central auditory processing.[5]

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- Palatal myoclonus (bilateral)
- Jugular vein venous hum (unilateral or bilateral)
- Idiopathic intracranial hypertension (bilateral)
- Vascular (arteriovenous malformation, turbulent flow) (unilateral or bilateral)
- Glomus jugulare paraganglioma (unilateral or bilateral)

Medications

- Loop diuretics (furosemide, ethacrynic acid) (bilateral)
- Aspirin, nonsteroidal anti-inflammatory drugs (bilateral)
- Hydroxychloroquine (bilateral)
- Quinine (cinchonism) (bilateral)
- Quinidine (bilateral)
- Cisplatin (bilateral)
- Methotrexate (bilateral)
- Gentamicin (bilateral)
- Azithromycin, clarithromycin (bilateral)
- Angiotensin-converting enzyme inhibitors (bilateral)

Metabolic

- Diabetes mellitus (bilateral)
- Hyperinsulinemia (bilateral)
- Hyperviscosity syndromes (bilateral)
- Pregnancy (bilateral)

[a] This condition is associated with autophony (ie, the increased hearing of one's own voice or internal bodily sounds).

TABLE 9-2 Causes and Common Descriptive Features of Tinnitus

Descriptive feature	Common patient descriptions	Causes/disorders
Unilateral tinnitus		
Constant continuous	Ringing, crickets, chirping, hissing, machine sound, cicadalike for most conditions; continuous roaring or humming for venous hum	Ceruminous impaction Unilateral sensorineural hearing loss Unilateral noise exposure Ménière disease Barotrauma Vestibular schwannoma Venous hum
Pulse synchronous	Throbbing, whooshing, thumping	Unilateral conductive hearing loss Idiopathic intracranial hypertension Carotid artery sounds transmitted (bruit, turbulence, dissection) Cardiac sounds transmitted such as valvular sound High-riding jugular bulb Superior canal dehiscence syndrome Glomus tumors Dural arteriovenous fistula, other arteriovenous malformation High-blood-flow states (pregnancy, hypertension, hyperthyroidism, anemia)
Pulse asynchronous	Sound of ocean waves, echoey, reverberating for patulous eustachian tube; manual typewriter, Morse code, machine gun for typewriter tinnitus; clicking, tapping, fluttering, or drumlike thumping for middle ear myoclonus	Patulous eustachian tube Typewriter tinnitus Middle ear myoclonus
Bilateral tinnitus		
Constant continuous (even if intermittent initially)	Ringing, crickets, chirping, hissing, machine sound, cicadalike	Idiopathic tinnitus Bilateral sensorineural hearing loss
Pulse synchronous	Throbbing, whooshing, thumping	Bilateral conductive hearing loss Many of the causes of pulse synchronous unilateral tinnitus may be reported by patients as being present bilaterally
Pulse asynchronous	Rhythmic clicking, tapping, or thumping	Palatal myoclonus/oculopalatal myoclonus

That is, in some individuals, the thalamic-paralimbic inhibitory feedback loop is insufficient to suppress the tinnitus. Over a period usually of days to weeks, the loss of thalamic inhibitory gating permits the auditory signals to cause synchronization and reorganization of cortical networks in the frontal, parietal, and limbic regions which may lead to perpetuation of chronic tinnitus based on central generators.[6,7] Consequently, chronic tinnitus cannot be eliminated even by severing the auditory nerve(s).[8] In addition, other medical and psychological factors influence how an individual may react to and cope with tinnitus. Functional MRI (fMRI) in 11 patients with tinnitus and 11 control subjects showed hyperactivity especially in the nucleus accumbens, a part of the corticostriatal pathways involved in reward and aversion responses and emotion. This system influences which sounds are important and which may be suppressed from consciousness. Tinnitus may in some ways mirror what happens with certain mood disorders and chronic pain.[5]

The mechanisms for objective forms of tinnitus are often easier to understand because mechanical processes generate the sound. For example, turbulent flow in a vessel or arteriovenous malformation (AVM) is transmitted to the cochlea via bone conduction. Although this type of tinnitus is the least common, it is often the most concerning because it is sometimes due to an underlying serious structural lesion such as a dural arteriovenous fistula (AVF), AVM, glomus tumor, or carotid artery dissection.

KEY POINTS

- Among the most serious causes of unilateral pulse-synchronous tinnitus are a dural arteriovenous fistula, arteriovenous malformation, or a glomus tumor arising from the jugular foramen or middle ear.
- Many forms of unilateral and bilateral tinnitus are more bothersome to patients with coexistence of depression, insomnia, stress, anxiety, and excessive caffeine use.

History and Examination of Tinnitus

In taking the history of a patient with tinnitus, the neurologist should ascertain the character and duration, whether the tinnitus is unilateral or bilateral, whether it is constant or intermittent, and whether it is triggered or modulated by any circumstances. It is also potentially helpful to have a description of the character of the tinnitus such as ringing, crickets, chirping, hissing, cicadalike, or low pitched (TABLE 9-2). The character description may have some use but does not often distinguish the mechanism. The clinician should inquire whether the tinnitus is caused or modulated in pitch or loudness by jaw clenching, neck movements, eye movements, or pressure on certain areas of the head or neck. When tinnitus is strongly linked to somatic triggers, such as jaw clenching, muscular contraction, or mechanical activities, it is referred to as *somatic tinnitus*.

Finally, determining the perceived severity and life impact of the tinnitus may aid in gauging psychosocial stressors and the severity of the symptom. Patient-rated scales, such as the Tinnitus Handicap Inventory that is scored on a scale of 0 to 100, help rate the impact on the patient's life as the patient perceives it. Nearly all forms of tinnitus are made more noticeable by comorbidities such as depression, insomnia, stress, anxiety, and excess caffeine use. In musical ear syndrome, a patient with hearing loss hears music or melodies. Although the precise cause is not known, it may be that sound deprivation results in a substitution with musical sounds from auditory association areas of the brain. This may be analogous to Charles Bonnet syndrome in which visual deprivation results in visual hallucinations. Hearing well-formed words and voices suggests auditory hallucinations due to psychosis rather than tinnitus.

Examination for tinnitus should include otoscopy to assure that the ear canal is not impacted with cerumen, which can be a cause of tinnitus and reduced

hearing. Limitation of jaw opening to less than 4 cm with clicking or popping or TMJ joint crepitus may suggest TMJ syndrome, which is associated with tinnitus. Funduscopic examination for papilledema can be a clue for idiopathic intracranial hypertension which may cause pulsatile tinnitus.

Pulsatile tinnitus may be pulse-synchronous or pulse-asynchronous. Whether the tinnitus is subjective (the examiner does not hear it) or objective (audible to the examiner) can depend on the examiner's own hearing, the location (s) auscultated, the environmental quietness, and the quality of the listening tools. For example, cursory auscultation with a poor-quality stethoscope in a noisy clinic setting may miss what is evident when using an electronic noise-canceling stethoscope in a quiet room. Particularly for pulse-synchronous tinnitus, auscultation should include the skull, temples, periauricular regions, carotid arteries, and heart because a dural AVF or other AVM may produce an audible bruit. The distinction between subjective and objective tinnitus seems prone to misclassification depending on the quality of the auscultation effort.

Classification of Tinnitus

Widely accepted classifications of tinnitus are few, but TABLE 9-3 offers one simple classification, and TABLE 9-2 outlines some of the common descriptions and related causes of tinnitus. In addition, as mentioned earlier, tinnitus can be pulse-synchronous or pulse-asynchronous; constant or intermittent; acute, subacute, or chronic; and unilateral or bilateral.

Other categorizations are divided among primary (idiopathic) and secondary (due to an identified condition); based on the grade of psychological or functional severity; or peripheral (originating in the cochlea) versus central (dysfunction within the central nervous system auditory processing pathways) causes. By far, the most common form of tinnitus is bilateral high-pitched ringing, crickets, or

TABLE 9-3 Classification of Tinnitus[a]

Unilateral

- **Constant or intermittent nonrhythmic (most common type)**
- **Pulse-synchronous (pulsatile)**
- **Pulse-asynchronous (rhythmic but not tied to heartbeat)**
  - Clicking/staccato
  - Hum/low-pitched

Bilateral

- **Constant or intermittent nonrhythmic (most common type)**
- **Pulse-synchronous (pulsatile)**
- **Pulse-asynchronous (rhythmic but not tied to heartbeat)**
  - Clicking/staccato
  - Hum/low-pitched

[a] Within this scheme, tinnitus may be intermittent or constant. Duration may be further characterized as acute (less than 3 months), subacute (3 to 6 months), or chronic (longer than 6 months).

cicada-sounding tinnitus. From the neurologist's point of view, the unilateral and pulse-synchronous forms are of greatest concern for a structural or neurologic origin.

KEY POINT

● Otologic causes of unilateral pulse-synchronous tinnitus include ceruminous impaction and middle ear disorders leading to conductive hearing loss such that bone-conducted sounds from vascular and other internal structures are heard more loudly.

Constant or Intermittent Nonrhythmic Tinnitus

Most tinnitus is a steady, sustained sound that has no rhythmic or staccato features. It may come and go and be more noticeable in quiet environments but does not pulsate or have interruptions in the sound. This category accounts for the large majority of tinnitus experienced by humans. It is often associated with some degree of high-frequency hearing loss but sometimes occurs in patients with normal hearing or precedes hearing loss by several years.

Pulsatile Tinnitus

Pulsatile tinnitus refers to tinnitus that has a rhythmic, periodic sound that alternates between louder and softer or that has discrete interruptions with periods of silence between the sounds. Pulse-synchronous means the pattern of pulsation occurs with the same timing and tempo as the individual's pulse and heartbeat. Pulse-asynchronous refers to a less common category in which tinnitus has a rhythmic recurring quality but does not correlate with heart rate or pulse. Many of the causes of pulse-synchronous tinnitus are benign and simply related to turbulent flow in arteries or transmitted heart sounds. Sometimes, however, serious conditions may present in this manner, and it is important not to overlook these cases. Pulsatile tinnitus can be unilateral or bilateral.

UNILATERAL CONSTANT OR INTERMITTENT NONRHYTHMIC TINNITUS. The most common cause of unilateral tinnitus is ceruminous impaction of the external ear canal. Once this is excluded by otoscopy, other common causes include prior unilateral excessive noise exposure, barotrauma, unilateral hearing loss, sudden sensorineural hearing loss, or viral labyrinthitis as examples. Structural lesions such as vestibular schwannoma may present with unilateral tinnitus, but it is most commonly also associated with slowly progressive ipsilateral hearing loss, as well. Ménière disease may sometimes present with fluctuating hearing or ear fullness and unilateral tinnitus that may vary in pitch and loudness (CASE 9-1).

Unilateral tinnitus not infrequently is simply the asymmetrical onset of what will eventually be bilateral tinnitus after several years. This same pattern may occur or develop in conjunction with unilateral hearing loss that later becomes bilateral.

A venous hum may cause unilateral tinnitus that may be high or low pitched and sometimes roaring or rough tinnitus that is often unilateral but occasionally described as bilateral. It may occur in adults or children and results from turbulent flow in the internal jugular vein. The hum is usually present throughout the cardiac cycle but may fluctuate and can be made to abate by compression of the internal jugular vein, the Valsalva maneuver, turning the head, or lying flat. It may be audible with auscultation of the neck just above the clavicle, more often on the right than on the left. A venous hum may be annoying to the patient, but it is not a harmful condition and needs no treatment.

UNILATERAL PULSE-SYNCHRONOUS TINNITUS. Unilateral conductive hearing loss sometimes results in pulsatile tinnitus due to the lack of masking effect from air-conducted sound in the ear affected with hearing loss. The absence of

external sounds heard by that ear may make one's own voice and internal heart and vascular sounds more apparent. Idiopathic intracranial hypertension and obstructive hydrocephalus (eg, due to aqueductal stenosis) are commonly accompanied by unilateral and, less often, bilateral pulsatile tinnitus.[9,10] It is caused by elevated intracranial pressure creating changes in arterial flow and venous draining leading to turbulence audible to the patient.

Carotid artery blood flow is another fairly common cause of pulse-synchronous tinnitus as irregular flow creates sound that is transmitted to the ear so that the patient perceives the sound. Atherosclerosis-related carotid bruits, carotid artery dissections, tortuous carotid siphon, and, less commonly, fibromuscular dysplasia may all be causes. Occasionally, conditions that cause high-flow states such as pregnancy, hypertension, anemia, or hyperthyroidism can cause

CASE 9-1

A 37-year-old man was referred by another neurologist because of 3 years of constant tinnitus that periodically fluctuates in pitch and loudness along with reduced hearing in the left ear. Although he seemed most focused on the tinnitus, when queried about other symptoms, he mentioned about eight episodes of vertigo over the previous 3 years usually lasting several hours. He had a history of migraine managed adequately with onabotulinumtoxinA, verapamil, and nortriptyline.

Brain MRI of the internal auditory canal with and without contrast performed 3 years ago because of migraine headaches and the left-sided tinnitus with hearing loss was normal.

Examination was normal except for reduced hearing in the left ear during bedside screening tests. His audiogram (FIGURE 9-1) showed low-frequency left-sided hearing loss characteristic of left Ménière disease.

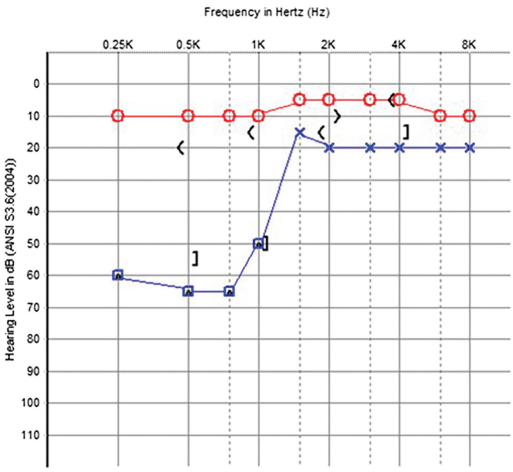

FIGURE 9-1
The audiogram for the patient in CASE 9-1 demonstrates low-frequency left-sided hearing loss characteristic of Ménière disease.
ANSI = American National Standards Institute, which sets specifications for audiometers.

COMMENT

This patient has left-sided Ménière disease and was referred primarily for the left-sided tinnitus. Most patients with Ménière disease are mainly concerned with the vertigo attacks if they have had them, so it is unusual that the patient did not mention it until he was asked if he had ever had vertigo. This patient also had unilateral constant left-sided tinnitus, but it would periodically fluctuate in pitch and loudness, which is a clue for Ménière disease.

increased flow through the heart or carotid arteries. Less often, heart sounds such as murmurs are loud enough to be perceived by patients. In superior canal dehiscence, patients have a heightened sense of hearing of internal bodily sounds (autophony), which may cause pulse-synchronous tinnitus. Jugular bulb abnormalities, such as a high-riding jugular bulb due to the proximity between the jugular bulb and the cochlea, may also cause unilateral pulse-synchronous tinnitus.[11]

Glomus tumors are rare paragangliomas that are derived from embryonic neural crest cells that make up chemoreceptors such as the carotid bodies. These tumors most commonly affect women in their forties and fifties; they are vascular and usually benign but may be locally invasive and may erode through bone. Those that present with hearing loss and pulsatile tinnitus usually originate in the jugular foramen (ie, glomus jugulare) or the middle ear (ie, glomus tympanicum). Sensorineural or conductive hearing loss and pulsatile tinnitus are common presenting symptoms.

Dural AVFs and, to a far lesser extent, other AVMs may cause and even present solely with unilateral pulse-synchronous tinnitus.[12] Dural AVFs are abnormal connections between branches of dural arteries and dural veins or venous sinuses. The most common locations of dural AVFs that result in pulsatile tinnitus are in the transverse, sigmoid, and cavernous sinuses. Most of the time, the tinnitus is unilateral (CASE 9-2), but if the dural AVF is near the midline, tinnitus may be reported as seeming to be bilateral. The most common cause in adults with an acquired dural AVF is venous sinus thrombosis and venous hypertension.[13] Treatment is neurosurgical, often consisting of endovascular coiling or embolization.[14]

CASE 9-2

A 57-year-old woman presented with 1 year of left-sided pulsatile tinnitus that correlated with her heartbeat along with reduced hearing in the left ear, all with gradual onset and progression. Auscultation of the left ear and periauricular region was unrevealing, and the otologic and neurologic examinations were normal except for left-sided reduced hearing. Digital compression of the left internal jugular vein just above the clavicle and just lateral to the carotid artery pulsation diminished the tinnitus.

An audiogram that had been conducted before the visit showed low-frequency conductive hearing loss on the left. Brain MRI showed a left occipitotemporal dural arteriovenous fistula (AVF) in which the middle meningeal artery and transosseous branches of the left occipital artery drained into the sigmoid sinus and petrous sinuses. She was referred to neurosurgery, and endovascular embolization occluded the dural AVF with resolution of the tinnitus.

COMMENT

This case illustrates how a dural AVF may present with unilateral pulsatile tinnitus. Dural AVFs commonly present with pulsatile tinnitus and are important to recognize because they confer an annual mortality rate from hemorrhage approaching 10%.

UNILATERAL PULSE-ASYNCHRONOUS TINNITUS. A patulous (distended or wide-open) eustachian tube can lead to rhythmic roaring tinnitus that is usually synchronous with respiration. The eustachian tube is a small mucosa-lined fibrocartilaginous tube connecting the nasopharynx to the middle ear that serves to equilibrate pressure between the middle ear and the outside world. It is approximately 3.5 cm long and with a lumen of approximately 1.5 mm to 2.5 mm. The eustachian tube is usually closed but opens with swallowing and yawning by contraction of the tensor veli palatini muscle. Failure of this tube to open leads to increased or decreased middle ear pressure resulting in muffled hearing and often a feeling of fullness or pressure. A patulous or persistently open eustachian tube leads to a hollow roaring kind of sound that modulates with respiration and swallowing. Sometimes, when the eustachian tube fails to open regularly or completely (the opposite of a patulous eustachian tube), cracking sounds or the sound of fluid can be heard in the ear on the affected side.

So-called *typewriter tinnitus* is a form of unilateral random and intermittent tinnitus that produces a sharp distinct series of staccato sounds likened by patients to a "machine gun," "Morse code," or a "tapping" sound.[15] Possible causes include microvascular loop compression, Ehlers-Danlos syndrome, and basilar invagination due to conditions such as osteogenesis imperfecta or Paget disease. This type of unilateral rhythmic tinnitus may respond to carbamazepine, oxcarbazepine, gabapentin, or pregabalin.[16]

Middle ear myoclonus is an uncommon cause of unilateral pulse-asynchronous tinnitus, often described by patients as intermittent salvos of unilateral fluttering, thumping, or buzzing (CASE 9-3). It is thought that the stapedius and tensor tympani muscles rhythmically contract, causing these sounds typically at a rate of 1 to 2 per second. That can cause visible movement of the tympanic membrane.[17] The stapedius muscle has its origin on the posterior wall of the temporal bone in the middle ear and attaches to the neck of the stapes; it dampens sounds transmitted through the ossicular chain and is innervated by the facial nerve. The tensor tympani muscle has its origin in the cartilaginous and bony edges of the auditory tube and inserts on the handle of the malleus and, like

CASE 9-3

A 38-year-old woman developed bothersome clicking/thumping tinnitus in her right ear over the past 2 years; it was initially intermittent but was daily over the last year. The clicking would momentarily diminish with mouth-opening or swallowing, but it was otherwise not influenced by activity; however, sometimes when lying with her right ear down, the clicking was bilateral. Brain MRI was normal, she had received several normal otolaryngology evaluations, and several audiograms were normal.

Neurologic examination was normal and synchronous movements of the tympanic membrane were not seen. Nevertheless, clicking near the right ear could be heard by the examiner in a quiet room.

COMMENT

This case illustrates a patient with palatal or middle ear myoclonus, but it is indeterminate whether it was due to myoclonus of the stapedius, tensor tympani, or tensor veli palatini muscle.

the stapedius muscle, dampens sounds through the ossicular chain and moves the tympanic membrane during yawning and swallowing. It is innervated by the medial pterygoid nerve of the trigeminal nerve. Treatment regimens for these conditions are poorly delineated in the literature but might include trials of clonazepam, baclofen, carbamazepine, oxcarbazepine, or onabotulinumtoxinA administration into the muscle affected assuming it can be ascertained by visual observation.

A similar phenomenon may also occur with the tensor veli palatini and is referred to as *palatal myoclonus*, which can be unilateral or bilateral. Palatal myoclonus is discussed in more detail later in this article.

BILATERAL CONTINUOUS OR INTERMITTENT, NONRHYTHMIC. This category accounts for the tinnitus that most people experience. It may begin as intermittent bilateral tinnitus and, over time, become constant. It is always subjective tinnitus, meaning it is audible only to the patient. The tinnitus may consist of high-pitched ringing, crickets chirping, hissing, or a cicadalike or machinelike sound or, less commonly, a low-pitched or musical sound. This type of tinnitus is very commonly associated with sensorineural hearing loss but may precede hearing loss yet to come by 1 or 2 years.

MANAGEMENT. Although this is a very common problem, evidence-based effective treatments are limited, and many remedies offered are not supported by evidence, despite claims to the contrary. Of course, the first step in treatment is to minimize aggravating factors such as medications that cause tinnitus, including those listed in TABLE 9-1. Patients should also reduce exposure to loud noises, and insomnia and stress should be managed through counseling, lifestyle changes, and possibly pharmacotherapy. Medications that are sometimes used with anecdotal success include antidepressant medications and anxiolytic medications such as benzodiazepines, which probably work mostly because they control anxiety or mood disorders. Pentoxifylline, gabapentin, pregabalin, papaverine, and clopidogrel have also been used anecdotally but have not been found effective by controlled trials. If somatic tinnitus contributions such as neck pain are present, these can be managed, sometimes indirectly improving tinnitus. TMJ syndrome may be treated if present, and massage relaxation techniques or acupuncture may be tried. Counseling, tinnitus retraining therapy, and cognitive-behavioral therapy are supported by weak evidence that they can reduce the negative impact on quality of life for patients with tinnitus.[18,19] Combined hearing aids and sound generators are commonly used but appear to offer little or no difference in tinnitus symptom severity by evidence-based standards.[20] Masking devices may result in some temporary improvement, but studies published to date have not found a significant change in the severity of tinnitus compared with the use of relaxation and other coping strategies.[21] Deep brain stimulation or neuromodulation has been preliminarily explored to treat tinnitus but further study is needed.[22]

BILATERAL PULSE-SYNCHRONOUS. Several of the conditions that may cause pulse-synchronous unilateral tinnitus may sometimes present as bilateral tinnitus, including intracranial hypertension, glomus tumors, and conditions that result in increased blood flow states. Superior canal dehiscence can also cause patients to perceive bilateral pulsatile tinnitus. Bilateral conductive hearing loss

KEY POINTS

- Unilateral pulse-synchronous tinnitus may be caused by sounds transmitted from the carotid artery (bruit) or the heart (murmur), increased intracranial pressure, or conditions that cause high-flow states such as pregnancy, anemia, and hyperthyroidism.
- "Typewriter tinnitus" is a term for a pulse-asynchronous tapping or Morse code–like staccato tinnitus that may respond to treatment with carbamazepine, oxcarbazepine, gabapentin, or pregabalin.
- Myoclonus of the stapedius or tensor tympani may cause a benign type of unilateral fluttering or thumping tinnitus that is rhythmic but not synchronous with the heart rate.
- Bilateral high-pitched subjective tinnitus that is constant but varies with ambient noise is the most common form of tinnitus and is often associated with some degree of sensorineural hearing loss.

KEY POINT

● Palatal myoclonus and oculopalatal myoclonus cause an objective clicking sound audible to the patient and others and that persists during sleep.

may also result in pulsatile tinnitus because air-conducted sound no longer masks internal heart sounds.

BILATERAL PULSE-ASYNCHRONOUS. Some of the causes for unilateral pulse-asynchronous tinnitus may rarely be perceived by the patient as bilateral and simultaneous, but this is quite uncommon.

Palatal myoclonus and oculopalatal myoclonus comprise a rhythmic involuntary movement of the uvula and soft palate, and often a clicking sound is heard both by patient and examiner, usually at a frequency of 0.5 Hz to 2 Hz. Because it is midline, it is often perceived by the patient as bilateral. In the case of oculopalatal myoclonus, the soft palate movements are synchronous with pendular torsional or vertical bobbing eye movements in one or both eyes and may be seen after stroke or with degenerative disorders, encephalitis, neoplasm, or other conditions that disturb the Guillain-Mollaret triangle (FIGURE 9-2). Oculopalatal myoclonus persists during sleep. Isolated palatal myoclonus in the absence of structural causes and eye movement findings is referred to as *essential palatal myoclonus*. Some individuals with essential palatal myoclonus can exert some volitional control over the movements.[23] Treatment for oculopalatal myoclonus and essential myoclonus has variable success using benzodiazepines, valproic acid, baclofen, or onabotulinumtoxinA injected into portions of the soft palate and in the tensor veli palatini.

Diagnostic Evaluation of Tinnitus

A workup for tinnitus should include an audiogram. Matching tinnitus pitch and loudness can be achieved by having the patient choose from among various sound frequencies that most resemble the tinnitus in pitch and loudness. Patients can also match the tinnitus through a variety of online tinnitus-matching websites. Although this is of interest, it rarely helps management strategies except perhaps for some sound-based interventions. Serum laboratory tests may include thyroid function, antinuclear antibody, fluorescent treponemal antibody absorption, complete blood cell count, fasting glucose or hemoglobin A_{1c}, and lipid panel and an assessment for serum zinc deficiency.

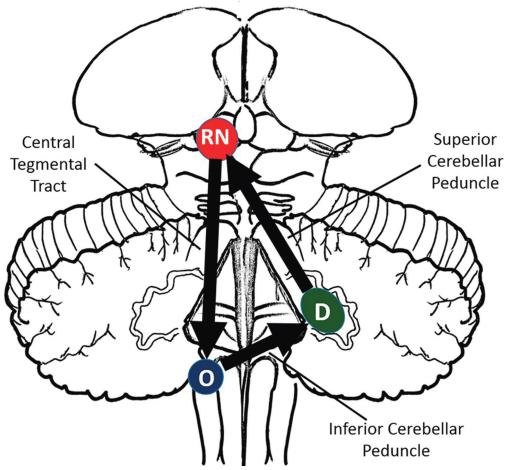

FIGURE 9-2
Guillain-Mollaret triangle structures and pathways.
D = dentate nucleus of the cerebellum; O = olivary nucleus of the medulla; RN = red nucleus.

Guidelines on imaging studies are outlined in TABLE 9-4.[24] As noted in TABLE 9-4, the workup for pulse-synchronous tinnitus should be directed at excluding dural AVF, AVM, and glomus jugulare and glomus tympanicum usually by CT angiography of the head and neck or even cervicocerebral digital subtraction angiography. This is in contrast to constant bilateral tinnitus for which imaging has low diagnostic value.[24]

In the absence of vestibular symptoms or imbalance, vestibular testing such as videonystagmography is rarely helpful. Vestibular-evoked myogenic potential (VEMP) testing (cervical or ocular) may be helpful if pulse-synchronous tinnitus is associated with other forms of autophony and superior semicircular canal dehiscence syndrome is suspected. For more information about superior semicircular canal dehiscence syndrome, refer to the article "Selected Otologic Disorders Causing Dizziness" by Gail Ishiyama, MD,[25] in this issue of *Continuum*.

HYPERACUSIS

Hyperacusis is an abnormally low tolerance for ordinary environmental sounds. A small subset of patients who develop hyperacusis have no known underlying

Imaging Studies for Tinnitus[a]

TABLE 9-4

Subjective or objective pulsatile tinnitus (not due to myoclonus)

- **Usually indicated (favorable risk-benefit ratio for patients)**
  - CT angiography (CTA) head and neck with contrast
  - Cervicocerebral CT and head venogram with contrast
- **Sometimes indicated (equivocal risk-benefit ratio for patients)**
  - Cervicocerebral angiography
  - Temporal bone CT without contrast
- **Usually not indicated (risk-benefit ratio for patients is likely to be unfavorable)**
  - Temporal bone CT with and without contrast

Asymmetric or unilateral subjective nonpulsatile tinnitus[b]

- **Sometimes indicated (equivocal risk-benefit ratio for patients)**
  - Temporal bone CT with or without contrast
  - Head and neck CTA with contrast
- **Usually not indicated (risk-benefit ratio for patients is likely to be unfavorable)**
  - Temporal bone CT with and without contrast
  - Cervicocerebral angiography

Symmetric bilateral subjective nonpulsatile tinnitus[c]

- **Usually not indicated (risk-benefit ratio for patients is likely to be unfavorable)**
  - Temporal bone CT with and without contrast
  - Head CT venogram with contrast
  - Head and neck CTA with contrast
  - Head CTA with contrast
  - Cervicocerebral angiography
  - Temporal bone CT without contrast

CT = computed tomography.
[a] Data from Expert Panel on Neurologic Imaging, J Am Coll Radiol.[24]
[b] Normal otologic examination, no hearing asymmetry, no neurologic deficits, no history of trauma.
[c] No hearing loss, no neurologic deficit, no history of trauma.

cause but nevertheless find ordinary sound excessively loud and bothersome, depending on pitch and loudness. For patients with hyperacusis, any sound beyond some threshold evokes negative reactions. Reactions to sound may include discomfort, dislike, irritation, anxiety, panic, fear, and ear pain and may result in depression and anxiety. Sometimes, this sound threshold is so low that it approaches the hearing threshold. A hallmark feature of hyperacusis is that the loudness of sounds that bother these patients does not bother most other people (CASE 9-4). Its cause is not clear but is believed to be due to a disorder of the central auditory pathways with effects on and reinforcement by the limbic and autonomic systems and not due to any primary inner ear process.[26] The best treatment strategies have not been established but have included sound avoidance, cognitive-behavioral therapy and desensitization, and cognitive-behavioral therapy combined with management of any comorbid anxiety and depression.

Not all aversion to loud sounds is hyperacusis. Many people with bilateral hearing loss, and particularly those who wear hearing aids, may find that some sounds seem excessively loud whereas others are not heard well, but this is not hyperacusis in the strict sense. Medical disorders have been linked to increased sound sensitivity including Williams syndrome, Bell's palsy

CASE 9-4

A 34-year-old woman was seen in consultation for 4 years of bilateral severe hypersensitivity to sound. She had a history of falling on the ice while figure skating near the time of the onset of symptoms but landed on her buttocks and did not strike her head. She recalled a reverberating sensation go through her ears, but she seemed to recover uneventfully. The sound sensitivity started gradually and had gotten much worse, causing her to seek accommodation from her employer to work from home. She had seen four otolaryngologists, an otologist, and three neurologists. She wore noise-canceling headphones most of the time after trying sound desensitization and finding it intolerable. Sounds such as people talking, background voices, clanging dishes and silverware, traffic, and doors opening and closing, as well as some air conditioner sounds and electrical hums, all bothered her, causing ear pain, anxiety, and depression.

Her examination was normal except for some erosion of the auricle and bulbous deformity of the ear cartilage on both sides (cauliflower ears) from the continued headphone use. MRI, multiple audiograms, temporal bone CT, vestibular-evoked myogenic potential testing, otoacoustic emissions, auditory brainstem responses, and routine laboratory tests including thyroid values had all been normal.

COMMENT

This case illustrates the extreme disability encountered by some patients with hyperacusis. This patient was referred to both a psychiatrist and a psychologist for a combined treatment approach to improve her function and to help her discontinue her constant headphone use.

(unilateral, due to stapedius paresis), and Lyme disease. Sound sensitivity is found sometimes in the aftermath of concussion, during active migraine events, during benzodiazepine withdrawal, and in some patients with depression, but these are more in keeping with phonophobia, the fear of or aversion to loud sounds.

Misophonia is a form of selective dislike of a specific type of sound to the point that a negative emotional reaction to it occurs. Examples can be anxiety from the sound of balloons, clicking of a pen, foot-tapping, whistling, chewing, or swallowing. Unlike hyperacusis, which is sensitivity to sounds of certain frequencies and loudness, misophonia is usually the dislike of a specific type of sound and evokes emotional responses such as disgust, irritation, feeling trapped, rage, panic, anxiety, or fear. Most patients with misophonia learn to function, but a smaller number develop severe avoidance behaviors and functional limitations. The cause is not known, but one theory is that patients may become sensitized from previous experiences, and repeated exposure creates enhancement of synaptic connections to the limbic system and emotional memory.[27] Proneness to misophonia has been linked to higher personality traits for anxiety.[28] Treatment consists of cognitive-behavioral therapy (CASE 9-5).

IDIOPATHIC UNILATERAL OTALGIA

Occasionally, patients are referred to a neurologist due to idiopathic otalgia (ear pain), most commonly affecting one side. Otalgia may be primary, meaning it arises from ear structures, or secondary, meaning it is either referred from or generated elsewhere. Most such referrals initially go to primary care doctors and otolaryngologists, so referral to neurology usually means no explanation such as

CASE 9-5

A 24-year-old university graduate student returned home to live with his parents so he could take his classes online due to the COVID-19 pandemic. He was seen yearly by his neurologist for migraine headache and migrainous dizziness managed adequately with diet, exercise, and infrequent use of low-dose diazepam but mentioned another complaint. He noted that he was exceptionally bothered by the sound of other people swallowing. He did not seem affected by the sound of his own swallowing but was particularly bothered by hearing either of his parents swallow. He was sometimes annoyed by the sounds of his friends swallowing but felt his parents swallowed very loudly to the point that he had to leave the room because it made him feel nauseated. He periodically saw a psychologist for stress management who diagnosed this as misophonia.

COMMENT

This case illustrates a common form of misophonia in a young adult. This patient was provided with stress-reduction techniques and education, but no specific therapy was directed at the misophonia itself. The patient eventually returned to his university and took his classes online from a residence near the campus.

otitis or other evident structural cause can be found on examination or by temporal bone MRI or CT or both.

Anatomic Background

A variety of cranial nerves converges to supply sensation and function to the ear and periauricular region: (1) the auricle receives innervation from cranial nerves V, VII, X, and the second and third cervical nerve roots; and (2) the external ear canal, tympanic membrane, and middle ear are supplied from cranial nerves V, VII, and X.

After adequately excluding the common otologic causes such as eustachian tube dysfunction or inner, middle, or external ear disorders, causes of secondary or referred pain should be considered. Such causes may include oromandibular disorders including TMJ syndrome, hemicranial headaches, herpes zoster oticus, postherpetic neuralgia, giant cell arteritis, and neuralgia affecting trigeminal, geniculate, glossopharyngeal, sphenopalatine, occipital, and vagus nerves. Glossopharyngeal (vagoglossopharyngeal) neuralgia is a rare cause of severe pain in the back of the throat or side of the tongue or ear on one side, often in salvos of sharp pains similar to those experienced by patients with the far more common

TABLE 9-5 Selected Bedside Tests of Hearing and Signs or Phenomena

Test, sign, or phenomenon	How it is performed	Outcomes	Implication
Conversational hearing, finger rub	Note whether patient has trouble hearing	Normal: no evident hearing loss; abnormal: patient cannot hear examiner	Screening observation for hearing loss
Weber test[a]	Vibrating tuning fork is placed in the middle of the top of the patient's forehead	Normal: patient hears the tuning fork equally on each side; abnormal: patient hears the tuning fork louder on one side	Sound is louder on the side of conductive hearing loss and away from the side of sensorineural hearing loss
Rinne test[a]	Vibrating tuning fork is placed on the mastoid until no longer heard, then placed next to the ear on the same side	Normal: patient still hears the tuning fork when it is placed next to the ear (air conduction is better); abnormal: patient cannot hear the tuning fork when it is placed next to the ear (air conduction is worse than bone conduction)	If sound is heard better by bone than by air conduction, it indicates conductive hearing loss on that side
Tullio phenomenon	A loud sound or sustained sound is applied in the ear	Normal: no response; abnormal: vertigo/dizziness, possibly nystagmus	Seen in canal dehiscence, Ménière disease, perilymphatic fistula, luetic otitis
Hennebert sign	Application of positive or negative pressure to the sealed external ear canal	Normal: no response; abnormal: several beats of nystagmus	Seen in canal dehiscence, Ménière disease, perilymphatic fistula, luetic otitis
Autophony	Not elicited by the examiner but asked about in the history	Patient reports hearing internal bodily sounds	Increased hearing of one's own voice or internal bodily sounds (heartbeat, eye movements, chewing, joint sounds)

[a] A 512-Hz tuning fork, if available, is preferred over a 125-Hz or 256-Hz fork.

trigeminal neuralgia, but the pain is evoked by swallowing, talking, or coughing and sometimes associated with syncopal events when severe. Possible causes include microvascular compression, multiple sclerosis plaque at the root entry zone, tumors or nearby infections, local squamous cell carcinoma, or, less often, Eagle syndrome (stylalgia) in which the stylohyoid ligament stretches the glossopharyngeal nerve. Treatment is similar to that for trigeminal neuralgia with carbamazepine topping the list of medications and microvascular decompression or stereotactic radiosurgery being reserved for severe symptoms not responding to medications.

HEARING LOSS

When evaluating hearing loss, it is important to distinguish conductive hearing loss from sensorineural hearing loss.[29] Hearing loss can also be mixed (conductive and sensorineural). In conductive hearing loss, amplification and transmission of sound to the cochlea (external ear to the middle ear) are impeded. In sensorineural hearing loss, processing of neural impulses and their transmission centrally (inner ear, cranial nerve VIII, or rarely central pathways) are affected.

This section begins with otologic history and examination. Because vestibular and auditory pathways are anatomically closely located, vestibular examination is essential. Vestibular examination is discussed further in "Approach to the History and Evaluation of Vertigo and Dizziness" by Terry D. Fife, MD, FAAN, FANS,[30] and "Vestibular Testing" by Timothy C. Hain, MD, and Marcello Cherchi, MD, PhD, FAAN,[31] in this issue of *Continuum*. Conductive hearing loss and its common etiologies are briefly reviewed here. Sensorineural hearing loss is discussed in more detail, specifically how a neurologist should approach a patient with acute or chronic, unilateral or bilateral sensorineural hearing loss. These patients often present with dizziness or imbalance or both.

History and Examination of Hearing Loss

The clinician should start with a thorough history that includes the date of symptom onset and symptom description. Was the hearing loss gradual or sudden in onset; unilateral or bilateral; progressively worsening, stable, or fluctuating? Has dizziness been an associated symptom at any time? Does the patient have an associated history of ear pain, discharge, fullness, popping, autophony, hyperacusis, distortion of sound, or tinnitus? If tinnitus is present, is it bilateral or unilateral, high pitched or low pitched, pulsatile or nonpulsatile? If pulsatile, is it synchronous with the heartbeat or not? Past medical history should include prenatal, perinatal, and postnatal events; early childhood diseases; infections; head and acoustic trauma/surgery; noise exposure; and the use of ototoxic medications. Family history may be helpful, such as if a history of hearing loss or neurofibromatosis is noted.

TABLE 9-5 outlines some of the methods for bedside testing of hearing. Otologic examination starts with noting if the patient can hear the examiner's conversational voice in a quiet room and if any loss of hearing of a finger rub is present.

Weber and Rinne testing can then help with localizing unilateral hearing loss (conductive versus sensorineural). Weber testing is performed by placing a 512-Hz vibrating tuning fork in the middle of the patient's forehead. The stimulus travels through the bone to bilateral acoustic nerves. With normal hearing, the sound will not lateralize to either ear. The sound will radiate to the impaired ear with conductive hearing loss and away from the impaired ear with sensorineural hearing loss. Rinne testing is performed by placing a 512-Hz

KEY POINTS

- While phonophobia is an aversion to loud sounds as may occur during migraine headaches, hyperacusis is a rare disorder with constant intolerance to sounds of ordinary loudness that do not bother most people.
- Unilateral continuing otalgia without an evident structural cause can sometimes be attributable to hemicranial headache disorders, herpes zoster oticus, or postherpetic neuralgia, giant cell arteritis, or neuralgia of cranial nerves V, VII, IX, and X or of the sphenopalatine or greater occipital nerves.
- In a patient presenting with hearing impairment, the first step is to distinguish conductive from sensorineural hearing loss.

vibrating tuning fork on the mastoid process. When the sound is no longer heard by the patient, the tuning fork is placed in front of the auditory canal. If the sound is not heard in this position, bone conduction is better than air conduction, indicating conductive hearing loss. Air conduction would be expected to be stronger than bone conduction in an individual with normal hearing. It is recommended to mask the sound input to the ear that is not being tested by crinkling paper in front of that ear.

Otoscopic inspection of the auditory canal and tympanic membrane is essential. The examiner should assess for obstruction, otitis media, cholesteatoma, and vesicular eruptions suggestive of herpes zoster. If on otoscopy anything looks other than normal, a consultation with an otolaryngologist should be considered. Images of normal and pathologic tympanic membranes can be found at diagnosis101.welchallyn.com/otoscopy/educational-topics/ear-pathologies/.

Otologic examination is then followed by detailed vestibular examination. For more information about the vestibular examination, refer to "Approach to the History and Evaluation of Vertigo and Dizziness" by Terry D. Fife, MD, FAAN, FANS,[30] in this issue of *Continuum*.

TESTING. Audiometry, vestibular and laboratory testing, and imaging are used to confirm and further investigate the potential diagnosis.

AUDIOMETRY. Although bedside hearing tests have good specificity for detecting hearing loss, the sensitivity is poor when compared with an audiogram, so an audiogram is the optimal test for the assessment of hearing loss.[32] When ordering an audiogram, the suspected diagnosis should be mentioned, and air-bone gap testing should be requested because it is not always automatically included.

VESTIBULAR TESTING. Depending on the working diagnosis, videonystagmography with caloric testing and ocular and cervical VEMPs can reveal vestibular function information not detected during the bedside examination. The video head impulse test is a quantitative functional assessment of the function of each of the three semicircular canals on each side. The bedside head impulse test can help in detecting vestibular hypofunction related to the horizontal semicircular

TABLE 9-6 **Recommendations and Appropriateness of Initial Imaging for Acquired Sensorineural Hearing Loss[a]**

Procedure	Appropriateness category
Head and internal auditory canals MRI with and without contrast	Usually appropriate
Temporal bone CT with or without contrast	May be appropriate
Head CT with or without contrast, temporal bone CT with and without contrast, head CT angiography, magnetic resonance venography, head magnetic resonance angiography	Usually not appropriate

CT = computed tomography; MRI = magnetic resonance imaging.
[a] Data from Expert Panel on Neurologic Imaging, J Am Coll Radiol.[34]

canal on one or both sides. Vestibular testing is discussed in detail in the article "Vestibular Testing" by Timothy C. Hain, MD, and Marcello Cherchi, MD, PhD, FAAN,[31] in this issue of *Continuum*.

KEY POINT

● Vestibular schwannoma can occasionally present with sudden unilateral hearing loss, although it is usually more gradual and progressive.

LABORATORY TESTING. Most recent clinical practice guidelines for sudden hearing loss from the American Academy of Otolaryngology–Head and Neck Surgery do not recommend routine laboratory testing.[29] History and physical examination findings should guide a clinician to targeted laboratory testing, such as serologic testing when considering autoimmune and infectious diseases, and gap junction beta-2 protein for connexin 26 mutation, a common cause of autosomal recessive sensorineural hearing loss. Several other genes have been found to be associated with hearing loss.[33] For specific genes, refer to Human Phenotype Ontology Sensorineural Hearing Impairment at hpo.jax.org/app/browse/term/HP:0000407 and consult with a geneticist.

IMAGING. **TABLE 9-6**[34] outlines initial imaging studies for acquired sensorineural hearing loss as studies that are rated for appropriateness. Imaging should be performed to confirm or narrow the list of suspected diagnoses based on history and examination. Therefore, specific recommendations are listed in the discussion of different conductive and sensorineural hearing loss etiologies.

Conductive Hearing Loss

In conductive hearing loss, air transmission of sound waves to the inner ear endolymph is impeded. This results in an air-bone gap on the audiogram. With a gap of 15 dB or more, conductive hearing loss is suspected. Common etiologies of conductive hearing loss are listed in **TABLE 9-7**.[35-39]

Sensorineural Hearing Loss

In patients with sensorineural hearing loss, one should determine whether it is unilateral or bilateral, constant or episodic, or fluctuating. In patients with sudden sensorineural hearing loss, audiometry should be obtained as soon as possible (within 14 days of symptom onset). Retrocochlear pathology is best evaluated with brain MRI with attention to the internal auditory canal.[29] It is not recommended to order routine CT of the head in the initial evaluation or to obtain routine laboratory tests.[29,34]

SUDDEN UNILATERAL SENSORINEURAL HEARING LOSS. Most commonly, sudden unilateral sensorineural hearing loss is idiopathic. The most common etiologies of an abrupt-onset audiovestibular syndrome include labyrinthitis, Ménière disease, temporal bone fracture, and labyrinthine infarction. Hearing and vestibular loss are usually very gradual and insidious in cases of vestibular schwannoma but may be acute in approximately 1% of cases.[29]

IDIOPATHIC SUDDEN SENSORINEURAL HEARING LOSS. Isolated sudden sensorineural hearing loss without vestibular symptoms is referred to as idiopathic sudden sensorineural hearing loss and is most commonly unilateral. The cause is often undetermined, but most cases are believed to be due to viral infection. Despite the presumed viral cause, the majority of patients do not report preceding viral upper respiratory infection symptoms. Other less common causes include immune-mediated mechanisms; cochlear membrane rupture, as may occur with barotrauma; cochlear infarction; and possibly iron deficiency anemia. Treatment

with pulse methylprednisolone (1000 mg/d IV for 3 days) or intratympanic corticosteroids is recommended within the first 7 days of onset, but the earlier administration begins, the better.

LABYRINTHITIS. Labyrinthitis (vestibular neuritis plus unilateral sensorineural hearing loss) is typically of viral etiology and usually unilateral. Less often, bacterial, spirochetal, and autoimmune mechanisms may be involved. Systemic infections have greater likelihood of bilateral involvement. Bacterial labyrinthitis can be a complication of meningitis and otitis media. On examination, usually unilateral sensorineural hearing loss is present; nystagmus is unidirectional with the fast phase of nystagmus beating away from the impaired ear. The nystagmus adheres to the Alexander law, beating more intensely in the direction of the fast

TABLE 9-7 Etiology of Conductive Hearing Loss

Location	Etiology	Comments and/or typical presentations
External ear	Impacted cerumen	Common
	Foreign body	Immense ear discomfort and itchiness; on examination, a live spider lodged in the ear canal
	Auricular malformations	Congenital or posttrauma
	Infections	Long-standing left ear pain, itching, and poor hearing; on examination, edematous, erythematous, foul-smelling discharge; edema partially occluding ear canal and discharge occluding tympanic membrane
	Neoplasms	Poor hearing bilaterally; on examination, multiple sessile protrusions on the bony canal near the drum consistent with bony exostoses[35]
Middle ear	Effusion	Common; tympanic membrane often appears dull and yellow, with air fluid level (air bubbles) behind the drum seen during otoscopy
	Barotrauma	Can result in tympanic membrane perforation, hemotympanum
	Otitis media with or without cholesteatoma[36]	Chronic otitis media associated with eustachian tube dysfunction can result in acquired cholesteatoma; a cholesteatoma is an abnormal noncancerous but locally invasive collection of squamous cells behind the eardrum; these growths can extend into the middle ear and erode ossicles resulting in conductive hearing impairment or erode into the middle ear damaging the cochlea; on otoscopy, expect gray-white keratin mass on or behind the tympanic membrane; otorrhea is commonly purulent and can be foul smelling
	Neoplasm (rare)	Progressive left hearing loss and left pulsatile tinnitus; on examination, red pulsating mass behind the left tympanic membrane, a systolic bruit over the left mastoid, and left facial nerve involvement; diagnosis is glomus tympanicum (rarely metastasize; can also involve cranial nerves IX to XII); differential for red pulsating mass in the middle ear includes high-riding jugular bulb (conductive hearing loss from sound transmission through the middle ear), dehiscent jugular bulb, aberrant intratympanic carotid artery, and persistent stapedial artery[37,38]
	Otosclerosis	Abnormal bone remodeling of the otic capsule resulting in progressive fixation of stapes footplate in the oval window, with extension to the cochlea, where spiral ligament atrophies and hyalinizes with subsequent bilateral mixed hearing loss[39]; autosomal dominant with incomplete penetrance; vestibular symptoms sometimes occur after stapes surgery

phase and less during gaze away from the fast phase. Head impulse shows catch-up saccades ipsilesionally. No skew deviation, which is more suggestive of a central mechanism, should be observed.

LABYRINTHINE INFARCTION. The labyrinthine artery usually arises from the anterior inferior cerebellar artery (AICA) (rarely from the basilar artery) (FIGURE 9-3). The AICA also provides vascular supply to some vestibular nuclei, the lateral pons, middle cerebellar peduncle, flocculus, inferior lateral cerebellum, and facial nerve. The labyrinthine artery branches off at the labyrinth to supply the vestibulocochlear nerve, semicircular canals, utricle, and saccule. Depending on where in its course the AICA is occluded, it may cause a combination of symptoms, such as hearing loss, dizziness (central or peripheral vestibular), ipsilateral lower motor neuron facial palsy, ipsilateral limb ataxia, and postural imbalance, among other symptoms. Isolated labyrinthine ischemia is thought to be rare and may be difficult to distinguish from viral labyrinthitis (CASE 9-6). Sudden unilateral hearing loss thus could precede the impending infarction of the cerebellum or ipsilateral pontomedullary region. Occlusion of the posterior inferior cerebellar artery (PICA) can rarely cause similar audiovestibular symptoms. HINTS (Head Impulse, Nystagmus, Test of Skew) is sensitive and specific. When performed by an experienced clinician, it is superior to early MRI; early MRI may miss a small infarction because of slice thickness or because the MRI, if obtained early enough, may not yet show diffusion restriction.[40]

CHRONIC UNILATERAL SENSORINEURAL HEARING LOSS. This section focuses on adult-onset chronic unilateral sensorineural hearing loss. Ménière disease

KEY POINTS

- When acute unilateral hearing loss is present, labyrinthine ischemia should be considered, but a viral/inflammatory cause is considered to be more common.
- Occlusion of the posterior inferior cerebellar artery can rarely cause acute audiovestibular symptoms.
- If any part of HINTS (Head Impulse, Nystagmus, Test of Skew) suggests central localization, the etiology must be assumed to be central until proven otherwise.
- Isolated labyrinthine stroke is unlikely to be visible on standard MRI.

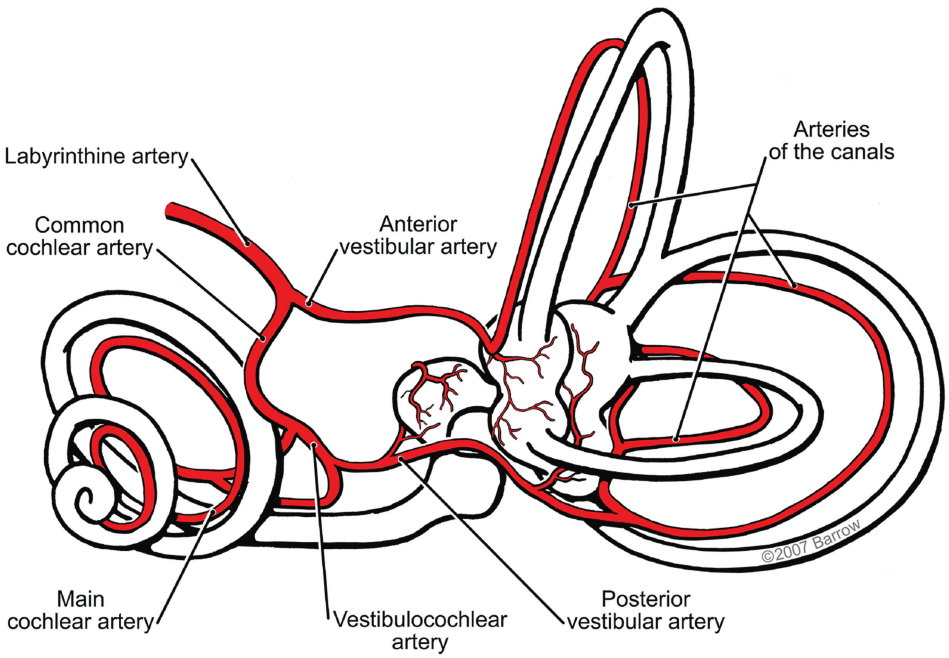

FIGURE 9-3
Vascular supply to the membranous labyrinth.
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(endolymphatic hydrops) and vestibular paroxysmia typically present with fluctuating auditory symptoms and episodic vertigo. Although Ménière disease symptoms typically last a few hours; in vestibular paroxysmia, the episodes are much shorter, lasting seconds to minutes at a time. Cochlear Ménière disease lacks vestibular symptoms, is rare, and is more difficult to diagnose. Systemic processes, such as autoimmune disease, may rarely cause endolymphatic hydrops, but it is more likely to be bilateral. In sarcoidosis, fluctuating sensorineural hearing loss and vertigo can occasionally be the only presenting symptoms. A chest radiograph to look for hilar adenopathy and referral to ophthalmology to look for uveitis should be considered. Hearing usually improves with steroid therapy. Acquired and congenital syphilis can present in adults as fluctuating or persistent unilateral or bilateral sensorineural hearing loss. Patients should be referred to ophthalmology where they can be evaluated for interstitial keratitis typically seen in congenital syphilis; patients should be evaluated for Hutchinson teeth, as well. Fluorescent treponemal antibody testing should be considered because venereal disease research laboratory testing is often normal. Testing and management of acquired syphilitic otitis are similar to that of neurosyphilis. Vestibular schwannoma, meningioma, ependymoma, cerebellar medulloblastoma, perineural spread of parotid gland adenoid cystic carcinoma and salivary duct carcinoma or cutaneous squamous cell carcinoma (in patients with a history of facial/scalp malignancy), and other cerebellopontine angle mass lesions more commonly present with slow progression of auditory symptoms and vertigo. Noise-induced hearing loss is not rare; middle and high frequencies are usually affected first, followed by sensorineural hearing loss across all frequencies with continued exposure. Although it is often bilateral, unilateral cases are possible.

CASE 9-6

A 68-year-old woman presented to the emergency department with an acute onset of vertigo, vomiting, imbalance, and muffled hearing on the left. Examination showed mild right-beating spontaneous nystagmus. Her head impulse test was without catch-up saccades (ie, normal). No skew deviation was present. The rest of the vestibular examination was normal. Tuning-fork testing was consistent with sensorineural hearing loss on the left. Her gait was ataxic. Brain MRI was normal. Her symptoms continued to persist, and, on reexamination the next day, she was found to have left upper extremity dysmetria, gaze-evoked nystagmus, and saccadic pursuit. Repeat MRI showed an acute cerebellar infarct consistent with an anterior inferior cerebellar artery (AICA) territory stroke.

COMMENT

This is a case of labyrinthine stroke (from the AICA occlusion) with additional posterior fossa ischemia that progressed beyond just the labyrinth by the next day to include the cerebellum. Completely isolated labyrinthine ischemia can occur, but depending on the location of the thrombosis, it may also involve the AICA distribution of the cerebellum and/or lateral pons.

MÉNIÈRE DISEASE (ENDOLYMPHATIC HYDROPS). For more information, refer to the article "Episodic Spontaneous Vertigo" by Scott D. Z. Eggers, MD,[41] and for the putative causes of Ménière disease, refer to the article "Selected Otologic Disorders Causing Dizziness" by Gail Ishiyama, MD,[25] in this issue of *Continuum*.

KEY POINT

● Ménière disease presents with an abrupt onset of recurrent vertiginous attacks with associated fluctuating unilateral low-frequency sensorineural hearing loss.

The typical presentation of Ménière disease includes recurrent spontaneous episodes of unilateral auditory symptoms such as hearing loss, ear fullness, and nonpulsatile tinnitus, with associated vertigo. Auditory symptoms may precede vertigo, occur during it, or follow it. Vertigo lasts at least 20 minutes and no longer than 24 hours. Although vertigo typically resolves within 2 to 6 hours, hearing loss may take longer to resolve, sometimes with incomplete recovery. In the early stages of disease, low- to midfrequency sensorineural hearing loss is more common. Hearing loss is usually progressive and permanent and eventually involves all frequencies. As auditory symptoms tend to fluctuate in the early stages of the disease, an audiogram should be done soon after the attack. The lack of sensorineural hearing loss on audiogram does not rule out Ménière disease, although, after a couple of years of recurrent symptoms, permanent sensorineural hearing loss on audiogram is expected. Delayed or absent cervical VEMPs can be seen in Ménière disease. Ordering antinuclear antibody, erythrocyte sedimentation rate, thyroid function, testing for Lyme disease, and fluorescent treponemal antibody tests should be considered because systemic processes may rarely cause endolymphatic hydrops.

In the majority of patients, vertiginous attacks will spontaneously remit. Preventive treatment is stepwise starting with a low-sodium diet, ideally less than 1500 mg/d, and a diuretic such as hydrochlorothiazide-triamterene 25 mg/37.5 mg. If vertigo attacks continue, consider high-dose betahistine (eg, 16 mg 3 times a day to 32 mg 3 times a day) therapy. Betahistine is used in Europe but is not approved in the United States; it can be ordered through a compounding pharmacy. At the onset of a typical Ménière attack, the patient can use promethazine, prochlorperazine, or low-dose benzodiazepines. Meclizine is less effective with severe symptoms. To allow central compensation, long-term use of vestibular suppressants is not recommended. Intratympanic dexamethasone shortly after the attack may reduce the frequency of the vertiginous attacks, although supportive evidence for this approach is weak. As the disease progresses, patients can experience abrupt drop attacks without loss of consciousness, as if someone pushed them to the ground (otolithic crises of Tumarkin). At this point of disease progression, intratympanic gentamicin injections may be considered; hearing loss is less likely with lower doses. In a minority of cases, the contralateral ear can become involved to cause bilateral Ménière disease, especially if caused by an autoimmune mechanism (TABLE 9-8). Most cases of Ménière disease remain unilateral. Of note, patients with Ménière disease are more likely to have coexistent vestibular migraine, especially with bilateral Ménière disease.

VESTIBULAR SCHWANNOMA. Vestibular schwannoma is a benign neoplasm derived from Schwann cells of the vestibular portion of cranial nerve VIII. Most vestibular schwannomas are sporadic and incidentally found on imaging. A few are associated with neurofibromatosis type 2, especially if bilateral vestibular schwannomas are present. In the internal auditory canal, a compressed cochlear

TABLE 9-8 Sensorineural Hearing Loss Etiologies

Toxic/metabolic

- Acetyl salicylic acid >2.5 g/d
- Alcohol
- Alkalizing agents
- Aminoglycosides (eg, gentamicin)
- Antivirals
- Benzodiazepines
- Bisphosphonates
- Chemotherapeutic agents
- Cocaine
- Ecstasy
- General anesthetics
- Heroin
- Immunosuppressive drugs
- Insecticides
- Loop diuretics
- Nonsteroidal anti-inflammatory drugs
- Organic mercury
- Pegylated interferon
- Retinoid
- Skeletal muscle relaxants
- Synthetic prostacyclin
- Uremia

Genetic (nonsyndromal are more prevalent)

- Alport syndrome
- Autosomal dominant cerebellar ataxia type 1
- Bone dysplasias
- Branchio-oto-renal syndrome
- Charcot-Marie-Tooth disease
- Chiari malformation
- Friedreich ataxia
- Gaucher disease
- *GJB2* mutation: the most common cause of nonsyndromic autosomal recessive hereditary hearing loss; *GJB2* encodes for the gap junction protein beta 2 connexin 26
- Hurler syndrome
- Jervell and Lange-Nielsen syndrome

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- **Mitochondriopathies:**
  - Mitochondrial encephalomyopathy with lactic acidosis and stroke like episodes
  - Progressive external ophthalmoplegia
  - Mitochondrial neurogastrointestinal encephalomyopathy
  - Myoclonic epilepsy with ragged red fibers
- **Neurofibromatosis type 2**
- **Pendred syndrome**
- **Perrault syndrome**
- **Refsum syndrome: hearing loss can present in adulthood**
- **Spinocerebellar ataxia 7**
- **Spinocerebellar ataxia 31**
- **Spinocerebellar ataxia 36**
- **Stickler syndrome**
- **Treacher Collins syndrome (sometimes hearing loss is due to microtia, atresia of the external ear)**
- **Usher syndrome**
- **Waardenburg syndrome**

Immune

- **Behçet disease: bilateral sensorineural hearing loss (rarely unilateral); vestibular symptoms can be the initial manifestation of the disease; mimics vestibular neuritis**
- **Cogan syndrome: recurrent episodic sensorineural hearing loss (unilateral or bilateral, fluctuating or progressively worsening over time, profound in half of the patients)**
- **Giant cell arteritis: unilateral sensorineural hearing loss can be an initial manifestation of the disease; it can progress in severity and can involve the other ear**
- **Granulomatosis with polyangiitis conductive hearing loss from obstruction of the eustachian tube by granulomas in the nasopharynx and/or sudden irreversible low-frequency sensorineural hearing loss, which can be an initial manifestation of the disease**
- **Guillain-Barré syndrome (rarely with hearing loss)**
- **Multiple sclerosis (rarely with hearing loss)**
- **Relapsing polychondritis: sudden unilateral or bilateral sensorineural hearing loss**
- **Rheumatoid arthritis: mostly sensorineural hearing loss**
- **Sarcoidosis: common sensorineural hearing loss (unilateral or bilateral), rare conductive hearing loss**
- **Sjögren syndrome: sensorineural hearing loss, rarely is an initial manifestation of the disease**
- **Systemic lupus erythematosus: sensorineural hearing loss, occasionally sudden in onset in patients with high titer of anticardiolipin antibodies**
- **Systemic sclerosis (scleroderma): sensorineural hearing loss, conductive hearing loss or mixed**
- **Vogt-Koyanagi-Harada disease: bilateral rapidly progressive sensorineural hearing loss**

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Infectious

- Bacterial meningitis (*Neisseria*, *Pneumococcus*, *Hemophilus*, *Mycobacterium tuberculosis*)
- Fungal (eg, cryptococcosis, coccidiomycosis)
- Spirochetal (eg, syphilis, *Borrelia*)
- Viral (eg, human immunodeficiency virus [HIV], herpes simplex virus, herpes zoster, cytomegalovirus), paramyxovirus (mumps)

Neoplastic

- Acoustic neurofibroma
- Cerebellopontine angle and petrous meningioma and metastasis
- Leptomeningeal carcinomatosis or lymphomatosis
- Meningeal carcinoma
- Myelodysplastic syndrome–associated hypercoagulability
- Neurolymphomatosis
- Vestibular schwannoma

Vascular

- Sickle cell crisis
- Stroke

Neurodegenerative

- Multiple system atrophy: unilateral or bilateral higher-frequency sensorineural hearing loss
- Parkinson disease: unilateral or bilateral higher-frequency sensorineural hearing loss

Iatrogenic

- Aminoglycoside or other ototoxic medication administration
- General anesthetic hemodynamic complication
- Microembolism from surgical complications
- Radiation

Other

- Bilateral Ménière disease
- Histiocytosis X: chronic bilateral
- Intracranial hypotension
- Superficial siderosis: chronic bilateral

portion of cranial nerve VIII results in progressive hearing loss and nonpulsatile tinnitus.[42] Facial paresthesia and weakness can be seen when larger tumors compress the nearby facial nerve. With cerebellopontine angle involvement, vestibular schwannomas may also compress the brainstem and cerebellum. Diagnosis is made through brain MRI with contrast with attention to the internal auditory canal, audiometry, and videonystagmography. Vestibular schwannomas are discussed further in the article "Vertigo Related to Central Nervous System Disorders" by Kamala Saha, MD,[43] in this issue of *Continuum*.

SUDDEN BILATERAL SENSORINEURAL HEARING LOSS (ONSET LESS THAN 1 MONTH). Bilateral sensorineural hearing loss is less often associated with vestibular symptoms and is less likely to recover compared with unilateral sudden sensorineural hearing loss.[44] Systemic processes, such as toxic, metabolic, immune-mediated, vascular, neoplastic, and infectious disease should be considered as a possible cause (TABLE 9-8).

Intracranial hypotension, whether spontaneous or from overshunting from a ventriculoperitoneal shunt, may be associated with audiovestibular symptoms. The sensorineural hearing loss can be unilateral or bilateral, or hearing may be distorted, but it is not usually a complete hearing loss.[45,46] Sensorineural hearing loss can involve all frequencies and is generally thought to be due to mild sagging of the brain and traction on the vestibulocochlear nerve(s) and disturbed perilymphatic-CSF dynamics within the labyrinth.

CHRONIC BILATERAL SENSORINEURAL HEARING LOSS. This is the most common category of bilateral sensorineural hearing loss. The three main causes of chronic sensorineural hearing loss in adults are (1) age-related hearing loss (FIGURE 9-4[47]), (2) heritable factors, and (3) noise exposure, which is the most common preventable cause. These three contributing causes are not mutually exclusive. In other words, as one ages, hearing diminishes in many people, possibly on a heritable basis, but noise exposure can accelerate the progression and severity of hearing loss. Nevertheless, some older people maintain excellent hearing, so it does not affect everyone equally as they age.

As mentioned earlier, sensorineural hearing loss is often associated with tinnitus. Hearing loss tends to make it more difficult to distinguish voices in the foreground from ambient background noise. Thus, it may be more difficult to hear on the telephone, in a moving car, in restaurants, and when many people are talking in a room, and certain sounds seem tinny and less rich and full. Certain parts of speech are harder to hear, such as consonant sounds including F, S, V, S, SH, and ZH (eg, the sound of the S in *measure*). Meanwhile, some sounds may seem especially loud or distracting. Finally, the direction from which a high-pitched sound emanates may be more difficult to ascertain.

The number of differential diagnoses for chronic bilateral sensorineural hearing loss is large (some are included in TABLE 9-8). Hereditary causes are many and beyond the scope of this article[48]; the Hereditary Hearing Loss website is a good source of information about nonsyndromic and less common syndromic etiologies of hereditary hearing loss (hereditaryhearingloss.org). The Human Phenotype Ontology bilateral sensorineural hearing impairment website is another helpful resource (hpo.jax.org/app/browse/term/HP:0008619).

KEY POINTS

- Intracranial hypotension can present with tinnitus, altered hearing, dizziness, or vertigo.
- Among the more prevalent etiologies of chronic bilateral sensorineural hearing loss are age-related hearing loss, heritable factors, and noise exposure.

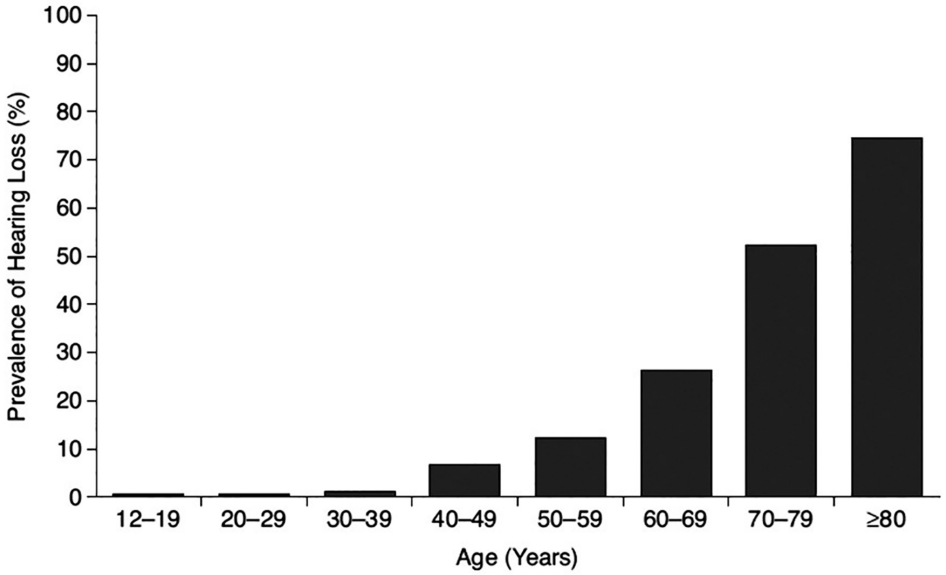

FIGURE 9-4
Relationship between age and hearing loss in the United States from 2001 to 2008. Hearing loss is defined as greater than 25-dB thresholds from 500 Hz to 4000 Hz in the better-hearing ear. Note that the prevalence of hearing loss approximately doubles every decade of life between the 12- to 17-year and 70- to 79-year age groups.
Modified with permission from Yamasoba T, et al, Hear Res.[47] © 2013 Elsevier B.V.

MANAGEMENT OF SENSORINEURAL HEARING LOSS. The management of disorders that cause hearing loss varies depending on its etiology.[29] Regarding the hearing loss itself, intratympanic dexamethasone injections by an otolaryngologist or high-dose IV corticosteroid therapy in patients with incomplete recovery from sudden sensorineural hearing loss should be considered as soon as possible after the onset of symptoms. No medications, diet, nutritional supplements, exercises, or therapies have been established to help chronic sensorineural hearing loss. Known or suspected ototoxic medications should be stopped. If an underlying autoimmune cause is identified, then immunotherapy can be instituted, although it is rarely successful if hearing loss has been present and is nonfluctuating for longer than 1 year.

Determining the frequency, severity, and evolution of sensorineural hearing loss is best done by obtaining an audiogram and monitoring with serial audiograms when indicated. The following sections are a neurologist's primer for interpreting an audiogram and include a simplified overview of options for hearing augmentation from hearing aids to cochlear implants. Patient education is valuable to help set expectations and advise on treatment risks and benefits and to allay any patient anxiety.

NEUROLOGIST'S PRIMER FOR INTERPRETING AN AUDIOGRAM

An audiogram is a graphical representation of human hearing (FIGURE 9-5). Across the top are the pure tone frequencies. Frequency is the pitch, measured in Hertz (Hz), that is, cycles per second of the sound vibration. For human hearing, the pure tones range from 250 Hz (low pitched sounds) to 8000 Hz (higher-pitched sounds) is assessed. Of course, sound frequencies are much broader, but this is what is used in standard hearing tests.

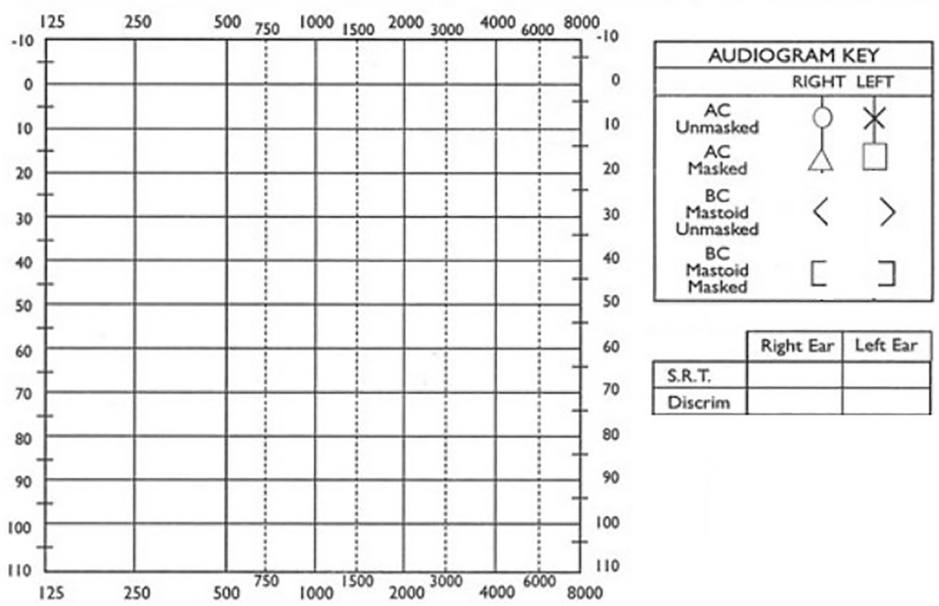

FIGURE 9-5

Blank audiogram form. Pure tone frequencies in hertz (Hz) are aligned across horizontally (x-axis), and decibels (dB hearing level) of hearing normalized to humans are on the vertical (y-axis). *Masked* refers to application of some noise to the ear opposite the one being tested to avoid the confounding effect of hearing from the ear not being tested.

AC = air-conducted hearing; BC = bone-conducted hearing; Discrim = speech discrimination; SRT = speech reception threshold.

Down the vertical axis of FIGURE 9-5 is the intensity, which refers to the loudness and is measured in decibels hearing level (dB HL) and often ranges from -10 dB to 120 dB in gradations of 10 dB. A decibel is 1/10 of a bel, a unit of intensity of sound. The dB HL measurements on the y-axis are normalized to human hearing, so 0 dB does not mean no sound is present; it simply means no amplification of sound above idealized human hearing.

The audiologist uses an audiometer to present the pure tones via headphones or inserted earphones in a sound-proof booth to test pure tone thresholds, that is, the quietest sound at a given frequency that the person can hear at least half the time. This is done sequentially in each ear at each of the specified frequencies and plotted as pure tone threshold values. By convention, *X* represents the left, and *O* represents the right ear for air-conducted sounds. To bypass the natural pathway of sound and test the hearing directly at the cochlea, the audiologist presents tones via bone conduction by placing a bone oscillator on the mastoid bone of each ear sequentially. A differential of 25 dB or more between the threshold for bone- and air-conducted sound thresholds is referred to as the air-bone gap and may suggest conductive hearing loss (as opposed to sensorineural hearing loss). Threshold values of 0 dB to 20 dB are considered normal, 25 dB to 40 dB mild, 41 dB to 55 dB moderate, 56 dB to 70 dB moderately severe, and 71 dB to 90 dB severe hearing loss; threshold values greater than 90 dB indicate hearing loss is profound at that frequency.

Masking is applying some narrow-band noise in the ear not being tested to prevent that ear from hearing sound applied to the ear being tested. Masking

usually is done when greater than 10 dB or more air-bone gap is present or when one ear hears much better than the other (eg, a difference of 40 dB or more). By convention *[* denotes the right ear (AD) and *]* denotes the left ear (AS) for bone-conducted thresholds.

Other tests used include speech reception threshold and speech discrimination score. With the speech reception threshold, a list of two-syllable words is given at the lowest reception threshold at which the individual can recognize speech and represents the threshold at which at least half the words are identified correctly in a quiet environment. The speech discrimination score measures how well the patient can hear speech that is loud enough to hear comfortably. A speech discrimination score of 100% means the individual understands all the words spoken.

HEARING AUGMENTATION

Hearing augmentation generally either amplifies the sound or provides some way to circumvent the dysfunctional part of the peripheral auditory system in those with hearing loss. TABLE 9-9[49,50] outlines some of the ever-growing list of ways to improve hearing in patients with hearing loss. A few consistent guidelines are available for objective measures of the degree of hearing loss that warrant a hearing aid, but for most people, it is tied to the ability to understand speech.

Cochlear Implants

A cochlear implant is a surgically implanted device that converts sound into electrical impulses that are delivered to the auditory nerve by electrodes implanted within the lumen of the cochlea. This bypasses the cochlear hair cells. The components include an external microphone, sound processor and transmitter, and an internal receiver and electrode array extending from the receiver through the round window into the cochlea.

Indications

For children with severe or profound hearing loss from birth, hearing augmentation is ideally initiated before language develops. If cochlear implants are placed sequentially, it is best to keep the time between placements to less than 18 months.[51] Because bilateral hearing improves overall hearing and spatial localization through binaural summation within central auditory processing pathways, it is often advised that cochlear implants and other hearing augmentation be applied in both ears even if bilateral hearing loss is asymmetrical. For adults, cochlear implant surgery has no upper age limit provided the patient does not have dementia. Adult candidates for cochlear implantation should have severe to profound bilateral sensorineural hearing loss that negatively impacts communication and cannot be sufficiently improved by hearing aids. Some insurance coverage policies add that hearing must be at least 70 dB or worse at 500 Hz, 1000 Hz, and 2000 Hz to qualify for payment coverage.

Cochlear implants allow patients with severe hearing loss to hear sound, which is different from hearing aids that just amplify sound. Because the sounds are translated by a sound processor and direct nerve stimulation, differences in sound quality occur, and it requires some practice hearing while seeing text to improve familiarity with how these sounds correlate to previously heard

language. Cochlear implants made by all manufacturers are approved as "MRI conditional," meaning that certain measures must be followed to assure the magnetic resonance study is done safely. In addition, the electrode leads do not become hot in ordinary magnetic resonance units because of the short length of electrode leads.

Two 2019 studies explored the use of a vestibular prosthesis either alone[52] or combined with a cochlear implant[53] with multichannel vestibular electrodes

Various Hearing Augmentation Options TABLE 9-9

Type of hearing aid	Description	Indication
Receiver-in-canal	A small receiver in the ear canal is connected by a thin wire to the electronic device that is located behind the ear; the receiver is in the ear canal but does not completely seal the canal; sound detected by the receiver in the ear canal is amplified	Mild to severe hearing loss adversely affecting quality of life; usually at least 40-dB hearing loss at three frequencies, especially in the 2000-Hz to 4000-Hz range
Behind-the-ear	Custom molds in the ear canal deliver sound transmitted by a tube from the electronic device that is located behind the ear	Mild to severe hearing loss adversely affecting quality of life; usually at least 40-dB hearing loss at three frequencies, especially in the 2000-Hz to 4000-Hz range
In-the-ear	Hearing devices that fill the external ear canal and are larger and more visible than invisible-in-canal and completely-in-the-canal types	Mild to severe hearing loss adversely affecting quality of life; usually at least 40-dB hearing loss at three frequencies, especially in the 2000-Hz to 4000-Hz range
Invisible-in-canal and completely-in-canal	Invisible-in-canal hearing devices placed deep in the ear canal and removed by tugging on a small plastic string; completely-in-canal devices are very similar	Mild to severe hearing loss adversely affecting quality of life; usually at least 40-dB hearing loss at three frequencies, especially in the 2000-Hz to 4000-Hz range
Direct-to-consumer	Mostly receiver-in-canal and behind-the-ear devices that may be purchased by consulting a doctor or audiologist; variability in quality; the US Food and Drug Administration (FDA) Reauthorization Act of 2017 developed FDA standards and package labels for over-the-counter hearing aids that as of the time of publication of this article are still pending[49]	Perceived mild to moderate hearing loss adversely affecting quality of life[50]
Contralateral routing of signals, bilateral routing of signals	Contralateral routing of signals is a system with a microphone in the deaf ear that wirelessly transmits sound from the deaf side to the normal-hearing ear; bilateral routing of signals is the same but with some amplification also in the better-hearing ear	Severe hearing loss in one ear and little or no hearing loss in the other ear (single-sided deafness)
Bone-anchored	A titanium screw is anchored into the bone behind the poor-hearing ear, and a titanium abutment attaches from the bone-anchored screw so it protrudes outside of the skin or via a subdermal magnet it attaches to a sound processor that transmits sound through the skull bone to the cochlea on both sides	Single-sided deafness or chronic conductive hearing loss as long as the cochlea on one side functions at least at a moderate hearing level

(FIGURE 9-6) for patients with severe bilateral peripheral vestibular loss without or with profound bilateral hearing loss. Preliminary results suggest that sustained canal-ocular vestibulo-ocular function can be demonstrated,[52,54] although it remains to be determined if this translates to meaningful clinical improvement for patients. Vestibular prosthesis, either alone or added to a cochlear implant, is still investigational at this time.

CONCLUSION

Tinnitus and hearing loss are common conditions affecting people of all ages, although more commonly older people. This article is intended to be a guide to approaching tinnitus, hyperacusis, idiopathic otalgia, and hearing loss and introduce neurologists to the basics of audiometry and hearing augmentation. It is hoped that this serves as a primer for organizing and understanding these conditions and for recognizing important neurologic disorders that might be associated with these symptoms.

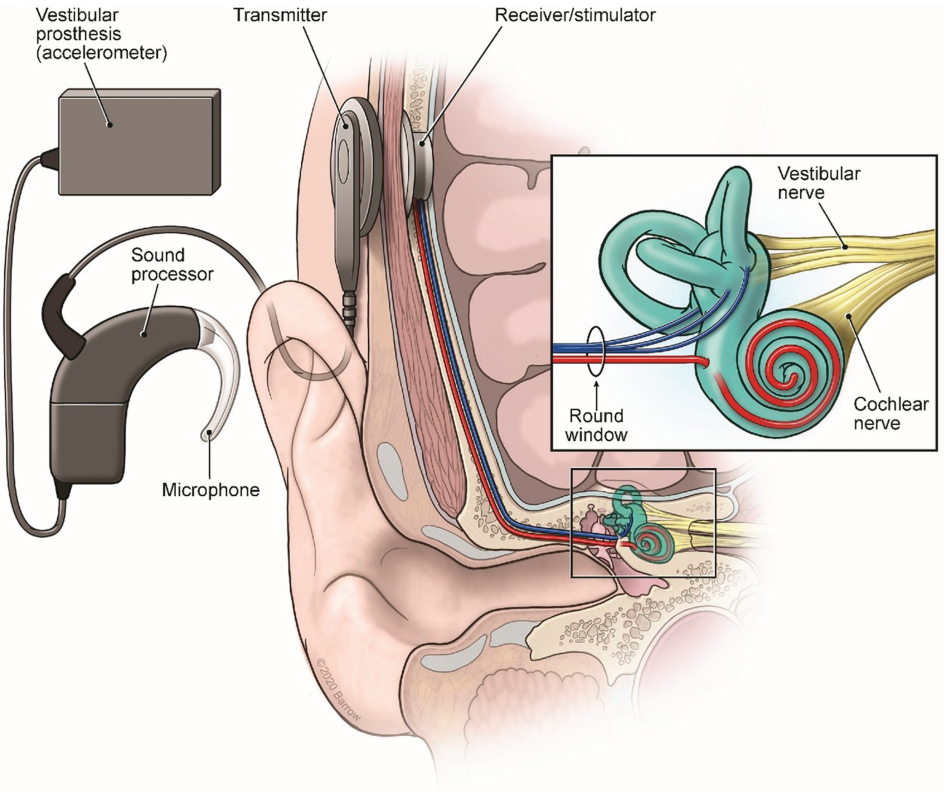

FIGURE 9-6

Depiction of how a combined cochlear implant and vestibular prosthesis system would work. Systems in development include a vestibular implant without or with a cochlear implant. Not all necessary ground and reference electrodes are shown. The electrode in red is for a cochlear implant, which is an established device; the electrode in blue for a hypothetical vestibular prosthesis is still investigational.

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USEFUL WEBSITES

BILATERAL SENSORINEURAL HEARING IMPAIRMENT
This Human Phenotype webpage provides a list of diseases and genes associated specifically with bilateral sensorineural hearing loss.
hpo.jax.org/app/browse/term/HP:0008619

HEREDITARY HEARING LOSS HOME PAGE
This site lists data and links for all known gene localizations and identifications for monogenic nonsyndromic hearing impairment.
hereditaryhearingloss.org

ONTOLOGY SENSORINEURAL HEARING IMPAIRMENT
This Human Phenotype webpage provides a list of diseases and genes associated with hearing loss.
hpo.jax.org/app/browse/term/HP:0000407

OTOSCOPIC PATHOLOGIES
This webpage briefly illustrates some important common pathologies and compares them with what will be observed in a normal ear canal and tympanic membrane.
diagnosis101.welchallyn.com/otoscopy/educational-topics/ear-pathologies/

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