ACCESS Medicine

Kaplan Medical Center

Access Provided by:

SILVERCHAIR INFORMATION / SYSTEMS

Chapter 34: Disorders of Hearing

Anil K. Lalwani

INTRODUCTION

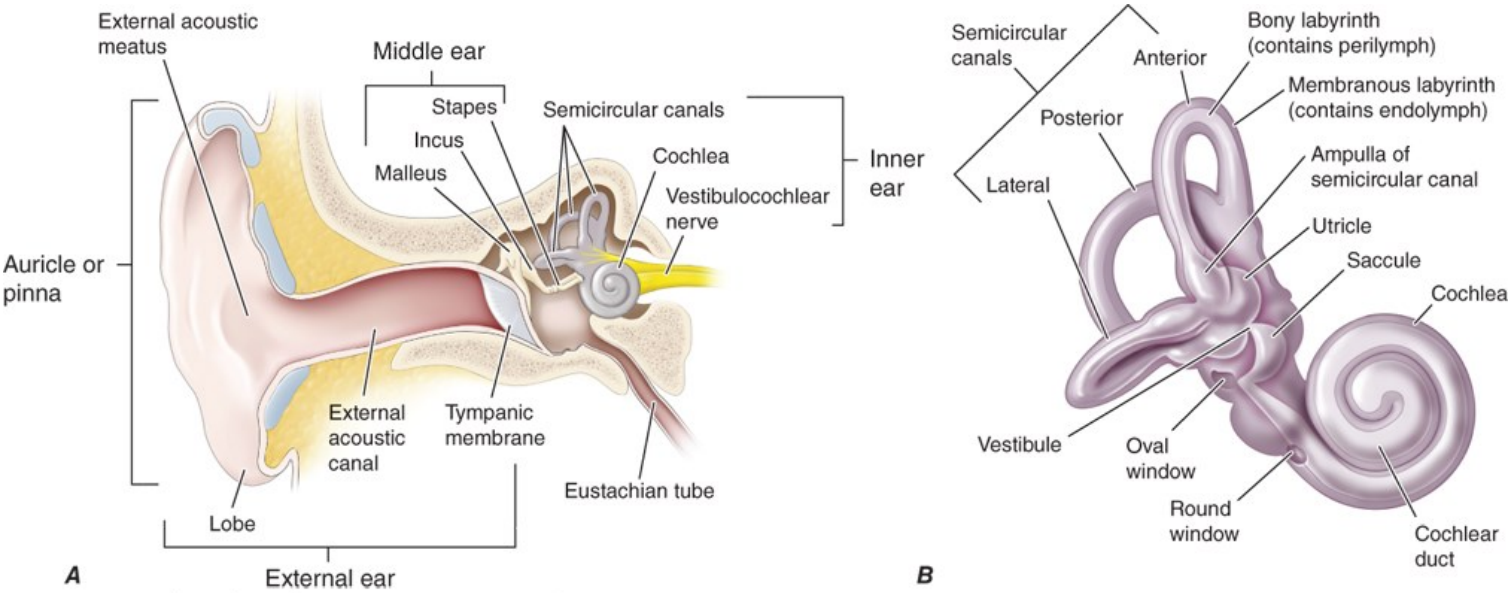
Hearing loss can present at any age and is one of the most common sensory disorders in humans. Nearly 10% of the adult population has some hearing loss, and one-third of individuals age >65 years have a hearing loss of sufficient magnitude to require a hearing aid.

PHYSIOLOGY OF HEARING

The function of the external and middle ear is to amplify sound to facilitate conversion of the mechanical energy of the sound wave into an electrical signal by the inner-ear hair cells, a process called mechanotransduction **(Fig. 34-1)**. Sound waves enter the external auditory canal and set the tympanic membrane (eardrum) in motion, which in turn moves the malleus, incus, and stapes of the middle ear. Movement of the footplate of the stapes causes pressure changes in the fluid-filled inner ear, eliciting a traveling wave in the basilar membrane of the cochlea. The tympanic membrane and the ossicular chain in the middle ear serve as an impedance-matching mechanism, improving the efficiency of energy transfer from air to the fluid-filled inner ear. In its absence, nearly 99.9% of the acoustical energy would be reflected and thus not heard. Instead, the eardrum and the ossicles boost the sound energy nearly 200-fold by the time it reaches the inner ear.

FIGURE 34-1

Ear anatomy. ***A.*** Drawing of modified coronal section through external ear and temporal bone, with structures of the middle and inner ear demonstrated. ***B.*** High-resolution view of inner ear.



Source: Joseph Loscalzo, Anthony Fauci, Dennis Kasper, Stephen Hauser, Dan Longo, J. Larry Jameson: Harrison's Principles of Internal Medicine, 21e
Copyright © McGraw Hill. All rights reserved.

Within the cochlea of the inner ear, there are two types of hair cells that aid in hearing: inner and outer. The inner and outer hair cells of the organ of Corti have different innervation patterns, but both are mechanoreceptors; they detect the mechanical energy of the acoustic signal and aid its conversion to an electrical signal that travels by the auditory nerve. The afferent innervation relates principally to the inner hair cells while the efferent

Downloaded 2024-2-17 9:35 A Your IP is 154.59.125.153
Chapter 34: Disorders of Hearing, Anil K. Lalwani
©2024 McGraw Hill. All Rights Reserved. Terms of Use • Privacy Policy • Notice • Accessibility

innervation relates principally to the outer hair cells. The outer hair cells outnumber the inner hair cells by nearly 6:1 (20,000 vs 3500). The motility of the outer hair cells alters the micromechanics of the inner hair cells, creating a cochlear amplifier, which explains the exquisite sensitivity and frequency selectivity of the cochlea.

Stereocilia of the hair cells of the organ of Corti, which rests on the basilar membrane, are in contact with the tectorial membrane and are deformed by the traveling wave. The deformation stretches tiny filamentous connections (tip links) between stereocilia, leading to opening of ion channels, influx of potassium, and hair cell depolarization and subsequent neurotransmission. A point of maximal displacement of the basilar membrane is determined by the frequency of the stimulating tone. High-frequency tones cause maximal displacement of the basilar membrane near the base of the cochlea, whereas for low-frequency sounds, the point of maximal displacement is toward the apex of the cochlea.

Beginning in the cochlea, the frequency specificity is maintained at each point of the central auditory pathway: dorsal and ventral cochlear nuclei, trapezoid body, superior olivary complex, lateral lemniscus, inferior colliculus, medial geniculate body, and auditory cortex. At low frequencies, individual auditory nerve fibers can respond more or less synchronously with the stimulating tone. At higher frequencies, phase-locking occurs so that neurons alternate in response to particular phases of the cycle of the sound wave. Intensity is encoded by the amount of neural activity in individual neurons, the number of neurons that are active, and the specific neurons that are activated.

There is evidence that the right and left ears as well as the central nervous system may process speech asymmetrically. Generally, a sound is processed symmetrically from the peripheral to the central auditory system. However, a “right ear advantage” exists for dichotic listening tasks, in which subjects are asked to report on competing sounds presented to each ear. In most individuals, a perceptual right ear advantage for consonant-vowel syllables, stop consonants, and words also exists. Similarly, whereas central auditory processing for sounds is symmetric with minimal lateral specialization for the most part, speech processing is lateralized. There is specialization of the left auditory cortex for speech recognition and production, and of the right hemisphere for emotional and tonal aspects of speech. Left hemisphere dominance for speech is found in 95–98% of right-handed persons and 70–80% of left-handed persons.

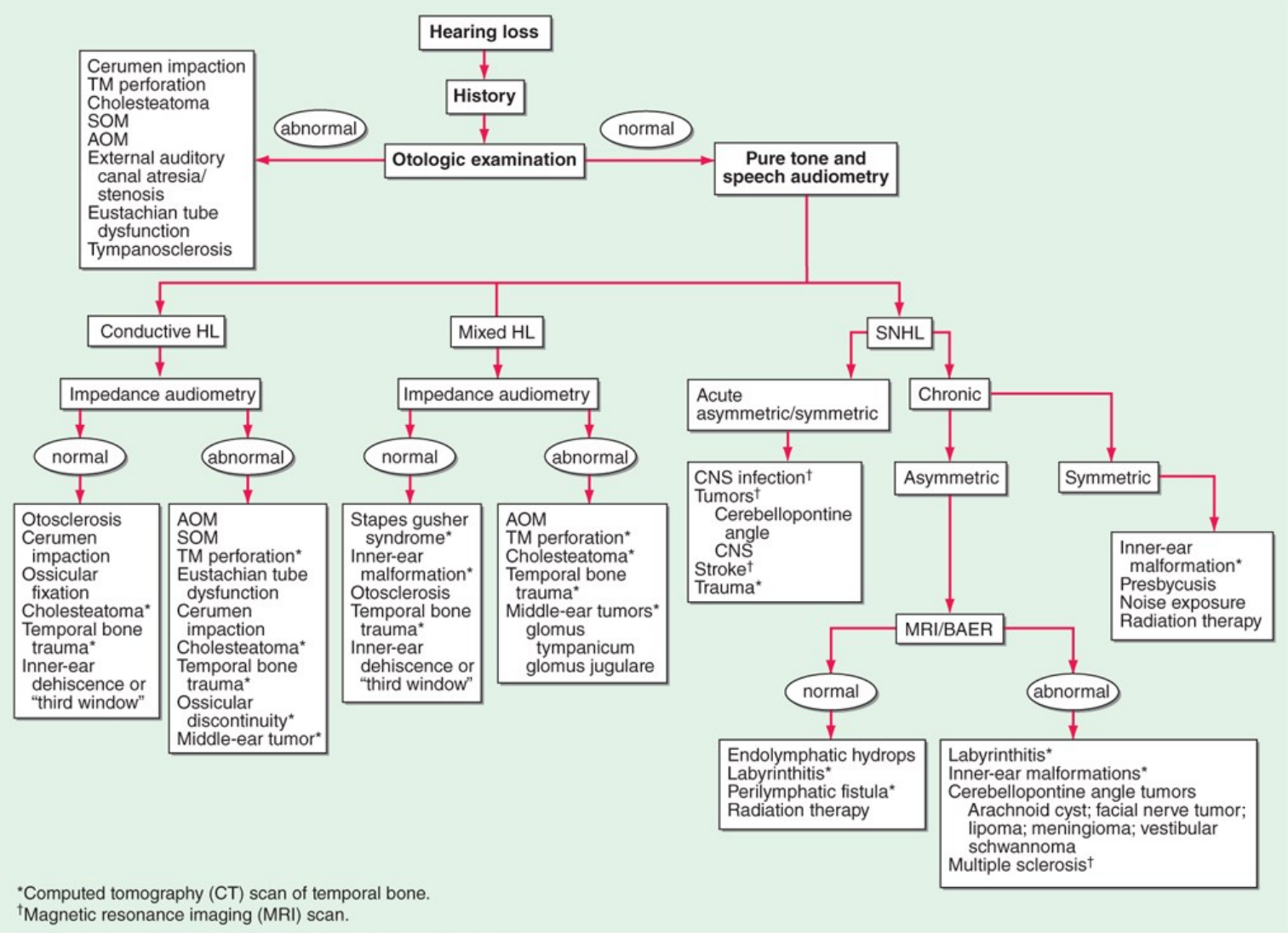
DISORDERS OF THE SENSE OF HEARING

Hearing loss can result from disorders of the auricle, external auditory canal, middle ear, inner ear, or central auditory pathways **(Fig. 34-2)**. In general, lesions in the auricle, external auditory canal, or middle ear that impede the transmission of sound from the external environment to the inner ear cause conductive hearing loss, whereas lesions that impair mechanotransduction in the inner ear or transmission of the electrical signal along the eighth nerve to the brain cause sensorineural hearing loss.

FIGURE 34-2

An algorithm for the approach to hearing loss. AOM, acute otitis media; BAER, brainstem auditory-evoked response; CNS, central nervous system; HL, hearing loss; SNHL, sensorineural hearing loss; SOM, serous otitis media; TM, tympanic membrane.

Downloaded 2024-2-17 9:35 A Your IP is 154.59.125.153
Chapter 34: Disorders of Hearing, Anil K. Lalwani
©2024 McGraw Hill. All Rights Reserved. Terms of Use • Privacy Policy • Notice • Accessibility



Source: Joseph Loscalzo, Anthony Fauci, Dennis Kasper, Stephen Hauser, Dan Longo, J. Larry Jameson: Harrison's Principles of Internal Medicine, 21e
Copyright © McGraw Hill. All rights reserved.

Conductive Hearing Loss

The external ear, the external auditory canal, and the middle-ear apparatus are designed to collect and amplify sound and efficiently transfer the mechanical energy of the sound wave to the fluid-filled cochlea. Factors that obstruct the transmission of sound or dampen the acoustic energy result in conductive hearing loss. Conductive hearing loss can occur from obstruction of the external auditory canal by cerumen, debris, and foreign bodies; swelling of the lining of the canal; atresia or neoplasms of the canal; perforations of the tympanic membrane; disruption of the ossicular chain, as occurs with necrosis of the long process of the incus in trauma or infection; otosclerosis; or fluid, scarring, or neoplasms in the middle ear. Rarely, inner-ear malformations or pathologies that create a "third window" in the inner ear such as superior semicircular canal dehiscence, lateral semicircular canal dysplasia, incomplete partition of the inner ear, and large vestibular aqueduct, are also associated with conductive hearing loss. This pathologic third window is associated with loss of mechanical energy associated with the sound wave leading to conductive hearing loss (see below).

Eustachian tube dysfunction is extremely common in adults and may predispose to acute otitis media (AOM) or serous otitis media (SOM). Recently, Eustachian tube balloon dilation has been shown to relieve acquired inflammatory obstruction of the Eustachian tube orifice and improve symptoms due to Eustachian tube dysfunction. Trauma, AOM, and chronic otitis media are the usual factors responsible for tympanic membrane perforation. While small perforations often heal spontaneously, larger defects usually require surgical intervention. Tympanoplasty is highly effective (>90%) in the

Downloaded 2024-2-17 9:35 A Your IP is 154.59.125.153
Chapter 34: Disorders of Hearing, Anil K. Lalwani
©2024 McGraw Hill. All Rights Reserved. Terms of Use • Privacy Policy • Notice • Accessibility

repair of tympanic membrane perforations. Otoscopy is usually sufficient to diagnose AOM, SOM, chronic otitis media, cerumen impaction, tympanic membrane perforation, and Eustachian tube dysfunction; tympanometry and Eustachian tube function testing can be useful to confirm the clinical suspicion of these conditions.

Cholesteatoma, a benign tumor composed of stratified squamous epithelium in the middle ear or mastoid, occurs frequently in adults, often in the setting of severe Eustachian tube dysfunction. This is a slowly growing lesion that destroys bone and normal ear tissue. Theories of pathogenesis include traumatic immigration and invasion of squamous epithelium through a retraction pocket of the tympanic membrane, implantation of squamous epithelia in the middle ear through a perforation or surgery, and metaplasia following chronic infection and irritation. A chronically draining ear that fails to respond to appropriate antibiotic therapy should raise suspicion of a cholesteatoma. On examination, there is often a perforation of the tympanic membrane filled with cheesy white squamous debris. The presence of an aural polyp obscuring the tympanic membrane is highly suggestive of an underlying cholesteatoma. Conductive hearing loss secondary to ossicular erosion is common. Bony destruction visualized on CT of the temporal bone is also highly suggestive of cholesteatoma. Surgery is required to remove this destructive process and reconstruct the ossicles.

Conductive hearing loss with a normal ear canal and intact tympanic membrane suggests either ossicular pathology or the presence of a "third window" in the inner ear (see below). Fixation of the stapes from *otosclerosis* is a common cause of low-frequency conductive hearing loss. It occurs equally in men and women and is inherited as an autosomal dominant trait with incomplete penetrance; in some cases, it may be a manifestation of osteogenesis imperfecta. Hearing impairment usually presents between the late teens and the forties. In women, the otosclerotic process is accelerated during pregnancy, and the hearing loss is often first noticeable at this time. A hearing aid or a simple outpatient surgical procedure (stapedectomy) can provide excellent auditory rehabilitation. Extension of otosclerosis beyond the stapes footplate to involve the cochlea (cochlear otosclerosis) can lead to mixed or sensorineural hearing loss. Fluoride therapy to prevent hearing loss from cochlear otosclerosis is of uncertain value.

Disorders that lead to the formation of a pathologic "third window" in the inner ear can be associated with conductive hearing loss. There are normally two major openings, or windows, that connect the inner ear with the middle ear and serve as conduits for transmission of sound; these are, respectively, the oval and round windows. A third window is formed where the normally hard otic bone surrounding the inner ear is eroded; dissipation of the acoustic energy at the third window is responsible for the "inner-ear conductive hearing loss." The superior semicircular canal dehiscence syndrome resulting from erosion of the otic bone over the superior circular canal can present with conductive hearing loss that mimics otosclerosis. A common symptom is vertigo evoked by loud sounds (Tullio phenomenon), by Valsalva maneuvers that change middle-ear pressure, or by applying positive pressure on the tragus (the cartilage anterior to the external opening of the ear canal). Patients with this syndrome also complain of fullness of the ear, pulsatile tinnitus, and being able to hear the movement of their eyes and neck. A large jugular bulb or jugular bulb diverticulum can create a "third window" by eroding into the vestibular aqueduct or posterior semicircular canal; the symptoms are similar to those of the superior semicircular canal dehiscence syndrome. Other inner-ear malformations such as lateral semicircular canal dysplasia, large vestibular aqueduct, or incomplete partition seen in stapes gusher syndrome can also be associated with inner-ear conductive hearing loss as a result of the third window. Low activation threshold on the vestibular-evoked myogenic potential test (VEMP test, see below) and inner-ear erosion on CT are diagnostic. Recalcitrant vertigo and dizziness may respond to surgical repair of the dehiscence.

Sensorineural Hearing Loss

Sensorineural hearing loss results from either damage to the mechanotransduction apparatus of the cochlea or disruption of the electrical conduction pathway from the inner ear to the brain. Thus, injury to hair cells, supporting cells, auditory neurons, or the central auditory pathway can cause sensorineural hearing loss. Damage to the hair cells of the organ of Corti may be caused by intense noise, viral infections, ototoxic drugs (e.g., salicylates, quinine and its synthetic analogues, aminoglycoside antibiotics, loop diuretics such as furosemide and ethacrynic acid, and cancer chemotherapeutic agents such as cisplatin), fractures of the temporal bone, meningitis, cochlear otosclerosis (see above), Ménière's disease, and aging. Congenital malformations of the inner ear may be the cause of hearing loss in some adults. Genetic predisposition alone or in concert with environmental exposures may also be responsible (see below).

Noise-Induced Hearing Loss

Exposure to loud noise, either a short burst or over a more prolonged period of time, can lead to noise-induced hearing loss. Acute exposure to noise can lead to either temporary or permanent threshold shifts, depending on the intensity and duration of sound, due to hair cell injury and/or death. Typically, with permanent hearing loss there is a "noise notch" with elevated hearing thresholds at 3000–4000 Hz. More recently, loud noise exposure

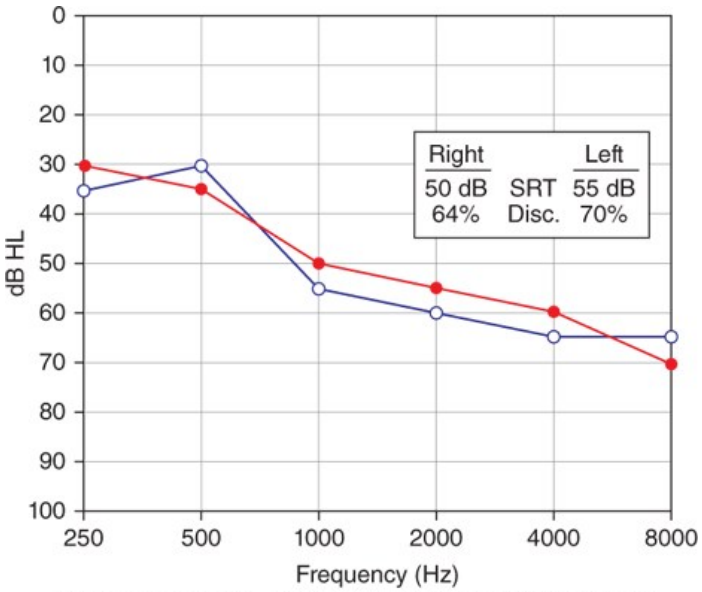
Downloaded 2024-2-17 9:35 A Your IP is 154.59.125.153
Chapter 34: Disorders of Hearing, Anil K. Lalwani
©2024 McGraw Hill. All Rights Reserved. Terms of Use • Privacy Policy • Notice • Accessibility

has also been associated with “hidden hearing loss”—hidden, because routine audiometry shows the pure tone hearing to be normal. Patients usually complain of not being able to hear clearly and are more bothered by the presence of background noise. In contrast to hair cell loss, hidden hearing loss is thought to be due to loss of auditory synapses on hair cells following noise exposure. In an increasingly noisy world, avoiding acoustic trauma with earplugs or earmuffs is highly recommended to prevent noise-induced or hidden hearing loss.

Presbycusis (age-associated hearing loss) is the most common cause of sensorineural hearing loss in adults. It is estimated to affect over half of adults aged >75 years in the United States, a population that is expected to double in size over the next 40 years. In the early stages, it is characterized by symmetric, gentle to sharply sloping, high-frequency hearing loss **(Fig. 34-3)**. With progression, the hearing loss involves all frequencies. More importantly, the hearing impairment is associated with significant loss in clarity. There is a loss of discrimination for phonemes, recruitment (abnormal growth of loudness), and particular difficulty in understanding speech in noisy environments such as at restaurants and social events. Poor hearing is also associated with an increased incidence of cognitive impairment, rate of cognitive decline, and falls. In the elderly, left untreated, hearing loss leads to diminished quality of life, and has been shown to increase overall morbidity and mortality through falls and accidents. Hearing aids are helpful in enhancing the signal-to-noise ratio by amplifying sounds that are close to the listener. Hearing aid use has been shown to reduce cognitive decline and risk of falls. Although hearing aids are able to amplify sounds, they cannot restore the clarity of hearing. Thus, amplification with hearing aids may provide only limited rehabilitation once the word recognition score deteriorates below 50%. Cochlear implants are the treatment of choice when hearing aids prove inadequate, even when hearing loss is incomplete (see below).

FIGURE 34-3

Presbycusis or age-related hearing loss. The audiogram shows a moderate to severe downsloping sensorineural hearing loss typical of presbycusis. The loss of high-frequency hearing is associated with a decreased speech discrimination score; consequently, patients complain of lack of clarity of hearing, especially in a noisy background. HL, hearing threshold level; SRT, speech reception threshold.



Source: Joseph Loscalzo, Anthony Fauci, Dennis Kasper, Stephen Hauser, Dan Longo, J. Larry Jameson: Harrison's Principles of Internal Medicine, 21e
Copyright © McGraw Hill. All rights reserved.

Ménière’s disease is characterized by episodic vertigo, fluctuating sensorineural hearing loss, tinnitus, and aural fullness. An absence of vertigo is inconsistent with the diagnosis of Ménière’s disease, and the presence of fluctuating sensorineural hearing loss, tinnitus, and fullness without vertigo is more suggestive of cochlear hydrops. Tinnitus and/or deafness may be absent during the initial attacks of vertigo, but invariably appear as the disease progresses and increases in severity during acute attacks. The annual incidence of Ménière’s disease is 0.5–7.5 per 1000; onset is most frequently in the fifth decade of life but may also occur in young adults or the elderly. Histologically, there is distention of the endolymphatic system (endolymphatic hydrops) leading to degeneration of vestibular and cochlear hair cells. This may result from endolymphatic sac dysfunction secondary to infection, trauma, autoimmune disease, inflammatory causes, or tumor; an idiopathic etiology constitutes the largest category and is most accurately referred to as Ménière’s disease. Endolymphatic sac tumors, often associated with von Hippel Lindau disease, may clinically mimic Ménière’s disease. Although any pattern of hearing loss can be observed, typically, low-frequency, unilateral sensorineural hearing impairment is

Downloaded 2024-2-17 9:35 A Your IP is 154.59.125.153
Chapter 34: Disorders of Hearing, Anil K. Lalwani
©2024 McGraw Hill. All Rights Reserved. Terms of Use • Privacy Policy • Notice • Accessibility

present. An abnormal VEMP test (see below) may be helpful in detecting Ménière's disease in a clinically unaffected contralateral ear. MRI should be obtained to exclude retrocochlear pathology such as a cerebellopontine angle tumor, endolymphatic sac tumor, or demyelinating disorder. Therapy is directed toward the control of vertigo. A 2-g/d low-salt diet is the mainstay of treatment for control of rotatory vertigo. Diuretics, a short course of oral glucocorticoids, intratympanic glucocorticoids, or intratympanic gentamicin may also be useful adjuncts in recalcitrant cases. Surgical therapy of vertigo is reserved for unresponsive cases and includes endolymphatic sac decompression, labyrinthectomy, and vestibular nerve section. Both labyrinthectomy and vestibular nerve section abolish rotatory vertigo in >90% of cases. Unfortunately, there is no effective therapy for hearing loss, tinnitus, or aural fullness from Ménière's disease.

Sensorineural hearing loss may also result from any neoplastic, vascular, demyelinating, infectious, degenerative disease, or trauma affecting the central auditory pathways. Characteristically, in hearing loss due to central nervous system pathology, a reduction in clarity of hearing and speech comprehension is much greater than the loss of the ability to hear pure tone. Auditory testing is consistent with an auditory neuropathy; normal otoacoustic emissions (OAEs) and an abnormal auditory brainstem response (ABR) are typical (see below). Hearing loss can accompany hereditary sensorimotor neuropathies and inherited disorders of myelin. Tumors of the cerebellopontine angle such as vestibular schwannoma and meningioma **(Chap. 90)** usually present with asymmetric sensorineural hearing loss with greater deterioration of speech understanding than pure tone hearing. Multiple sclerosis **(Chap. 444)** may present with acute unilateral or bilateral hearing loss; typically, pure tone testing remains relatively stable while speech understanding fluctuates. Isolated labyrinthine infarction can present with acute hearing loss and vertigo due to a cerebrovascular accident involving the posterior circulation, usually the anterior inferior cerebellar artery; it may also be the heralding sign of impending catastrophic basilar artery infarction **(Chap. 426)**. HIV **(Chap. 202)**, which can produce both peripheral and central auditory system pathology, is another consideration in the evaluation of sensorineural hearing impairment.

A finding of conductive and sensorineural hearing loss in combination is termed *mixed hearing loss.* Mixed hearing losses can result from pathology of both the middle and inner ear, as can occur in otosclerosis involving the ossicles and the cochlea, head trauma, chronic otitis media, cholesteatoma, middle-ear tumors, and some inner-ear malformations.

Trauma resulting in temporal bone fractures may be associated with conductive, sensorineural, or mixed hearing loss. If the fracture spares the inner ear, there may simply be conductive hearing loss due to rupture of the tympanic membrane or disruption of the ossicular chain. These abnormalities can be surgically corrected. Profound hearing loss and severe vertigo are associated with temporal bone fractures involving the inner ear. A perilymphatic fistula associated with leakage of inner-ear fluid into the middle ear can occur and may require surgical repair. An associated facial nerve injury is not uncommon. CT is best suited to assess fracture of the traumatized temporal bone, evaluate the ear canal, and determine the integrity of the ossicular chain and involvement of the inner ear. Cerebrospinal fluid leaks that accompany temporal bone fractures are usually self-limited; the value of prophylactic antibiotics is uncertain.

Tinnitus

Tinnitus is defined as the perception of a sound when there is no sound in the environment. It can have a buzzing, roaring, or ringing quality and may be pulsatile (synchronous with the heartbeat). Tinnitus is often associated with either a conductive or sensorineural hearing loss. The pathophysiology of tinnitus is not well understood. The cause of the tinnitus can usually be determined by finding the cause of the associated hearing loss. Tinnitus may be the first symptom of a serious condition such as a vestibular schwannoma. Pulsatile tinnitus requires evaluation of the vascular system of the head to exclude vascular tumors such as glomus jugulare tumors, aneurysms, dural arteriovenous fistulas, and stenotic arterial lesions; it may also occur with SOM, superior semicircular dehiscence, and inner-ear dehiscence. It is most commonly associated with some abnormality of the jugular bulb such as a large jugular bulb or jugular bulb diverticulum. In absence of demonstrated pathology on MRA/MRV or CT angiography, pulsatile tinnitus is usually attributed to turbulent venous blood flow through the transverse sinus, sigmoid sinus, and the jugular bulb.

GENETIC CAUSES OF HEARING LOSS

 More than half of childhood hearing impairment is thought to be hereditary; hereditary hearing impairment (HHI) can also manifest later in life. HHI may be classified as either nonsyndromic, when hearing loss is the only clinical abnormality, or syndromic, when hearing loss is associated with anomalies in other organ systems. Nearly two-thirds of HHIs are nonsyndromic. Between 70% and 80% of nonsyndromic HHI is inherited in an autosomal recessive manner and designated DFNB; another 15–20% is autosomal dominant (DFNA). Less than 5% is X-linked (DFNX) or maternally inherited via the mitochondria.

Downloaded 2024-2-17 9:35 A Your IP is 154.59.125.153
Chapter 34: Disorders of Hearing, Anil K. Lalwani
©2024 McGraw Hill. All Rights Reserved. Terms of Use • Privacy Policy • Notice • Accessibility

More than 150 loci harboring genes for nonsyndromic HHI have been mapped, with recessive loci outnumbering dominant ones; numerous genes have now been identified **(Table 34-1)**. The hearing genes fall into the categories of structural proteins (*MYH9, MYO7A, MYO15, TECTA, DIAPH1*), transcription factors (*POU3F4, POU4F3*), ion channels (*KCNQ4, SLC26A4*), and gap junction proteins (*GJB2, GJB3, GJB6*). Several of these genes, including *GJB2, TECTA*, and *TMC1*, cause both autosomal dominant and recessive forms of nonsyndromic HHI. In general, the hearing loss associated with dominant genes has its onset in adolescence or adulthood, varies in severity, and progresses with age, whereas the hearing loss associated with recessive inheritance is congenital and profound. Connexin 26, a product of the *GJB2* gene, is particularly important because it is responsible for nearly 20% of all cases of childhood deafness; half of genetic deafness in children is *GJB2* related. Two frameshift mutations, 35delG and 167delT, account for >50% of the cases; however, screening for these two mutations alone is insufficient, and sequencing of the entire gene is required to fully capture *GJB2*-related recessive deafness. The 167delT mutation is highly prevalent in Ashkenazi Jews; ~1 in 1765 individuals in this population is homozygous and affected. *GJB2* hearing loss can also vary among the members of the same family, suggesting that other genes or factors influence the auditory phenotype. A single mutation in *GJB2* in combination with a single mutation in *GJB6* (connexin 30) can also lead to hearing loss and is an example of digenic inheritance of hearing loss.

TABLE 34-1

Hereditary Hearing Impairment Genes

DESIGNATION	GENE	FUNCTION
Autosomal Dominant		
DFNA1	*DIAPH1*	Cytoskeletal protein
DFNA2A	*KCNQ4*	Potassium channel
DFNA2B	*GJB3*	Gap junction
DFNA2C	*IFNLR1*	Class II cytokine receptor
DFNA3A	*GJB2*	Gap junction
DFNA3B	*GJB6*	Gap junction
DFNA4A	*MYH14*	Class II nonmuscle myosin
DFNA4B	*CEACAM16*	Cell adhesion molecule
DFNA5	*GSDME/DFNA5*	Executioner of pyroptosis
DFNA6/14/38	*WFS1*	Transmembrane protein
DFNA7	*LMX1A*	Transcription factor
DFNA8/12	*TECTA*	Tectorial membrane protein
DFNA9	*COCH*	Unknown
DFNA10	*EYA4*	Developmental gene
DFNA11	*MYO7A*	Cytoskeletal protein

Downloaded 2024-2-17 9:35 A Your IP is 154.59.125.153
Chapter 34: Disorders of Hearing, Anil K. Lalwani
©2024 McGraw Hill. All Rights Reserved. Terms of Use • Privacy Policy • Notice • Accessibility

DFNA13	*COL11A2*	Cytoskeletal protein
DFNA15	*POU4F3*	Transcription factor
DFNA17	*MYH9*	Cytoskeletal protein
DFNA20/26	*ACTG1*	Cytoskeletal protein
DFNA22	*MYO6*	Unconventional myosin
DFNA23	*SIX1*	Developmental gene
DFNA25	*SLC17A8*	Vesicular glutamate transporter
DFNA27	*REST*	Transcriptional repressor
DFNA28	*GRHL2*	Transcription factor
DFNA34	*NLRP3*	Pyrin-like protein involved in inflammation
DFNA36	*TMC1*	Transmembrane protein
DNA37	*COL11A1*	Cytoskeletal protein
DFNA40	*CRYM*	Thyroid hormone–binding protein
DFNA41	*P2RX2*	Purinergic receptor
DFNA44	*CCDC50*	Effector of epidermal growth factor–mediated signaling
DFNA50	*MIRN96*	MicroRNA
DFNA51	*TJP2*	Tight junction protein
DFNA56	*TNC*	Extracellular matrix protein
DFNA64	*SMAC/DIABLO*	Mitochondrial proapoptotic protein
DFNA65	*TBC1D24*	ARF6-interacting protein
DFNA66	*CD164*	Sialomucin
DFNA67	*OSBPL2*	Intracellular lipid receptor
DFNA68	*HOMER2*	Stereociliary scaffolding protein
DFNA69	*KITLG*	Ligand for KIT receptor
DFNA70	*MCM2*	Initiation and elongation during DNA replication
DFNA73	*PTPRQ*	Member of type III receptor-like protein-tyrosine phosphatase (PTPase) family

Downloaded 2024-2-17 9:35 A Your IP is 154.59.125.153
Chapter 34: Disorders of Hearing, Anil K. Lalwani
©2024 McGraw Hill. All Rights Reserved. Terms of Use • Privacy Policy • Notice • Accessibility

	DMXL2	Regulator of Notch signaling
	MYO3A	Member of myosin superfamily
	PDE1C	Catalyze hydrolysis of cAMP and cGMP
	TRRAP	Transformation/transcription domain associated protein
	PLS1	Actin-bundling protein
	SCD5	Catalyzes formation of monounsaturated fatty acids from saturated fatty acids
	SLC12A2	Sodium-potassium-chloride transporter
	MAP1B	Microtubule binding protein
	RIPOR2/FAM65B	Membrane-associated protein in stereocilia
Autosomal Recessive		
DFNB1A	*GJB2*	Gap junction
DFNB1B	*GJB6*	Gap junction
DFNB2	*MYO7A*	Cytoskeletal protein
DFNB3	*MYO15A*	Cytoskeletal protein
DFNB4	*SLC26A4*	Chloride/iodide transporter
DFNB6	*TMIE*	Transmembrane protein
DFNB7/B11	*TMC1*	Transmembrane protein
DFNB8/10	*TMPRSS3*	Transmembrane serine protease
DFNB9	*OTOF*	Trafficking of membrane vesicles
DFNB12	*CDH23*	Intercellular adherence protein
DFNB15/72/95	*GIPC3*	PDZ domain–containing protein
DFNB16	*STRC*	Stereocilia protein
DFNB18	*USH1C*	Unknown
DFNB18B	*OTOG*	Tectorial membrane protein
DFNB21	*TECTA*	Tectorial membrane protein
DFNB22	*OTOA*	Gel attachment to nonsensory cell

Downloaded 2024-2-17 9:35 A Your IP is 154.59.125.153
Chapter 34: Disorders of Hearing, Anil K. Lalwani
©2024 McGraw Hill. All Rights Reserved. Terms of Use • Privacy Policy • Notice • Accessibility

DFNB23	*PCDH15*	Morphogenesis and cohesion
DFNB24	*RDX*	Cytoskeletal protein
DFNB25	*GRXCR1*	Reversible S-glutathionylation of proteins
DFNB26	*GAB1*	Member of insulin receptor substrate 1–like multisubstrate docking adapter protein family
DFNB28	*TRIOBP*	Cytoskeletal-organizing protein
DFNB29	*CLDN14*	Tight junctions
DFNB30	*MYO3A*	Hybrid motor-signaling myosin
DFNB31	*WHRN*	PDZ domain–containing protein
DFNB32/105	*CDC14A*	Protein phosphatase involved in hair cell ciliogenesis
DFNB35	*ESRRB*	Estrogen-related receptor beta protein
DFNB36	*ESPN*	Ca-insensitive actin-bundling protein
DFNB37	*MYO6*	Unconventional myosin
DFNB39	*HFG*	Hepatocyte growth factor
DFNB42	*ILDR1*	Ig-like domain–containing receptor
DFNB44	*ADCY1*	Adenylate cyclase
DFNB48	*CIB2*	Calcium and integrin binding protein
DFNB49	*BDP1*	Subunit of RNA polymerase
DFNB49	*MARVELD2*	Tight junction protein
DFNB53	*COL11A2*	Collagen protein
DFNB59	*PJVK*	Zn-binding protein
DFNB60	*SLC22A4*	Prestin, motor protein of cochlear outer hair cell
DFNB61	*SLC26A5*	Motor protein
DFNB63	*LRTOMT/COMT2*	Putative methyltransferase
DFNB66	*DCDC2*	Ciliary protein
DFNB66/67	*LHFPL5*	Tetraspan protein
DFNB68	*S1PR2*	Tetraspan membrane protein of hair cell stereocilia

Downloaded 2024-2-17 9:35 A Your IP is 154.59.125.153
Chapter 34: Disorders of Hearing, Anil K. Lalwani
©2024 McGraw Hill. All Rights Reserved. Terms of Use • Privacy Policy • Notice • Accessibility

DFNB70	*PNPT1*	Mitochondrial–RNA–import protein
DFNB73	*BSND*	Beta subunit of chloride channel
DFNB74	*MSRB3*	Methionine sulfoxide reductase
DFNB76	*SYNE4*	Part of *LINC* tethering complex
DFNB77	*LOXHD1*	Stereociliary protein
DFNB79	*TPRN*	Unknown
DFNB82	*GPSM2*	G protein signaling modulator
DFNB84	*PTPRQ*	Type III receptor-like protein-tyrosine phosphatase family
DFNB84	*OTOGL*	Otogelin-like protein
DFNB86	*TBC1D24*	GTPase-activating protein
DFNB88	*ELMOD3*	GTPase-activating protein
DFNB89	*KARS*	Lysyl-tRNA synthetase
DFNB91	*SERPINB6*	Protease inhibitor
DFNB93	*CABP2*	Calcium-binding protein
DFN94	*NARS2*	Mitochondrial asparaginyl-tRNA synthetase
DFNA97	*MET*	Oncogene/hepatocyte growth factor receptor
DFNB98	*TSPEAR*	Epilepsy-associated repeats containing protein
DFNB99	*TMEM132E*	Transmembrane protein
DFNB100	*PPIP5K2*	Diphosphoinositol-pentakisphosphate kinase
DFNB101	*GRXCR2*	Maintaining stereocilia bundles
DFNB102	*EPS8*	Epidermal growth factor receptor
DFNB103	*CLIC5*	Chloride ion transport
DFNB104	*FAM65B/RIPOR2*	Membrane-associated protein in stereocilia
DFNB106	*EPS8L2*	Actin remodeling in response to EGF stimulation
DFNB108	*ROR1*	Receptor tyrosine kinase-like orphan receptor
	WBP2	Transcriptional coactivator for estrogen receptor-alpha and progesterone receptor

Downloaded 2024-2-17 9:35 A Your IP is 154.59.125.153
Chapter 34: Disorders of Hearing, Anil K. Lalwani
©2024 McGraw Hill. All Rights Reserved. Terms of Use • Privacy Policy • Notice • Accessibility

ACCESS Medicine
Kaplan Medical Center
Access Provided by:
SILVERCHAIR INFORMATION SYSTEMS

	ESRP1	Modulates activation of G proteins
	MPZL2	Mediates epithelial cell-cell interactions in developing tissues
	CEACAM16	Cell adhesion molecule
	GRAP	Cytoplasmic signaling protein
	SPNS2	Sphingosine-1-phosphate (S1P) transporter
	CLDN9	Tight junctions
	CLRN2	Maintenance of transducing stereocilia in auditory hair cells
X-linked		
DFNX1	PRPS1	Catalyzes phosphoribosylation of ribose 5-phosphate to 5-phosphoribosyl-1-pyrophosphate
DFNX2	POU3F4	Transcription factor
DFNX4	SMPX	Small muscle protein
DFNX5	AIFM1	Mitochondrial flavin adenine dinucleotide (FAD)-dependent oxidoreductase
DFNX6	COL4A6	Collagen protein

In addition to *GJB2*, several other nonsyndromic genes are associated with hearing loss that progresses with age. The contribution of genetics to presbycusis is also becoming better understood and likely reflects a combination of genetic susceptibility impacted by environmental exposure to sound. Sensitivity to aminoglycoside ototoxicity can be maternally transmitted through a mitochondrial mutation. Susceptibility to noise-induced hearing loss may also be genetically determined.

There are >400 syndromic forms of hearing loss. These include Usher's syndrome (retinitis pigmentosa and hearing loss), Waardenburg's syndrome (pigmentary abnormality and hearing loss), Pendred's syndrome (thyroid organification defect and hearing loss), Alport's syndrome (renal disease and hearing loss), Jervell and Lange-Nielsen syndrome (prolonged QT interval and hearing loss), neurofibromatosis type 2 (bilateral acoustic schwannoma), and mitochondrial disorders (mitochondrial encephalopathy, lactic acidosis, and stroke-like episodes [MELAS]; myoclonic epilepsy and ragged red fibers [MERRF]; and progressive external ophthalmoplegia [PEO]) **(Table 34-2)**.

TABLE 34-2

Syndromic Hereditary Hearing Impairment Genes

SYNDROME	GENE	FUNCTION
Alport's syndrome	*COL4A3-5*	Cytoskeletal protein
BOR syndrome	*EYA1*	Developmental gene
	SIX5	Developmental gene
	SIX1	Developmental gene

Downloaded 2024-2-17 9:35 A Your IP is 154.59.125.153
Chapter 34: Disorders of Hearing, Anil K. Lalwani
©2024 McGraw Hill. All Rights Reserved. Terms of Use • Privacy Policy • Notice • Accessibility

ACCESS Medicine®
Kaplan Medical Center
Access Provided by:
SILVERCHAIR INFORMATION / SYSTEMS

Jervell and Lange-Nielsen syndrome	*KCNQ1* *KCNE1*	Delayed rectifier K^+ channel Delayed rectifier K^+ channel
Norrie's disease	*NDP*	Cell–cell interactions
Pendred's syndrome	*SLC26A4*	Chloride/iodide transporter
	FOXI1 *KCNJ10*	Transcriptional activator of *SLC26A4* Inwardly rectifying K^+ channel
Treacher Collins syndrome	*TCOF1* *POLR1D* *POLR1C*	Nucleolar-cytoplasmic transport Subunit of RNA polymerases I and III Subunit of RNA polymerases I and III
Usher's syndrome	*MYO7A*	Cytoskeletal protein
	USH1C	Unknown
	CDH23	Intercellular adherence protein
	PCDH15	Cell adhesion molecule
	SANS *CIB2*	Harmonin-associated protein Calcium- and integrin-binding protein
	USH2A	Cell adhesion molecule
	VLGR1	G protein–coupled receptor
	WHRN	PDZ domain–containing protein
	CLRN1 *HARS* *PDZD7*	Cellular synapse protein Histidyl-tRNA synthetase PDZ domain–containing protein
WS type I, III	*PAX3*	Transcription factor
WS type II	*MITF*	Transcription factor
	SNAI2	Transcription factor
WS type IV	*EDNRB*	Endothelin B receptor
	EDN3	Endothelin B receptor ligand
	SOX10	Transcription factor

Abbreviations: BOR, branchio-oto-renal syndrome; WS, Waardenburg's syndrome.

Downloaded 2024-2-17 9:35 A Your IP is 154.59.125.153
Chapter 34: Disorders of Hearing, Anil K. Lalwani
©2024 McGraw Hill. All Rights Reserved. Terms of Use • Privacy Policy • Notice • Accessibility

The goal in the evaluation of a patient with auditory complaints is to determine (1) the nature of the hearing impairment (conductive vs sensorineural vs mixed), (2) the severity of the impairment (mild, moderate, severe, or profound), (3) the anatomy of the impairment (external ear, middle ear, inner ear, or central auditory pathway), and (4) the etiology. The presence of signs and symptoms associated with hearing loss should be ascertained **(Table 34-3)**. The history should elicit characteristics of the hearing loss, including the duration of deafness, unilateral versus bilateral involvement, nature of onset (sudden vs insidious), and rate of progression (rapid vs slow). Symptoms of tinnitus, vertigo, imbalance, aural fullness, otorrhea, headache, facial nerve dysfunction, and head and neck paresthesias should be noted. Information regarding head trauma, exposure to ototoxins, occupational or recreational noise exposure, and family history of hearing impairment may also be important. A sudden onset of unilateral hearing loss, with or without tinnitus, may represent a viral infection of the inner ear, vestibular schwannoma, or a stroke. Patients with unilateral hearing loss (sensory or conductive) usually complain of reduced hearing, poor sound localization, and difficulty hearing clearly in the presence of background noise. Gradual progression of a hearing deficit is common with otosclerosis, noise-induced hearing loss, vestibular schwannoma, or Ménière's disease. Small vestibular schwannomas typically present with asymmetric hearing impairment, tinnitus, and imbalance (rarely vertigo); cranial neuropathy, in particular of the trigeminal or facial nerve, may accompany larger tumors. In addition to hearing loss, Ménière's disease may be associated with episodic vertigo, tinnitus, and aural fullness. Sound-induced vertigo, autophony, and being able to hear one's own neck or eye movement are highly suggestive of superior semicircular canal dehiscence. Hearing loss with otorrhea is most likely due to chronic otitis media or cholesteatoma.

TABLE 34-3

Signs and Symptoms Suggestive of Hearing Loss

Saying "huh" a great deal
Reduced clarity of hearing
Difficulty understanding conversations in background noise
Family complaining of hearing loss
Tinnitus
Turning the volume up on radio or television
Sensitivity to noises
Fullness in the ear
Avoiding social settings

Examination should include the auricle, external ear canal, and tympanic membrane. In the elderly, the external ear canal is often dry and fragile; it is preferable to clean cerumen with wall-mounted suction or cerumen loops and to avoid irrigation. Irrigation should also be avoided when a tympanic membrane perforation is present or the integrity of the eardrum cannot be established. In examining the eardrum, the topography of the tympanic membrane is more important than the presence or absence of the light reflex. In addition to the pars tensa (the lower two-thirds of the tympanic membrane), the pars flaccida (upper one-third of the tympanic membrane) above the short process of the malleus should also be examined for retraction pockets that may be evidence of chronic Eustachian tube dysfunction or cholesteatoma. Insufflation of the ear canal is necessary to assess tympanic membrane mobility and compliance. Careful inspection of the nose, nasopharynx, and upper respiratory tract is important. Unilateral serous effusion or unexplained otalgia should prompt a fiberoptic examination of the nasopharynx and larynx to exclude neoplasms. Cranial nerves should be evaluated with special attention to facial and trigeminal nerves, which are commonly affected with tumors involving the cerebellopontine angle.

The Rinne and Weber tuning fork tests, with a 512-Hz tuning fork, are used to screen for hearing loss, differentiate conductive from sensorineural hearing losses, and confirm the findings of audiologic evaluation. The Rinne test compares the ability to hear by air conduction with the ability to hear by bone conduction. The tines of a vibrating tuning fork are held near the opening of the external auditory canal, and then the stem is placed on the

Downloaded 2024-2-17 9:35 A Your IP is 154.59.125.153
Chapter 34: Disorders of Hearing, Anil K. Lalwani
©2024 McGraw Hill. All Rights Reserved. Terms of Use • Privacy Policy • Notice • Accessibility

mastoid process; for direct contact, it may be placed on teeth or dentures. The patient is asked to indicate whether the tone is louder by air conduction or bone conduction. Normally, and in the presence of sensorineural hearing loss, a tone is heard louder by air conduction than by bone conduction; however, with conductive hearing loss of ≥30 dB (see "Audiologic Assessment," below), the bone-conduction stimulus is perceived as louder than the air-conduction stimulus. For the Weber test, the stem of a vibrating tuning fork is placed on the head in the midline and the patient is asked whether the tone is heard in both ears or better in one ear than in the other. With a unilateral conductive hearing loss, the tone is perceived in the affected ear. With a unilateral sensorineural hearing loss, the tone is perceived in the unaffected ear. A 5-dB difference in hearing between the two ears is required for lateralization.

LABORATORY ASSESSMENT OF HEARING

Audiologic Assessment

The minimum audiologic assessment for hearing loss should include the measurement of pure tone air-conduction and bone-conduction thresholds, speech reception threshold, word recognition score, tympanometry, acoustic reflexes, and acoustic-reflex decay. This test battery provides a screening evaluation of the entire auditory system and allows one to determine whether further differentiation of a sensory (cochlear) from a neural (retrocochlear) hearing loss is indicated.

Pure tone audiometry assesses hearing acuity for pure tones. The test is administered by an audiologist and is performed in a sound-attenuated chamber. The pure tone stimulus is delivered with an audiometer, an electronic device that allows the presentation of specific frequencies (generally between 250–8000 Hz) at specific intensities. Air- and bone-conduction thresholds are established for each ear. Air-conduction thresholds are determined by presenting the stimulus in air with the use of headphones. Bone-conduction thresholds are determined by placing the stem of a vibrating tuning fork or an oscillator of an audiometer in contact with the head. In the presence of a hearing loss, broad-spectrum noise is presented to the nontest ear for masking purposes so that responses are based on perception from the ear under test.

The responses are measured in decibels (dBs). An *audiogram* is a plot of intensity in dBs of hearing threshold versus frequency. A dB is equal to 20 times the logarithm of the ratio of the sound pressure required to achieve threshold in the patient to the sound pressure required to achieve threshold in a normal-hearing person. Therefore, a change of 6 dB represents doubling of sound pressure, and a change of 20 dB represents a tenfold change in sound pressure. Loudness, which depends on the frequency, intensity, and duration of a sound, doubles with approximately each 10-dB increase in sound pressure level. Pitch, on the other hand, does not directly correlate with frequency. The perception of pitch changes slowly in the low and high frequencies. In the middle tones, which are important for human speech, pitch varies more rapidly with changes in frequency.

Pure tone audiometry establishes the presence and severity of hearing impairment, unilateral versus bilateral involvement, and the type of hearing loss. Conductive hearing losses with a large mass component, as is often seen in middle-ear effusions, produce elevation of thresholds that predominate in the higher frequencies. Conductive hearing losses with a large stiffness component, as in fixation of the footplate of the stapes in early otosclerosis, produce threshold elevations in the lower frequencies. Often, the conductive hearing loss involves all frequencies, suggesting involvement of both stiffness and mass. In general, sensorineural hearing losses such as presbycusis affect higher frequencies more than lower frequencies (Fig. 34-3). An exception is Ménière's disease, which is characteristically associated with low-frequency sensorineural hearing loss (though any frequency can be affected). Noise-induced hearing loss has an unusual pattern of hearing impairment in which the loss at 3000–4000 Hz is greater than at higher frequencies. Vestibular schwannomas characteristically affect the higher frequencies, but any pattern of hearing loss can be observed.

Speech recognition requires greater synchronous neural firing than is necessary for appreciation of pure tones. *Speech audiometry* tests the clarity with which one hears. The *speech reception threshold* (*SRT*) is defined as the intensity at which speech is recognized as a meaningful symbol and is obtained by presenting two-syllable words with an equal accent on each syllable. The intensity at which the patient can repeat 50% of the words correctly is the SRT. Once the SRT is determined, discrimination or word recognition ability is tested by presenting one-syllable words at 25–40 dB above the SRT. The words are phonetically balanced in that the phonemes (speech sounds) occur in the list of words at the same frequency that they occur in ordinary conversational English. An individual with normal hearing or conductive hearing loss can repeat 88–100% of the phonetically balanced words correctly. Patients with a sensorineural hearing loss have variable loss of discrimination. As a general rule, neural lesions produce greater deficits in discrimination than do cochlear lesions. For example, in a patient with mild asymmetric sensorineural hearing loss, a clue to the diagnosis of vestibular schwannoma is the presence of greater than expected deterioration in discrimination ability. Deterioration in discrimination ability at higher intensities above the SRT also suggests a lesion in the eighth nerve or central auditory pathways.

Downloaded 2024-2-17 9:35 A Your IP is 154.59.125.153
Chapter 34: Disorders of Hearing, Anil K. Lalwani
©2024 McGraw Hill. All Rights Reserved. Terms of Use • Privacy Policy • Notice • Accessibility

Tympanometry measures the impedance of the middle ear to sound and is useful in diagnosis of middle-ear effusions. A *tympanogram* is the graphic representation of change in impedance or compliance as the pressure in the ear canal is changed. Normally, the middle ear is most compliant at atmospheric pressure, and the compliance decreases as the pressure is increased or decreased (type A); this pattern is seen with normal hearing or in the presence of sensorineural hearing loss. Compliance that does not change with change in pressure suggests middle-ear effusion (type B). With a negative pressure in the middle ear, as with Eustachian tube obstruction, the point of maximal compliance occurs with negative pressure in the ear canal (type C). A tympanogram in which no point of maximal compliance can be obtained is most commonly seen with discontinuity of the ossicular chain (type A_d). A reduction in the maximal compliance peak can be seen in otosclerosis (type A_s).

During tympanometry, an intense tone elicits contraction of the stapedius muscle. The change in compliance of the middle ear with contraction of the stapedius muscle can be detected. The presence or absence of this *acoustic reflex* is important in determining the etiology of hearing loss as well as in the anatomic localization of facial nerve paralysis. The acoustic reflex can help differentiate between conductive hearing loss due to otosclerosis and that caused by an inner-ear "third window": it is absent in otosclerosis and present in inner-ear conductive hearing loss. Normal or elevated acoustic reflex thresholds in an individual with sensorineural hearing impairment suggest a cochlear hearing loss. An absent acoustic reflex in the setting of sensorineural hearing loss is not helpful in localizing the site of lesion. Assessment of *acoustic reflex decay* helps differentiate sensory from neural hearing losses. In neural hearing loss, such as with vestibular schwannoma, the reflex adapts or decays with time.

OAEs generated by outer hair cells only can be measured with microphones inserted into the external auditory canal. The emissions may be spontaneous or evoked with sound stimulation. The presence of OAEs indicates that the outer hair cells of the organ of Corti are intact and can be used to assess auditory thresholds and to distinguish sensory from neural hearing losses.

Evoked Responses

Electrocochleography measures the earliest evoked potentials generated in the cochlea and the auditory nerve. Receptor potentials recorded include the cochlear microphonic, generated by the outer hair cells of the organ of Corti, and the summating potential, generated by the inner hair cells in response to sound. The whole nerve action potential representing the composite firing of the first-order neurons can also be recorded during electrocochleography. Clinically, the test is useful in the diagnosis of Ménière's disease, in which an elevation of the ratio of summating potential to action potential is seen.

Brainstem auditory-evoked responses (*BAERs*), also known as ABRs, are useful in differentiating the site of sensorineural hearing loss. In response to sound, five distinct electrical potentials arising from different stations along the peripheral and central auditory pathway (eighth nerve, cochlear nucleus, superior olivary complex, lateral lemniscus, and inferior colliculus) can be identified using computer averaging from scalp surface electrodes. BAERs are valuable in situations in which patients cannot or will not give reliable voluntary thresholds. They are also used to assess the integrity of the auditory nerve and brainstem in various clinical situations, including intraoperative monitoring, and in determination of brain death.

The *VEMP test* investigates otolith and vestibular nerve function by presenting a high-level acoustic stimulus and evoking a short-latency electromyographic potential; cVEMP (or cervical VEMP) and oVEMP (or ocular VEMP) have been described. The cVEMP elicits a vestibulocollic reflex whose afferent limb arises from acoustically sensitive cells in the saccule, with signals conducted via the inferior vestibular nerve. cVEMP is a biphasic, short-latency response recorded from the tonically contracted sternocleidomastoid muscle in response to loud auditory clicks or tones. cVEMPs may be diminished or absent in patients with early and late Ménière's disease, vestibular neuritis, benign paroxysmal positional vertigo, and vestibular schwannoma. On the other hand, the threshold for VEMPs may be lower in cases of superior canal dehiscence, other inner-ear dehiscence ("third window"), and perilymphatic fistula. The oVEMP, in contrast, is a response involving the utricle primarily and superior vestibular nerve. The oVEMP excitatory response is recorded from the extraocular muscle. The oVEMP is abnormal in superior vestibular neuritis.

Imaging Studies

The choice of radiologic tests is largely determined by whether the goal is to evaluate the bony anatomy of the external, middle, and inner ear or to image the auditory nerve and brain. Axial and coronal CT of the temporal bone with fine 0.3-mm cuts is ideal for determining the caliber of the external auditory canal, integrity of the ossicular chain, and presence of middle-ear or mastoid disease; it can also detect inner-ear malformations. CT is also ideal for the detection of bone erosion with chronic otitis media and cholesteatoma. Pöschl reformatting in the plane of the superior semicircular canal is required for the identification of dehiscence or absence of bone over the superior semicircular canal. MRI is superior to CT for imaging of retrocochlear pathology such as vestibular schwannoma, meningioma, other lesions of the cerebellopontine angle, demyelinating lesions of the

Downloaded 2024-2-17 9:35 A Your IP is 154.59.125.153
Chapter 34: Disorders of Hearing, Anil K. Lalwani

©2024 McGraw Hill. All Rights Reserved. Terms of Use • Privacy Policy • Notice • Accessibility

brainstem, and brain tumors. Both CT and MRI are equally capable of identifying inner-ear malformations and assessing cochlear patency for preoperative evaluation of patients for cochlear implantation.

TREATMENT OF DISORDERS OF THE SENSE OF HEARING

In general, conductive hearing losses are amenable to surgical correction, whereas sensorineural hearing losses are usually managed medically. Atresia of the ear canal can be surgically repaired, often with significant improvement in hearing. Alternatively, the conductive hearing loss associated with atresia can be addressed with a bone-anchored hearing aid (BAHA). Tympanic membrane perforations due to chronic otitis media or trauma can be repaired with an outpatient tympanoplasty. Likewise, conductive hearing loss associated with otosclerosis can be treated by stapedectomy, which is successful in >95% of cases. Tympanostomy tubes allow the prompt return of normal hearing in individuals with middle-ear effusions. Hearing aids are effective and well tolerated in patients with conductive hearing losses.

Patients with mild, moderate, and severe sensorineural hearing losses are regularly rehabilitated with hearing aids of varying configuration and strength. Hearing aids have been improved to provide greater fidelity and have been miniaturized. The current generation of hearing aids is nearly invisible, thus reducing stigma associated with their use. In general, the more severe the hearing impairment, the larger the hearing aid required for auditory rehabilitation. Digital hearing aids lend themselves to individual programming, and multiple and directional microphones at the ear level may be helpful in noisy surroundings. Because all hearing aids amplify noise as well as speech, the only absolute solution to the problem of noise is to place the microphone closer to the speaker than the noise source. This arrangement is not possible with a self-contained, cosmetically acceptable device. A significant limitation of rehabilitation with a hearing aid is that although it is able to enhance detection of sound with amplification, it cannot restore clarity of hearing that is lost with presbycusis.

The cost of a single hearing aid (~$2300 US) is a significant obstacle for many hearing-impaired individuals and usually bilateral amplification is recommended. To reduce cost and spur innovation, a new category of over-the-counter amplification devices that can be purchased similar to reading eyeglasses by simply walking into a store has recently been approved by the US Food and Drug Administration. By reducing the cost of amplification devices to consumers, promoting innovation, and increasing competition, this new class of devices could fundamentally change the way hearing rehabilitation is delivered.

Patients with unilateral deafness have difficulty with sound localization and reduced clarity of hearing in background noise. They may benefit from a contralateral routing of signal (CROS) hearing aid in which a microphone is placed on the hearing-impaired side, and the sound is transmitted to the receiver placed on the contralateral ear. The same result may be obtained with a BAHA, in which a hearing aid clamps to a screw integrated into the skull on the hearing-impaired side. Like the CROS hearing aid, the BAHA transfers the acoustic signal to the contralateral hearing ear, but it does so by vibrating the skull. Patients with profound deafness on one side and some hearing loss in the better ear are candidates for a BICROS hearing aid; it differs from the CROS hearing aid in that the patient wears a hearing aid, and not simply a receiver, in the better ear. Unfortunately, while CROS and BAHA devices provide benefit, they do not restore hearing in the deaf ear. Only cochlear implants can restore hearing (see below). Increasingly, cochlear implants are being used for the treatment of patients with single-sided deafness; they show great promise in not only restoring hearing and reducing tinnitus, but also improving sound localization and performance in background noise.

In many situations, including lectures and the theater, hearing-impaired persons benefit from assistive devices that are based on the principle of having the speaker closer to the microphone than any source of noise. Assistive devices include infrared and frequency-modulated (FM) transmission as well as an electromagnetic loop around the room for transmission to the individual's hearing aid. Hearing aids with telecoils can also be used with properly equipped telephones in the same way. Bluetooth technology has revolutionized connectivity between hearing aids and other devices such as smart phones.

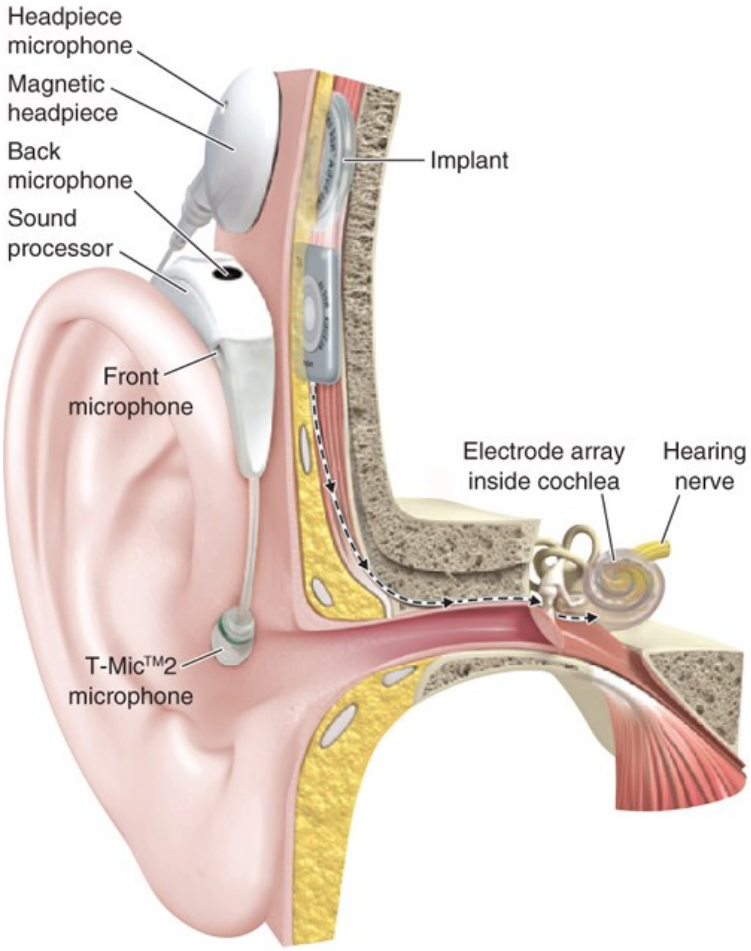
In the event that the hearing aid provides inadequate rehabilitation, cochlear implants may be appropriate **(Fig. 34-4)**. Criteria for implantation include severe to profound hearing loss with open-set sentence cognition of ≤40% under best-aided conditions. Worldwide, >600,000 hearing-impaired individuals have received cochlear implants. Cochlear implants are neural prostheses that convert sound energy to electrical energy and can be used to stimulate the auditory division of the eighth nerve directly. In most cases of profound hearing impairment, the auditory hair cells are lost but the ganglionic cells of the auditory division of the eighth nerve are preserved. Cochlear implants consist of electrodes that are inserted into the cochlea through the round window, speech processors that extract acoustic elements of speech for conversion to electrical currents, and a means of transmitting the electrical energy through the skin. Patients with implants experience sound that helps with speech reading, allows open-set word recognition, and helps in modulating the person's own voice. Usually, within the first 3–6 months after implantation, adult patients can understand

Downloaded 2024-2-17 9:35 A Your IP is 154.59.125.153
Chapter 34: Disorders of Hearing, Anil K. Lalwani
©2024 McGraw Hill. All Rights Reserved. Terms of Use • Privacy Policy • Notice • Accessibility

speech without visual cues. With the current generation of multichannel cochlear implants, nearly 75% of patients are able to converse on the telephone. Bilateral cochlear implantations are commonly performed, especially in children; these patients perform better in background noise, have better sound localization, and are less fatigued by the "work" compared to monaural hearing.

FIGURE 34-4

A cochlear implant is composed of an external microphone and speech processor worn on the ear and a receiver implanted underneath the temporalis muscle. The internal receiver is attached to an electrode that is placed surgically in the cochlea.



Source: Joseph Loscalzo, Anthony Fauci, Dennis Kasper, Stephen Hauser, Dan Longo, J. Larry Jameson: Harrison's Principles of Internal Medicine, 21e
Copyright © McGraw Hill. All rights reserved.

Hybrid cochlear implants are indicated for the treatment of high-frequency hearing loss in patients who do not have profound hearing loss and yet do not benefit from hearing aids. Patients with presbycusis typically have normal low-frequency hearing while suffering from high-frequency hearing loss associated with loss of clarity that cannot always be adequately rehabilitated with a hearing aid. However, these patients are not candidates for conventional cochlear implants because they have too much residual hearing. The hybrid implant has been specifically designed for this patient population; it has a shorter electrode than a conventional cochlear implant and can be introduced into the cochlea atraumatically, thus preserving low-frequency hearing. Individuals with a hybrid implant use their own natural low-frequency "acoustic" hearing and rely on the implant for providing "electrical" high-frequency hearing. Patients who have received the hybrid implant perform better on speech discrimination tests in both quiet and noisy backgrounds.

For individuals who were born without cochlea or have had both eighth nerves destroyed by trauma or bilateral vestibular schwannomas (e.g., neurofibromatosis type 2), brainstem auditory implants placed near the cochlear nucleus may provide auditory rehabilitation. Currently, brainstem implants provide sound awareness but unfortunately speech understanding remains elusive.

Downloaded 2024-2-17 9:35 A Your IP is 154.59.125.153
Chapter 34: Disorders of Hearing, Anil K. Lalwani
©2024 McGraw Hill. All Rights Reserved. Terms of Use • Privacy Policy • Notice • Accessibility

Tinnitus often accompanies hearing loss. Similar to background noise, tinnitus can degrade speech comprehension in individuals with hearing impairment. Patients with tinnitus should be advised to minimize caffeine ingestion, avoid high dosage of nonsteroidal anti-inflammatory drugs (NSAIDs), and reduce stress. Therapy for tinnitus is usually directed toward minimizing the appreciation of tinnitus. Relief of the tinnitus may be obtained by masking it with background music or white noise. Hearing aids are also helpful in tinnitus suppression, as are tinnitus maskers, devices that present a sound to the affected ear that is more pleasant to listen to than the tinnitus. The use of a tinnitus masker is often followed by several hours of inhibition of the tinnitus. Antidepressants have also been shown to be beneficial in helping patients cope with tinnitus.

Hard-of-hearing individuals often benefit from a reduction in unnecessary noise in the environment (e.g., radio or television) to enhance the signal-to-noise ratio. Speech comprehension is aided by lip reading; therefore, the impaired listener should be seated so that the face of the speaker is well illuminated and easily seen. Although speech should be in a loud, clear voice, one should be aware that in sensorineural hearing losses in general and in hard-of-hearing elderly in particular, recruitment (abnormal perception of loud sounds) may be troublesome. Above all, optimal communication cannot take place without both parties giving it their full and undivided attention.

PREVENTION

Conductive hearing losses may be prevented by prompt antibiotic therapy of adequate duration for AOM and by ventilation of the middle ear with tympanostomy tubes in middle-ear effusions lasting ≥12 weeks. Loss of vestibular function and deafness due to aminoglycoside antibiotics can largely be prevented by careful monitoring of serum peak and trough levels.

Some 10 million Americans have noise-induced hearing loss, and 20 million are exposed to hazardous noise in their employment. Noise-induced hearing loss can be prevented by avoidance of exposure to loud noise or by regular use of earplugs or fluid-filled ear muffs to attenuate intense sound. **Table 34-4** lists loudness levels for a variety of environmental sounds. High-risk activities for noise-induced hearing loss include use of electrical equipment for wood- and metalworking, and target practice or hunting with small firearms. All internal-combustion and electric engines, including snow and leaf blowers, snowmobiles, outboard motors, and chainsaws, require protection of the user with hearing protectors. Virtually all noise-induced hearing loss is preventable through education, which should begin before the teenage years. Programs for conservation of hearing in the workplace are required by the Occupational Safety and Health Administration (OSHA) whenever the exposure over an 8-h period averages 85 dB. OSHA mandates that workers in such noisy environments have hearing monitoring and protection programs that include a preemployment screen, an annual audiologic assessment, and the mandatory use of hearing protectors. Exposure to loud sounds above 85 dB in the work environment is restricted by OSHA, with halving of allowed exposure time for each increment of 5 dB above this threshold; for example, exposure to 90 dB is permitted for 8 h; 95 dB for 4 h, and 100 dB for 2 h **(Table 34-5)**.

Downloaded 2024-2-17 9:35 A Your IP is 154.59.125.153
Chapter 34: Disorders of Hearing, Anil K. Lalwani
©2024 McGraw Hill. All Rights Reserved. Terms of Use • Privacy Policy • Notice • Accessibility

ACCESS Medicine®
Kaplan Medical Center
Access Provided by:
SILVERCHAIR INFORMATION SYSTEMS

TABLE 34-4

Decibel (Loudness) Level of Common Environmental Noise

SOURCE	DECIBEL (dB)
Weakest sound heard	0
Whisper	30
Normal conversation	55–65
City traffic inside car	85
OSHA Monitoring Requirement Begins	**90**
Jackhammer	95
Subway train at 200 ft	95
Power mower	107
Power saw	110
Painful Sound	**125**
Jet engine at 100 ft	140
12-gauge shotgun blast	165
Loudest sound that can occur	194

Abbreviation: OSHA, Occupational Safety and Health Administration.

Downloaded 2024-2-17 9:35 A Your IP is 154.59.125.153
Chapter 34: Disorders of Hearing, Anil K. Lalwani
©2024 McGraw Hill. All Rights Reserved. Terms of Use • Privacy Policy • Notice • Accessibility

TABLE 34-5

OSHA Daily Permissible Noise Level Exposure

SOUND LEVEL (dB)	DURATION PER DAY (h)
90	8
92	6
95	4
97	3
100	2
102	1.5
105	1
110	0.5
115	≤0.25

Note: Exposure to impulsive or impact noise should not exceed 140-dB peak sound pressure level.

Source: From *https://www.osha.gov/pls/oshaweb/owadisp.show_document?p_table=standards&p_id=9735*.

FURTHER READING

Carlson ML: Cochlear implantation in adults. N Engl J Med 382:1531, 2020. [PubMed: 32294347]

Espinosa-Sanchez JM, Lopez-Escamez JA: Menière's disease. Handb Clin Neurol 137:257, 2016. [PubMed: 27638077]

Moser T, Starr A: Auditory neuropathy—neural and synaptic mechanisms. Nat Rev Neurol 12:135, 2016. [PubMed: 26891769]

Patel M et al: Intratympanic methylprednisolone versus gentamicin in patients with unilateral Ménière's disease: a randomised, double-blind, comparative effectiveness trial. Lancet 388:2753, 2016. [PubMed: 27865535]

Tikka C et al: Interventions to prevent occupational noise-induced hearing loss. Cochrane Database Syst Rev 7:CD006396, 2017. [PubMed: 28685503]

Wilson BS et al: Global hearing health care: new findings and perspectives. Lancet 390:2503, 2017. [PubMed: 28705460]

Downloaded 2024-2-17 9:35 A Your IP is 154.59.125.153
Chapter 34: Disorders of Hearing, Anil K. Lalwani
©2024 McGraw Hill. All Rights Reserved. Terms of Use • Privacy Policy • Notice • Accessibility