

Hearing Loss

Hearing loss occurs when there is loss of sound sensitivity produced by an abnormality anywhere in the auditory system. A wide variety of conditions can cause hearing loss, including otosclerosis, cholesteatoma, and others. While physicians can sometimes identify the causes of hearing loss, in some cases the causes are unknown, or idiopathic.

Conductive Hearing Loss: Conductive hearing loss occurs when sound waves are prevented from passing from the air to the fluid-filled inner ear. This may be caused by a variety of problems including buildup of earwax (cerumen), infection, fluid in the middle ear, a punctured eardrum, or fixation of the ossicles, as in otosclerosis. Other causes include scarring, narrowing of the ear canal, tumors in the middle ear, and perforation of the tympanic membrane. Once the cause is found and removed or treated, hearing usually is restored.

Sensorineural Hearing Loss: Sensorineural hearing loss develops when the auditory nerve or hair cells in the inner ear are damaged. The source may be located in the inner ear, the nerve from the inner ear to the brain, or in the brain. Sensorineural hearing loss, commonly referred to as "nerve deafness," frequently occurs as a result of the aging process in the form of presbycusis, which is a gradual loss occurring in both ears. Tumors such as acoustic neuromas can lead to sensorineural hearing losses, as can viral infections, Meniere's disease, meningitis, and cochlear otosclerosis. Sensorineural hearing loss can also be the result of repeated, continuous loud noise exposure, certain toxic medications, or an inherited condition. Generally, it is non-reversible. Scientists have, however, made great progress in uncovering the genes responsible for a number of forms of congenital hearing impairments/ deafness, and this genetic research may in time lead to therapies for some congenital causes of hearing loss.

Sensorineural hearing loss may be further differentiated as **sensory** or **neural**. Sensory hearing loss refers to loss caused by abnormalities in the cochlea, such as by damage from noise trauma, viral infection, drug toxicity, or Meniere's disease. Neural loss stems from problems in the auditory (eighth cranial) nerve, such as tumors or neurologic disorders. While tumors in this nerve may be life threatening, they are also often curable.

Mixed Hearing Loss is a combination of both conductive and sensorineural hearing loss.

Hearing loss may be partial or total. It may develop gradually or suddenly. People with hearing loss may experience difficulties hearing conversations, especially if there is background noise. Hissing, roaring, or ringing in the ears (tinnitus) occurs in some conditions, as may dizziness or problems with balance (vertigo).

Evaluating Hearing Loss

We will perform a number of examinations to determine the presence, extent, location, magnitude, and qualities of any hearing loss. A physical exam, using an otoscope, will evaluate the ear canal and tympanic membrane. The nose, nasopharynx, and upper respiratory tract will also be examined.

Air conduction tests, which stimulate the ear using air, test the hearing ability of the external auditory canal, the middle ear, and the integrity of the inner ear, eighth cranial nerve, and central auditory pathways.

Bone conduction tests use vibrating tuning forks placed in contact with the head. By bypassing the external auditory canal and middle ear, bone conduction tests can help distinguish problems in the inner ear, eighth cranial nerve, and central auditory pathways.

The **Weber and Rinne tuning fork tests** can distinguish between conductive and sensorineural hearing losses.

The **Weber test** may be performed using a 256 or 512 Hz fork. During this test, the stem of a vibrating tuning fork is placed on the head in the midline. If the tone is perceived in the affected ear, this indicates a unilateral conductive hearing loss. In the case of unilateral sensorineural hearing loss, the tone is heard in the unaffected ear instead. In the **Rinne test**, air and bone conduction tests are compared. In normal hearing tones are louder by air conduction than by bone conduction. In conductive hearing loss, however, the bone-conduction stimulus is perceived as louder. In sensorineural hearing loss both air and bone conduction sounds are diminished, but the air conduction sound is perceived as louder. The Rinne test is most sensitive in detecting mild conductive hearing losses if a 256 Hz fork is used.

Audiometry involves the presentation of tones or speech to each ear. A range of frequencies is used, and the patient's pattern of response is analyzed. Patients raise their hands in response to the sound, or in the case of children, conditioned play audiometry (CPA) is used.

Acoustic Reflex testing measures the contraction of a tiny ear muscle that responds to sounds at different volumes. The loudness at which the reflex occurs, or the absence of the acoustic reflex, provides important information.

Tympanometry measures the impedance of the middle ear to sound. It uses an airtight seal and a microphone to deliver sound into the ear canal. The amount of sound that is absorbed or reflected from the middle ear is measured at the microphone at normal, positive, and negative air pressures. Tympanometry is useful in identifying middle-ear effusions in children, and it may also detect facial nerve paralysis and may differentiate sensory from neural hearing losses.

Otoacoustic Emissions (OAE) testing uses sensitive microphones inserted into the external auditory canal. The presence of otoacoustic emissions indicates that the outer hair cells are intact, and this can help distinguish sensory from neural hearing losses. OAE are especially robust in infants, and this painless test is routinely performed on newborns.

Auditory Brainstem Response (ABR) testing is also used to test newborns, although ABR fails to detect half of infants with profound hearing impairment. ABR is, however, the most powerful method of detecting the site of sensorineural hearing loss. In this painless test, electrodes are placed on the scalp and sounds are presented. Distinct waveforms are recorded by computer, and these waveforms reflect conditions such as lesions in the eighth cranial nerve or brainstem.

Other tests: patients with conductive hearing losses may require **computed tomography (CT) scans** of the temporal bones. Those with unilateral or asymmetric sensorineural hearing loss should have **magnetic resonance imaging (MRI)** of the head. Many patients with sensorineural hearing loss should receive **electronystagmography and caloric testing** of the vestibular system.