10.2.8

Disorders of the Ear - Otosclerosis, Tinnitus, and Hearing Loss

Otosclerosis

Otosclerosis involves formation of new spongy bone around the stapes and oval window. It results in progressive conductive hearing loss due to immobilization of the middle ear bones. It can occur in one or both ears. This condition is genetic and typically begins between ages 15-35. Otosclerosis can cause mild to severe hearing loss, but it rarely leads to complete loss of hearing. The most effective treatment is reconstruction or replacement of the stapes, but hearing aids can be used as well.

Tinnitus

Tinnitus is the perception of ringing, buzzing, hissing, or whistling sounds in one or both of the ears. This noise isn't from an external sound, and usually other people can't hear it. It is often associated with an underlying problem such as hearing loss, ear injury, or a problem with the circulatory system. It can be constant or come and go. Tinnitus can be divided into two categories:

- **Objective tinnitus** is potentially detectable by another observer. It can be caused by turbulent blood flow due to vascular disorders. This type of tinnitus is also called pulsatile tinnitus and often occurs in a rhythmic whooshing sound that is in sync with the heartbeat.
- **Subjective tinnitus** is tinnitus that only the patient can hear. It can be caused by a buildup of cerumen, medications, noise-induced hearing loss, HTN, atherosclerosis, or a head injury. Low noise generators are often used as treatment to facilitate auditory adaptation to the tinnitus.

Hearing Loss

Conductive hearing loss is caused when stimuli is not conducted from the eardrum and ossicles to the inner ear. It can be due to impacted cerumen, fluid, foreign bodies, otosclerosis, and Paget's disease.

Sensorineural hearing loss happens after inner ear damage or because of issues with the nerve pathways between the inner ear and the brain. Sensorineural hearing loss may have a genetic cause or be due to exposure to loud noises, an infection (e.g., bacterial meningitis), trauma, or illness. Some drugs can also cause this type of hearing loss. Antibacterial drugs known as aminoglycosides, chemotherapeutic agents, certain antimalarial drugs, some loop diuretics, and salicylates (e.g., aspirin) are all associated with ototoxicity and potential hearing loss. The damage to the inner ear structures due to these drugs in some cases is permanent. It depends on the dose and duration of the drug treatment.

There are two screening techniques used to assess hearing in neonates:

1. **Transient otoacoustic emissions (TOAEs):** an otoacoustic emission (OAE) is a sound that is generated by the cochlea as a part of hearing. This sound is assumed to be related to the amplification function of the cochlea's outer rows of hair cells. The function of the cochlear outer hair cells is to contract up and down (via the protein prestin) to amplify the movement on the basement membrane and lead to increased sensation of sound by the inner hair cells. The movement of the outer row of hair cells can produce a sound wave that is transferred back through the cochlea and through the middle ear and can be picked up by a very sensitive microphone on the ear canal. Trained professionals in OAE testing will use a series of clicks to assess the expected response by the cochlea. Unexpected results on this test may suggest damage to the cochlea or cranial nerve VIII or both.
2. **Auditory brainstem response (ABR):** this test is done by using an electroencephalogram (EEG) to measure waves generated by the brain after applying a stimulus like tones or clicks to the hearing apparatus. The EEG waves will be analyzed to assess the amplitude, frequency, and latency (speed of transmission) of neuronal activity. This information gives some clues as to how well the neural-sensory side of hearing is working, including the conduction of signals in the central nervous system.

A **cochlear implant** can be used to help restore some hearing function for those who have hearing loss or impaired hearing. It is a small electronic device that has an external component and an internal component. The external component sits near the ear and contains one or more microphones that pick up sound. The device may also contain some sound filters to more accurately isolate sound frequencies in the audible voice range. This helps with speech recognition. The microphone then passes the sound wave energy to a receiver that passes an electrical current to an array of electrodes surgically implanted in the cochlea. As the electrical signals stimulate the cochlear tissue, auditory neurons are depolarized in an area of the cochlea that approximates a section normally stimulated by a similar but natural sound wave frequency.

An implant does not restore normal hearing. Instead, it can give a hearing impaired person a useful representation of sounds in the environment and help him or her to understand speech. Studies have shown that when young children get a cochlear implant, they are able to comprehend music, speech, and environmental sounds at a faster rate than children with comparable hearing deficiencies who do not get an implant. Some adults who have severe hearing loss may also choose to get a cochlear implant. For these adults, the device will help them as they learn to associate the signals from the implant with sounds they remember, including language.