

Sensorineural Hearing Loss

By David Foyt, M.D.

This paper covers basic information on sensorineural hearing loss.

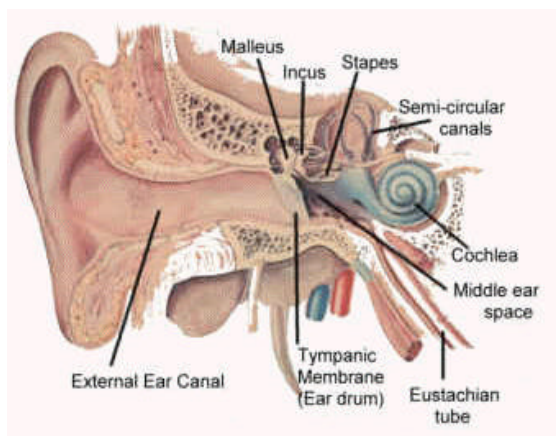
Sensorineural hearing loss, also commonly referred to as nerve deafness, affects 31 million Americans every yearⁱⁱⁱ. One in ten individuals is affected with a communication disorder which creates an enormous financial burden to the community. Sensorineural hearing loss not only involves a reduction in sound level, or ability to hear faint sounds, but also affects speech understanding, or ability to hear clearly.

How the Ear Works

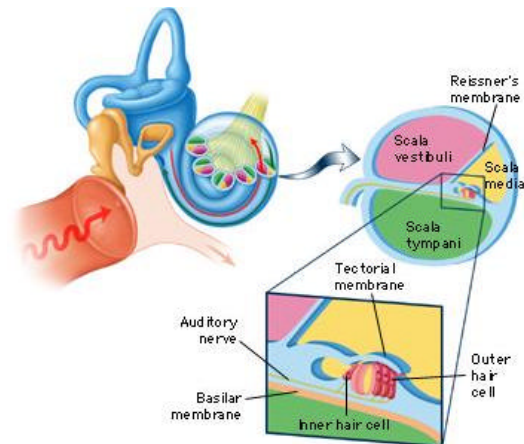
In order to understand hearing loss one must first have an understanding of how the ear perceives sound. While the intricate details of this process is still not known, the basic process is understood. The ear can be broken down into three units:

The **outer ear** (auricle, ear canal, eardrum) ; and **middle ear** (eardrum, hearing bones) function to transmit the physical vibrations of sound to the delicate nerve endings of the inner ear (**cochlea**).

Hearing loss caused by the first two units is called a conductive hearing loss since sound cannot be properly conducted to the nerves of the inner ear. Hearing loss caused by any process after the outer and middle ears is called a sensorineural, or nerve, hearing loss.

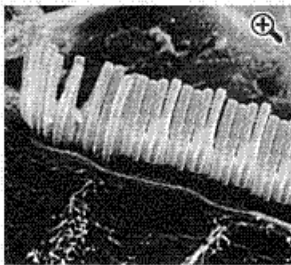


The process by which the ear translates the physical vibrations of sound to an electrical nerve



signal is complex and still only superficially understood.

The structure within the cochlea that receives sound is called the Organ of Corti after a nineteenth century anatomist. Within this structure are cells that have cilia (not really "hair" cells) that come out of one end. These cilia cause an electrochemical discharge in the cell when they are bent. Sound vibrations within the cochlea cause bending of these cilia which then causes a nerve impulse to travel to nerve endings called ganglion cells. These ganglion cells send the signals to the brain.



Sensorineural hearing loss can be caused by disorders of the cochlear nerve (Cranial nerve VIII), the inner ear, or central processing centers of the brain. The most common form is a progressive age related hearing loss called presbycusis. This type of loss is caused by the death and loss of the delicate hair cells within the inner ear. The cause for this hair cell loss is unknown. There are many other causes of Nerve Deafness as outlined bellow:

Common Causes of Nerve Deafness

Presbycusis. For unknown reasons many people start to lose some hearing as they age. This is a neural-type hearing loss believed to be the result of death of the inner ear hair cells. There are three patterns of age-related hearing loss that have been described. The most common is a high frequency

Noise Exposure. Excessive exposure to noise is known to cause damage to the delicate inner ear hair cells causing hearing loss. The Occupational Safety and Health Administration (OSHA) has issued guidelines for safe noise exposure. Acceptable exposure is dependent on the amount of time that the person is exposed to the noise. These guidelines can be found at their web site: http://www.osha.gov/pls/oshaweb/owadisp.show_document?p_table=STANDARDS&p_id=9735.

Congenital (born with). Contrary to popular thought most types of congenital hearing loss is not caused by problems with hair cells or nerve endings. The most common nonsyndromal hearing loss, the connexin 26 gene mutation) is actually caused by a defect in the ability of the ear to produce energy. Congenital hearing loss may present at birth but also may be delayed for several years.

Acoustic Neuroma. Acoustic Neuroma (properly called Vestibular Schwannoma) is a benign growth of the covering of the balance nerve. It typically causes hearing loss in only one ear. Any asymmetry in hearing loss or tinnitus requires an MRI (magnetic resonance imaging) to confirm that a neuroma does not exist.

Medication. The list of medications that can cause hearing loss is quite extensive. A list can be found at: <http://www.hearinglossweb.com/Medical/Causes/oto/oto.htm>. Generally the most common are cancer fighting drugs and some antibiotics that affect rapidly repairing cells such as found in the inner ear.

Other Diseases Other diseases can affect hearing are: Diabetes, atherosclerosis, hyperlipidemia (high cholesterol), and heart disease. These disorders affect the blood flow to the inner ear.

Severity of Hearing loss

Hearing loss is graded according to what a person can hear at multiple frequencies (pitches) during a hearing test or audiogram. The value, called the pure tone average, is given as an average of three or four of the most important frequencies in decibels (dB). Below is a chart of how hearing loss is graded.

Degree of hearing loss	Hearing loss range (dB HL)
Normal	-10 to 15
Slight	16 to 25

Mild	26 to 40
Moderate	41 to 55
Moderately severe	56 to 70
Severe	71 to 90
Profound	91+

Hearing loss is not simply a problem with hearing the sound intensity, but also a problem with the intelligibility of sound. This is called speech discrimination and is graded as a percentage score of words correctly heard during a hearing test. A typical complaint that people with hearing impairment have is that they can hear, but they cannot understand.

The Options

There are now many different options available to people who have hearing loss. If a doctor tells you nothing can be done about “nerve deafness” or “sensorineural hearing loss,” he or she is wrong. See further help by seeing an otologist (ear doctor) or otolaryngologist (ear-nose-throat doctor). After a medical exam to rule out medical causes, see a certified audiologist or hearing specialist for a hearing evaluation.

Hearing Aids: A hearing aid is a small electronic device that you wear in or behind your ear. It makes some sounds louder so that a person with hearing loss can listen, communicate, and participate more fully in daily activities. A hearing aid can help people hear more in both quiet and noisy situations. However, only about one out of five people who would benefit from a hearing aid actually uses one.

A hearing aid has three basic parts: a microphone, amplifier, and speaker. The hearing aid receives sound through a microphone, which converts the sound waves to electrical signals and sends them to an amplifier. The amplifier increases the power of the signals and then sends them to the ear through a speaker

Hearing aids have come a long way from simple ear trumpets available at the turn of the century to high powered miniature digital computers that can be fine tuned to the person’s hearing loss. By New York State law hearing aids cannot be dispensed in a physician’s office. A medical clearance is required prior to fitting an aid.

Cochlear Implant: A cochlear implant is a complex electronic device that can help to provide a sense of sound to a person who is profoundly deaf or severely hard-of-hearing. The implant consists of an external portion that sits behind the ear and a second portion that is surgically placed under the skin. An electrode wire is placed into the cochlea. The electrical signal bypasses the cochlear hair cells and directly stimulates the ganglion nerve cells of the inner ear. According to the Food and Drug Administration (FDA), at the end of 2006, more than 112,000 people worldwide had received implants. In the United States, roughly 23,000 adults and 15,500 children have received them.

Baha implant: The Baha, an implantable device, can be used to allow patients with single-sided deafness to perceive sound from their “dead ear” side. The device is comprised of a small titanium screw that is implanted into the bone behind the ear. Bone is allowed to grow into the screw after which a bone vibrating hearing device is attached.

CROS, BiCROS, and Transear Hearing aids: CROS (Contralateral Routing of Signal) are also used for single sided hearing loss. They work by sending sound from a microphone situated in the “dead” ear to the good ear. The transear is a deep fitting hearing device that sends signals to the good ear by bone vibration much like the BAHA.

Vibrant Soundbridge: The Vibrant Soundbridge is an implanted hearing device that is FDA approve for mild to moderate hearing loss. The device has two components: one is implanted under the skin and attached to the hearing bones, the other is worn externally. Surgery takes about an hour with people going home the same day. The Soundbridge is nearly invisible and since there is nothing sitting in the ear canal ear infections and wax are not issues. Better speech discrimination scores over traditional hearing aids have been documented.ⁱⁱⁱ

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ⁱ Mohr, P.E., Feldman, J.J., Dunbar, J.L., et. al. (2000). The societal costs of severe to profound hearing loss in the United States. *International Journal of Technology Assessment in Health Care*, 16(4): 1120-1135.

ⁱⁱ Benson, V., & Marano, M.A. (1995). Current estimates from the National Health Interview Survey, 1993. *National Center for Health Statistics. Vital Health Stat* 10(190).

ⁱⁱⁱ Truy E, et al. Vibrant Soundbridge Versus Conventional Hearing Aid in Sensorineural High-Frequency Hearing Loss: A Prospective Study, *Otol Neurotol*, 29(5) 684-7,2008