

markedly improved. This, in turn, has led to a broadening of criteria for implant patients. Select patients with severe hearing loss who receive some benefit from hearing aids are considered possible implant candidates.

—*Marcia J. Weiss, J.D.*

See also Aging; Audiology; Ear surgery; Ears; Hearing loss; Hearing tests; Otorhinolaryngology; Sense organs.

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HEARING LOSS

DISEASE/DISORDER

ANATOMY OR SYSTEM AFFECTED: Ears, nervous system

SPECIALTIES AND RELATED FIELDS: Audiology, geriatrics and gerontology, neurology, occupational health, otorhinolaryngology

DEFINITION: Loss of sensitivity to sound pressure changes as a result of congenital factors, disease, traumatic injury, noise exposure, or aging.

KEY TERMS:

aging process: the process in which physiological, neurological, and biological changes affect behavior and function

auditory: referring to the ear and to the sense of hearing
auditory cortex: that portion of the temporal lobe in the human brain where the ascending auditory pathway terminates

auditory nerve: the cochlear branch of the eighth cranial nerve

neural hearing loss: hearing impairment caused by a loss of the neural tissue that constitutes the ascending pathway of the auditory system

ossicles: three small bones located in the middle ear that convey sound pressure changes from the tympanic membrane (eardrum) to the oval window of the cochlea; the bones are commonly referred to as the hammer (malleus), anvil (incus), and stirrup (stapes)

phonology: the study of the sounds that make up any verbal language system

sensory hearing loss: hearing impairment caused by a loss of sensory cells (nerve cells) in the cochlea

THE PHYSIOLOGY OF THE EAR

The normal, young ear is capable of detecting frequencies (tones) from as low as 20 hertz to as high as 20,000 hertz. (Hertz is the current notation for cycles per second.) Actually, in terms of frequency, humans can hear as low as 2 hertz, but about 20 hertz is required for a perception of “tonality.” This is an amazing range. At the very low end of the frequency scale, one is not certain whether a tone is being “heard” or whether the sensation is a “tactile” one. (There is some neuroanatomical speculation that the organ of hearing is a very specialized tactile sensor.) At the very high end of the frequency scale, one can detect the highest strings of the violin, the rustle of leaves as they are disturbed by the wind, and the distinctive cry of birds and animals. A host of other sounds in between the lowest and highest frequencies can also be perceived. The subjective, psychological correlate of frequency is pitch. In general, the higher the frequency of a sound, the higher is the perceived pitch.

The ear performs an amazing feat in dealing with the broad range of intensities. Intensity is directly proportional to the magnitude of sound pressure change. The ear is so sensitive to small changes in sound pressure that the normal ear is capable of detecting pressure changes no greater than the diameter of a hydrogen molecule. At the other end of this continuum, the human ear can withstand great amplitude changes in sound pressure without damage. In terms of sound pressure units, the range from the weakest sound detected to the loudest sound tolerated represents a ratio of 10,000,000:1. In other words, the most intense sound pressure that is bearable is on the order of 10,000,000 times as great as the softest one that is perceptible under optimum listening conditions.

Intensity is expressed in decibels. Essentially, the higher the decibel value, the more intense or louder the sound. The ear is sensitive to a range of intensities from about 0 decibels to about 135 decibels. At the very extreme of this range, pain is experienced and permanent damage can occur if the sound intensity is prolonged. The psychological correlate of intensity is loudness. Under ideal conditions, if the intensity is increased by 9 decibels, the signal is perceived as being twice as loud.

The ability to process acoustic information correctly depends on the critical relationship between frequency and intensity. The critical frequency range needed to un-

INFORMATION ON HEARING LOSS

CAUSES: Buildup of earwax, perforated eardrum, disease, congenital malformation, swelling of external ear canal, otitis media, traumatic injury, long-term exposure to loud and continuous noise, tumors

SYMPTOMS: Difficulty understanding speech, difficulty hearing in presence of background noise, social isolation

DURATION: Ranges from short-term to chronic

TREATMENTS: Rehabilitation programs, use of hearing aids, surgery

derstand the English phonological system extends from about 300 hertz to 4,000 hertz. Theoretically, if one heard no other sounds above 4,000 hertz and below 300 hertz, one would experience no difficulty in understanding the intended message. Relative to intensity, a listener would have no difficulty in information processing if speech were presented at 65 decibels at a distance of one meter. In addition to the listener's distance from the speech source, speech understanding is also influenced by ambient background noise. For adequate processing to occur, speech must be louder than the ambient noise.

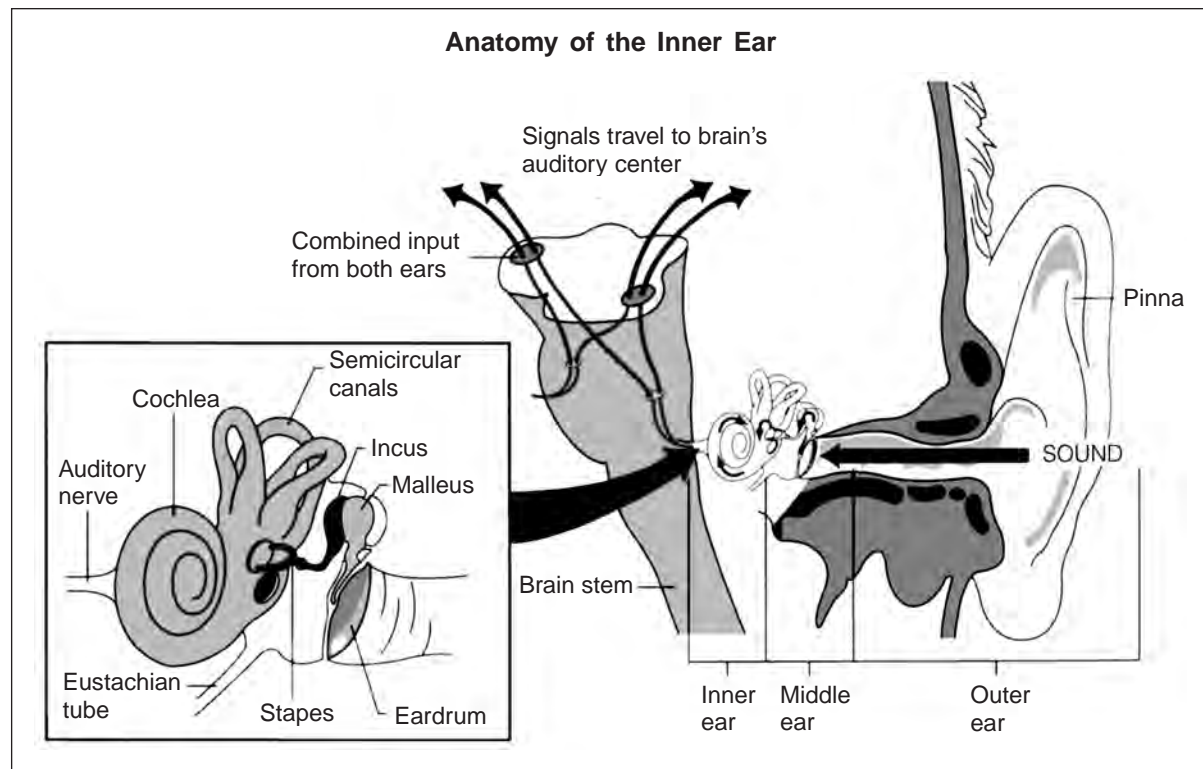
Sound is produced by a generating source such as a musical instrument, sounds of nature, or the vibrations of the human vocal cords that create a voice. The resulting series of sound pressure changes represents all the frequency and intensity characteristics generated at the source. These sound pressure changes travel through the atmosphere at a constant speed (approximately 335 meters per second at sea level). They are channeled through the external ear canal and strike the eardrum (tympanic membrane). The eardrum is sensitive to negative and positive changes in sound pressure and moves in concert with these changes. The greater the sound pressure, the greater is the magnitude of movement. Similarly, the higher the frequency, the more rapid are the movements of the eardrum. In the middle ear of the hearing system are three small bones, called ossicles. The first of these, the hammer (malleus), is attached to the eardrum. The second, the anvil (incus), is attached by ligaments to the malleus and to the third ossicle, the stirrup (stapes), which in turn attaches to the oval window membrane of the cochlea. As the eardrum is displaced in a negative or positive direction by sound pressure changes in the external ear canal, there is an absolute and corresponding movement of the ossicles.

In the cochlea, there are three anatomical divisions: the scala vestibuli, the scala tympani, and the scala media. Each of these spaces is filled with a fluid. The fluid of the scala media is chemically different from the fluid in the other two spaces. Housed in the scala media are all the specialized nervous tissues that respond to the movement of the fluid. These specialized nerve cells are situated on top of an anatomical structure called the basilar membrane. The wave forms generated within the fluid are caused by the movement of the stapes bones, as the oval window membrane communicates directly with the fluid of the cochlear space. The rate at which the stapes moves will cause the basilar membrane to be displaced at a site-specific location along its length. High-frequency sounds generate amplitude changes of the membrane at the apical portion. The movement of the membrane causes the small nerve cells (hair cells) to "fire," creating a neural discharge that travels from the cochlea to the temporal cortex of the brain. The neural events generated by sound pressure changes are interpreted by the brain.

CAUSES, SYMPTOMS, AND TREATMENTS

In the truest sense, hearing loss is any reduction in threshold sensitivity for any frequency, including those below or above the range for the normal hearing of speech. The real issue, however, is whether minor changes in sensitivity create significant problems in understanding speech and other information-bearing acoustic signals. For example, it is known that loss of threshold sensitivity below 300 hertz and above 4,000 hertz has a minimal effect on understanding speech information. It is when hearing loss exists within this critical frequency range that an individual may experience appreciable difficulty in understanding intended messages. The question becomes, then, "What conditions may cause a permanent or temporary loss of hearing, and how is such a loss managed by medical, surgical, or rehabilitative intervention?"

Conductive Hearing Loss. Any barrier or impedance that keeps sound from reaching the cochlea of the human auditory system at its intended loudness is termed "conductive hearing loss." A very common cause of conduction loss is a buildup of earwax (cerumen) in the external ear canal. The production of earwax in the ear canal is essential. It prevents the skin of the ear canal from drying and sloughing off, and it may serve to trap minute foreign particles and keep them from causing damage to the external canal. Normally, earwax will migrate out of the ear and create no conduction prob-



lem. It is when the earwax accumulates to an amount sufficient to block sound from entering the ear that something needs to be done. In most cases, earwax can be removed by irrigation. A physician washes out the earwax using a special liquid solution that does not damage the tissue of the ear canal or the eardrum itself.

Another cause of conductive hearing loss is a hole (perforation) in the eardrum, which can be created by a number of conditions, including injury. Depending on the size and location of the hole, surgery (tympanoplasty) is often successful in restoring normal hearing function. For some persons, otosclerosis (a disease causing hardening and fixing of the three small bones in the middle-ear space) results in significant conductive hearing impairment. Otosclerosis prevents these tiny bones from moving efficiently as the eardrum moves, and hearing sensitivity is reduced. Fortunately, advances in surgical procedures have allowed the surgeon to replace the stapes bone with a suitable prosthesis, re-instating relatively normal activity of the ossicles and greatly improving hearing ability.

Congenital malformation of the pinna or the ear canal, known as atresia, is an infrequent cause of conductive hearing loss. Often, when the pinna is malformed, there is no opening into the ear canal. In some cases, the

ear canal has failed to develop. Depending on the severity of the malformation, of either the pinna or the ear canal, surgery may be successful in restoring function. Other causes of conductive hearing loss include Eustachian tube malfunction, disruption of the ossicular chain (the three tiny bones in the middle ear), and swelling (edema) of the external ear canal.

A frequently occurring cause of conductive pathology is otitis media. Otitis media may refer to inflammation involving the middle-ear space or to a disorder in which the middle ear is filled with a watery fluid. In some cases, the fluid may harbor bacteria, creating significant medical problems if this condition is not treated early. Such middle-ear effusions are more common in children than in adults. If fluid is present in the middle ear, its mass will restrict the movement of the ossicles and create hearing impairment. Generally, patients with otitis media can be successfully treated through medical or surgical intervention.

It must be noted that conductive pathology does not affect the behavior of inner-ear structures; that is, the inner ear is capable of normal auditory performance. If the conductive pathology is eliminated by appropriate treatment, normal hearing will be restored. Severity of the condition may prevent the restoration of hearing,

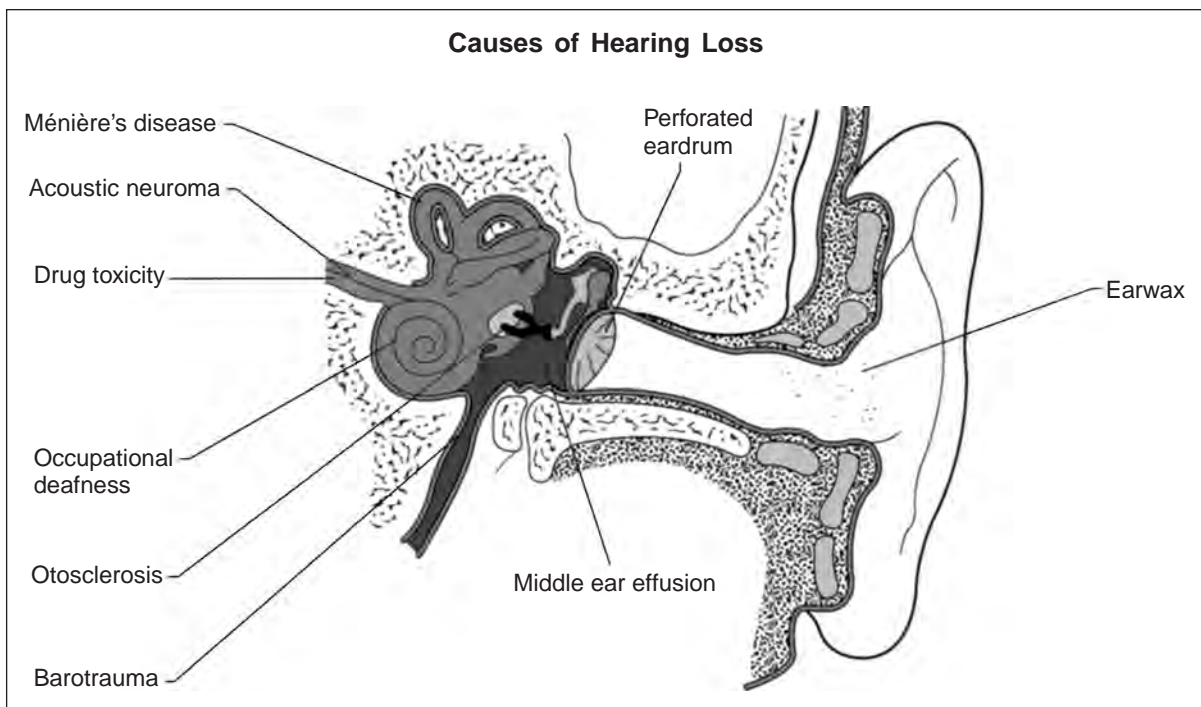
however, even if an aggressive treatment program is followed.

Sensorineural Hearing Loss. As a major classification of hearing loss, this term is somewhat misleading. Actually, there are two types of hearing loss within this classification. One is a sensory loss which involves the destruction of nerve cells (hair cells) in the cochlea. The other is a neural loss which involves neural cells in the ascending auditory pathway from the cochlea to the brain. It is possible to experience one type of loss without the other. Some examples of sensory loss include the following: loss of nerve cells resulting from traumatic injury to the cochlea, such as from whiplash, sharp blows to the head, or sudden, brief, and intense noises; loss of sensory cells from viral infections such as measles; loss of sensory cells caused by ototoxic drugs such as those in the mycin group (such as streptomycin or kanamycin); congenital problems associated with a lack of embryonic development; and exposure to loud and continuous (long-term) noise, a very common cause of hearing loss in adults. This last type of impairment is different from traumatic injury resulting from sudden, intense noise because it may take months or years for hearing loss caused by long-term noise to manifest itself. Research has also established a clear correlation between the normal aging process and sensory hearing impairment.

When sensory hearing impairment occurs, it is permanent. At the moment, there is no way of regenerating sensory tissue after the cell body has died. The only exception to this rule is found in those patients suffering from Ménière's disease. This disorder is often characterized by vertigo, dizziness, vomiting, and hearing loss. In the initial stages, however, the loss of sensitivity to sound is the result of changes in cellular physiology rather than of necrosis (death) of the nerve cells.

Examples of neural hearing loss are found among those hearing-impaired individuals with tumors, acoustic neuromas (benign tumors), cysts, and other anomalous conditions affecting the transmission of nerve impulses from the cochlea to the brain. Depending on the magnitude of the disorder, neural hearing loss has a much more devastating effect on speech understanding and signal processing than does sensory loss. As with sensory hearing impairment, neural hearing loss is a rather frequent occurrence associated with the aging process. For a sizable portion of those who experience hearing impairment, components of both sensory and neural loss are present. If the cause of the hearing deficit is entirely neural in nature, then the impairment is referred to as a retrocochlear loss.

For some types of neural pathology, medical or surgical intervention can be undertaken successfully. Acoustic neuromas are often removed after they have



been confirmed by audiologic, otologic, radiologic, and other diagnostic modalities. The size and location of the neuroma or tumorous growth will often dictate whether hearing can be preserved following surgery.

For those millions suffering from hearing impairment, it is the loss of speech discrimination ability that is of greatest concern. Thousands of studies have been undertaken to investigate the correlation between the magnitude, type, and length of hearing loss and the degree of speech recognition difficulty. One of the essential findings of these studies indicates that, in general, hearing loss is more pronounced for the high frequencies (above 1,000 hertz), whether the loss is caused by disease, drugs, noise, or the aging process. Another major finding is that one's ability to identify vowel and consonant information is frequency-dependent. Vowel identification is dependent on frequencies from about 200 hertz to 1,000 hertz, while consonant identification is dependent on frequencies above 1,000 hertz. A listener understands about 68 percent of speech sounds if nothing above 1,500 hertz is heard and about 68 percent of speech if nothing below 1,500 hertz is heard.

PERSPECTIVE AND PROSPECTS

Hearing loss is quite common and affects some twenty million Americans, ranging from infants to the elderly. The primary reason for preserving hearing is to maintain social adequacy in communication skills. Hearing conservation programs have been instituted by public and private schools, industry, military installations, construction organizations, and more recently, the U.S. government. In 1970, the Occupational Safety and Health Act (OSHA) was passed, making it mandatory for employers to provide safe work areas for workers exposed to noise levels exceeding government standards.

For the hearing impaired, understanding of speech is related to the degree of loss and the type of impairment. Because medical or surgical care cannot always ameliorate the loss, rehabilitation programs may take the form of speech or lipreading to improve communication skills. These rehabilitative programs constituted the treatment of choice until the introduction of wearable electric hearing aids.

Before the advent of electric hearing aids, however, the early ear trumpets were very effective for some in restoring speech recognition ability. Through ingenious design, some of the ear trumpets were "acoustically tuned" to provide more amplification in the higher frequencies than in the lower, but the volume of sound

was determined by the person talking—often a major problem for the hearing impaired.

With the development of the vacuum tube in the early part of the twentieth century, electric hearing aids became the treatment of choice when medical or surgical intervention was not indicated in resolving the hearing loss. It was possible not only to control the loudness of the hearing aid sound (volume) but also to shape the frequency response of the instrument to match the acoustic needs of the patient. In the second half of the twentieth century, there were significant advances in hearing aid technology. Transistor technology makes it possible to reduce the size of the hearing aid device without sacrificing performance. Computer science has also been used in the design of hearing aids. With digital technology, it is now possible to program electroacoustic characteristics into the hearing aid, which extends its utility.

Recently, major emphasis has been given to hearing conservation. Such continuing efforts have been instrumental in conserving the hearing of tens of thousands who might otherwise suffer from hearing losses sufficient to create problems in speech understanding. Although such programs are too late for millions of hearing-impaired individuals, advances in rehabilitative practices and the scientific application and use of hearing aids have provided them with a quality of life that was not possible only a generation ago.

—Robert Sandlin, Ph.D.

See also Aging; Audiology; Ear infections and disorders; Ear surgery; Ears; Hearing aids; Hearing tests; Ménière's disease; Nasopharyngeal disorders; Neuralgia, neuritis, and neuropathy; Otorhinolaryngology; Sense organs; Speech disorders.

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Ferrari, Mario. *Ear, Nose, and Throat Disorders*. New York: Elsevier, 2002. A clinical yet accessible reference text that provides a comprehensive list of disorders, with a summary of the condition, background, diagnosis, treatment, outcomes, prevention, and resources.

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- Hearing Exchange. <http://www.hearingexchange.com/>
An online community designed to foster the exchange of information and the provision of support for the hearing impaired.
- Katz, Jack, ed. *Handbook of Clinical Audiology*. 5th ed. Philadelphia: Lippincott Williams & Wilkins, 2002. Text that examines advances in the scientific, clinical, and philosophical understanding of audiology. Sections of the book cover behavioral tests, physiologic tests, special populations, and the management of hearing disorders.
- Pascoe, David. *Hearing Aids: Who Needs Them?* St. Louis: Big Bend Books, 1991. This easy-to-read text presents an abundance of data relative to hearing, hearing aid devices, and their use. Answers many questions that may arise concerning hearing aid use in direct and simple terms. One of the most significant aspects of this book is that it explains, in reasonable detail, how to use and evaluate hearing aids.
- Yost, William. *Fundamentals of Hearing*. 4th ed. San Diego, Calif.: Academic Press, 2000. This text describes, in easy-to-understand terms, the organ of hearing and its contribution to an individual's behavior. Simple auditory theory is examined, as is the nature of the ear's response to acoustic energy.

HEARING TESTS

PROCEDURE

ANATOMY OR SYSTEM AFFECTED: Ears, nervous system

SPECIALTIES AND RELATED FIELDS: Audiology, neurology, otorhinolaryngology, speech pathology

DEFINITION: Evaluation techniques for determining the type and severity of hearing loss in children.

KEY TERMS:

auditory brainstem response: measurement of the nervous discharge produced by the central auditory system as a response to sound stimulation; also known as brainstem auditory evoked response (BAER) or auditory brainstem potentials (ABR)

auditory nerve: the nerve that conducts sound stimuli to the brain for interpretation

behavioral audiometry: a technique that the audiologist employs to evaluate hearing in infants, toddlers, or uncooperative patients (both children and adults) with developmental deficits

cochlea: the organ localized in the inner portion of the auditory system that detects sound

mastoid: referring to the bone behind the ear

middle ear: the part of the auditory system, consisting of the ossicular chain and the auditory tube, that serves as a conductor of and transducer of sound

otoacoustic emissions: sound produced in the middle ear as a response to the vibration produced by the cochlea when it is stimulated by external sounds

INDICATIONS AND PROCEDURES

Hearing tests are done to establish the presence, type, and severity of hearing impairment in children and adults. Such tests are conducted by an audiologist, although screening tests can also be done by a technician under the supervision of an audiologist. The severity of hearing loss is classified as mild, moderate, moderately severe, severe, and profound. It is also classified according to the anatomic region affected: conductive, sensorineural, or mixed hearing loss.

The selection of tests to evaluate hearing will depend on the patient's age and ability to follow directions and the ability of the audiologist to elicit responses from the patient. When a patient cannot follow instructions such as lifting a hand or pressing a button, a test that does not require the patient's cooperation is used. Two tests that do not require the patient's cooperation are the auditory brainstem potential (ABR) test and the evoked otoacoustic emissions (EOAE) test. Both tests require only that the patient be quiet. For this purpose, the patient may need sedation if normal sleep cannot be induced.

The ABR test requires the placement of four electrodes in the child's head: in both mastoid regions and in the mid forehead and upper center of the head. A stimulus is sent through a small microphone placed in the patient's external ear canal or via headphones. The instrument records the average of the electrical discharges generated by the auditory nerve in response to sound stimuli and produces a tracing of waves that correspond to the different electrical potentials generated in response to the stimuli. Analysis of the waves can determine the presence of hearing loss and measure its severity. The ABR test may be used for screening, to determine whether the subject can hear, or for the clinical evaluation of hearing loss. It can be done at any age. An automated method of ABR testing is available for screening newborn infants for hearing loss; it automatically determines if the patient has passed or failed. The clinical ABR test requires specially trained personnel and takes from forty-five to fifty minutes to perform. The automated method can be applied by a technician.