

International Encyclopedia of Rehabilitation

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Hearing Impairment: Definitions, Assessment and Management

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Definitions

Hearing loss is considered to be the most prevalent congenital abnormality in newborns and is more than twice as prevalent as other conditions that are screened for at birth, such as sickle cell disease, hypothyroidism, phenylketonuria, and galactosaemia (Finitzo & Crumley, 1999). It is one of the most common sensory disorders and is the consequence of sensorineural and/or conductive malfunctions of the ear. The impairment may occur during or shortly after birth (congenital or early onset or may be late onset) caused post natal by genetically factors, trauma or disease. Hearing loss may be pre-lingual (i.e., occurring prior to speech and language acquisition) or post-lingual (i.e., occurring after the acquisition of speech and language).

Since hearing loss in infants is silent and hidden, great emphasis is placed on the importance of early detection, reliable diagnosis, and timely intervention (Spivak et al., 2000). Even children who have mild or unilateral permanent hearing loss may experience difficulties with speech understanding, especially in a noisy environment, as well as problems with educational and psycho-social development (Bess et al., 1988; Culbertson & Gilbert 1996). Children with hearing loss frequently experience speech-language deficits and exhibit lower academic achievement and poorer social-emotional development than their peers with normal hearing.

The period from birth to 3-5 years is often considered as the "critical period" for the development of normal speech and language. Normal hearing in the first six months of life is also considered critical for normal speech and language skills. Hence, early identification and appropriate intervention within the first six months of life have been demonstrated to prevent or reduce many of the adverse consequences and to facilitate language acquisition (Yowhinaga-Itano et al., 1998). Consequently, in developed countries with a high standard of health care, primary services include the early detection of congenital hearing loss and the initiation of auditory habilitation before six months of age.

The prevalence of congenital and early-onset hearing loss in most developed countries is estimated to range between 2-4 infants with moderate-severe hearing loss in every 1000 births. In contrast, only limited information is available on developing regions, including the Middle East especially in the Arab countries, where the prevalence is estimated to be markedly higher than in Israel or European and North American countries (Attias et al.,

2006). In developing countries, more than 10 infants in every 1000 births are estimated to be affected by a severe profound hearing loss. Of the 62 million deaf children younger than 15 years old worldwide, two-thirds reside in developing countries (Smith, 2003).

Presbycusis (age-related hearing loss) is the loss of hearing that gradually occurs in most individuals as they grow older. Hearing loss is a common disorder associated with aging and is ranked as the third most prevalent chronic condition in elderly people after hypertension and arthritis. Its prevalence and severity increase with age, rising from about 30-35 percent of adults aged 65 and older to an estimated 40-50 percent of adults aged 75 and older (Cruikshanks et al., 1998). The loss associated with presbycusis is usually greater for high-pitched sounds. For example, it may be difficult for someone to hear the nearby chirping of a bird or the ringing of a telephone, and it is most difficult to understand speech in a noisy background. However, the same person may be able to clearly hear the low-pitched sound of a truck rumbling down the street. Presbycusis most often occurs in both ears, affecting them equally. Because the process of loss is gradual, people who have presbycusis may not realize that their hearing is diminishing.

Schuknecht (1974) has described four types of human presbycusis: (1) sensory, mainly affecting the cochlear hair cells and supporting cells; (2) neural, typified by the loss of afferent neurons in the cochlea; (3) metabolic, where the lateral wall and stria vascularis of the cochlea atrophy; and (4) mechanical, where there seemed to be a so-called “stiffening” of the basilar membrane and organ of Corti. There are many causes of presbycusis, though it is most commonly the result of changes in the inner ear as a person ages. It can also stem from changes in the middle ear or from complex changes along the nerve pathways leading to the brain.

The negative impact of hearing loss on older adults is significant (LaForge et al., 1992). Hearing loss is associated with depression, social isolation, poor self-esteem, and functional disability (Mulrow et al., 1990a), particularly for those suffering from hearing impairment who have not yet been evaluated or treated for hearing loss.

Hearing impairment is a broad term that refers to hearing losses of varying degrees, ranging from hard-of-hearing to total deafness. As the general population continues to age, the prevalence of hearing impairment can be expected to increase. Since the use of hearing aids or surgical intervention to improve hearing loss has been shown to have a positive impact on quality of life (Mulrow et al., 1990b, 1992; Weinstein, 1991), more screening programs for elderly adults should be established, followed by appropriate referral to audiologists based on individual needs. Without early diagnosis and treatment of hearing impairment, quality of life and functional status are likely to decline in the aging population.

The major challenge facing people with hearing impairment is communication. Hearing-impaired persons vary widely in their communication skills. Among the conditions that affect the development of communication skills by persons with hearing impairments are personality, intelligence, nature and degree of deafness, degree and type of residual hearing, degree of benefit derived from amplification by hearing aid, family environment,

and age of onset. Age of onset plays a crucial role in the development of language. Persons with pre-lingual hearing loss (present at birth or occurring before the acquisition of language and the development of speech patterns) are more functionally disabled than those who lose some degree of hearing after the acquisition of language and speech.

When describing hearing impairment, three attributes are considered:

1. Type of hearing loss (part of the hearing mechanism that is affected).
2. Degree of hearing loss (range and volume of sounds that are not heard).
3. Configuration (range of pitches or frequencies at which the loss has occurred).

Types of hearing loss

A hearing loss can be classified as a conductive, sensory, neural, or mixed hearing loss, based on the anatomic location of the problem (site of lesion, i.e., middle or inner ear). A hearing loss may also be labeled as unilateral or bilateral, depending on whether the loss is in one (unilateral) or both (bilateral) ears. The degree of loss might be the same in both ears (symmetrical hearing loss) or it could be different for each ear (asymmetrical hearing loss).

Conductive hearing loss is characterized by an obstruction to air conduction that prevents the proper transmission of sound waves through the external auditory canal and/or the middle ear. It is marked by an almost equal loss of all frequencies. The auricle (pinna), external acoustic canal, tympanic membrane, or bones of the middle ear may be dysfunctional. Conductive hearing loss may be congenital or caused by trauma, severe otitis media, otosclerosis, neoplasms, or atresia of the ear canal. Some conductive hearing loss can be treated surgically with tympanoplasty or stapedectomy, and the use of hearing aids and assistive listening devices may also be beneficial.

Sensorineural hearing loss occurs when the sensory receptors of the inner ear are dysfunctional. Sensorineural deafness is a lack of sound perception caused by a defect in the cochlea and/or the auditory division of the vestibulocochlear nerve. This type of hearing loss is more common than conductive hearing loss and is typically irreversible. It tends to be unevenly distributed, with greater loss at higher frequencies. Sensorineural hearing loss may result from congenital malformation of the inner ear, intense noise, trauma, viral infections, ototoxic drugs (e.g., cisplatin, salicylates, loop diuretics), fractures of the temporal bone, meningitis, ménière's disease, cochlear otosclerosis, aging (i.e., presbycusis), or genetic predisposition, either alone or in combination with environmental factors. Many patients with sensorineural hearing loss can be habilitated or rehabilitated with the use of hearing aids. Patients with profound bilateral sensorineural hearing loss (e.g., at least 90 dB) who derive no benefit from conventional hearing aids may be appropriate candidates for the cochlear implant device, which bypasses the damaged structures of the cochlea and stimulates the function of the auditory nerve. Auditory brainstem implants, which are similar to multichannel cochlear implants, are used in patients with neurofibromatosis type 2 following vestibular schwannoma removal, especially those individuals who have lost integrity of the auditory nerves.

Auditory Neuropathy (AN) is a type of sensorineural hearing loss that can be congenital or acquired. Unlike other types of sensorineural hearing loss where both Otoacoustic Emissions (OAE) and Auditory Brainstem Response (ABR) tests are likely to be abnormal, Auditory Neuropathy is characterized by normal OAE results and significantly abnormal ABR responses, even when measured with very loud sounds. The combination of normal OAE responses and severely impaired ABR responses is thought to reflect normal outer hair cell (OHC) function in the cochlea and abnormal auditory nerve function. The site of lesion for AN is often unknown, but possibilities include cochlear inner hair cells, cochlear spiral ganglia, synapse and/or eighth nerve fiber disorders. Audiograms of children with AN vary from hearing in the normal range with complaints of difficulty hearing in background noise to profound hearing loss.

Individuals with **mixed hearing loss** have both conductive and sensory dysfunction. Mixed hearing loss is due to disorders that can affect the middle and inner ear simultaneously, such as otosclerosis involving the ossicles and the cochlea, head trauma, middle ear tumors, and some inner ear malformations. Trauma resulting in temporal bone fractures may be associated with conductive, sensorineural, and mixed hearing loss.

Auditory Processing Disorder (APD) is a deficit in neural processing of auditory stimuli that is not due to higher order language, cognitive, or related factors. However, APD may lead to or be associated with difficulties in higher order language, learning, and communication functions. This type of auditory problem affects more complex auditory processes, such as understanding speech when there is background noise. The results of hearing sensitivity and physiological tests, such as otoacoustic emissions (OAE) and auditory brainstem response (ABR) are normal in children with a central auditory disorder.

A great many, including what exactly constitutes an APD, with some professionals being still unconvinced that it exists as a separate clinical entity, poor understanding of the boundaries and overlap between APD and language or other developmental disorders, and lack of uniform accepted guidelines regarding testing and management of APD.

Degree of hearing loss

- **Deaf/Deafness** refers to a person who has a profound hearing loss and uses sign language.
- **Hard of hearing** refers to a person with a hearing loss who relies on residual hearing to communicate through speaking and lip-reading.
- **Hearing impaired** is a general term used to describe any deviation from normal hearing, whether permanent or fluctuating, and ranging from mild hearing loss to profound deafness.
- **Residual hearing** refers to the hearing that remains after a person has experienced a hearing loss. It is suggested that greater the hearing loss, the lesser the residual hearing.

The level of severity of hearing loss, as used in this guideline, is defined as follows:

-10 to 15 dB HL	Normal Hearing
16-25 dB HL	Slight Hearing Loss
26-40 dB HL	Mild Hearing Loss
41-55 dB HL	Moderate Hearing Loss
56-70 dB HL	Moderate-Severe Hearing Loss
71-90 dB HL	Severe Hearing Loss
>90 dB HL	Profound Hearing Loss

(Average threshold level **re** for 0.5, 1 and 2 kHz, Clark (1981))

Configuration of hearing loss

There are four general configurations of hearing loss:

1. **Flat:** thresholds essentially equal across test frequencies.
2. **Sloping:** lower (better) thresholds in low-frequency regions and higher (poorer) thresholds in high-frequency regions.
3. **Rising:** higher (poorer) thresholds in low-frequency regions and lower (better) thresholds in higher-frequency regions.
4. **Trough-shaped** (“cookie-bite” or “U” shaped): greatest hearing loss in the mid-frequency range, with lower (better) thresholds in low- and high-frequency regions.

Assessment

Hearing loss is confirmed using a battery of audiologic tests, with the specific tests and measures selected according to the age of the patient. However, in general, a comprehensive hearing assessment designed to confirm hearing loss usually includes a hearing history, physiological procedures, and behavioral procedures (see Table 1).

Table 1

Components of a Comprehensive Hearing Assessment and Hearing History

- General concern about hearing and communication
- Auditory behaviors (reacting to and recognizing sounds)
- History of otological diseases and other risk factors for hearing loss

Physiological procedures or acoustic admittance measurements

Otoacoustic emissions (OAE)

OAE are low-level sounds produced by the sensory hair cells of the cochlea (primarily the outer hair cells of the inner ear) as part of the normal hearing process. Hair cells that are normally functioning emit acoustic energy, which can be recorded by placing a small probe (containing a microphone) attached to a soft ear tip in the external ear canal

opening. The earphone delivers test signals into the ear canal that evoke an acoustic response from the hair cells, and the responses are recorded by a second microphone in the probe. These responses are called evoked otoacoustic emissions (EOAE).

Auditory brainstem response (ABR)

Using clicks or tones, this test can estimate hearing threshold sensitivity and determine the integrity of the auditory pathway from the cochlea to the level of the brainstem. Small disc electrodes are pasted on the scalp, and repetitive stimuli are delivered by an earphone. The auditory potentials (electrical/neural activity generated by the auditory nerve and brainstem) that are evoked by the repetitive stimuli are then recorded by a computer.

Middle ear muscle reflexes

Involuntary middle-ear muscle reflexes to sounds, usually elicited by moderately loud tones or noises, are recorded.

Tympanometry

This procedure is used to assess the function of the middle ear by placing a small probe attached to a soft, plastic ear tip at the ear canal opening and varying the air pressure released into the ear canal. Tympanometry is not a hearing test.

Behavioral audiometry testing

The goal of behavioral audiometric testing is to obtain a valid measure of hearing threshold sensitivity for each ear in the speech-frequency range, ideally from 250 through 8,000 Hz. Results of the audiometric assessment are displayed on an audiogram. Behavioral audiometric tests are used to:

- Determine whether or not a patient has a hearing loss.
- Determine the degree, configuration, and type of hearing loss if hearing loss does exist.
- Monitor the patient's hearing over time.
- Provide information for the fitting of hearing aids or other sensory devices.
- Help determine the functional benefit of hearing aids or other sensory devices.

For children, behavioral audiometric test methods are selected to be appropriate for the developmental age of the infant or young child. The tests can be divided into two general categories: unconditioned and conditioned behavioral response procedures.

Behavioral Observation Audiometry (BOA) is an unconditioned behavioral response procedure in which an observation of general awareness of sound (e.g., mother's voice, environmental sounds and music) is used to determine a general level of auditory responsiveness or function.

Visual Reinforcement Audiometry (VRA) is a conditioned behavioral response procedure used to determine threshold sensitivity in infants beginning at approximately six months of age (i.e., developmental age). A head-turn response to the presentation of

an audiometric test stimulus is rewarded by the illumination and activation of an attractive animated toy.

Conditioned Play Audiometry (CPA) is a conditioned behavioral response procedure used to determine threshold sensitivity in young children beginning at approximately two years of age (i.e., developmental age). A play response (block-drop, ring stack) to the presentation of an audiometric test stimulus is rewarded, usually by giving the child social praise.

Speech Audiometry is used to assess the ability to detect, discriminate, identify, and comprehend speech. Several test procedures are used for speech audiometry in infants and young children, including speech sounds (syllables), words, phrases, and sentences. The tests can be conducted by using earphones in each ear or through a loudspeaker. In infants, the conditioned head-turn response can be used to estimate speech detection thresholds for words or individual syllables. In young children, speech identification ability is determined at a listening level that is comfortable for the child, well above threshold. Usually, young children are asked to identify body parts (e.g., “Where’s your nose?”) or familiar objects (e.g., “ball,” “spoon”) by pointing to or picking up the object. Older children may be asked to repeat the stimulus words or point to pictures in order to determine their speech identification ability. The final speech identification score (percent of words or simple sentences identified correctly) is sometimes referred to as a “speech discrimination score” or “word recognition score”. The limitation of speech audiometry is that tests of speech identification require that the stimulus items be within the child’s receptive vocabulary. Speech audiometry test results can be useful in determining intervention goals, in monitoring auditory skill development, and in examining the functional benefit of hearing aids or other assistive technology.

Management

Interventions for most infants and young children with hearing loss are primarily focused on the following goals:

- Preventing or reducing the communication problems that typically accompany early hearing loss.
- Improving the child’s ability to hear.
- Facilitating family support and confidence in parenting a child with a hearing loss.

Interventions focused on developing a child’s communication skills and abilities differ according to the type of communication approach that will be used by the child and family. Communication approach options for young children with hearing loss range from sign language alone to auditory/verbal (spoken language) or various combination approaches. Often parents must make an initial decision about a communication approach soon after their child has been diagnosed with a hearing loss.

Parents also must choose a means for improving their child’s access to sound. The assistive devices most commonly used to amplify sound are hearing aids. Other assistive

devices include FM systems and tactile aids. Some children with severe to profound hearing loss who have demonstrated little benefit from conventional hearing aids may receive a cochlear implant, an electronic device that is surgically placed in the inner ear.

An aural rehabilitation consultation is considered medically necessary as part of the hearing impairment evaluation. Aural rehabilitation is considered medically necessary for the treatment of a hearing impairment when ALL of the following criteria are met:

- The hearing impairment is the result of trauma, tumor, or disease or following implantation of a cochlear or auditory brainstem device.
- An evaluation that includes standardized speech and/or hearing tests has been completed by a certified speech-language pathologist or licensed audiologist.
- The treatment being recommended has the support of the treating physician.
- The therapy being ordered requires one-to-one intervention and supervision by a speech-language pathologist or audiologist.
- The therapy plan includes specific tests and measures that will be used to document significant progress.
- Meaningful improvement is expected from the therapy.
- The treatment includes a discharge plan for the transition from one-to-one supervision to maintenance provided by an individual or caregiver.

Aural rehabilitation refers to the services and procedures needed to facilitate adequate receptive and expressive communication in individuals with hearing impairments [American Speech-Language-Hearing Association (ASHA), 1984]. It is also called auditory or audiologic rehabilitation. Aural rehabilitation is typically an integral component used in the overall management of individuals with hearing loss and is often an interdisciplinary endeavor involving physicians, audiologists, and speech-language pathologists. For school-age children, therapy may also be coordinated with the school system. In general, services may be initiated as soon as a patient has been identified as having a hearing impairment or following the fitting of a hearing device or implantation of a cochlear device.

Services involved in the provision of aural rehabilitation include

- Identification and evaluation of sensory capabilities, including the extent of impairment and the fitting of auditory aids.
- Interpretation of the audiological findings, plus counseling and referral.
- Development and provision of an intervention program for communicative disorders in order to facilitate expressive and receptive communication.
- Re-evaluation of the patient's status.
- Evaluation and modification of the intervention program.

Aural rehabilitation should be structured, systematic, individualized, and goal-directed (i.e., both long- and short-term goals). Although aural rehabilitation programs are accepted and widely used in the management of hearing-impaired individuals, the role of aural rehabilitation in overall treatment and its impact on health outcomes has not been

clearly delineated, other than for patients with cochlear device implants. For patients who acquire a hearing loss post-lingually, treatment would be considered rehabilitative and restorative in nature. Although the term “rehabilitation” is commonly used in association with services provided to pre-lingually hearing-impaired patients, treatment would more accurately be described as “habilitation” in nature, as it does not involve restoring a lost function.

Audiologists and speech-language pathologists certified by ASHA (2006) are qualified to provide aural rehabilitation components. The audiologist is typically responsible for the fitting, dispensing, and management of a hearing device, for providing counseling about hearing loss and ways to enhance communication skills, and for providing training on environmental modifications that will facilitate the development of receptive and expressive communication. The speech-language pathologist is typically responsible for evaluating receptive and expressive communication skills and for providing services to improve them, as well as for providing training and treatment in communication strategies (e.g., assertive listening tactics), speech-perception training (e.g., speech-reading, auditory training, and auditory-visual-speech-perception training), speech and voice production, and comprehension of oral, written, and sign language.

Initially, aural rehabilitation records should be provided to substantiate the need for this therapy. Records should include clinical narrative notes from the attending physician or referring provider, with a description of expressive and/or receptive speech impairments and reports of standardized speech and hearing tests, if applicable. An evaluation and treatment plan, including assessment of function level, measurable long- and short-term goals, progress toward achieving goals, anticipated timeframe for achieving goals, and anticipated frequency and duration of therapy, should also be provided. For continued treatment, it is necessary to obtain follow-up evaluations, auditory therapy notes, documentation of progress toward goals, and treatment plan revisions.

Both group therapy and computer-based training are often used in aural rehabilitation; however, these methods are not individualized to specific patient needs. The primary focus of many aural rehabilitation components and interventions may be training (e.g., speech-reading training, vocational training), held in either group or individual sessions. Group-based therapy, computer-based learning programs, and school-based programs are considered as training in nature and not medically necessary, as they do not involve the formal interaction of one-to-one supervision with a speech-language pathologist or audiologist and are not tailored to meet individual needs. Furthermore, maintenance programs of routine, repetitive drills and exercises that do not require the skills of a speech-language therapist or audiologist and that can be reinforced by the individual or caregiver are also considered to be outside the realm of medical prescription.

Computer-assisted programs used in aural rehabilitation include, but are not limited to:

- Computer-Assisted Speech Perception Testing and Training at the Sentence Level (CASPERSent)

http://www-rohan.sdsu.edu/~aboothero/files/CASPERSENT/CasperSent_preprint.pdf

- The Computer-Assisted Speech Training (CAST) software system, which was developed to help accelerate the auditory rehabilitation process of patients with cochlear implants.
http://www.tigerspeech.com/tst_cast.html
- Sound and WAY Beyond™ (Cochlear Americas, Centennial, CO).
<http://www.cochlearamericas.com/PDFs/soundandwaybeyond.pdf>
- Listening & Communication Enhancement (LACE)™ (Neurotone, Inc., Redwood City, CA).
<http://www.neurotone.com/>

Aural rehabilitation following cochlear device and auditory brainstem implantation

Aural rehabilitation following implantation of cochlear and auditory brainstem devices is considered an integral part of the overall management of implant patients. Although programs vary widely, both with regard to treating disciplines and to the duration and scope of treatment, the general consensus is that some type of post-implantation aural therapy maximizes the benefit of the device. Sound recognition and speech intelligibility are evaluated prior to and just after implantation. Hearing capabilities are assessed by an audiologist, both with and without the assistance of a hearing aid. A speech-language pathologist evaluates and categorizes the patient's pre-implantation speech and language skills. Post-cochlear implantation rehabilitation programs generally include the following components: sound awareness (e.g., recognition of novel auditory signals); visual/auditory processing, including speech-reading training (e.g., lip-reading, facial expression, gestures, and body language); speech recognition; mechanical training (e.g., use of the device and telephone); and voice, speech production, and language therapy.

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