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Hearing Impairment

98

Peter Chung, Shibani Kanungo, and Dilip R. Patel

Abstract

Hearing impairment or loss may occur in isolation or in association with other developmental disorders and can be due to numerous causes. The screening and early diagnosis of hearing impairment represents an important public health concern as early intervention is associated with better long-term outcomes. With advances in technology, interventions for hearing impairment are becoming more sophisticated; however, available treatments also have to contend with definitions of health and cultural sensitivity. Hearing impairment is a chronic condition with multi-dimensional characteristics and its management requires a well-coordinated interdisciplinary approach.

Introduction

Hearing impairment or loss is the partial or total inability to hear sounds experienced in the environment. According to the World Health Organization (WHO), there are approximately 360 million people worldwide with hearing impairment [1]. The screening, diagnosis and management of hearing impairment is complex with multiple stakeholders. Hearing impairment as a diagnosis encompasses a wide spectrum of presentations and disease processes, has a vital impact on human development, and is a major public health concern. It is a battleground for cultural identity and conception of health and a growing research field rich with ongoing questions.

In this chapter, the terms "hearing loss" and "hearing impairment" are used interchangeably. Following the convention in the educational literature, the term "deaf and hard-of-hearing"

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(DHH) is also utilized to describe this population. Finally, in accordance with the field of Deaf studies, the term "Deaf" will be used when discussing Deaf culture and aspects of the Deaf community.

Physiology and Pathophysiology

The structure of the human ear is shown in Fig. 98.1 and is divided into outer, middle, and inner components.

The external ear canal is connected to the tympanic membrane, behind which is a series of small bones (ossicles) that connect to the cochlea. Within the cochlea is the organ of Corti, which encapsulates a series of hair cells that connect to the auditory nerve. Sound is transmitted as vibrations in the air that are then channeled by the external ear structures (e.g. the pinna) into the canal, causing movement of the tympanic membrane. This motion, transmitted through the linked ossicles, results in shifts in the fluid of the cochlea that stimulate the hair cells in the organ of Corti. The number and pattern of hair cell activation transmit information regarding amplitude (volume) and frequency (pitch). Signals are transmitted through the spiral ganglion and relay nuclei in the pons and midbrain and are eventually processed in the auditory cortex in the temporal lobe. The temporal lobe also connects to the limbic system for processing of emotion and the hippocampus for processing of learning and memory.

Hearing Loss and Impairment

Hearing loss or impairment can be described objectively by use of an audiogram (see Figs. 98.2, 98.3, and 98.4), which displays information regarding the frequency, severity, and type of hearing loss. An audiogram displays hearing frequencies on the x-axis and the severity as decibels (dB) on the y-axis for each ear.

Frequency: An individual may have normal hearing for some frequencies with significant impairment in others. Although the human ear can detect frequencies ranging from 20 Hz (low pitch) to 20,000 Hz (high pitch), conventional audiology typically focuses on frequencies from 250 to 8000 Hz, which is the acoustic range of normal human speech.

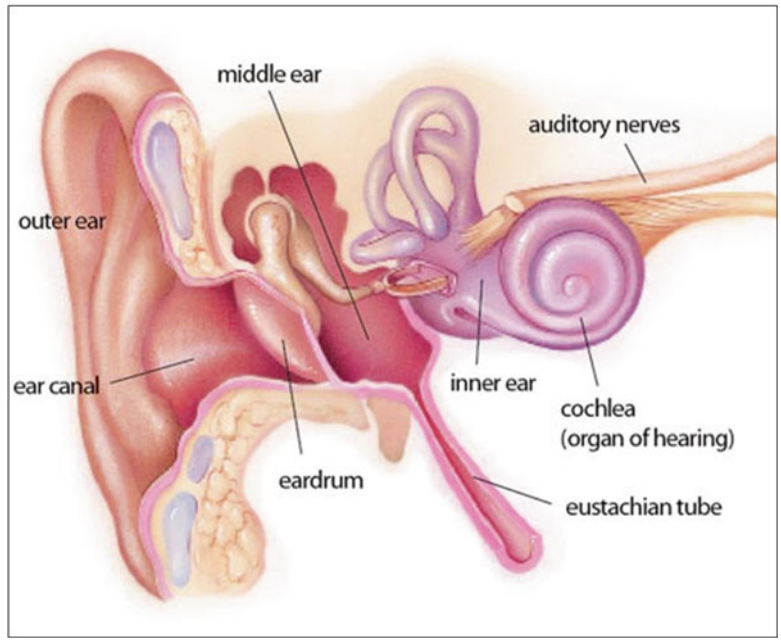

Fig. 98.1 Anatomy of the human ear

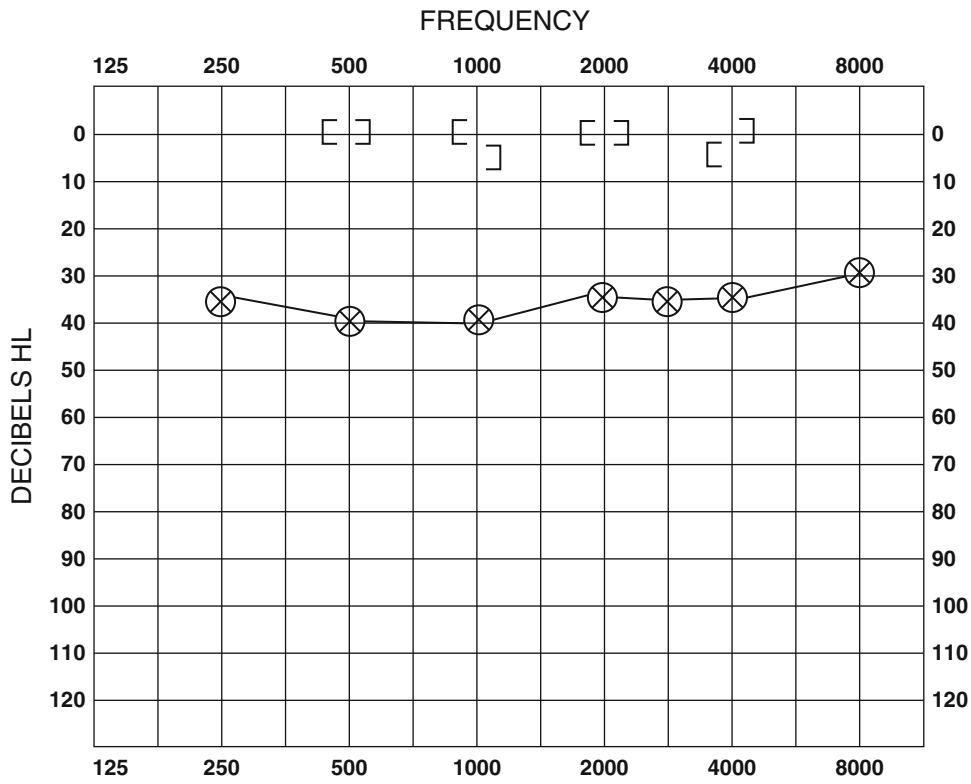

Fig. 98.2 Bilateral mild conductive hearing loss

Severity: The lowest threshold at which an individual can hear a sound expressed as decibels (dB). The decibel is a logarithmic unit describing intensity of sound, with each 10 dB increase signifying a tenfold increase in the power of sound.

The degree of hearing impairment is typically described as the "three frequency pure tone average," which is the mean threshold for 500, 1000, and 2000 Hz. Hearing impairment severity is divided into mild (21–40 dB), moderate (41–55 dB), moderately severe (56–70 dB), severe (71–90 dB), and profound (90+dB). See Table 98.1 for the impact of hearing impairment based on severity.

Type Disturbances in the external ear canal, the tympanic membrane, or the structures of the middle ear result in conductive hearing impairment. Problems with the cochlea, hair cells, auditory nerve, and higher processes result in sensorineural hearing impairment. Mixed hearing impairment consists of both conductive and sensorineural components. The different types of hearing impairment can be documented on the audiogram (see Figs. 98.2, 98.3, and 98.4).

- Air conduction thresholds are obtained through the use of earphone testing. They are indicated by "O" for the right ear and "X" for the left ear and generally demonstrate conductive hearing ability. The audiogram in Fig. 98.2 demonstrates mild bilateral conductive hearing loss.
- Bone conduction thresholds are obtained through the use of an oscillator which induces the tones directly into the skull via the mastoid bone, bypassing the conductive structures of the middle ear. These thresholds are indicated by "<" for the left ear and ">" for the right ear. In general, they indicate sensorineural hearing ability. The audiogram in Fig. 98.3 demon-

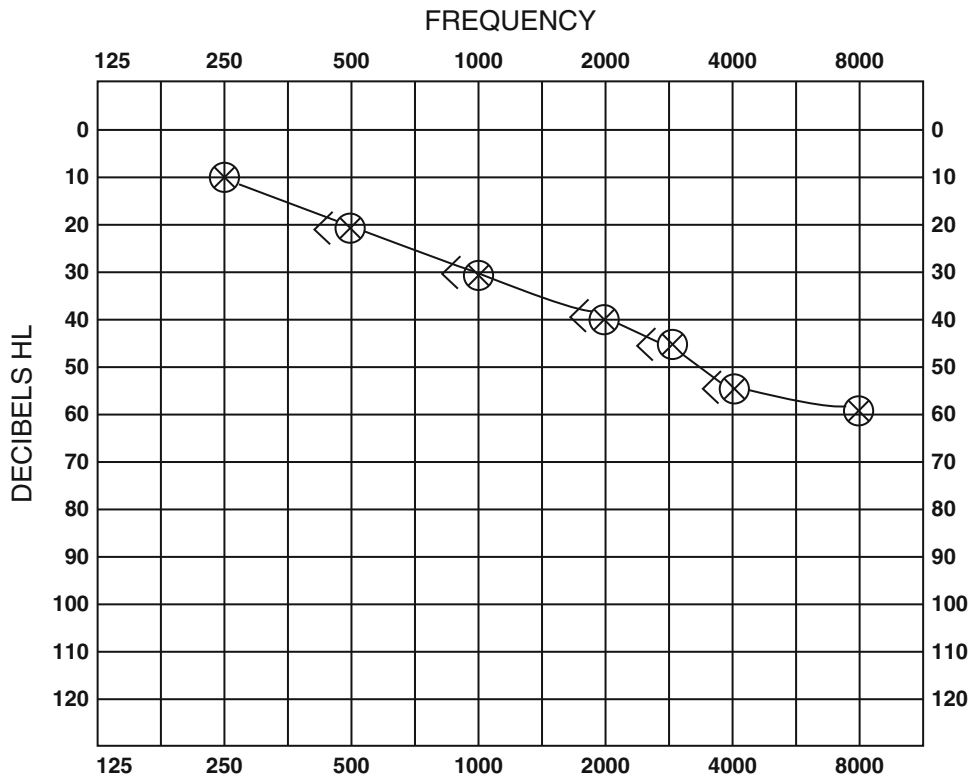

Fig. 98.3 Bilateral mild-to-moderate sensorineural hearing loss

strates mild-to-moderate sensorineural hearing loss in both ears.

- The air-bone gap is the amount of hearing impairment between the air and bone conduction thresholds. An individual with pure sensorineural hearing impairment should demonstrate relatively equivalent bone and air conduction. A gap greater than 10 dB is suggestive of mixed hearing impairment, as is seen in Fig. 98.4.
- Sound field thresholds are recorded when an individual is not able to tolerate testing of individual ears. Thresholds are recorded as "S" and only indicate the hearing ability of the better ear.
- Speech reception threshold is the lowest intensity at which the individual can repeat familiar two-syllables words with 50 % accuracy. It is usually within 5 dB of the three frequency pure tone average level.

Development of Hearing

The formation of hearing structures begins early in embryologic development and is refined throughout early childhood, which has important implications for hearing impairment. The anatomic structures in the middle ear are developed by 15 weeks gestation and are functional by 20 weeks. However, the auditory system is not functional until the hair cells of the cochlea become connected to the brain stem at around 25–29 weeks. Auditory pathways to the temporal lobe are functional around 28–30 weeks. By 27 weeks, most fetuses will respond to vibroacoustic stimuli; as they develop in an aquatic environment, their hearing is thought to be due to head vibration and not transmission through the conductive systems of the middle ear. Research suggests that noise experienced by the fetus in-

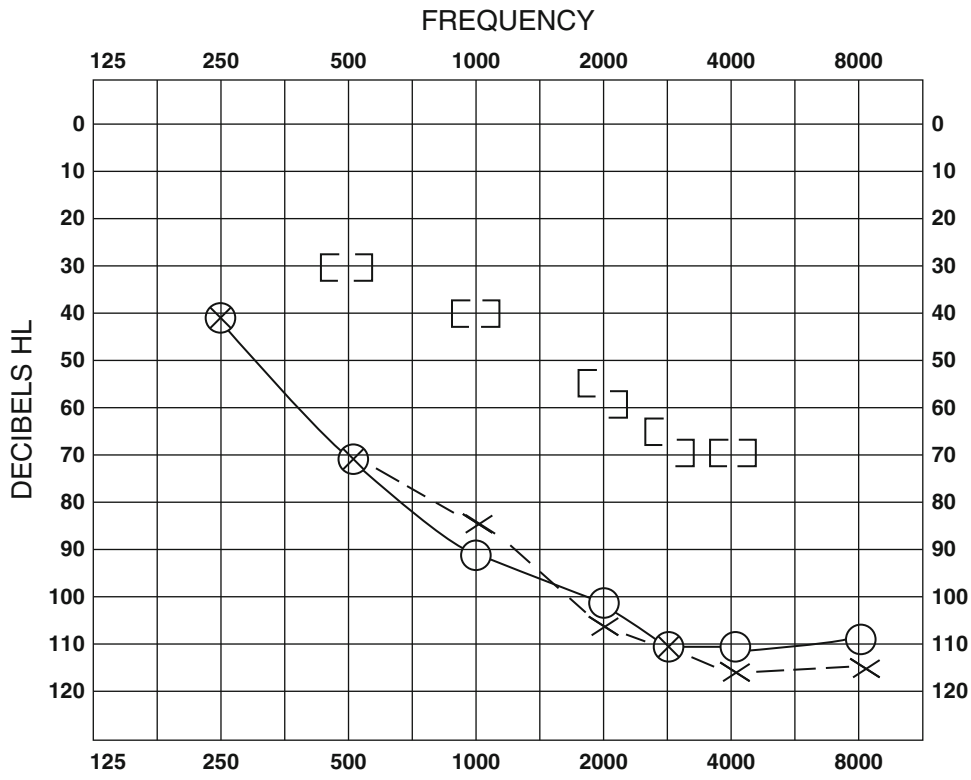

Fig. 98.4 bilateral moderate-to-profound mixed hearing impairment

utero consists mostly of sound from the mother's gastrointestinal, cardiovascular, and respiratory systems as well as her body movements and speech [2].

Auditory structures that are created in-utero continue to be refined by environmental influences. For example, hair cells and ganglion cell nerve fibers are tuned for specific frequencies between 28 weeks and the first few months of life, and loud ambient noise (e.g. in the NICU) can interfere with this process [3]. Although the cochlea appears to be functionally mature by 35 weeks, additional developments on the cellular level can affect hair cell motility beyond that time period [4]. Likewise, although the synapses of the auditory cortex are completed by term, synaptogenesis and myelination continue in early childhood in response to environmental noise. Finally, sounds that are experienced in the developing brain not only enhance connections within the memory circuits of the auditory cortex but also shape connections to the limbic system, facilitating the association of emotions with memory [3]. The influence of environmental sound on the developing brain is an important consideration for noise control in the care of the preterm infant [5].

Sarah is a 4 month old girl who failed her newborn hearing screen and was subsequently diagnosed with bilateral profound hearing loss. Her father brings her to the follow-up visit you had scheduled to make sure they followed through with their audiology evaluation and to discuss the results of the evaluation. He is noticeably upset at the diagnosis and asks, "How could this happen? We don't have deaf people in our family. My wife took all the vitamins she was supposed to."

Epidemiology

The prevalence of newborn hearing loss in the United States is reported to be 1.1–1.2 per 1000 infants screened [6]. The prevalence of perma-

Table 98.1 Impact of various degrees of hearing loss (From Neault [70], Table 17.1–17.2)

Degree of loss in better ear	Effect of hearing speech	Impact on language, education, and vocation
Mild (21–40 dBHL)	The individual can understand speech one-to-one at 3 ft if language development is already established. He or she hears parts of utterance at greater distance. The person misses or mishears word endings. He or she benefits from hearing aids	The condition causes mild delay in language development. The individual benefits from acoustical treatments of the classroom. He or she attends inclusive educational placement if hearing loss is the only issue
Moderate (41–55 dBHL)	The person can understand speech one-to-one at 2–3 ft only with clear delivery and with lip-reading. He or she may not hear 75 % of speech sounds without hearing aids. The person benefits from hearing aids	The condition causes delayed language development and speech articulation errors if present in early childhood. The individual needs educational supports to learn in inclusive classrooms. On the job with hearing aids, the individual needs to look at the talker's face when listening. He or she may use an amplified telephone
Moderately severe (56–70 dBHL)	The individual understands only loud speech close to the ear and catches occasional loud words in an utterance. He or she benefits from hearing aids	Significant language delay is expected if the condition if present in childhood and not remediated beginning in early infancy. Educational placement depends on spoken language skills. An FM educational amplification system is needed to hear the teacher. On the job, the individual needs read back/feedback of instructions and modified communication strategies to hear in a group
Severe (71–90 dBHL)	The individual may hear a loud voice close to the ear. Without hearing aids, he or she does not detect conversational speech. The person needs early detection, hearing aids, and aggressive therapy to avoid significant language delay. He or she has difficulty monitoring the loudness and clarity of his or her own voice. Cochlear implantation is an option if speech recognition with hearing aids is poor despite training	Delay in spoken language development is expected if the condition is not remediated beginning in early infancy. The individual needs small specialized class placement unless his or her language skills are robustly normal. Many individuals develop both spoken language and sign language. They should have friends and mentors with hearing loss. On the job, the supervisor and co-workers should be taught how to communicate successfully. The person many be able to use an amplified telephone with a telephone switch on the hearing aid

(continued)

Table 98.1 (continued)

Degree of loss in better ear	Effect of hearing speech	Impact on language, education, and vocation
Profound (>90 dBHL)	The person is aware of a few loud environmental sounds. He or she does not develop speech without early use of hearing aids and intensive therapy. The individual may rely on sign language to communicate and is unlikely to understand words through hearing aids without visual cues. Cochlear implantation is an option if is psychoeducationally appropriate for the person	The individual typically needs a small self-contained class for deaf children unless cochlear implants in infancy resulted in rapid spoken language development, in which case an inclusive placement with support services may succeed. The individual needs deaf friends and mentors. With hearing aids, the person is unlikely to be able to use the telephone; he or she needs a TTY (tele-typewriter) for phone use. E-mail is preferred to telephone use

Table 98.2 Prevalence of newborn hearing loss

Country	Prevalence
United States	1.2 per 1000
United Kingdom	1.8 per 1000
Switzerland	1.2 per 1000
Ireland	1.3 per 1000
Israel	1.3 per 1000
Belgium	1.5 per 1000
Turkey	2.2 per 1000
Brazil	0.96 per 1000
Brazil	2 per 1000
UAE	2.6 per 1000
India	8 per 1000

nent SNHL increases from 2.7 per 1000 children under the age of 5 years to 3.5 per 1000 adolescents [7]. Prevalence of hearing loss in different countries is shown in Table 98.2.

The etiology of hearing impairment or loss depends on the age at presentation (congenital versus acquired) and the type of impairment (conductive, sensorineural, or mixed). Various causes of hearing loss are listed in Table 98.3.

Audiologic Screening and Diagnostic Testing

The early interactions between parent and child are critical in the development of speech perception and language outcomes [8]. Therefore, early detection of hearing impairment and provision of appropriate intervention strategies is associated with improved language acquisition and decreased burden of disease [9, 10]. Newborn hearing screens typically consist of one test or two tests in sequence. Evoked otoacoustic emissions (OAE) measure the cochlea's response to generated clicks or tones and can be used to screen for moderate hearing loss. The auditory brainstem response (ABR) measures the neural response to a range of frequencies but requires minimal motion to produce accurate results. In one study, the combination of these tests demonstrated an estimated sensitivity of 100 %, specificity at 99.7 %, and a positive predictive value of 83 % [11].

Jenny is a 2 year old girl who presents for her 24 month well child check. A developmental screening test demonstrates isolated language delay, although she is otherwise doing well. Upon reviewing her chart, you note that she was a full term infant who had an uncomplicated pre-natal and immediate post-natal course, including a passed newborn hearing screen. Although her parents are concerned about her language, they question the referral to an audiology evaluation. "She passed her newborn test, didn't she?" the mother asks. "And we think she can hear things perfectly fine."

Table 98.3 Causes of hearing loss

Type of hearing loss	Selected conditions and comments
Congenital conductive hearing loss	Malformations of the outer and middle ear apparatuses, such as is seen in craniofacial syndromes like hemifacial microsomia and Treacher-Collins syndrome. Tympanic membrane abnormalities or ossicular malformations
Acquired conductive hearing loss	Acute otitis media, otitis media with effusions, cerumen impaction, foreign bodies, trauma, cholesteotomas, and tumors. Down syndrome – may have conductive, sensorineural or mixed hearing loss
Congenital sensorineural hearing loss (SNHL)	Genetic for more than 50 % of individuals, of which 80 % are autosomal recessive and 70 % are non-syndromic (i.e. unaccompanied by visible birth defects)
	Less than 1 % of genetically caused hearing impairment is X-linked or mitochondrial. Genetic syndromes associated with hearing loss include Usher syndrome, Pendred syndrome, Jarvell Lange-Nielsen syndrome, Waardenburg syndrome, and CHARGE syndrome
	In non-syndromic individuals, the most prominent genetic mutation is in gap junction beta-2 (GJB2). This gene is responsible for the protein connexin 26 and accounts for 30–50 % of congenital non-syndromic hearing impairment
	Congenital SNHL can also be due to prenatal infections with cytomegalovirus (CMV), herpes simplex virus, rubella, syphilis, and toxoplasma or post-natal infections with meningitis. CMV has replaced rubella as the most significant infectious agent associated with hearing loss, with 14 % of children with congenital CMV infection developing SNHL
Acquired SNHL	Untreated neonatal hyperbilirubinemia and the use of ototoxic medications like aminoglycoside antibiotics and antineoplastic agents
	Prematurity and perinatal anoxia are associated with early-onset hearing impairment
	In the older population, age-related degeneration and toxic effects of noise exposure can result in hair cell loss and sensorineural hearing impairment
Mixed hearing loss	Many children, particularly ones with chronic medical problems such as extreme prematurity, malignancy, immunodeficiency, HIV infections, and genetic syndromes, can present with mixed hearing impairment. As such, it is vital that adequate evaluations are completed so physicians can make accurate predictions of hearing ability and optimize the child's treatment plan

In the United States, the Joint Committee on Infant Hearing recommends that screening should be completed by 1 month of age, diagnosis should be made by 3 months, and intervention should begin by 6 months [11]. Universal hearing screening in the United States has been successful at reaching the vast majority of newborns; however, the rates of follow-through for those who did not pass hearing screens are not as favorable. The Center for Disease Control and Prevention (CDC) reports that almost 98 % of newborn infants in United States are appropriately screened for hearing impairment, although 35 % of infants who did not pass their hearing screen were lost to follow-up [12].

In early childhood, risk factors for hearing impairment should be assessed regularly at well-child checks, with formal hearing screens to occur periodically after 4 years of age [13]. The

American Academy of Pediatrics also recommends a formal hearing evaluation between 24 and 30 months for children with risk factors for hearing impairment (see Table 98.4).

The choice of hearing screening test depends on the age and cooperation of the child. Common screening tests in the very young child generally focus on evaluating the structural integrity of the hearing apparatus. Tympanometry measures the movement of the tympanic membrane in response to air pressure and can provide important information about potential conductive hearing loss. Tools used for newborn hearing screens (ABR and OAE) can also be utilized beyond the neonatal stage, although sedation may be required if there is significant motion artifact. In the older child, behavioral audiometry can be used to screen the hearing threshold at specific frequencies; however, they should only be considered diagnostic if administered in a soundproof room with a compliant patient. Children between the ages of 2 and 4 years can usually cooperate with play audiometry, in which they are asked to complete a task (e.g. picking a puzzle piece, dropping a block, etc.) when a sound is heard. Conventional screening audiometry (i.e. raising a hand in response to sound) can usually be completed in children over the age of 4 years. Whatever the modality of hearing screen, any failed screen

Table 98.4 Risk factors associated with neurosensory hearing loss in children

Family concerns	Any concerns from family members or caregivers of the child about hearing, language or speech
Family history	Family history of hearing loss during childhood or later progressive hearing loss
Medical history	Bacterial or viral meningitis
	Treatment with ototoxic drugs
	History of head trauma
	History of chemotherapy
	Conditions requiring treatment in the neonatal intensive care unit
	History of exchange transfusion
	History of extracorporeal membrane oxygenation (ECMO)
	In-utero infections
Physical examination findings	Facial dysmorphic features associated with syndromes with high risk for hearing loss
	Congenital craniofacial anomalies
Genetic syndromes	Waardernburg
	Alport
	Pendred
	Jervell
	Lange-Neilson
	Usher
	Down (conductive, neurosensory or mixed)
	Cockayne syndrome
Neurodegenerative conditions and inborn errors of metabolism	Sensory motor neuropathies
	Wolfram syndrome (DIDMOAD)
	Mucopolysaccharidoses
	Mitochondrial metabolism disorders
	Peroxisomal disorders
	Vitamin responsive disorders
	Canavan disease
	PRPP synthetase deficiency

should be followed-up with a formal diagnostic evaluation.

Appropriate diagnostic evaluations of hearing depend on the developmental age of the child, and hearing loss can be diagnosed even in infants and children who are unable to comply with conventional audiometry. Diagnostic ABR differs from screening ABR in that it provides frequency-specific information and can differentiate conductive versus sensorineural hearing loss. It can be performed in natural sleep but may require sedation for accurate results. This testing modality is typically utilized in infants younger than 6 months or those children who fail less-invasive modalities. Visual reinforced audiometry (VRA) is usually utilized in children between 6 and 24 months, in which children are trained to turn towards an object of interest such as a lighted toy in response to frequency-specific sounds. For older children, play or conventional audiometry can be used to diagnose hearing loss so long as they are completed with a trained audiologist in a sound proof room.

Hearing Impairment and Intellectual Disability

Recent surveys of children with hearing impairment have shown that they are more likely than their hearing peers to have additional disabilities, with up to 40 % of deaf children carrying additional diagnoses such as developmental delays, intellectual disability, autistic spectrum disorder, and vision impairment [14]. The Annual Survey of Deaf and Hard of Hearing Children and Youth from 2009 to 2010 demonstrated that the prevalence rate of reported autism was 1 in 53, which is above the rate of 1 in 68 seen in the general population [15]. In these surveys, more severe hearing loss was associated with more severe autism and intellectual disability. Population surveys of children with cerebral palsy suggest that between 4 and 12 % of them have hearing loss, with higher rates of hearing loss in children with more severe cerebral palsy as evidenced by lower motor function, vision impairment, and intellectual disability [16, 17]. Children with sensorineural hearing impairment are also more likely to have motor deficits, balance problems, vestibular control, and sensory processing [18]. Adults with hearing impairment have been shown to have a higher rate of mood disorders, decreased social participation, increased social avoidance, and increased psychosocial stress, although quality-of-life measures in children with hearing impairment have more mixed results for mental health and well-being [19–22].

Possible explanations for these connections include shared genetic mutations, underlying etiologies, and risk factors (e.g. in-utero infection, pre-term delivery); it is unlikely that the auditory deprivation from hearing loss directly causes autism or other neurodevelopmental disorders.

Management of Hearing Impairment

The deaf community represents a subset of individuals with hearing impairment that constitute a unique ethnic minority with its own language, art, history, and culture that exists within the majority "hearing world." The term "Big-D" deaf has been utilized by anthropologists as well as Deaf individuals to refer to the ethnocultural identity of deafness rather than the medical-disability conception of hearing impairment. In fact, many in the deaf community have rejected terms like "disability" and "impairment" because they suggest a deficiency [23]. Membership in the deaf community is more complex than the diagnosis of hearing impairment alone; it has been defined as an audiological, social, political, and linguistic identity [24]. Deaf studies highlight the "90 % rule" of deafness: 90 % of deaf children are born to hearing parents, 90 % of deaf individuals marry other deaf individuals, and 90 % of deaf couples have hearing children [25]. As such, the culture typically exists horizontally within one generation, passing from peer-to-peer rather than from parent to child. However, the effects of deafness may extend across three generations–the hearing parent of the deaf child, the deaf individual himself, and the child of the deaf individual [26].

As a minority culture group that is often unrecognized, the deaf community has experienced unique struggles and impacts resulting from development in the medical and educational fields. In fact, United States federal surveys do not make distinctions between hearing impairment as a diagnosis and deafness as a cultural identity [27]. These difficulties are particularly unique as very few deaf individuals are born into deaf families; it is a common story for an individual with hearing impairment to discover the shared-culture of the deaf community in his or her young adulthood. Early intervention for hearing impairment and aggressive mainstreaming of children in public schools poses challenges to cultural conceptions of deafness as identity and sign language as the primary mode of communication.

Systems of communication represent an important issue in the field of deaf studies. Historically, the "oralist" method, championed by Alexander Graham Bell and Horace Mann, encouraged children with hearing impairment to speak and read lips instead of signing. Conversely, the "manualist" method, headed by educators like Thomas Gallaudet, emphasized the use of manual signs as the main method of communication. Signing systems range from word-for-word translation of English words and phrases (Signed Exact English, or SEE) to a unique system of communication utilizing visual grammar and "natural" syntax (American Sign Language, or ASL), with "pidgin" variants in between. The decision for a primary communication modality has important ramifications for cultural identity (i.e. deaf versus mainstream) as each method is closely connected to self and cultural identity. ASL is the primary language of the deaf community and has been described as a "natural language" with grammar, syntax, and complexity equivalent to spoken language [28]. In fact, those children with hearing impairment who are raised in a setting with native signers have a language developmental trajectory similar to children without hearing impairment [29]. ASL and English ability are positively correlated with the benefits of ASL/English bilingualism paralleling bilingualism for two spoken languages [30, 31].

Today, many educators endorse the "total communication" method, wherein the child with hearing impairment is educated through a combination of lip reading, speech, and sign language [32]. Early immersion in sign language can provide important grammatical frameworks for the child with severe hearing impairment. However, most of children with hearing impairment are not surrounded by fluent signers in early childhood and miss out on language-building activities like nursery rhymes, songs, and storybooks that are typically found in the hearing environment. Parents who are interested in pursuing exposure to sign language may need to contact their local school district to find appropriate resources. If not available in the school setting, parents and educators may wish to use electronic or digital resources to provide this stimulation (e.g. http://www.signingtime.com/). Internet resources are also available for connecting with local deaf communities through social networks.

Hearing Aids

In general, unilateral hearing loss can be managed with education/awareness, preferential seating, and other behavioral strategies to minimize the impact of impairment. With bilateral hearing loss, assistive devices such as hearing aids may be considered. Hearing aids in general amplify sound but do not completely correct hearing loss. For maximum efficacy, they may also require the reduction of background noise below 35 dB and reverberation time (the amount of time before sound will subside) below 0.6 ms. Hearing aids may be found in a variety of formulations, including those that fit behind the ear, over the ear, in the ear, in the canal, or completely in the canal (see Fig. 98.5).

For those with external ear malformations, a bone anchored hearing aid can be surgically implanted in the skull to facilitate sound conduction. In the school environment, modifications may need to be made to the classroom such as the use of drapes, carpet, and acoustic wall panels to reduce background noise and echo. Alternatively, hearing aids can be paired with FM radios in the

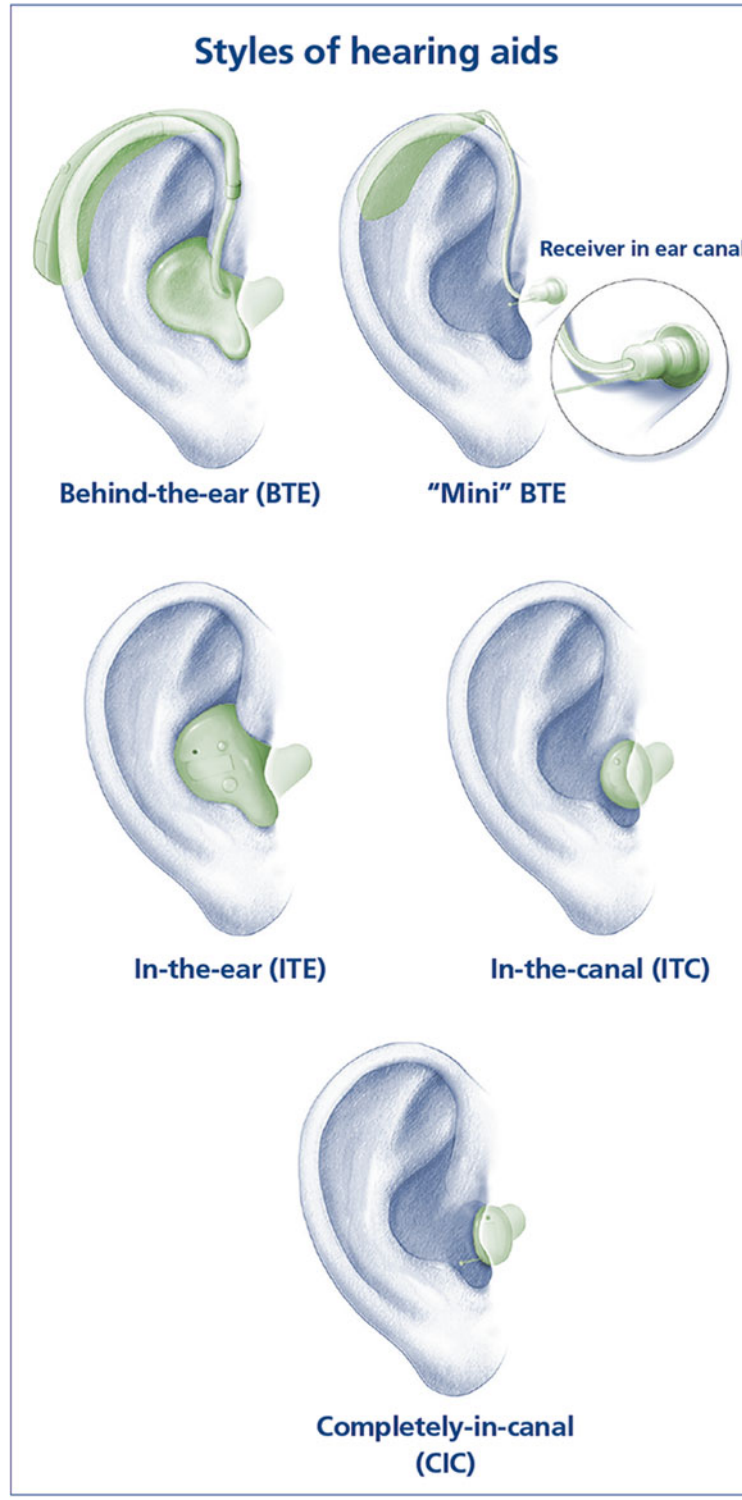

Fig. 98.5 Styles of hearing aids

school environment in which the sound from a teacher's microphone is directly transmitted to the child's hearing aid.

The benefits of hearing aids are contingent upon their regular use, which can be challenging if the individual has difficulty in accepting the aid. Behavioral techniques such as graduated practice and positive reinforcement strategies may be required before they can be used on a consistent basis. Hearing aids should be kept as dry as possible–removed when swimming or bathing, protected in damp environments (e.g. in the bathroom while showering), and stored with a dehumidifying container when not in use.

Cochlear Implants

Cochlear implants represent a major advancement in the field of hearing impairment for those who do not achieve adequate gains with hearing aids. The implants consist of an external microphone and a speech processor that receive noises from the environment; a transmitter that sends information through the skin; and a surgically implanted receiver/stimulator that communicates to the cochlea via an electrode array. The stimulator therefore bypasses the hair cells of the cochlea, which is often the limitation of a hearing aid (see Fig. 98.6).

Henry is a 2.5 year old boy with CHARGE syndrome who was diagnosed with profound bilateral mixed hearing impairment within the first weeks of life by ABR. Despite intensive early intervention services including bilateral hearing aids, he has not made dramatic gains in his language progress. His parents are eager to do "whatever they need to do" to maximize his communication skills and ask for your advice regarding the appropriate educational setting as well as the consideration for a cochlear implant, which they believe will fix many of his hearing difficulties. They ask that you write them a letter to the insurance company stating that the cochlear implant is medically necessary for his condition.

Cochlear implants are approved by the US Food and Drug Administration (FDA) for use in children as young as 12 months; additionally, "off-label" use in children under a year of age is not uncommon [33]. The drive to implant younger children stems from a growing body of research describing a critical period of neuroplasticity within the first 4 years of life, with the end of the sensitive period around 7 years of age [34]. Evidence suggests that children who receive cochlear implants in the preschool years have improved academic performance as high schoolers, although their literacy skills still lag behind those of their hearing colleagues [35]. A systematic review of the literature on outcomes following cochlear implantation suggested that earlier implantation and longer duration of cochlear implant use were associated with improved speech perception and overall quality of life [36]. However, diagnostic hearing assessments and adequate trials of hearing aids are difficult to conduct in the young child, and care must be taken that implantation is not done inappropriately [37]. Historically, unilateral cochlear implantation has been the standard with bilateral implantation done in only select cases. However, recent research has suggested that bilateral cochlear implantation to allow binaural hearing may induce better auditory development and potentially better speech outcomes, and further research in this area may change the standard of care [38].

Children with intellectual and developmental disabilities (IDD) and hearing impairment were once excluded from cochlear implantation, but there is a growing trend of considering these children for implantation. The research outcomes on these children are limited by the heterogeneity of the population and the paucity of quality studies. Children with IDD and hearing impairment have been shown to make language gains following cochlear implant but below the rates of their peers with hearing impairment alone, although a sub-population of these children may make very little gains at all [39, 40]. Similarly mixed findings have been shown in children with hearing impairment and vision impairment, with developmental levels serving as an important mediator in language outcomes following implantation

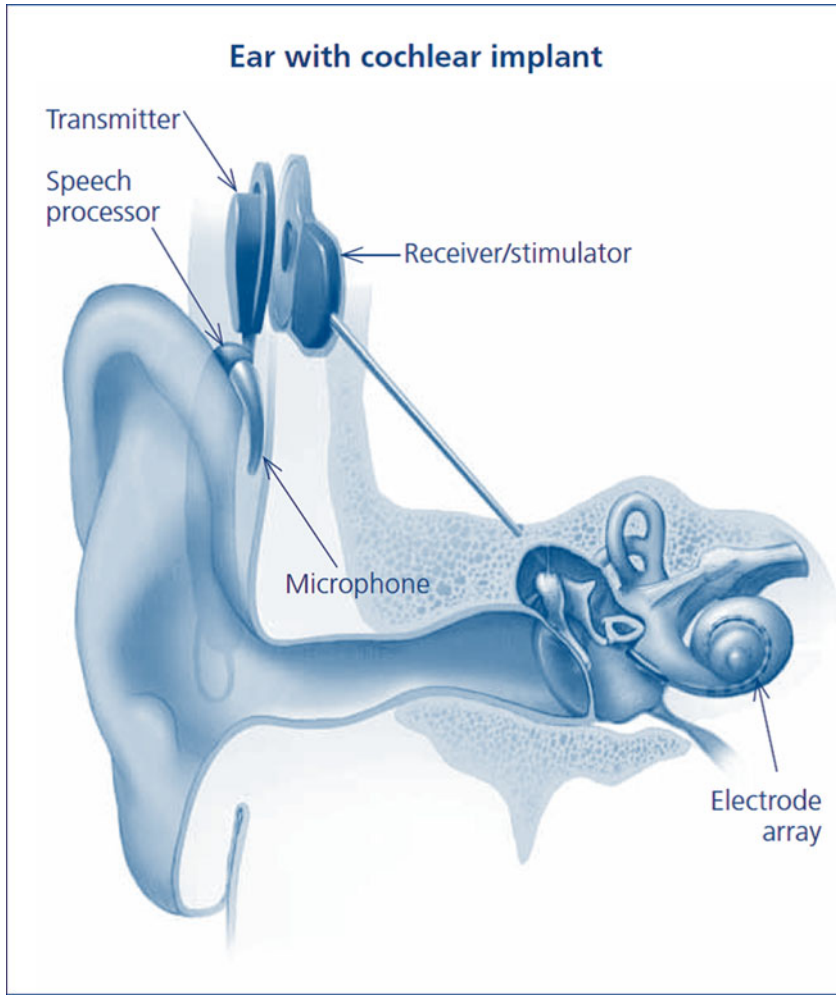

Fig. 98.6 Ear with cochlear implant

[41]. Other important predictors of outcome in cochlear implantation in children with multiple disabilities include maternal education and degree of hearing loss; however, these mediators may also depend on the nature of the additional disabilities, with disorders such as autism playing an important role in mediating poorer language and behavioral outcomes [42, 43]. Additional disabilities may increase the rates of limited or non-use following implantation [44].

Cochlear implants have posed an interesting ethical dilemma when placed in the context of the Deaf community. As mentioned previously, the majority of children with congenital SNHL are born to hearing parents who may not be members of the Deaf community. When the FDA approved the use of cochlear implants in children in 1990, the National Association of the Deaf (NAD) issued a statement that cochlear implants threatened a linguistic and cultural minority and recommended an ethics conference to discuss cochlear implantation in children. However, when a National Institutes of Health (NIH) conference was held in 1995 to discuss this issue, no representative from the deaf community was present. This led to a greater schism between health professionals and supporters of deaf culture. However, likely due to the influence of moderates in the deaf community and emerging outcomes research on cochlear implant use, the NAD withdrew its earlier position paper and replaced it with a new one in 2000, recognizing cochlear implants as one of many treatment options. Parents of children with hearing impair-

ment and the health care professionals that treat them should be cognizant of issues surrounding deaf culture when considering a cochlear implant.

Therefore, the decision to place a cochlear implant should include a thorough discussion of the potential benefits as well as risks of the procedure. The most common side effects of the procedure include taste disturbance, device failure, wound complications, and facial nerve stimulation [45]. In addition, children with cochlear implants have a small but documented increased risk of bacterial meningitis even years following implantation [46]. Because of the device's invasive nature, implantation causes irreversible changes in the inner ear which will disrupt any natural hearing ability. Parents and caregivers who make the decision on behalf of the child should have realistic expectations on the anticipated gains from the cochlear implant, and continued therapy and follow-up are necessary to maximize the benefits of the device. As such, the evaluation for cochlear implant candidacy should fall under the purview of a multidisciplinary team operating in a methodical, step-wise fashion [47, 48].

Educational Interventions and Outcomes

Hearing impairment can have repercussions across multiple dimensions of functioning even if it is not accompanied by additional disabilities. Even a mild hearing impairment has been associated with lower academic outcomes and behavioral problems; and many children with profound hearing impairment never achieve more than a fourth grade reading level [49–51]. However, research has demonstrated that earlier intervention is associated with improved outcomes, underlying the importance of early identification and intervention [52, 53].

Federal and state-funded educational institutions play an important role in the intervention for a child with hearing impairment. Children with documented hearing impairment are eligible for Early Intervention services under Part C of the Individuals with Disabilities Education Act (IDEA). The referral patterns demonstrated that 86 % of infants diagnosed with hearing loss as a result of newborn screening procedures were referred appropriately to intervention services [34]. For school age children, deafness or hearing impairment is a low-incidence disability, comprising around 1 % of students who receive services under the IDEA (http://tadnet.public.tadnet.org/). However, children with "hearing impairment" may differ in age at hearing loss, age at intervention, level of non-verbal IQ, presence of additional disabilities, parental perspectives, socioeconomic status, proficiency of language, use of assistive devices or technology, and others [54]. As such, it can be difficult to generalize findings in education literature regarding interventions for children with hearing impairment.

The public schools tasked with creating Individualized Education Plans (IEPs) for children utilize standardized assessments to inform their plans. However, in the research literature, there is little standardization of accommodations provided in academic assessments for individuals with hearing impairment despite the fact that these assessments are crucial in determining the educational plan [55]. Moreover, the services provided under the IEP can vary widely depending on the resources of the school and the strengths and challenges of the individual child. Broadly speaking, children with hearing impairment have shifted from primarily sign-language schools to general education settings as "schools for the deaf" violated the federal mandate for the least restrictive environment under the IDEA [56]. Accommodations for children with hearing impairment can include extended time for testing or access to a resource facility, which are common to individuals with other disabilities. A child who communicates primarily in sign language may require an interpreter to receive directions and record responses. Parents may wish to investigate residential, more restrictive school environments that emphasize sign language and Deaf culture, which are scattered throughout the United States.

In some studies, DHH students educated in general education settings appear to do better than those in self-contained classrooms; however, as an observational finding, this only demonstrates association, not causation [56, 57]. A growing body of evidence suggests that a bilin-

gual model combining ASL and English exposure may result in greater academic gains in reading and mathematics, even in the presence of additional disabilities, perhaps because of the unique skills required in sign language [58]. DHH children who communicate primarily in sign language may have weaknesses in sequential working memory, processing speed, increased memory load, and possible deficiencies in attention when compared to hearing individuals. Visuospatial recall, imagery, and dual encoding seem to be stronger in signing individuals than hearing individuals, suggesting that educational strategies may need to be adjusted to optimally address their learning capabilities [59].

Studies on the academic achievement of DHH children have demonstrated poorer outcomes when compared to their typically developing peers, with consistent annual academic gains that may be as low as one-third of expected [60]. Children and individuals with hearing impairment have higher rates of school drop-out and around half the rates of high school, college, and post-graduate education when compared to the hearing population. They also have lower rates of health insurance, lower family incomes, and higher rates of unemployment [61]. However, there are some DHH children that maintain the expected level of academics with early academic success predicting later academic success [62–64]. More research needs to be done to determine the various predictors of academic achievement and the relative strengths and weaknesses of self-contained, mainstream, and bilingual environments.

The impact of hearing impairment can continue through adulthood with lasting effects on employment and quality of life. Around 30 % of young adults with isolated hearing impairment stated they experienced at least one limitation in an activity of daily functioning, with more significant impairment for those with additional disabilities [65]. In another study, young adults with isolated childhood-onset hearing impairment were at increased risk of lacking an adult social role, with almost 18 % of not being employed, a student, or a caregiver [66]. A multivariable analysis of these data suggested that degrees of impairment and educational impairment can mediate the acquisition of adult social roles. On another note, young adults with isolated hearing impairment have been shown to have lower rates of smoking and alcohol abuse when compared with individuals with other disabilities [67–69].

Conclusions

Hearing impairment is a complex and multifaceted diagnosis that can have long-reaching impacts on health, education, and quality of life. Throughout different outcomes research, the predominant common theme is that early diagnosis and intervention result in the best outcomes. As the population with hearing impairment is heterogeneous with a wide spectrum of disease burden, treatment plans must be personalized and optimized for each affected individual. Family-centered and culturally-sensitive care are equally important when discussing different treatment options, especially to enable long-term follow-up and optimize favorable lifelong outcomes.

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