Introduction

Parents of young children who are identified as deaf or hard of hearing (DHH) are suddenly thrust into a world of new concepts and a bewildering array of terms. What's a decibel or hertz? What does sensorineural mean? Is a moderate hearing loss one to be concerned about, since it's only moderate? What's a tympanogram or a cochlear implant? These are just a few of the many questions that a parent whose child has been identified as DHH may have. In addition to parents, questions also arise from professionals and paraprofessionals who work in the field of early hearing detection and intervention (EHDI) and are not audiologists. The purpose of this chapter is to provide basic answers to these and other important questions about the field of audiology.

What is an audiologist?

An audiologist is a specialist in hearing and balance who typically works in either a medical, private practice, or an educational setting. The primary roles of an audiologist include the identification and assessment of hearing and balance problems, the habilitation or rehabilitation of hearing and balance problems, and the prevention of hearing loss. When working with infants and young children, the primary focus of audiology is hearing. Audiologists are licensed by the state in which they practice and may be members of the American Speech-Language-Hearing Association (ASHA), American Academy of Audiology (AAA), Academy of Doctors of Audiology (ADA), or the Educational Audiology Association (EAA). Some audiologists hold the...
American Board of Audiology’s Pediatric Audiology Specialty Certification. Others may hold the ASHA Certificate of Clinical Competence in Audiology (CCC-A).

Several online search tools are available to find an audiologist:

- Early Hearing Detection and Intervention Pediatric Links to Services (EHDI-PALS)
- Early Hearing Detection and Intervention (EHDI) program, also known as the Newborn Hearing Screening Program, in each state
- ASHA
- AAA

What are the parts of the ear?

A basic understanding of the parts of the auditory system and how they work is helpful to understanding the different types of hearing loss. There are four main parts of the auditory system:

- Outer ear
- Middle ear
- Inner ear
- Central auditory system

Each part plays an important role in transferring and processing sound, so that the brain can recognize and interpret what a particular sound means.

The outer ear consists of three major parts:

- Pinna
- Ear canal
- Eardrum (tympanic membrane)

The pinna is the part of the ear that we see and contributes slightly to locating a sound. The ear canal, which is about an inch long and S-shaped, channels sound toward the eardrum. The ear canal produces ear wax which helps to clean the ear canal of debris. The last part of the outer ear is the eardrum which is the boundary between the outer and the middle ears. The eardrum is a very thin membrane, consisting of layers of skin and fiber tissue. It is attached to the wall of the ear canal and also to one of the bones of the middle ear. The eardrum seals the middle ear from the environment. Sound vibrates the eardrum and is changed to mechanical energy.

The middle ear is a hollow space that is separated from the ear canal by the eardrum and contains the three smallest bones in the body. Sounds travel through the outer ear and are transferred to the inner ear by these bones (ossicles). The three bones are the hammer (malleus), anvil (incus), and stirrup (stapes). They are connected by ligaments, and two of the bones have tiny muscles attached. The shape and arrangement of the ossicles increases the strength of the mechanical energy. When loud sounds are present, the tiny muscles contract and reduce the strength of those sounds. This helps protect the ear from damage due to loud sounds.

The Eustachian tube is also part of the middle ear system and connects the middle ear space to the back of the throat. The Eustachian tube is normally closed but opens periodically to keep the air pressure in the middle ear space the same as the surrounding environment. An example of the Eustachian tube working occurs when a yawn or a swallow “unstuffs the ears”
If a child is born with a significant malformation of the outer and/or middle ear that prevents or reduces the sounds being conducted to the inner ear, surgery may be possible. The child’s hearing ability may or may not improve after surgical treatment.

When flying in an airplane, sometimes the tissues of the Eustachian tube become swollen from a cold or upper respiratory infection, and it doesn’t open and close well—causing problems in the middle ear. It’s possible for infection to spread from the nose and throat area through the Eustachian tube to the middle ear, which is one of the causes of middle ear infections.

The third part of the auditory system is the inner ear. The inner ear has two sections: one that is responsible for balance and the other for hearing. The hearing part of the inner ear is the cochlea. The cochlea is a snail-shaped space in the skull that contains very tiny structures that convert mechanical energy into electrical impulses needed for the nervous or central auditory system. The cochlea is divided by tissue structures into three channels, each of which is filled with fluid. There are many thousands of tiny hair cells (stereocilia) that are embedded in the tissue that divides the three sections. The hair cells bend slightly in response to different kinds of sounds, depending on where they are located in the cochlea. When bent, the hair cells create electrical signals that are then sent to the central auditory nervous system.

The last part of the auditory system consists of the auditory nerve and the central auditory system in the brain. The electrical nerve impulses produced in the cochlea by the hair cells are transmitted and processed along the auditory nerve that consists of about 25,000 nerve fibers. The signal continues through the brain stem to the auditory cortex of the brain. It is in the cortex that sounds are interpreted based on experience and association and that meaning is assigned to sounds that travel through the outer, middle, and inner ears.

Auditory Transduction by Brandon Pletsch is an excellent 7-minute animation on YouTube about the different structures of the auditory system, how they work, and how each contributes to hearing.

What are the types of hearing loss?

Hearing losses can be categorized by when they occur. A hearing loss that is present at birth is called a congenital hearing loss. But hearing loss can and does occur at any time and can be called later-onset or acquired. If a hearing loss continues to get worse, it is called a progressive hearing loss.

One of the ways that an audiologist describes a hearing loss is by how many ears are involved. If a hearing loss is only in one ear, the loss is called a unilateral (one-sided) hearing loss. If there is hearing loss in both ears, it is described as a bilateral (two-sided) hearing loss.

The different types of hearing loss are primarily based on what part of the ear is preventing a sound from being transferred and processed effectively.

Conductive Hearing Loss

A conductive hearing loss occurs because problems in the outer and/or middle ear keep the sound from being “conducted” well. Conductive hearing losses can be either temporary (transient) or permanent. Medical treatment of the underlying cause of the temporary conductive hearing loss may result in the hearing returning to normal or near normal. For example, if the ear canal is plugged with earwax or an object of some sort, some hearing loss will occur until the blockage is removed. The amount of hearing loss would be similar to having an earplug in your ear canal.

At least 80% of children have three or more episodes of ear infections (otitis media) before 3 years of age (Roberts & Hunter, 2002). These ear infections can be painful, and if the middle ear space fills with fluid, a temporary conductive hearing loss can occur. Middle ear infections left untreated can cause some other middle ear problems that may result in a permanent loss.

If a child is born with a significant malformation of the outer and/or middle
ear that prevents or reduces the sounds being conducted to the inner ear, surgery may be possible. The child’s hearing ability may or may not improve after surgical treatment.

**Sensorineural (or Sensory) Hearing Loss**

A sensorineural (or sensory) hearing loss from problems in the cochlea or inner ear is almost always permanent. There are many different causes of a sensorineural hearing loss. Some losses can be genetic or syndromic—the result of some medications, infections, high fevers, or head trauma.

Auditory neuropathy spectrum disorder (ANSD) describes a disordered transmission of the electrical signal along the acoustic nerve. ANSD—sometimes called auditory neuropathy/dyssynchrony—is relatively rare in the well-baby population and somewhat more prevalent in babies who have spent time in the neonatal intensive care unit.

**Mixed Hearing Loss**

A mixed hearing loss has both a conductive component and a sensorineural component. The conductive component is the result of a problem in the outer and/or middle ear, while the sensory or sensorineural portion results from a problem in the inner ear.

**Central Hearing Loss**

Central auditory processing refers to how well the central nervous system transmits and uses auditory information. Disorders of central auditory processing can include problems with determining where a sound is coming from and excessive difficulty understanding speech and auditory signals in poor listening conditions, such as noisy settings. These disorders may coexist with other disorders, such as language impairment and learning disorders, but is not the result of those disorders.

How often does hearing loss occur in young children?

Many children have a temporary conductive hearing loss due to ear infections, but how many have a permanent hearing loss? Each year, state EHDI programs send the results of their newborn hearing screening program to the Centers for Disease Control and Prevention (CDC). In 2013, over 97% of the babies born in the United States had their hearing screened within the first few days of life, and 5,253 of them were born with a permanent congenital hearing loss. This is a prevalence of 1.5 per thousand (15 of every 10,000) babies who received a newborn hearing screening. Those are only the babies with a confirmed hearing loss. Nationally, almost one-third of the newborns who don’t pass the newborn hearing screening aren’t getting the recommended follow-up evaluations, so there are likely more congenital hearing losses than are reported (CDC, 2015).

About 40% of the newborns reported by the state EHDI programs to the CDC for 2013 had a unilateral hearing loss, while 60% of them had a bilateral hearing loss. Only 14% of the babies born with a permanent hearing loss had a conductive hearing loss. About 62% of the permanent congenital hearing losses reported to the CDC were sensorineural—by far the most common type. Permanent congenital mixed hearing losses are somewhat rare—about 8% of the results reported to CDC in 2013. ANSD is also rare, accounting for only 5% of the permanent congenital losses (CDC, 2015).

Hearing loss can and does occur at any time in a person’s life. Eiserman et al. (2008) found that 1.5 per thousand (15 of every 10,000) children up to 3 years of age who had been screened using otoacoustic emissions technology in Early Head Start programs had a permanent hearing loss that hadn’t been identified earlier. Similarly, Bhatia et al. (2013) identified 2.5 per thousand (25 of every 10,000) with a newly identified permanent hearing loss in children birth to 3 years in a screening program at federally-funded...
The rate of permanent hearing loss continues to climb as children get older. It is estimated that as many as 14.9% of school-age children have a hearing loss.

In 2006, Morton and Nance published *Newborn Hearing Screening - A Silent Revolution*, which identified the causes of permanent hearing loss at birth and also at 4 years of age. As can be seen in Figure 2, the incidence of permanent hearing loss at birth was nearly 2 per thousand (186 per 10,000). By 4 years of age, as shown in Figure 3, the incidence of permanent hearing loss increased to about 3 per thousand (270 per 10,000).

The rate of permanent hearing loss continues to climb as children get older. It is estimated that as many as 14.9% of school-age children have a hearing loss (Niskar et al., 1998).

**What causes permanent hearing loss?**

Morton and Nance (2006) reported that a mutation of the GJB2 gene was responsible for 21% of congenital permanent hearing losses (see Figure 2). That, however, was only one of the genetic causes of congenital hearing loss. Causes of hearing loss associated with various syndromes, including Pendred's syndrome, accounted for 17% of congenital hearing loss, and an additional 30% of hearing loss at birth was due to unspecified nonsyndromic genetic factors. Overall, genetic factors accounted for 68% of the congenital permanent hearing losses.

**Figure 2**

*Causes of Hearing Loss at Birth*

- Incidence at Birth (186 per 100,000)
  - Clinically apparent infection, 10%
  - CMV, 21%
  - GJB2 mutation, 21%
  - Other genetic causes, 44%
  - Nonsyndromic, 30%
  - Pendred's syndrome (SLC26A4), 3%
  - Clinically inapparent infection, 11%

**Figure 3**

*Causes of Hearing Loss at 4 Years of Age*

- Prevalence at 4 Years (270 per 100,000)
  - Other genetic causes, 33%
  - CMV, 25%
  - Nonsyndromic, 22%
  - Other environmental causes, 14%
  - Syndromic, 11%
  - mtA1555G, 1%
  - GJB2 mutation, 15%
  - EVA, SL26A4, 7%
  - Late-onset, 10%
  - Clinically inapparent infection, 8%
  - Clinically apparent infection, 7%
Congenital cytomegalovirus (CMV) is a leading cause of congenital hearing loss, accounting for 21% of the hearing losses present at birth. About half of children with hearing loss due to CMV show other complications of congenital CMV, such as vision loss; small head size; or problems with the liver, spleen, or lungs.

By 4 years of age, over 50% of the hearing losses were due to genetic factors (see Figure 3). Congenital CMV still accounted for about one-fourth of all permanent hearing losses, but there were some late-onset hearing losses due to CMV.

Why is it important to screen hearing as early as possible?

It is critical to understand the importance of discovering a hearing loss as soon as possible. The sooner we identify a hearing problem, the earlier the intervention to minimize the impact of the hearing loss and strategies to maximize use of the remaining hearing sensitivity can be implemented. Simply stated, the sooner we can find a hearing problem, the sooner we can start to help, and the greater the success of language and communication development.

There are formal recommendations for the minimum ages and time periods for each step in the process for the identification and diagnoses of hearing loss and the necessary intervention and followup. The Joint Committee on Infant Hearing (JCIH, 2007) 2007 Position Statement recommends the following newborn hearing screening guidelines:

1 month. By 1 month of age, a hearing screening is completed.

3 months. By 3 months of age, the child failing or referring a hearing screening will have a complete diagnostic hearing evaluation with audiology and otolaryngology examinations. If a hearing loss is diagnosed, the child will be fit with hearing aids as per the parents choice.

6 months. By 6 months of age, the child will be enrolled in early intervention services.

Why are these targets so important? Because the rate of growth and development in the first year of a baby’s life is unmatched at any other time during postnatal (after birth) development. These findings are supported by various brain-imaging techniques. Imaging studies, such as this Positron Emission Tomographic (PET) scan (see Figure 4), show that the brain rapidly matures in an orderly fashion during the first years of life. The orange-red color represents the rapid growth from 1 month to 1 year of age.

During this period, the infant brain is developing, and tiny synapses, which are biological electrical connections, are forming. The amount of stimulation a child receives directly impacts the number of synapses formed within the brain. This includes the hearing, speech, and language centers of his or her brain. The creation of synapses is virtually complete after the first 3 years of life, thus those years are the most important in brain development.
How is a hearing loss diagnosed?

A hearing loss is detected by screening and testing the auditory system through use of age-appropriate hearing tests. Hearing tests are used to determine four things:

These tests are sequential—moving from the outer part of the ear (peripheral) to more internal (inner ear) to more central and then to the whole auditory system. It is important to remember that no one test can stand alone. It takes an assessment of all the elements of the auditory system—or a “battery of tests”—to confirm hearing status.

Significance of the Hearing Loss

This means determining if the hearing loss is mild, moderate, moderate-to-severe, severe, or profound in nature.

The Kind or Type of Hearing Loss

This means determining if the hearing loss is caused by problems getting the sound into the inner ear where it can then be heard (conductive hearing loss), or if it is a problem in the inner ear or beyond in the pathways of the auditory system (sensorineural hearing loss).

Configuration of the Hearing Loss

This means determining if hearing is better or worse at some pitches (frequencies). Hearing loss can be equal or flat across all the pitches or better at either the low or high pitches.

Make Intervention Recommendations

And decisions on treatment strategies that most benefit the child.

What tests are used to determine hearing?

There are four primary tests for assessing hearing. They are:

1. **Immittance Audiometry.** A test of middle ear function.
2. **Otoacoustic Emissions (OAE).** A test of inner ear function.
3. **Auditory Brainstem Response (ABR).** A test of neural pathways of the auditory system.
4. **Behavioral Testing.** Assessment of the function of the whole auditory system from initial sound stimulation to an intentional response.

Immittance Audiometry (Tympanometry and Acoustic Reflexes)

Tympanometry is a measure of middle ear function. It provides information on the status (condition and function) of the middle ear system. It evaluates the normal occurring pressures of the middle ear system (the pressure you feel change when you “pop” your ears), as well as the needed mobility or movement of the ear drum or tympanic membrane. It is conducted by placing a small probe with a soft rubber tip in the ear canal and introducing pressure changes along with a sound “tone” into the ear canal. An abnormal result is consistent with a problem in the transfer of sound into the auditory system—known as a conductive hearing loss. You may have experienced a conductive hearing loss when you had a head cold or an ear infection where sound was muffled.

Tympanometry helps identify middle ear conditions, such as:

- A hole or perforation of the eardrum.
- Fluid behind the ear drum.
- Negative air pressure behind the ear drum.
- Normal ear drum movement.

In summary, tympanometry is an objective test of middle ear function. It is not a test...
of hearing sensitivity but rather a measure of acoustic (sound) energy transmission through the middle ear. As such, it is not used to assess the sensitivity of hearing but the function of the middle ear system and its resulting impact on hearing.

The middle ear system not functioning properly can lead to a problem in the transmission of sound energy and result in a conductive hearing loss. The presence or absence of acoustic reflexes can also be very helpful in the diagnosis of the nature of a hearing loss. The results of these tests should always be viewed in conjunction with the other hearing tests.

**OAE Testing**

OAE testing is a measure of inner ear function. Measurement of OAEs are a relatively recent addition to the audiologic test battery. Even though the existence of emissions was discovered by David Kemp in England in the late 1970s, it was not seen as a routine part of clinical testing until the late 1990s.

OAEs are a measurement of normally produced sound responses generated by very small hair cells in the cochlea. These responses are measured and recorded in the ear canal by placing a small probe with a soft rubber tip into the ear and providing sound stimulation. A very small microphone records and measures the tiny response (emission) obtained in direct response to the stimulation. Most normal healthy inner ears have an OAE response.

The presence of OAEs indicates that the middle ear system is most likely functioning appropriately (sound was transmitted normally), and the inner ear (outer hair cells) are functioning normally. Conversely, if there is no recordable OAE, then there may be a problem with one or both (middle ear and/or inner ear) systems (see “How OAEs Work” for a quick tutorial on OAE testing).

OAE testing can be done with people of any age, but the response is very robust in infants and young children. Children are easiest to test when they are very young or over the age of 18 to 24 months. Testing children between 6 months of age and 2 years of age may require the use of distractions with appropriate toys or using other strategies. As with all tests, OAE testing has some limitations—one of which is that it does not provide information about the degree or severity of a hearing loss. Another limitation is that, depending on test settings or parameters, it may not detect minimal or slight hearing losses.

**ABR Test (Auditory Evoked Response)**

ABR tests assess the function of the higher auditory system by measuring the reaction of the parts of a child's nervous system that affect hearing (auditory pathways). More simply put, ABR testing measures the hearing nerve's response to sounds. The ABR test is safe, can be automated for screening purposes, and is painless.

Notice that we are progressing from the “outside in.” In other words from the outer and middle ear system, to the inner ear's function, and then to the auditory pathways in the brain. ABR testing is completed by placing three to four small recording discs (electrodes that are connected to a computer) on the child's head and near his or her ears. Small earphones are placed into the child's ear canal, and sounds (usually clicks) are presented to stimulate the auditory system. Small waveforms that constitute responses to the stimuli at certain locations within the brain are recorded by the computer. The presence or absence of waveforms at specific sound levels and frequencies can confirm and help describe a hearing loss. We can simulate the ear and record responses from the brainstem that can confirm either normal hearing or a hearing loss.

The ABR test is sensitive to movement. Therefore the child being tested must be still. The ABR test can be completed only if the child is sleeping or lying still—relaxed and with eyes closed. Some factors to consider in this regard are:
ABR testing is the most commonly used auditory-evoked potential test. The ABR is valuable for use with infants and young children.

- Newborns are easily tested during normal and natural sleep. If a child is younger than 6 months of age, the ABR test usually can be done while he or she naps.

- For children between the ages of 6 months and 7 years, the ABR test is done under sedation, which means that the child will need medication to help him or her sleep through the test. ABR tests requiring sedation are most often done in a same-day outpatient surgery center.

- If the child is older than 7 years, the ABR test can often be done while the child is awake, relaxed, and lying still. The test is usually done by the audiologist in a quiet setting, such as a special sound-treated suite.

When sedation is needed, there are special restrictions or rules for eating and drinking that must be followed in the hours before the test. The test itself takes about 1 to 1.5 hours, but the entire appointment will take about 2 hours without anesthesia, and up to 4 hours due to the recovery time if the child needs sedation.

There are different types of auditory-evoked potentials that audiologists will use depending upon the situation. ABR is the most commonly known and is used in both automated newborn screening and diagnostics. While it is not in the scope of this chapter to go into detail on the various types of evoked potentials, they are named or labeled based on where in the auditory pathway the response occurs. The responses are expected to occur within certain specific timeframes and are measured in milliseconds. The further "up" in the system, the longer it takes to see the response. The amount of time lapsed or how long it takes for the response to occur is called latency.

ABR testing is the most commonly used auditory-evoked potential test. The ABR is valuable for use with infants and young children, because it provides the following information:

- A close estimate of the child's hearing levels (thresholds). The softest intensity level at which the ABR responses appear roughly correspond to the child's hearing level for each frequency range tested. This electrophysiologic response is slightly higher than the actual hearing levels.

- Evaluation of nerve conduction delays (timing) provide additional information on how the sound signal is processed for meaning.

**Behavioral Testing**

Behavioral testing requires an observable response to sound from the child. The child and the parent (or caregiver) is seated in a sound-treated booth. Sounds of varying intensity are presented through calibrated speakers or earphones. The sounds may consist of speech sounds as well as specific tones of different frequencies that are critical to hear speech sounds. The audiologist records the child's responses to the softest sounds and plots them on a graph called an audiogram.

Behavioral hearing tests include the following methods for specific developmental ages:

- Behavioral observation audiometry (BOA) is used for developmental ages of 0 to 5 months. The audiologist observes and records the child's responses to sounds. Responses may consist of quieting, eye widening, startle, etc. These responses must be consistent, repeatable, and appropriately correlated to the presentation of a sound.

- Visual reinforcement audiometry (VRA) is used for developmental ages of 6 months to 2 years. The audiologist observes and records when the child turns to the sound stimulus and gives a visual reinforcement or reward that is timed to the response. The reward is typically either a toy or puppet that lights up and/or moves to reinforce the child's response.
During a hearing test, the softest levels (thresholds) that sounds or tones of different frequencies can be heard are measured and recorded for each ear on the audiogram.

During a hearing test, the softest levels (thresholds) that sounds or tones of different frequencies can be heard are measured and recorded for each ear on the audiogram. Frequency is measured in Hertz, often abbreviated as Hz. The frequencies on the audiogram range from 125 Hz to 8000 Hz. Sounds on the left portion of the audiogram are lower-pitched sounds, with the lowest pitch on the audiogram being 125 Hz. As you move to the right, toward 8000 Hz, sounds get higher in pitch.

The intensity or loudness is shown along the left side of the audiogram. It is measured in decibels—often abbreviated as dB. As you move down the audiogram, the louder a sound must be made to obtain a response and establish the threshold. For example, a 10 dB sound is softer than a whisper for a person with normal hearing, while a 120 dB sound is as loud as a jet airplane. Familiar sounds are plotted on the audiogram for demonstration purposes, indicating the approximate pitch and loudness levels that these sounds occur. For example, a lawn mower is a very loud, low-pitched sound, while a bird’s chirp is a soft, high-pitched sound.

**Figure 5**

**Audiogram with Speech and Environmental Sounds**

Hearing loss is not “all or nothing” but has various degrees—just as vision does. Remember the loudness level that a sound can just barely be heard is the threshold. The audiogram is set up, so that if a sound is presented at a very soft level and heard, this threshold or detection level is marked at the
In this variation of the audiogram, the yellow-shaded area represents where the sounds of speech at a soft conversational level take place. If you look closely at this area—sometimes called the “speech banana”—you will notice that the sounds of speech occur in the loudness range from approximately 15-50 dB and the frequency range from approximately 250 to 8000 Hz. Vowels tend to be lower pitched and louder than consonants.

What are the degrees of hearing loss?

Normal hearing for children is in the range below 15 dB, corresponding to the orange band on the audiogram (see Figure 6). In other words, for a person with normal hearing at the different pitches (frequencies), the tone can be detected at very soft levels. A person with normal hearing would be able to easily hear all of the louder sounds, such as all of the speech sounds represented in the “speech banana,” and the other noises in Figure 5, such as a bird chirp.

The range of thresholds in the aqua band represents a minimal hearing loss. Some of the speech sounds on the “speech banana,” such as the “f” and “th,” are no longer detectable. So a child with a minimal loss wouldn’t be able to hear the difference between “fin” and “thin” based on an auditory signal alone, even in the best of listening environments. All the other speech sounds are being heard at a softer level.

A mild hearing loss—shown with the lavender band—occurs when the hearing thresholds are between 25-40 dB. We may think that “mild” is, well, mild . . . but notice how many speech sounds displayed in the speech banana are not being heard. So “mild” has a significant impact on understanding, especially for a very young child who has not yet acquired language and can’t “fill in the blanks” like adults with a long history of access to speech and language.

The green band shows the range of hearing thresholds for a moderate hearing loss. Notice how the speech sounds for soft conversational speech are nearly all inaudible, and that normal conversational levels (red dotted line) are perceived as a whisper. Moderate? Not when you consider the impact this degree of hearing loss would have on a child’s access to everyday speech during critical periods of language development.

The pink bar on this audiogram shows the threshold levels for a moderate-to-severe hearing loss, which is 55 dB to 70 dB. Most if not all of typical conversational speech would not be detected with this degree of hearing loss.

The blue area indicates a severe hearing loss with thresholds in the 70 to 90 dB range. The tan band at the bottom of the audiogram shows thresholds in the profound hearing loss range.
With these degrees of hearing loss, typical conversation as well as many environmental sounds would be inaudible without amplification.

Many hearing losses are not “flat” with similar thresholds across the frequency range but instead have different thresholds at various frequencies.

*Figure 7* is an example of a common configuration of hearing loss with different degrees of hearing loss at various frequencies. This is an example of a mild-to-severe sloping hearing loss. Some speech sounds are audible to this person, but many are not, so there’s a lack of clarity with many words and conversations. People with a long history of accessing speech and language can fill in some of the gaps, but very young children don’t have the advantage of that experience.

There are many unique configurations of hearing loss. Some have more hearing loss at the lower frequencies, while others have more loss in the highs. Others have about the same degree of hearing loss at all frequencies. Some may have normal hearing at some frequencies and a significant hearing loss at others. Every hearing loss makes some sounds in everyday conversations more difficult to access and therefore makes understanding more challenging. Here are two resources that simulate different degrees of hearing loss and are helpful for a person with normal hearing to experience the effects of hearing loss on understanding speech:

- Flintstones Cartoon by House Ear Institute
- Demonstrations by Success for Kids with Hearing Loss

**What are the treatment and intervention options for children with hearing loss?**

Once a child has been diagnosed with a hearing loss, it is necessary to begin treatment and intervention as quickly as possible. Intervention includes three key areas:

1. **Surgical Intervention.**
   - Evaluation for any medical/surgical intervention that may fix or repair a structural problem contributing to the hearing loss.

2. **Amplification/Cochlear Implants.**
   - Maximizing any residual hearing through amplification or cochlear implants.

3. **Early intervention for communication development**
   - via the parents’ choice of communication modalities (verbal, manual, combination, etc.).
Surgical Intervention

The first thing to consider is whether medical intervention, including surgery, may be helpful. In some cases, surgical intervention could assist or repair a problem, such as a closed ear canal (atresia) where all the other parts of the ear are normal. In these cases, the child would be evaluated for surgery, and, if appropriate, the surgery would be completed. The child’s hearing would be evaluated afterwards and compared to the prior results to quantify improvement and determine if further intervention is indicated.

Amplification

The second intervention to consider is the fitting of amplification (hearing aids) to enable the child to make maximum use of residual hearing capacity. The vast majority of children with hearing loss have some level of residual hearing, and amplification with hearing aids is the appropriate intervention. The JCIH (2007) recommends all infants diagnosed with permanent hearing loss should be fit with amplification within 1 month of confirmation of the hearing loss.

It is important to understand that it is possible to proceed with amplification as soon as a diagnosis of hearing loss is made, but the parents may not be emotionally prepared for such a step.

Cochlear Implants

When hearing loss is profound with little or no residual hearing, and hearing aids have not provided any benefit, a cochlear implant may be considered. There are specific criteria that must be met for a child to receive an implant. The criteria for candidacy has changed over time. However, the current criteria is as follows:

- Lack of benefit from amplification.
- A minimum age of 12 months but sometimes younger with approval.
- A minimum of bilateral severe-to-profound hearing loss.
- Must have no medical contraindications.
- Must have available appropriate educational and intervention support services for post-cochlear implant aural re/habilitation.
- Evaluation of family factors, such as motivation to followup. and provide the required post-implant education and intervention services, and that the family has realistic expectations.

Parents and primary health care providers should confer with a cochlear implant team to ask questions and determine a child’s candidacy. Current Federal Drug Administration (FDA) information and candidacy requirements can be found at:

- http://www.fda.gov/MedicalDevices/ProductsandMedicalProcedures/ImplantsandProsthetics/CochlearImplants/default.htm

How does a cochlear implant work?

A cochlear implant uses special electronic technologies to take the place of the nonworking parts in the inner ear and is designed to mimic natural hearing. Here is a brief overview of the parts of an implant and how it works. For more detailed information, see the Cochlear Implant and Cochlear Implant Candidacy chapters of this publication.

A cochlear implant has several parts (see Figure 8) that work as follows:
Early intervention is crucial for communication and overall development. Audiologists work closely with a team of professionals that often includes early interventionists, deaf mentors, parent infant programs, educators of the deaf and hard of hearing, speech language pathologists, and therapists with different communication specialties. Treatment and intervention should be focused on the following:

- Meeting overall developmental milestones.
- Communication modalities, such as oral, auditory verbal, American Sign Language, cued speech, total communication, and others.
- Emotional development for the child and support for the family.
- Social development.
- Cognitive development.

To assist early interventionists and contribute to the success of treatment and education plans, audiologists provide education on how to troubleshoot the child’s hearing aids or cochlear implant to ensure the child is hearing optimally. In addition, the audiologist provides very important information on what the child can or cannot hear with either hearing aids or cochlear implant. This detailed information includes describing what frequencies and speech sounds the child hears and how well he or she hears them.

Information regarding audibility of speech is important for many reasons. For instance, some speech sounds are louder (voiced), and others are quieter (no voicing). An example of this would be the sounds “B” and “P.” The sound “B” in isolation is made with the lips with sound vibration from the vocal cords. In contrast, the sound “P” is made the same way with the lips but has no voicing from the vocal cords. Try saying both in an alternating fashion a few times! If one were relying on visual cues, they would look the same. We differentiate the two sounds by hearing the voicing. In this example, the audiologist can help the family and early intervention team understand if...
the child can hear the difference and at what distances from the speaker or sound source.

Distance information is crucial. Sound travels and works in a predictable way. For example, every time you decrease the distance between a sound and its source by half, the sound increases in loudness by 6 dB. There is a saying in the world of habilitation of hearing loss that goes, “Come closer to me by 6 dB,” suggesting that we interact at optimal distances for hearing. The converse is also true in that every time you increase the distance by half, the sound decreases in loudness.

The audiologist can help determine the best distance from the sound source, which in most cases is where the communication partner, such as the child’s parents or the early interventionist, should be located. If one were to draw a circle around the child where the majority of speech sounds are audible, this would be referred to as the child’s “listening bubble.” All important communication should occur within the child’s listening bubble.

One can assess and determine a child’s listening bubble (optimal hearing range) by using what is referred to as the “Ling 6 Sound Test” (named after Dr. Daniel Ling, a pioneer in the area of aural habilitation). To do the test, you teach the child to imitate or provide a response to sound, specifically the speech sounds of “mm,” “ah,” “ooh,” “ee,” “shhh,” and “sssss,” which phonetically are written as /m, a, u, I, sh, s/. The sounds go from the lowest frequency speech sounds to the highest. The speech sounds move to the front of the mouth, and the last two (“shhh” and “sssss”) are unvoiced.

Once the child provides an observable response to the sounds, use the sound test to find the distances the child hears the sounds. Start at 20 inches from the child’s hearing aids, then move to 3 feet, 6 feet, and finally to 9 feet. Record the distances at which all sounds are heard. This will allow you to determine the optimal distance at which most communication happens. Ensure you are interacting at the optimal distance for the child.

What other intervention strategies can audiologists help facilitate?

Other intervention strategies to consider include maximizing the learning environments of hearing-impaired children. Especially important are the auditory and visual environments—an audiologist can help with structuring these environments. Some examples include:

Maximize the Auditory Environment

- Ensure the amplification system is working.
- Ask the audiologist to train early interventionists, educational staff, and others about the devices (hearing aids and or cochlear implants).
- Thoughtful placement to learning centers.
- Be aware of and reduce background noise.
- Make sure the child’s attention is focused on the speaker and talk naturally and clearly.

Highlight the Visual Environment

- Position children with hearing loss, so they can easily focus on activities.
- Be sure lighting is appropriate.
- Direct to auditory language information.
- Ensure that child positioning in relationship to the teacher promotes positive social relationships while enhancing learning.

Conclusion

Audiology is the study of hearing. Audiologists screen, diagnose, and manage those with hearing loss or other disorders of hearing.

Contact your local audiologists for help whenever you have a question or a need for expertise in hearing. You can engage your community audiologists by making outreach efforts either individually or as a group. Invite participation in your public health screening efforts in newborn hearing screening and early childhood hearing screenings. Work collaboratively, and always, always keep asking questions!
References


