Facts About Hearing Loss

Trying to understand a medical diagnosis of any kind can be overwhelming. Professionals will give you information and you may read about it but you still may not understand it. Do not feel bad. Remember that the professionals have had many years of schooling to understand the things they are trying to explain to you.

The parent of one deaf child stated, “When our son was diagnosed with a hearing loss, we had to learn two new languages. Sign language and the medical jargon that the professionals used to describe the daily events happening in his life. OAE, ABR, amplification, audiogram, sensorineural, bilateral, visual communication, and the list goes on and on. I felt like we needed a translator with us at every appointment.”

It is okay if you do not understand something. Ask the professional to explain it another way. You can ask them to explain it as many times as is necessary for you to feel comfortable with the new information. Remember to be patient. Understanding will come.

In the Facts About Hearing Loss section of this handbook you will find the following information:

• Description of the Ear
• Types of Hearing Loss
• Degrees of Hearing Loss & Potential Effects

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Description of the Ear

The ear is made up of three parts:

1. **Outer Ear**
2. **Middle Ear**
3. **Inner Ear**

1. **The Outer Ear**
   
   This part of the ear includes the **auricle** or pinna (**part of the ear on the outside of the head**) and the ear canal. It is also called the external ear. Sound travels through the ear canal and moves or vibrates the eardrum (**tympanic membrane**).

2. **The Middle Ear**
   
   This part of the ear is between the eardrum and the inner ear. The middle ear contains three tiny bones called the ossicles. The eardrum vibrates causing the middle ear bones to move and send the sound through the middle ear to the inner ear. There is also a tube that runs from the middle ear space to the back of the throat called the **Eustachian tube**.

3. **The Inner Ear**
   
   The inner ear includes the cochlea (**the spiraled organ of hearing**), the semi-circular canals (**the organ of balance**) and the auditory nerve (**8th cranial nerve leading from the ear to the brain**). When sound vibrations enter the cochlea from the middle ear, the hair cells send nerve impulses to the brain via the auditory nerve. Once the brain receives the nerve impulses, there is a sensation of hearing.
Types of Hearing Loss

Hearing loss can happen in any part of the ear. This includes the outer ear, middle ear and the inner ear. Hearing loss can happen in one of these places or in more than one place. Each type of hearing loss has a different name and has different possible treatments.

Conductive Hearing Loss in Children

This type of hearing loss involves the outer and/or middle ear. A conductive loss prevents sound from moving effectively through the outer and/or middle ear to the inner ear.

The simplest way to perceive conductive hearing loss is to plug your ears. Sounds that normally enter the canals are reduced.

Causes of conductive hearing loss in children are:
- Atresia – closure of the ear canal
- Malformations of the ossicles
- Middle ear infections, otitis media, or fluid in the middle ear
- Obstruction of the ear canal by ear wax or foreign objects
- Microtia – smallness of the auricle

Treatment of conductive hearing loss

Most conductive hearing losses can be treated and corrected with medication, surgery or by amplifying sound through a hearing aid. Hearing aids are very effective in compensating for a conductive hearing loss when surgical or medical treatment is not an option.

Sensorineural Hearing Loss in Children

This type of hearing loss is the most common permanent hearing loss in children and involves the inner ear. The primary cause of sensorineural hearing loss is damage or deformity of the hair cells in the cochlea. Sensorineural hearing loss may also occur when the auditory nerve does not function properly, though this is less common.

Auditory Neuropathy Spectrum Disorder (ANSD) is a type of hearing loss that causes poor transmission of nerve impulses along the auditory nerve although some of the hair cells are intact.

Treatment

Sensorineural hearing loss is generally permanent and cannot be treated by medication or corrective surgery. The most effective treatment to compensate for this type of hearing loss are hearing aids or cochlear implants.

Several factors (social, emotional or audiological) affect the decision for a child to use hearing aids, get a cochlear implant or do neither. Depending on the degree of hearing loss, hearing
aids can be very effective in providing children improved sound awareness. However, children with severe to profound hearing loss have a more difficult time understanding speech, despite improved sound awareness and may qualify for a cochlear implant. There are standard audiological considerations for choosing between aids or an implant.

**Mixed Hearing Loss**

A mixed hearing loss occurs in both the outer/middle ear and in the inner ear. It is a combination of a conductive loss and sensorineural hearing loss.

**Unilateral Hearing Loss**

A unilateral hearing loss occurs in only one ear. It can be conductive, sensorineural or mixed. Although a child with this loss has good hearing in one ear, he/she will have difficulty knowing where sound is coming from, hearing in noisy environments, and hearing on the affected side.

**Progressive Hearing Loss**

A progressive hearing loss occurs when a child loses their hearing over time. A baby may be able to hear at birth and gradually lose their hearing. It is important to note that even if a baby passes their newborn hearing screening, this does not ensure that they will always have normal hearing. In fact, gradual losses may occur anytime after birth and it is important for parents to tell their pediatrician if they have concerns about their child's hearing.

**Fluctuating Hearing Loss**

This type of hearing loss refers to hearing that changes, or fluctuates. A child may have better hearing on some days and poorer hearing on other days. Fluctuating hearing loss can be conductive or sensorineural. A common cause of fluctuating hearing loss is otitis media, or fluid in the middle ear.

**Syndromic Hearing Loss**

Sometimes an infant or young child who is deaf or hard of hearing may have other signs or symptoms as well. When multiple congenital malformations appear together, they may be described as a syndrome. This is important because if hearing loss is detected early, then hearing specialists, such as genetic professionals, may be able to test for certain syndromes that may not be physically identifiable by appearance alone. Several examples of such syndromes are:

- Usher Syndrome which is associated with loss of vision
- Jervell and Lange-Nielsen Syndrome which is associated with heart defects
- Pendreds Syndrome is associated with an enlarged thyroid and balance issues
- Waardenberg Syndrome features distinctive facial characteristics
Other common causes of sensorineural hearing loss

• Faulty development of the inner ear
• Genetic or family history of hearing loss (Connexin 26, CHARGE Syndrome)
• Damage to the inner ear or hearing nerve from illness before birth
• Rubella
• Toxoplasmosis
• Cytomegalovirus (CMV)
• Meningitis
• Lack of oxygen at birth
• Treatment with certain drugs such as streptomycin, kanamycin, garamycin,
• Quinine that reaches toxic levels
• Premature birth with NICU admission
• Damage to ear from loud noises
• Head injuries
• Rh Factors
• Measles
• Severe jaundice
Types of Hearing Tests

Working with an experienced audiologist is critical in getting valid assessment of a young child’s hearing. Tests of hearing function in children can be accomplished with a broad range of techniques available to pediatric audiologists.

Hearing testing is done to find out how well a child can hear. Usually an audiologist will do the testing. If a child does have a hearing loss the audiologist and/or otolaryngologist may do other tests to find out more specific information about:

- The **type of hearing loss**: Conductive, Sensorineural or Mixed
- The **degrees of the hearing loss**: Mild, Moderate, Severe or Profound
- The **reason for the hearing loss**:
  - The audiologist or otolaryngologist may discuss with the family the option of referral for genetic testing.
  - The otolaryngologist may complete additional lab or imaging tests.

Pediatric audiologists employ multiple tests (described below) to assess hearing sensitivity. Some procedures are better suited for a particular child, based on age, ability to participate in the testing, medical condition of the child, etc. A typical method of pediatric hearing assessment employs the “cross-check principle”. That is, the results of a single test are cross-checked by an independent test measure. The audiologist chooses and performs different tests to get the most accurate determination of a child’s hearing.

**OBJECTIVE TESTS**

**Otoacoustic Emissions Testing**

*Also known as: OAE, DPOAE, TEOAE*

**How it is done:** A small probe is placed in the child’s ear canal. A sound, generated by the testing equipment is sent to the cochlea (inner ear). If the hair cells in the cochlea are functioning normally, they generate an otoacoustic emission that is generated by the testing equipment.

**What it will show:** If an emission is present, it suggests normal cochlear function. If an emission is not present then further testing is needed. Generally, children with normal hearing or mild hearing loss will have otoacoustic emissions, present otoacoustic emissions alone are not enough to diagnose normal hearing.

**Who is it for:** This test is used for infants, for children who cannot respond to other types of hearing tests, and for children with disabilities. OAEs can be used: regardless of a client's age at testing; as another way to cross-check test results; to help diagnose auditory neuropathy and to monitor hearing when an individual has had noise exposure.
Auditory Evoked Potentials

*Different types of testing may be known as: Brainstem Audiometry Evoked Response, BSER, BAER, ABR, ASSR*

*How it is done:* This tests hearing from the level of the outer ear through the lower brainstem. This test can be done if the child is either sedated or asleep. Electrodes are attached to the child’s head and earphones are placed on the child’s ears. Sounds are played through the earphones and the electrodes measure how the child’s brain responds. This test gathers specific information about the child’s hearing at different pitches and loudness levels. This test is completely painless.

*What it will show:* This test gives a close approximation of the child’s hearing sensitivity.

*Who is it for:* This test is used for infants, for children who cannot respond to other types of hearing tests, and for children with severe disabilities.

Acoustic Immittance

*Also known as: Impedance testing, immittance testing, or tympanometry*

*May also include: acoustic reflexes*

*How it is done:* A probe is placed in your child’s ear and a signal is presented. There may be a change in pressure depending on what information the audiologist is trying to gather. The signal bounces off the eardrum and back to the probe. It only takes between 3-30 seconds per ear.

*What it will show:* Tympanometry will chart the way sound enters into the middle ear which shows how the middle ear is functioning. It can help determine if there is a hole in the ear drum or if there is fluid behind the eardrum. This is part of one test that audiologist use to assess eardrum movement, but is not always clear-cut.

*Who is it for:* Any child.

**SUBJECTIVE TESTS**

Subjective tests of hearing require a response from the child. This response can range from a widening of the eyes, to a turning of the head, to a raising of the hand. These responses provide the best indication of what sounds the child responds to and provide the data for the audiogram. Subjective tests can be done using a speaker in the test room (a soundfield system) or earphones. Tests done in the soundfield cannot separate the hearing status of the left ear from that of the right ear. Tests done with earphones provide the best ear-specific data, allowing for diagnosis of hearing loss for each ear separately.

**Behavioral Observation Audiometry (BOA)**

*How it is done:* An audiologist observes the child’s reaction to different frequencies
and loudness levels. Reactions may include a sucking reflex or head turn.

**What it will show:** The test relies heavily on parent and provider interpretation. Therefore, the test will only give a rough estimate of the degree of hearing loss. Behavioral observation audiometry must be part of a larger test battery.

**Who is it for:** This test is usually done with very young babies, especially when no other tests are available.

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**Visual Reinforced Audiometry (VRA)**

**How it is done:** The child will either sit in a chair or on the lap of an adult in the sound booth. When the sound is introduced a toy will light up. Children will learn to look at the toy in response to the sound, even when the sound is the quietest the child can hear.

**What it will show:** This test can tell the type and degree of hearing loss at different pitches.

**Who is it for:** This test is used for infants and young children with adequate neck control to turn towards a stimulus.

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**Play Audiometry**

**How it is done:** Children will drop a block or perform some other game when they hear a sound. The child is rewarded for a correct response. Some listening games may include stringing beads, building block towers, putting pegs into a peg board, putting pennies in a bank or doing a puzzle. Earphones are usually used with this test.

**What it will show:** This test can tell the type and degree of hearing loss at different pitches.

**Who is it for:** Young children, about 3 - 4 years of age.

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**Pure Tone Audiometry**

**How it is done:** Tones of different pitch and loudness levels are introduced to your child. Your child will indicate if they have heard the tone, usually by raising their hand. The tones are presented through earphones or through a bone conduction vibrator that works by sending signals through vibrations in the skull to the inner ear.

**What it will show:** This test will give information about how your child hears different pitches at different loudness levels. Earphones are used to collect individual ear information. If earphones are not used, this information will reflect the better ear.

**Who is it for:** Children (approximately 4-5 years of age) through adults.
Audiograms

What is an Audiogram?

An audiogram is a graph of the softest levels at which your child can hear sound. It is a picture of the results of a test that is done by an audiologist. Your child’s audiogram will often be used to describe his/her hearing loss.

The audiogram shows two things: **Intensity and Frequency.** Intensity, the loudness of sound, is measured in decibels (dB). Loudness levels are located along the lines on the audiogram that are drawn up and down. Intensities usually go from 0 dB to 110 dB; with 0 dB being very quiet and 110 dB being very loud. Frequency, the pitch of the sound, is measured in Hertz (Hz). The different frequencies are found along the vertical lines of the audiogram. Frequencies range from 250 Hz to 8000 Hz, although other frequencies may be tested at the audiologist's discretion. 250 Hz is a very low-pitched sound and 8000 Hz is a very high-pitched sound.

What does an audiogram look like?

As the audiologist tests your child’s hearing they will make marks using different symbols on the audiogram that represent the softest levels at which your child consistently responds. This level of sound is called the threshold. The location of each symbol will tell you how loud a certain pitch has to be for your child to hear it.

What do the symbols and drawn lines mean on the audiogram?

If your child is tested with earphones, it is called Air Conduction Testing. Because sound is presented to each individual ear, information can be gathered about hearing in each ear, separately. The symbols used to represent Air Conduction testing are an X for the left ear and an O for the right ear. Sometimes colors are used for all of the different symbols: red for right and blue for left.

If the child does not hear the sound at the loudest level of the audiometer (the machine used to test hearing), it may be indicated several different ways, with a NR (no response), a squiggly downward line or an arrow downward from the X or O.
If your child is tested using a bone conduction vibrator (a vibrating piece of plastic placed behind the ear) then brackets will be used. The symbol > is used to show the left ear results and < for right ear.

After the audiologist has information about various pitches, they will connect the symbols to make a line on the graph for each ear. This line is the configuration of the audiogram. Configurations vary due to each child’s individual hearing loss. Sometimes configurations go somewhat straight across. These are called flat hearing losses. Some configurations will angle downward, either gently or sharply. These are called sloping losses. Professionals may use configurations to describe your child’s hearing loss.

Due to the relationship of hearing and speech, an audiologist may try to get some additional information. She may try to find out the softest level at which your child can perceive speech. This is called a speech detection threshold.

The audiologist may also try to find out the softest level at which your child understands speech. This is the speech reception threshold (SRT) and is typically used in children older than 30 months. The audiologist may read a list of two syllable spondee words (such as baseball, hot dog, ice cream) to the child and have the child repeat the words or point to a picture. This is usually recorded in decibels.

Word recognition or speech discrimination testing may also be done using hearing alone or using hearing and looking at the audiologist’s face. A variety of stimuli are used with this type of test and it is usually recorded in percentages (how many words are repeated correctly).

Many of the speech sounds are made in the pitches between 250 and 5000 Hz and are spoken at a loudness of 20 to 60 dB. Sometimes an audiogram will have shading on it that resembles a banana and falls in between pitch and loudness levels. This is put on the audiogram to show where speech sounds typically occur. If the audiologist fits a hearing aid on your child, they will try to make sure that your child can hear sounds in this area.

The audiologist may also use the audiogram to chart what sounds your child can hear with hearing aids on. The softest sounds your child can hear with hearing aids is called the aided threshold. Many times the letter A will be the symbol used to represent aided thresholds. Ideally, these A’s will be within the banana lines. See the last graph in this section.
Speech Intelligibility Index (SII)

The audiologist may quantify the amount of speech your child can hear using the speech intelligibility index (SII). This number varies from 0 to 100. A child with an SII of 100 has access to all the speech cues and a child with an SII of 0 has access to no cues of speech. This number is useful for measuring the change in access to speech provided by a hearing aid. For example, a child may have an SII of 12 without their hearing aids, but with the hearing aids on, they may have an SII of 75.

Children are able to understand speech very well even with an SII of 80. That is because not every speech cue needs to be heard for a person to understand the message. However, when environments become noisy, every speech cue counts, so children with hearing loss tend to have more difficulty in noisy places.

If you want to know what your child’s SII is without hearing aids, you can plot their hearing thresholds on a “count-the-dot” audiogram. The number of dots that are below the audiogram is approximately equal to the child’s SII.

What can you find out from an Audiogram?

Although your child’s audiogram will be referred to often through the years and may even be used to describe your child’s hearing loss, it is not a predictive measure. An audiogram can be compared to a growth chart. A growth chart will give you some indication as to how big the child may become as an adult, but it is not a precise indicator of who that child will become. An audiogram can give you an idea of what that child’s usable (residual) hearing is, but is not a precise indicator of how your child will use their hearing to learn speech or how your child will process sound. Remember, your child is very unique! More than anything, a child is NOT his or her audiogram!
## Degree of Hearing Loss and Potential Effects

The following chart was created to help explain what sounds your child may and may not hear without amplification based on the degree of hearing loss. It identifies how amplification may help your child and the potential effects the hearing loss might have on your child’s ability to hear and recognize spoken conversation and environmental sounds. However, this is only a guide. Each child has unique potential. Only time will tell how your child will use his/her hearing potential and how they may or may not benefit from use of amplification.

<table>
<thead>
<tr>
<th>Degree of Loss</th>
<th>Decibels</th>
<th>Potential Effects</th>
</tr>
</thead>
<tbody>
<tr>
<td>Minimal/Slight</td>
<td>16 - 25 dB</td>
<td>A minimal loss of some sounds. May have difficulty hearing quiet or distant conversations especially in noisy environments.</td>
</tr>
<tr>
<td>Mild</td>
<td>26 - 40 dB</td>
<td>Without amplification the child can hear most conversations up close and in quiet environments, but is likely to miss parts of words. The child may appear to be “hearing when she wants to.” Amplification and lip-reading may supplement understanding of what is said. The child will require support services to learn language.</td>
</tr>
<tr>
<td>Moderate</td>
<td>41 - 55 dB</td>
<td>Without amplification, the child will have difficulty hearing spoken conversation. 50 – 100% of spoken conversation may be missed. Proper amplification and intervention should enable the child to hear and recognize all sounds. The child will require support services to learn language.</td>
</tr>
<tr>
<td>Moderately Severe</td>
<td>56 - 70 dB</td>
<td>Conversation must be very loud to be heard without amplification. Proper amplification will help the child to develop awareness of spoken language. Age of amplification, consistent use of hearing aids, and intervention are important to help the child learn to use his/her hearing. The child will require support services to learn language.</td>
</tr>
<tr>
<td>Severe</td>
<td>71 - 90 dB</td>
<td>Without amplification, the child may hear loud voices and sounds close to the ear. With early and consistent use of hearing devices, many children will be able to detect sounds such as speech. The child will require support services to learn language.</td>
</tr>
<tr>
<td>Degree of Loss</td>
<td>Decibels</td>
<td>Potential Effects</td>
</tr>
<tr>
<td>---------------</td>
<td>---------</td>
<td>------------------</td>
</tr>
<tr>
<td>Profound</td>
<td>91 dB +</td>
<td>Without amplification, the child may perceive sounds as vibrations. The child will require support services to learn language.</td>
</tr>
</tbody>
</table>

Sample: An Audiogram is produced from dB/Hz measurements and illustrates degrees of a child's hearing loss.

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