

Getting the Facts



Trying to understand a medical diagnosis of any kind can be very overwhelming. Professionals will talk with you about this information, and you may read about it. And you may still not understand it. Do not feel bad. Remember that the professionals have had many, many years of schooling to understand the things they are trying to explain to you now. It is OK if you do not understand something. Please ask the professional that you are working with 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. Be patient with the new information.

In the ***Getting the Facts*** section of this notebook you will find information that will help you better understand the ear, the types of tests used to diagnose hearing loss, the types of hearing loss, and the terminology that is often used when describing each. The terminology that is underlined in this section is also located in the ***Glossary*** found at the back of the notebook.

“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. The list goes on and on. I felt like we needed a translator with us at every appointment.”

Description of the Ear



The ear is made up of three parts:

- Outer ear
- Middle ear
- Inner ear

Outer Ear

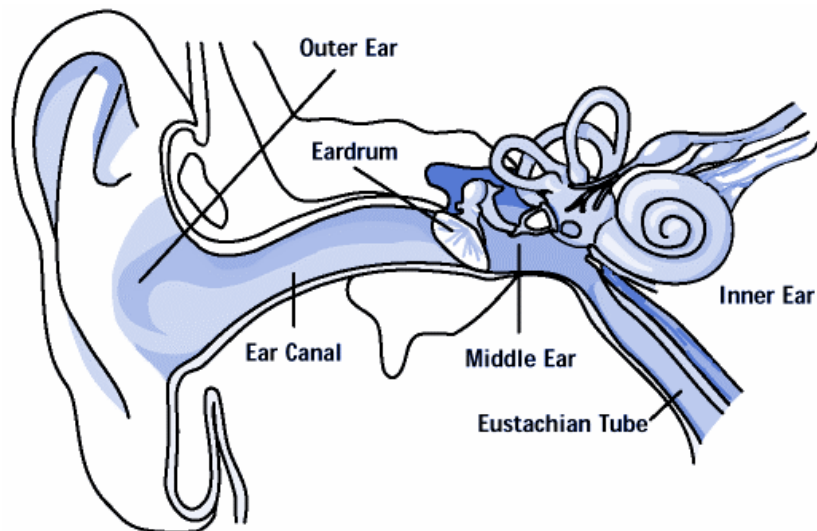
This part of the ear includes the auricle or pinna (ear on the outside of the head) and the ear canal. It is also called the external ear. Sound travels through the ear canal and moves or vibrates the eardrum (tympanic membrane).

Middle Ear

This part of the ear is between the eardrum and the inner ear. The middle ear contains three tiny bones (the ossicles). The eardrum vibrates causing the middle ear bones (ossicles) 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.

Inner Ear

The inner ear includes the cochlea (snail shaped organ), the semi-circular canal (balance mechanism), and the auditory nerve (8th cranial nerve leading from the ear to the brain). When the sound vibrations enter the inner ear from the middle ear, the cochlea sends nerve impulses to the brain through 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, the middle ear, and the inner ear. Hearing loss can happen in any one of these places or in more than one place. Each type of hearing loss has a different name and has different possible treatments or therapeutic strategies.

Conductive Hearing Loss:

This type of hearing loss occurs in the outer ear and/or in the middle ear. Something is preventing sound waves from reaching the inner ear. This blockage can be partial or complete. Conductive hearing losses don't cause total hearing loss but do cause a loss of loudness. When the sound is made louder, the sound is heard clearly.

Most conductive hearing losses can be treated with medication, surgery, or environmental change. Sometimes hearing aids or other amplification devices can help by making sound louder.

Common causes of conductive hearing loss are:

- Faulty development of the outer ear and/or middle ear
- Blockage of the ear canal (wax or foreign object)
- Damage to the ear drum
- Damage to the three bones in the middle ear
- Failure of the Eustachian tube to let air into the middle ear space
- Infection in the middle ear
- Fluid in the middle ear
- Growth of extra bony material around the three bones in the middle ear
- Cysts

Sensorineural Hearing Loss:

This type of hearing loss occurs in the inner ear, the auditory nerve or the parts of the brain that receives the nerve impulses from the auditory nerve. This type of hearing loss is sometimes called “nerve deafness.” Hearing loss due to trouble with the auditory nerve is called “auditory neuropathy” or auditory dyssynchrony syndrome.

Sensorineural hearing loss can range from mild to profound. Not only is there a loss of loudness but there is also a loss of clarity. Almost all children with this type of loss will have some degree of loudness and/or clarity left. What hearing is left is called residual hearing. This residual hearing can be amplified and the child may be able to be taught to pick up sounds and/or speech. This type of hearing loss cannot be treated by medication or surgery. It is considered permanent because nerve cells cannot grow back or be replaced.

Common causes of sensorineural hearing loss are:

- Faulty development of the inner ear
- Family history of hearing loss
- Damage to the inner ear and/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, ethacrynic acid
- Quinine that reaches toxic levels
- Premature birth with NICU admission
- Damage to ear from loud noises
- Head injuries
- High fever
- Rh factors
- Measles

Mixed Hearing Loss:

A mixed hearing loss occurs in both the outer/middle ear and in the inner ear. It is a combination of conductive 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 because a baby passes their newborn hearing screening does ensure that they will always have normal hearing. This loss can be conductive, sensorineural or mixed.

Fluctuating Hearing Loss:

This type of loss is one that changes frequently by improving or worsening. Some conductive hearing losses are fluctuating.

An example of this is hearing that worsens when a child has fluid in the middle ear, caused by an ear infection, and the hearing improves when the infection is resolved.

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 specialists, such as genetic professionals, may be able to test for certain syndromes that may not be physically identifiable by appearance alone. Two examples of such syndromes are:

- 1) Usher's Syndrome which is associated with progressive loss of vision
- 2) Jervell and Lange-Nielsen Syndrome, which is associated with heart defects.



Degrees 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 and uses that potential differently. 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.

<u>Degree of Loss</u>	<u>Decibels</u>	<u>Potential Effects</u>
Minimal Hearing Loss	16-25 dB	A minimal loss of some sounds. May have difficulty hearing quiet or distant conversations especially in noisy environments.
Mild Hearing Loss	26-40 dB	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/he wants to." Amplification and lip-reading may supplement understanding of what is said.
Moderate Hearing Loss	41-55 dB	Without amplification, the child will have difficulty hearing spoken conversation. 50-100% of spoken conversations may be missed. Proper amplification and intervention should enable the child to hear and recognize all sounds.
Moderate to Severe	56-70 dB	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, consist use of hearing aids, and intervention are important to help the child learn to use his/her hearing.

<u>Degree of Loss</u>	<u>Decibels</u>	<u>Potential Effects</u>
Severe Hearing Loss	71-90 dB	Without amplification, the child may hear loud voices and sounds close to the ear. With early and consistent use of hearing aids, many children will be able to detect sounds such as speech. Most children will use vision in addition to or in place of hearing.
Profound Hearing Loss	91 dB or	Without amplification, the child will be more aware of sounds as vibrations. The child may rely on vision rather than hearing as the primary means for communication and learning. Amplification may or may not be useful in hearing spoken conversation.
Unilateral Hearing Loss (one side)		May have difficulty hearing faint or distant spoken conversations. Usually have difficulty knowing where sounds are coming from. May have difficulty understanding spoken conversations coming from the side of the head that has the hearing loss.

Types of Hearing Tests



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 may do other tests to find out more specific information about:

- what kind of hearing loss it is
- how severe the hearing loss or the “degree” of hearing loss
- what the reason for the loss is

The audiologist and the family will discuss different ways to help the child and family to communicate. When looking for an audiologist, it is important to find one who has experience with infants and children. If you are looking for an audiologist, contact Wisconsin First Step (800-642-STEP/7837). When first contacting the audiologist, ask what experience they have had with young children.

Different hearing tests may be done depending on the age of the child and the information the audiologist is looking for. The following is a description of different tests and the ages of children they work best with.

Otoacoustic Emissions Testing

Also known as: OAE, DPOAE, TEOAE

How it is done: A small earphone is placed in the ear canal and sound is sent to the eardrum. When the inner ear hears the sound, an echo is sent back. A microphone in the earphone listens for the echo. A strong echo means the ear is hearing normally.

What it will show: It can find a hearing loss that occurs in the inner ear.

Who is it for: This test is used for infants up to 6 months of age, for children who cannot respond to other types of hearing tests, and for children with severe handicaps. It is also used for people of all ages.

Auditory Brainstem Response

Also known as: Brainstem Audiometry Evoked Response, BSER, BAER, ABR.

How it is done: This is a more thorough test than the OAE described above because it tests both the ear and the brain’s response to sound. This test can only be done if the child is either asleep or sedated. Electrodes are attached to the child’s head and tiny earphones are placed over or in the child’s ears. Sounds are given through the ear piece and the electrodes measure how the child’s brain responds. The audiologist will compare your child’s hearing response to information gathered on infants and children with normal hearing. This test gathers specific information about the child’s hearing at different pitches and loudness levels.

What it will show: This test gives an approximation of the amount of hearing. If there is a loss, the type of hearing loss can be found.

Who it is for: This test is used for infants up to 6 months of age, for children who cannot respond to other types of hearing tests, and for children with severe handicaps.

Behavioral Observation Audiometry

How it is done: A trained person in behavioral responses observes the child's reactions to different frequency and loudness that are introduced. Reactions may include a sucking reflex or a head turn.

What it will show: The test relies heavily on parent and provider interpretation. Therefore, this test will only give an approximation of the degree of hearing loss. Earphones are not used which means that information about each ear is not available.

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

Visual Reinforced Audiometry

How it is done: The child will sit either in a chair or on the lap of an adult in the sound booth. A toy that is of interest to the child is near the speaker where the sound will come from. When sound is introduced the toy will light up. Children will learn to look at the toy in response to the sound. Children naturally turn to the sound source and this process uses that tendency. Earphones may or may not be used for this test.

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, the information will reflect the better ear.

Who it is for: This test is used for children of about 6 months to about 2 years of age.

Play Audiometry

How it is done: Children learn to drop a block or perform some other task when they hear a sound. The child is rewarded for a correct response. Some listening activities may include stringing beads, building block towers, putting pegs in a peg board, putting pennies in a bank, or doing a puzzle. Earphones may or may not be used with this test.

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, the information will reflect the better ear.

Who it is for: This test is used with children older than 17 months.

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 either through earphones or through a vibrator placed behind the 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, the information will reflect the better ear.

Who it is for: This test is used with children older than 30 months.

Tympanometry

Also known as: Impedance testing, immitance testing, compliance and acoustic reflexes.

How it is done: A probe is placed in your child's ear and a signal presented. The signal can be a sound or 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 the eardrum is moving which shows how the middle ear is functioning. It determines if there is fluid in the middle ear or if the middle ear bones are working properly. Compliance testing will measure the point at which the eardrum moves best. It can determine if there is a hole in the eardrum or if the child's tubes are open. Acoustic reflex measure how loud the sound must be to cause the middle ear muscles to contract. Individuals with severe to profound hearing losses do not have these reflexes.

Who it is for: This test is used for any child where a middle ear problem is suspected.

What is an Audiogram?

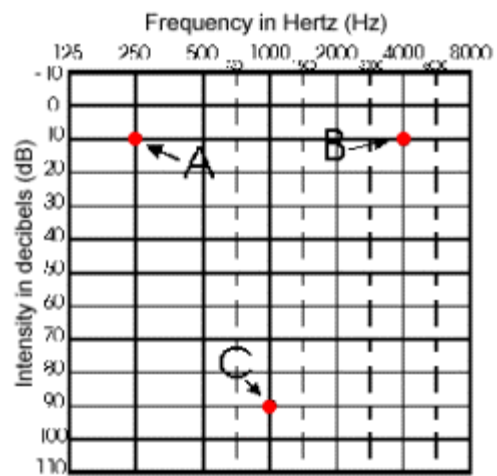


An audiogram is a graph of the softest levels at which your child can hear sound. It is a picture of the results of a test that is done by an audiologist. Your child's audiogram will often be used to describe his/her hearing loss.

The audiogram shows two things: intensity and frequency. Intensity (loudness) 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, which is another word for pitch, is measured in Hertz (Hz). The different pitches are found along the lines drawn left to right on the audiogram. Pitches range from 125 Hz to 8000 Hz. 125 Hz is a very low sound and 8000 Hz is a very high sound.

What does an audiogram look like?

Every point on an audiogram represents a different sound. For example, point A on the audiogram to the right represents a soft low-pitch sound and point B represents a soft high-pitch sound. Point C represents a loud mid-pitch sound.



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 is aware of sound. This softest level of sound awareness is called the threshold. By looking at the point where the pitch and loudness lines cross (where the mark is made), you can see what loudness levels the child can detect at various pitches.

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

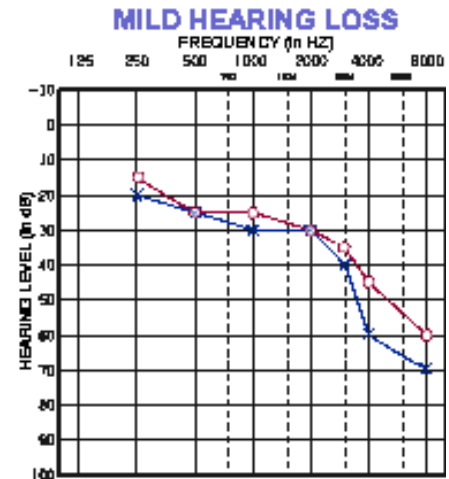
If your child is tested with earphones, it is called Air Conduction testing. Because sound is presented to each individual ear, information can be gathered about the hearing in each ear, separately. The symbols used to represent Air Conduction testing are an X for the left ear and an O for the right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

Graphs were included with permission from Boys Town National Research Hospital www.babyhearing.org

If your child is tested using a bone conduction vibrator, (a vibrating piece of plastic that is placed behind their ear rather than in it) different symbols will be used. A > is used to show the left ear results and < for the 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. Some 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.

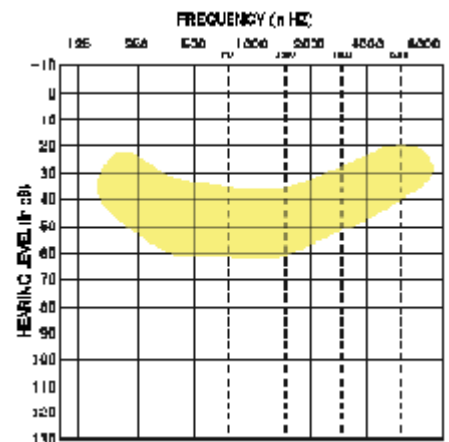


Due to the relationship of hearing and speech, an audiologist may try to get some additional information. The audiologist may try to find out the softest level at which your child can *perceive* speech. This is called the speech detection threshold (SDT).

The audiologist may also try to find out the softest level at which your child *understands* speech. The speech reception threshold (SRT) is typically used with children older than 30 months. The audiologist may read a list of two syllable spondee words (such as baseball, ice cream, and bathtub) 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 were repeated correctly).

Many of the speech sounds are made in the pitches between 250 and 5000 Hz and are spoken at a loudness level of 20 dB to 60 dB. Sometimes an audiogram will have shading on it that resembles the outline of a banana and falls in between the pitch and loudness levels mentioned above. 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 instruments on is called aided thresholds. Many times the letter A will be the symbol used to represent aided thresholds. Ideally, these A's will be within the "banana" lines.

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 hearing (residual hearing) is, but it is not a precise indicator of how your child will use their hearing to learn speech or how your child will process sound. Remember that every child is unique.