

Understanding Deafness

An introductory guide to different types of deafness and hearing tests



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THE EAR AND HOW IT WORKS

The ear has two main functions.

- It receives sound and converts it into signals that the brain can understand.
 - It helps us to balance.
- The two functions are closely related.

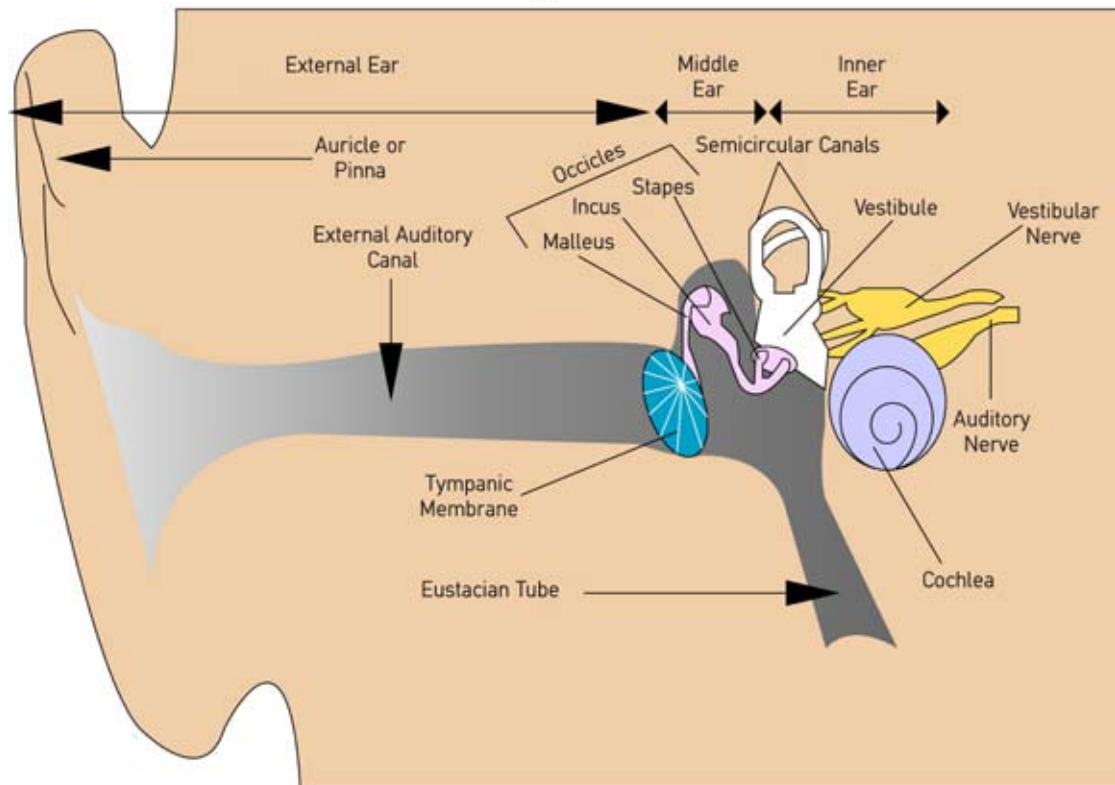
The ear

The ear is the first part of the hearing system. The pinna (the outside part of the ear) catches sound waves and directs them down the ear canal. The waves then cause the eardrum to vibrate. These vibrations are passed across the middle ear by three tiny bones: the malleus, incus and stapes (sometimes known as the hammer, anvil and stirrup, known together as the ossicles). The bones increase the strength of the vibrations before they pass through the oval window into the cochlea. The cochlea looks like a snail's shell. It is filled with fluid and contains thousands of tiny sound-sensitive cells. These cells are known as hair cells. The vibrations entering the cochlea cause the fluid and hair cells to move, much like the movement of seaweed on the seabed when waves pass over it. As the hair cells move, they create a small electrical charge or signal. The auditory nerve carries these signals to the brain where they are understood as sound. For an ear to work fully and allow us to pick up sound, all of these parts must work well. Hearing loss happens when one or more parts of the system are not working effectively.

Balance

The brain uses information from the eyes (what we see), our body (what we feel) and the inner ear to balance. The semicircular canals in the inner ear are three tubes, filled with liquid and movement-sensitive hair cells. As we move, the fluid moves. This creates signals that are sent to the brain about balance.

Diagram of a Human Ear



TYPES OF HEARING LOSS

Conductive hearing loss is when sound cannot pass efficiently through the outer and middle ear to the cochlea and auditory nerve. The most common type of conductive hearing loss in children is caused by 'glue ear'. Glue ear (or otitis media) affects about one in five children at any time. Glue ear is a build-up of fluid in the middle ear. For most children, the glue ear clears up by itself and does not need any treatment. For some children with long-term or severe glue ear, hearing aids may be provided; or the child may need surgery to insert grommets into the eardrums. Grommets are tiny plastic tubes which are inserted into the eardrum. They allow air to circulate in the middle ear and help to prevent fluid from building up.

Sensori-neural (or nerve) hearing loss is when there is a fault in the inner ear (most often because the hair cells in the cochlea are not working properly) or auditory (hearing) nerve. Sensori-neural hearing loss is permanent. Children who have a sensori-neural hearing loss can also have a conductive hearing loss such as glue ear. This is known as **mixed hearing loss**.



CAUSES OF PERMANENT HEARING LOSS

There are many reasons why a child can be born deaf or become deaf early in life. It is not always possible to identify the reason, but you may be offered further tests to try and establish the cause of your child's hearing loss.

Causes before birth (pre-natal causes)

Around half the children with hearing loss born in every year have a hearing loss because of a genetic (inherited) reason. Hearing loss can be passed down in families, even though there appears to be no family history of hearing loss. For about 70% of these deaf children, no other problems will occur. For the other 30%, the gene involved may cause other disabilities or health problems. Hearing loss can also be caused by complications during pregnancy. Infections such as rubella, cytomegalovirus (CMV), toxoplasmosis and herpes can cause a child to be born with a hearing loss. There are also a range of medicines, known as ototoxic drugs, which can damage a baby's hearing system before birth.

Causes in early childhood (post-natal causes)

Being born prematurely can increase the risk of a child having a hearing loss. Premature babies are often more prone to infections that can cause hearing loss. Severe jaundice or a lack of oxygen at some point can also cause hearing loss. Infections during early childhood, such as meningitis, measles and mumps, can be responsible for a child having a hearing loss. Occasionally, a head injury or exposure to loud noise can damage the hearing system.

HEARING TESTS

There are a variety of tests that can be used to find out how much hearing your child has. The tests used will depend on your child's age and stage of development. It is possible to test the hearing of all children from birth onwards. Screening tests are normally done first to see if it is likely that there is a hearing loss and the child needs to be referred to an audiologist. The audiologist will then perform more detailed tests to build up an accurate picture of your child's hearing. You can ask your audiologist for a copy of your child's test results. You may like to keep them in your own file at home or take them with you when you visit the audiologist or the ear, nose and throat doctor.

Objective hearing tests

Otoacoustic emissions (OAE)

The otoacoustic emission test is commonly used as part of the screening tests carried out on babies shortly after birth. It works on the principle that a healthy cochlea will produce a faint response when stimulated with sound. A small earpiece (containing a speaker and microphone) is placed in the child's ear. A clicking sound is played and if the cochlea is working properly, the earpiece will pick up the response. This is recorded on a computer and tells the tester whether the child needs to be referred for further tests. A poor response to an OAE test does not necessarily mean that a child has a hearing loss. Background noise, an unsettled baby, or fluid in the ear from the birth can all make it difficult to record the tiny response.

Auditory brainstem response (ABR)

The audiologist will place three small sensors and a set of headphones on the child's head. For an accurate result, the child must be very still and quiet throughout the test. In young babies the test can be carried out while they are sleeping. In slightly older children, a light sedative or an anaesthetic may be offered.

This test measures whether sound is being sent from the cochlea and through the auditory nerve to the brain. It can be used as a screening test (Automated Auditory Brainstem Response – AABR – where the computer judges whether a response is present at quiet levels of sound) or as a more detailed test where different levels of sound are used and the audiologist interprets the results to find the quietest level of sound being picked up by the hearing nerves. In very young children or children who are not developed enough to have behavioural hearing tests, the results of the ABR test can be used to accurately fit hearing aids if these are necessary. In older children this test may be used to confirm the results of their behavioural test.

Behavioural Tests

As your child grows older, their audiologist will get more information about your child's hearing through behavioural tests. These tests use toys and play as part of the assessment and involve your child listening for a variety of sounds as part of a game.

Visual response audiometry (VRA)

Visual response audiometry is suitable for children from six months to about two-and-a-half years. Using a machine called an audiometer, sounds of different frequencies and loudness are played through speakers. When the child hears the sound, they will turn their head when a visual 'reward' is activated, such as a toy lighting up or a puppet. The test can check the full range of hearing but does not give specific information about each ear. If your audiologist feels it is important to get information about each ear individually, this test can be done with small earphones.

Pure tone audiometry

From about the age of three, children are actively

involved in testing by using a technique known as conditioning. Younger children are shown how to move a toy (for example, putting a peg into a board) each time they hear a sound. Older children are asked to respond to sounds by saying yes or pressing a button. The sounds come through headphones, earphones placed inside the child's ear, or sometimes through a speaker (when the test is known as soundfield audiometry).

Bone-conduction

All of the tests above are described as testing using air conduction (that is, sounds passing through the ear canal and middle ear before reaching the cochlea). ABR, VRA and PTA can also be tested using bone-conduction. A small vibrating device is placed behind the child's ear. This passes sound directly to the inner ear through the bones in the head. This technique is useful for identifying whether a hearing loss is conductive or sensori-neural.

Speech discrimination tests

Speech discrimination tests check the child's ability to hear words at different listening levels. The tester asks the child to identify toys or pictures, or to repeat words spoken by themselves or from a recording. From this the tester can assess the quietest level at which the child can correctly identify the words used. This test can also be used to assess lipreading and signing skills.

Hearing tests and children with additional or complex needs

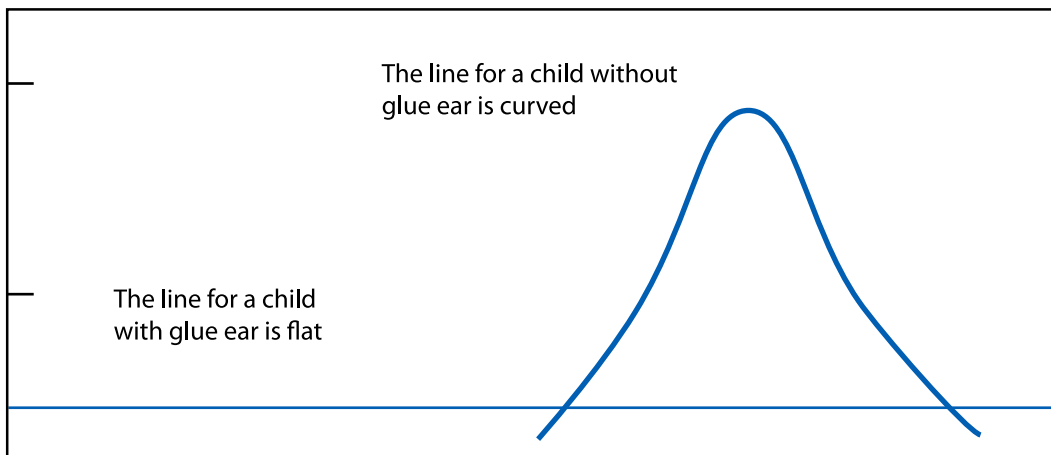
The tests used will depend on your child's age as well as their stage of development. It is more likely that several different tests will need to be done to get a

clear picture of any hearing difficulty. Objective tests (such as OAE and ABR) do not need a child to respond to a sound in order to get a result. However, the child needs to be very still and quiet throughout the test, which may mean they need a light sedative or an anaesthetic. Some children with additional needs may have to be tested using techniques that are normally used with younger children. If your child has additional needs you can ask to be assessed by a specialist with experience of testing children with complex needs.



Tympanometry

Tympanometry is used to check how well the moving parts of the middle ear are working. A small earpiece is held gently in the ear canal. A pump causes the pressure of the air in the ear canal to change. The eardrum should move freely in and out with the change in pressure. The earpiece measures this by checking the sound reflected by the eardrum. If the eardrum is not moving freely, there is likely to be some fluid or another problem with the middle ear. This build-up of fluid can cause temporary conductive hearing loss. Tympanometry is not a test of hearing, but of middle ear function.

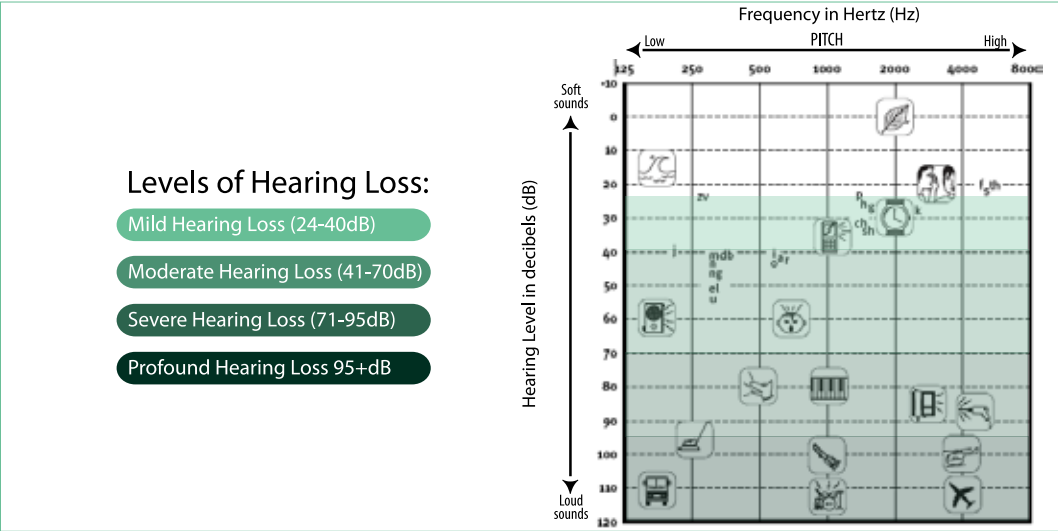


AUDIOGRAMS

Some of your child’s test results will be written on a chart like the one below, known as an audiogram. It shows you how loud a sound has to be, and at what frequency, before your child can hear it. Your child’s test results may be plotted on one chart (as below) or two charts, side by side, for each ear separately. Crosses always indicate results for the left ear, and circles for the right ear. A child may have a hearing loss in one ear (unilateral hearing loss) or both ears (bilateral hearing loss). If your child has hearing loss in both ears, the hearing loss may be similar in both ears (symmetrical hearing loss) or different in each ear (asymmetrical hearing loss).

Your child’s hearing loss may also be described as high frequency or low frequency, measured in hertz (Hz). We commonly think of frequency as the pitch of a sound. A piano keyboard runs from low-pitch on the left to high-pitch sounds on the right and the audiogram is the same. There are different levels of hearing loss. These can be described as a decibel (dB) hearing level (how loud a sound has to be for your child to hear it) or described using terms such as ‘mild’, ‘moderate’, ‘severe’ or ‘profound’. The very quietest sounds are at the top of the chart, getting louder as you look down the page.

Picture of Audiogram of common sounds



On the audiogram there are pictures of common sounds that give us an idea of loudness and frequency. There are also speech sounds drawn on the chart, and you can see that all the sounds of speech cover a range of frequencies. Try saying some of the speech sounds out loud while looking at the chart. The sounds m, b, and d are on the left-hand side and part way down the chart, meaning that they are lower

frequency and slightly louder than say f, s, and th, which are higher in frequency and much quieter. So it is important to be able to hear sounds at a quiet level, across the frequency range, to be able to hear all the sounds of speech clearly. Ask your child's audiologist to explain your child's hearing test results to you and how they will affect your child's ability to hear speech.



HEARING TESTS WITH HEARING AIDS

If your child does have a hearing loss, they may be fitted with hearing aids. VRA, soundfield audiometry and speech testing can all be used while wearing hearing aids, and the results provide some information about what your child can hear with them. When these results are written down, they are called 'aided responses'. 'Real ear measurements' will also be used to make sure the hearing aid's settings are as close as possible to your child's hearing loss.

Verification of Hearing Aid performance

Your child's hearing aids will be programmed for each child's hearing loss. Two children with identical hearing losses and identical hearing aids will have slightly different prescriptions. This is because the size of each child's ear canal will vary, and this can alter the signal (or frequency response) coming from the hearing aid. The audiologist will use a 'probe tube microphone' to take measurements in your child's ear canal to make sure that the hearing aid is set correctly. (This type of testing is not suitable for children who use bone-conductor hearing aids.)

Other methods of assessing the benefit of hearing aids

Your audiologist or teacher of deaf children will go through a questionnaire or checklist with you and your child to find out how well your child listens in different situations with the hearing aid (for example, how they are at identifying different sounds at home, working in groups at school, or using the phone). If your child is very young, your observations may be used to fine-tune the settings of the hearing aids if necessary.



MEDICAL TESTS USED TO HELP DIAGNOSE THE CAUSE OF PERMANENT HEARING LOSS

The process to find out why a child has a hearing loss is sometimes called an 'aetiological investigation'. The tests listed in this section can find the reason for a child's hearing loss in 40% to 50% of cases. For the other 50% to 60% of cases it is not possible to find out why a child is deaf. If it is not possible to find out the cause of your child's hearing loss, it may be helpful for you to know what did not cause it. Doctors sometimes may suggest tests on other parts of your child's body, like the kidneys or heart, to help identify the cause or rule out certain conditions that can be associated with hearing loss. Hearing loss can be part of a 'syndrome' (syndrome is a medical term meaning a collection of symptoms or signs that commonly appear together). It is important to know about any associated medical conditions so you can consider appropriate treatment or ways of managing the hearing loss or condition. However, these conditions are relatively rare in deaf children and extremely rare in the population as a whole. The doctor will take details of your child's medical history. This will include questions about the pregnancy, including any medication that was taken during the pregnancy and the mother's health before, during, and after the birth, and family hearing history.

Physical examination

The doctor will look at your child's head and face area and may also take some measurements. The doctor will also look at your child's neck, skin, nails, arms, legs, chest, abdomen (tummy), eyes, mouth, palate (roof of the mouth) and ears. They are looking for any minor differences or signs (for example, tiny holes in the skin known as 'pits'), that may help to diagnose the cause of the hearing loss.

Imaging

Imaging is a general term covering different ways of looking at parts inside the body (such as bones or major organs) and how they are working.

The doctor uses an MRI (magnetic resonance imaging) scan or a CT (computerised tomography) scan to look at the structure of the ear and hearing nerve as well as other parts of the body that may be associated with the hearing loss. Both types of scan are commonly used with children who have hearing loss.

An MRI scan will show soft tissues including the brain and hearing nerve. It will show if the hearing nerve has developed normally. MRI scans use magnets and radio waves to produce detailed pictures of the inside of the body. There are no known side effects associated with this type of scan. An MRI scan can be carried out on a child from birth.

A CT scan will show the bony parts of the ear including the 'ossicles' (the three tiny bones in the middle ear) and the 'cochlea' (the inner ear). A CT scan will show if the bony parts have developed normally. A CT scan uses radiation in the form of X-rays. The amount of radiation your child is exposed to depends on the number of images taken.

A further type of scan that may sometimes be used is a renal ultrasound. This is a scan that uses sound waves to create images of the kidneys. It is similar to the scans used during pregnancy. It is only likely to be used to rule out a rare syndrome or if there is a family history of kidney problems. There are no risks associated with this type of scan.

Electrocardiography (ECG)

An ECG is a recording of the rhythm and electrical activity of the heart, because there is a very rare syndrome linking severe and profound hearing loss to a heart problem. There are no risks associated with having this test.

Blood and urine tests

These tests can help doctors to identify the cause of the hearing loss.

Ophthalmology (eye test)

All children learn from what they see and hear around them. Children who are deaf rely on their eyesight even more than other children do. Up to 40% of children with sensori-neural hearing loss also have an eye problem. This may simply mean the child will need to wear glasses when they are older, but an eye test can also help to diagnose a syndrome associated with hearing loss. As babies can't tell us what they can see, special eye tests are used. It is recommended that all children diagnosed with hearing loss are referred for an eye test and have regular eye tests throughout their childhood.

Genetic counselling

Just as children inherit features such as hair colour or eye colour from their parents, sometimes hearing loss is inherited.

Genetic counselling gives families information about:

- the cause of a range of inherited conditions
- how an inherited condition might affect the child and family in the future
- how likely you are to have another child with the same condition.

A genetic test involves your child, and possibly other family members, having a blood test. The blood sample will be used to look for the gene or genes known to be involved with hearing loss. Not all the genes related to hearing loss have been identified and for most there is not yet a routine test. This means that even if the hearing loss is inherited, it may not be possible to confirm this with a genetic test at the moment. About 50% of children with permanent

hearing loss have a genetic cause. In about 30% of children with a genetic hearing loss, the hearing loss is part of a syndrome.

WHAT NEXT ?

Some or all of the tests in this section will be offered to your child, not necessarily in the order given here. Your doctor and your audiologist will give you more information which will help you decide how to proceed. Test results that may offer immediate benefit to a baby are best done at an early stage, some routine genetic testing) will not offer an immediate or direct benefit to a baby and may be delayed until a later stage. It is important that you feel comfortable with your child having the various tests.