

Understanding your child's hearing tests



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We use the term 'deaf' to refer to all types of hearing loss from mild to profound. This includes deafness in one ear or temporary hearing loss such as glue ear.

We use the term 'parent' to refer to all parents and carers of children.

Throughout this resource we use the following terms.

- We use 'your doctor' to mean the doctor in charge of your child's audiological care. This may be an audiovestibular physician, ear, nose and throat (ENT) specialist, or community paediatrician in audiology.
- 'Your family' includes your child's grandparents on both sides.

Acknowledgements

Our thanks to the audiology team at Sheffield Children's Hospital for providing facilities and expertise for the photos in this resource, and thank you to the patients and their families who agreed to demonstrate the processes shown.

1 Introduction

Finding out your child has, or might have, hearing loss can be an emotional time. You may be feeling overwhelmed with information about hearing tests and what happens next.

This resource will give you a greater understanding of the different tests your child may be offered. It explains:

- the different types and levels of deafness
- the different tests that can check a child's hearing
- about the ear and how it works
- what audiograms are (a chart showing your child's hearing test results)
- the different medical tests or investigations that are used to help diagnose the cause of permanent deafness

We are the National Deaf Children's Society, the leading charity for deaf children. We help families give the best possible support to their deaf child every step of the way, and we are here for you. For more information and support, contact our Helpline on **0808 800 8880**, email **helpline@ndcs.org.uk** or see other ways to contact us at **ndcs.org.uk/helpline**.



Find out more about childhood deafness at **ndcs.org.uk**.



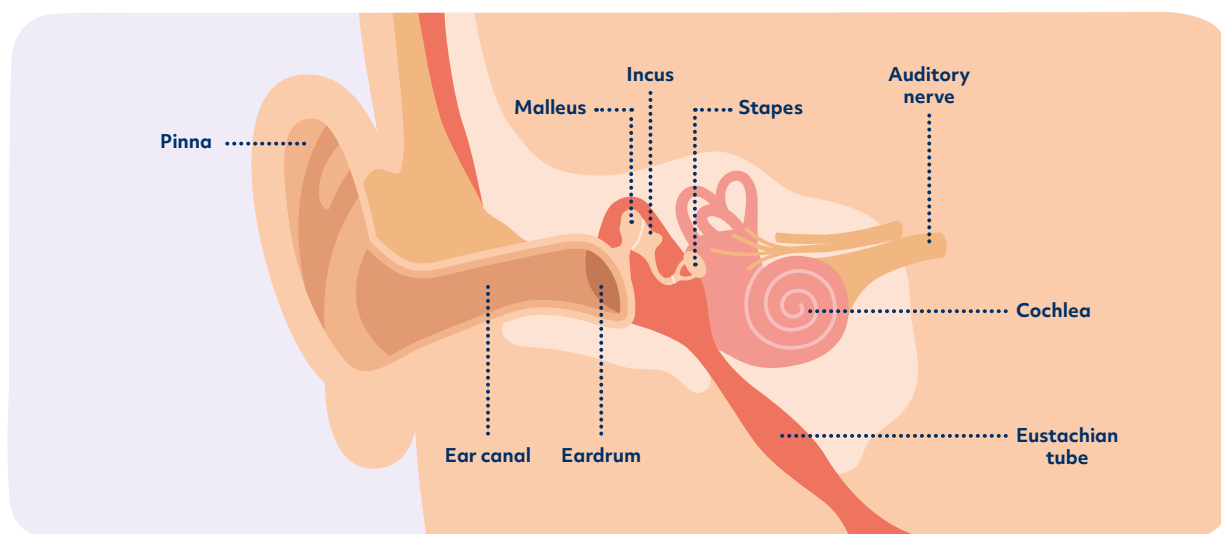
2 The ear and how it works

Before we explain the different types of hearing tests, it may be useful for you to find out more about the ear and how it works.

The ear has two main functions:

- it receives sound and changes 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 outside part of the ear (known as the pinna) collects sound waves, directing 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, or 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 and is about the size of a thumbnail. It's filled with fluid and contains thousands of tiny, sound-sensitive 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 (hearing) 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. Deafness 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 semi-circular 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.

In addition to the semi-circular canals, the inner ear also contains tiny organs called the saccule and utricle, or otoliths. The otolith organs help us understand where our bodies are in relation to gravity and acceleration, such as whether we're speeding up or slowing down.

You child's balance system may be tested if:

- they have a history of dizziness
- they have some difficulty with balance
- there has been delay in learning to sit or walk
- their deafness is one-sided (unilateral) or a different level in each ear (asymmetrical)
- their level of deafness is changing
- a cochlear implant is being considered
- the cause of the deafness is being examined in detail

The tests can:

- help to find out the cause of balance problems and/or dizziness
- give more information about the function of the inner ear
- help find the cause of your child's deafness

Even if a deaf child appears to have good balance, it can still be useful to look more closely at their balance system. Children are clever at compensating for weaknesses in the balance system and it may not be obvious until full testing is done.

Identifying balance problems can help your doctor understand more about your child and can sometimes help to identify the cause of their deafness.



For more information
about balance visit
ndcs.org.uk/balance.



3

Different types of deafness

There are four types of deafness: conductive, sensorineural, central, and mixed. It can be helpful for you to know and understand your child's type of deafness. You can ask your child's audiologist if you're not sure.

Conductive deafness is when sound can't pass efficiently through the outer and middle ear to the cochlea and auditory nerve. Conductive deafness may be temporary or permanent. In these cases, the inner ear is healthy.

Temporary causes of conductive deafness include impacted wax (when wax hardens deep in the ear canal), ear infections and glue ear. In many cases, treatment such as medication or surgery can improve hearing levels.

Glue ear is the most common type of conductive deafness in children. Also known as otitis media with effusion (OME), glue ear is caused by a build-up of fluid in the middle ear. 80% of children will have experienced at least one episode of glue ear before they start school.

For most children, glue ear clears up by itself and doesn't need any treatment.



For more information, visit ndcs.org.uk/glueear.

Permanent causes of conductive deafness include microtia (under-development of the outer ear), atresia (the absence of the ear canal) and diseases that permanently damage the eardrum or bones in the middle ear, such as otosclerosis. In these cases, it may not be possible to restore hearing fully with surgery or medication.



For more information about microtia and atresia, visit ndcs.org.uk/microtia-atresia.

Sensorineural (or nerve) deafness is when there's a problem in the inner ear (most often because the hair cells in the cochlea are not working properly) or auditory nerve. Sensorineural deafness is permanent.

The term 'sensorineural' is used to cover a wide range of different types of deafness, and audiologists might use different terminology. For example, one type of sensorineural deafness is auditory neuropathy spectrum disorder (ANSD), where sound is able to reach the acoustic nerve, but there's an issue with how sound is transmitted to the brain. Some professionals may use the term 'retrocochlear' to describe deafness caused by a problem beyond the inner ear, such as ANSD. Sensorineural deafness can have a range of causes, including genetic conditions or infections.

Central deafness is caused by a dysfunction in the central auditory processing system. It's most commonly caused by strokes affecting the auditory regions in the brain but can also be associated with other conditions.

Mixed deafness is when there's a combination of sensorineural and conductive deafness, such as when a child has glue ear and a permanent sensorineural deafness.



Find out more about causes of deafness at ndcs.org.uk/causes.



Questions to ask your doctor

- Which part of the auditory pathway is affected?
- Which parts appear to be working normally?
- Is the deafness temporary or permanent?



4 Hearing tests

You may have heard that there are different types of hearing tests and be unsure which ones will be used to test your child's hearing, and what they will show. The tests used will depend on your child's age and stage of development.

It's possible to test the hearing of all children from birth. Newborn babies are offered a hearing test (known as newborn hearing screening) to identify hearing loss at a young age. This test is normally done a few days or weeks after your baby is born, either before discharge from hospital (if that's where your child was born) or by a health professional, healthcare assistant or health visitor. If it's possible that your child has hearing loss, they will be referred to an audiologist immediately and will be given an appointment at a clinic within four weeks. The appointment will take around one to two hours, and the audiologist will carry out objective and/or behavioural tests to build up an accurate picture of your child's hearing.

Objective hearing assessments measure how well the ear and hearing nerve are working. They don't require the child to show that they have heard the sound.

Behavioural hearing assessments involve the audiologist recording the child's response to sound. For babies, the response could be a startle, look or head turn. For older children, a response might be moving a toy or saying 'yes' when they hear a sound.

You can ask your audiologist for a copy of your child's test results. You may like to take them with you when you visit the audiology department or ear, nose and throat (ENT) doctor.



Our comic 'Going to the hearing clinic' can help to prepare children for hearing tests. Visit ndcs-bookshop.mysshopify.com/products/hearing-clinic.



Objective hearing tests

Otoacoustic emissions (OAE)

The OAE test works on the principle that a healthy cochlea (inner ear) will give a faint response when stimulated with sound. The cochlea contains outer hair cells which produce this response to sound. The OAE test gives information about how well the outer hair cells are working.

A small, soft-tipped ear piece, containing a speaker and a microphone, is placed in the ear. A gentle clicking sound is played. If the cochlea is working properly, the microphone in the ear piece will pick up the response, which is shown on a computer screen. The test is very quick, painless and gives the result straightaway.

Although it's a test of how the inner ear is working, the OAE test can't give information about levels of hearing.

A poor response to an OAE test doesn't necessarily mean that a child is deaf. It can be difficult to get a response if a baby or child is unsettled at the time of the test, if the room is noisy, or if there's any fluid in the ear.

Equally, a present response does not rule out all types of hearing loss. OAE testing may not detect milder levels of hearing loss or some variations of hearing loss, such as if a child has good hearing in low frequencies but hearing loss in the high frequencies. OAE testing may not detect rarer types of hearing loss, such as those caused higher up the hearing pathway.

Audiologists typically use OAE testing as a diagnostic tool to help determine the nature of a child's deafness. It is usually interpreted in conjunction with other test results.

Automated otoacoustic emissions (AOAE) testing is a variant of the OAE test that is fully automated. It's primarily used in newborn hearing screening, which is offered to all newborn babies. In AOAE testing, the computer, rather than an audiologist, interprets the results.

If the test records strong responses from the baby's ear then they will not need any more tests.

If there isn't a clear response from one or both ears, your baby will have a second screening test called an automated auditory brainstem response (AABR). If your baby has spent more than 48 hours in a special care baby unit (SCBU) or neonatal intensive care unit (NICU) then they will also have an AABR test routinely.

As parents, you should always be asked for consent before your child has any procedure and be given the opportunity to say 'no.' If you need time to think about it, newborn hearing screening can be done in England, Scotland and Wales until a baby is three months old. In Northern Ireland, newborn hearing screening can be done until your baby is six months old. If you change your mind about having your child's hearing tested, speak to your GP or health visitor.



**Automated
otoacoustic
emissions
(AOAE) testing**

Automated auditory brainstem response (AABR)

The AABR measures whether sound is being sent from the cochlea and through the auditory nerve to the brain. A computer judges whether there's a response to quiet sounds.

The screener will place three small sensors on your baby's head and neck and a set of soft headphones on their ears. For an accurate result, the baby must be very still and quiet throughout the test, so it's usually carried out while they are sleeping. The AABR test takes 5 to 15 minutes, is painless and isn't harmful to your baby.

A present AABR usually rules out significant hearing losses but may not detect milder levels of hearing loss or some configurations of hearing loss, such as if a baby has good hearing in low frequencies but hearing loss in the high frequencies. The AABR may not detect rarer types of hearing loss.

Most babies are considered to have passed their newborn hearing screening if they have passed either an AOA or an AABR. If they do not pass either test, they will be referred to audiology.

The guidelines are slightly different for babies who have spent more than 48 hours in a SCBU or NICU. If a baby who has spent more than 48 hours in a SCBU or NICU passes the AABR but does not pass the AOA, they should be seen again at 8 months old. If they pass the AOA but not the AABR, they should be referred to audiology for immediate assessment.

Auditory brainstem response (ABR)

If deafness is suspected in a child, or your baby's newborn hearing screening didn't give a clear result, they will be referred to audiology for more tests. The audiologist will carry out a more detailed version of the AABR test, known as a diagnostic ABR. Diagnostic ABRs usually involve either three or four sensors, depending on equipment, and may use either headphones or a small, soft-tipped earpiece appropriate for a baby's ear canal. The audiologist can test the hearing at different frequencies and at different volumes and will interpret the results to find the quietest level of sound being picked up by the hearing nerves. A diagnostic ABR takes about 30 to 90 minutes and may take multiple appointments, depending on the results. Each appointment will normally take 1.5 to 2 hours.

If the diagnostic ABR test shows that your child has a permanent hearing loss, the audiologist will offer them hearing aids. If the hearing loss is caused by glue ear, the audiologist will probably follow up with them later to see if the glue ear clears up.

In babies and children who aren't developed enough for behavioural hearing tests, the results of the diagnostic ABR test can be used to accurately fit their hearing aids. In older children, ABR may be used to confirm the results of their behavioural test. For an accurate result, the child must be very still and quiet throughout the test, so if they no longer sleep during the day they might be offered a light sedative or an anaesthetic.



For more information about the different types of hearing aids available, read our resource 'Hearing aids: Information for families.' Visit ndcs-bookshop.myshopify.com/products/hearing-aids-guide.

Auditory steady state response (ASSR)

ASSR testing works similarly to ABR testing, in that it looks at how sounds are transmitted to the brain via the auditory nerve. It uses the same equipment as ABR testing (sensors and earphones), but a key difference is that both ears and multiple frequencies can be tested at the same time. A computer uses statistical analysis to determine whether there are responses at given levels.

This is a relatively new technology and is not usually used by itself, but it can be useful in addition to the ABR results.

There are national guidelines for ABR, ASSR, OAE, CERA, tympanometry, PTA and VRA to ensure the tests are done to a high standard. The results may also go through a peer review process (where another audiologist from the same or different hospital reviews the results) to ensure there is agreement in the test analysis and interpretation.

Cortical evoked response audiometry (CERA)

Cortical evoked response audiometry (CERA) is similar to an ABR test. While ABR looks at how sounds are transmitted to the brain via the auditory nerves, CERA looks at the part of the brain that recognises sound, the auditory cortex. Sensors are placed on the head while sounds are played, to measure the brain's responses to sound. CERA can be performed while a child is awake, typically while sat reading or watching a TV programme. The test can be useful for children where behavioural assessment is more challenging. It is not always suitable for very young children as their neural responses may not have fully matured.





"George failed his newborn hearing test, so we were referred for an outpatient appointment. A few days after leaving hospital, we took George to his appointment feeling somewhat frustrated. We just wanted to enjoy our newborn bubble and didn't need this 'pointless' appointment as our baby could hear perfectly well. He failed the hearing test again and we were referred to our local hospital for a different type of test. I remember saying to the woman, "Do we have to have this testing? I'd know if my baby was deaf." We were given the same line... that it's probably fluid, but it's better to have early intervention if there is a problem.

We had to wait a few weeks for the next appointment, and during this time we were constantly watching for any reaction to sounds, as the reality set in that maybe our baby did have a hearing loss. We noticed that his left ear seemed to be worse than his right, but we were still confident that he was responding to sounds.

At the appointment, they wired George up and began the testing. They went through different frequencies etc, giving us the result each time... "no response". Our hearts shattered. At the end, the audiologist told us he has a severe to profound bilateral sensorineural hearing loss."

- Louise



Questions to ask your audiologist

- Has this test been performed in accordance with national standards?
- Will the test results go through a peer review process?
- What happens if the peer reviewer disagrees with the test analysis?



Behavioural tests

As your child develops, their audiologist will assess their hearing using behavioural tests. These tests use toys and play as part of the assessment and involve your child listening for different sounds as part of a game. Most children will have had objective tests at a younger age before having behavioural tests. However, some children may not have been referred with a suspected hearing loss until they were older, or may not have been screened at birth.

Visual reinforcement or response audiometry (VRA)

Visual reinforcement audiometry is suitable for children from 6 months to about 2 and a half years. During the test, your child can sit on your lap or a chair. Using a machine called an audiometer, sounds of different frequencies and loudness are then played through speakers. When the child hears the sound, they will turn their head to see a visual 'reward', such as a toy lighting up or a puppet. The test can check the full range of hearing but doesn't give specific information about each ear. If your audiologist feels it's important to get information about each ear individually, this test can be done with small earphones.



Visual reinforcement audiometry (VRA) testing



"Unlike auditory brainstem response (ABR) tests, the behavioural tests aimed to determine Lucas's conscious response to sounds by observing him turning his head when a sound was played into his ears from the audiologist's computer. He seemed to enjoy the tests and happily sat on my lap the whole time. The tests confirmed that Lucas's hearing was 80-100dB across the four main speech frequencies which put him in criteria for cochlear implantation."

- Tess



Pure tone audiometry (PTA)

From about the age of three, children are actively involved in testing by using a technique called conditioning. Younger children are shown how to move a toy (for example, putting a peg into a board) each time they hear a sound – you may hear this referred to as 'play audiometry' or 'performance testing'. Older children are asked to respond to sounds by saying "yes" or pressing a button. The sounds come through headphones, earphones, or sometimes through a speaker (when the test is known as soundfield audiometry). By changing the level of the sound, the tester can work out the quietest sounds your child can hear.

In both PTA and VRA, the audiologist will try to condition your child in different ways if one way is not working. For VRA, the audiologist might switch up the rewards or the type of sound to make it more interesting for your baby. During play audiometry, the audiologist can change the game, introduce competition, or try other things to make it fun. Your audiologist should try different things and communicate with you to find the best way to engage your child in the test.

PTA, visual reinforcement audiometry (VRA), otoacoustic emissions (OAE) and auditory brainstem response (ABR) testing all use air conduction – sounds passing through the ear canal and middle ear before reaching the cochlea. ABR, VRA and PTA can also be tested using bone conduction.



Pure tone audiometry (PTA) testing



"At audiology, two lovely doctors initially conducted an inner ear pressure test to see if there was congestion. Having seen nothing visibly blocking the ear, the test confirmed that there was some congestion in the inner ear. They then did a play audiometry test to check Robin's hearing at different volumes and frequencies.

The result confirmed that her hearing levels had dropped. It was a relief to know that what we thought we'd noticed was real."

- Emma



Bone conduction testing

Sounds are heard both through the ear canal (air conduction) and through the skull bone (bone conduction). Bone conduction testing is performed in a similar way to air conduction testing. A small vibrating device is placed behind the child's ear. This allows sound vibrations to pass directly to the inner ear through the bones in the head, bypassing the outer and middle ear. This technique is useful for identifying whether a hearing loss is conductive or sensorineural.

For a child who has conductive deafness, the results from bone conduction testing will be much better than air conduction results. Sound can get to the inner ear more easily through the skull bone by bypassing any blockage or other problem with the outer or middle ear.

Bone conduction and air conduction results are similar in a child with sensorineural deafness because the inner ear detects sound at the same level whether through the ear or through the bone.

Bone conduction testing can sometimes be used as a conditioning tool. If a child is not responding to any sounds, loud sounds are played through bone conduction to see if the child responds to the vibrations in a VRA test or through play techniques. If a child responds well to the vibrations, it means they are able to take part in the task. If the child then doesn't respond to sound, it can suggest difficulty hearing the sounds. If a child does not respond well to vibrations, it can be an indication they are not ready for the test.



Bone conduction testing



Questions to ask

If the audiologist says your child isn't ready for this hearing test, you could ask:

- What have you tried to condition them?
- Have you tried a vibrotactile stimulus (a vibration which the child can feel)?

Speech discrimination tests

Speech discrimination tests check how well the child can hear words at different listening levels. Depending on the child's age and stage of development, the tester will ask the child to name a toy or picture, or to copy 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 lip-reading and signing skills.

An example of a speech discrimination test for younger children is the McCormick Toy Test. This test can be used for children aged around two years and over. It's made up of 14 paired words in the form of toys, which a child will easily recognise (see below list). Each pair contains some similar speech sounds. This means that if the child is not able to hear the full range of speech sounds they may choose the other toy in the matched pair by mistake because they missed the beginning or end sound of the word. The test can therefore provide extra information to other hearing tests.

- tree – key
- shoe – spoon
- cow – house
- plane – plate
- horse – fork
- duck – cup
- man – lamb

A different version of this test is available for children with English as an additional language (EAL), which uses slightly different language pairs. This test is called the EAL Toy Test.

Before the test starts, the tester asks the child to identify each toy. All the toys will be on the table, but any toy that isn't well known to the child is taken away (along with the other toy in the pair) and isn't included in the test. The test can be carried out with and without lip-reading, and the words are spoken at conversational level. The child is taught to point to each toy when asked "show me the ...", "where is the ...".

The tester then removes any visual clues by covering their nose and mouth with a small screen and repeats the test, lowering their voice until the child can correctly identify four out of five toys. The level of the tester's voice when the child got four out of five correct answers is then measured and recorded.

Sometimes speech tests are performed using an automated system where the audiologist can select difference voices (male/female) or add background noise to see how well a child can discriminate speech in noisy environments.



**Speech discrimination testing
– the McCormick Toy Test**

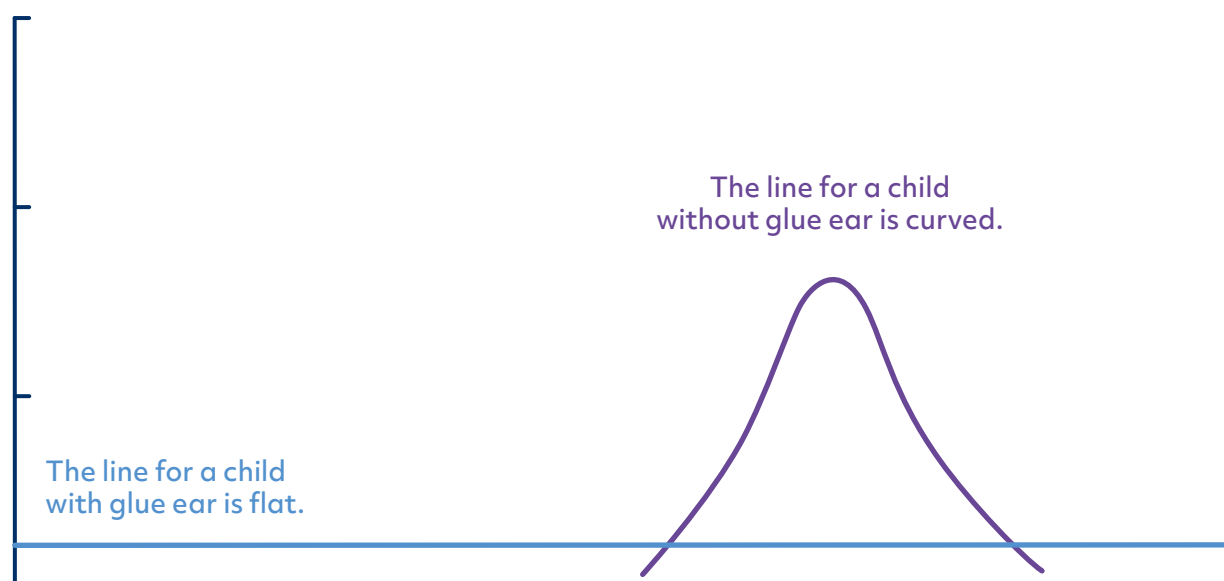
Tympanometry

Tympanometry is not a test of hearing. It's used to check how well the moving parts of the middle ear are working.

A small ear piece is placed gently in the ear. 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 isn't moving freely, there's likely to be some fluid or another problem with the middle ear. This build-up of fluid is usually glue ear (also known as otitis media with effusion – OME). Glue ear can cause temporary conductive deafness. Younger children will usually have this test as part of their hearing assessment because glue ear is common in the early years.



Tympanometry testing



Tympanogram – a chart that shows how well the middle ear is functioning



For more information, visit ndcs.org.uk/glueear.

Your audiologist might use a new technology called wideband tympanometry, which measures middle ear function at several different frequencies. Wideband tympanometry can be useful in identifying various middle ear conditions.

Acoustic reflex testing

The ear has a natural defence mechanism against very loud sounds. Tiny muscles (the stapedius and tensor tympani) reflexively stiffen the eardrum, reducing its ability to vibrate. This action helps protect the inner ear from potential damage from loud noises.

Acoustic reflex testing checks whether these muscles are working effectively. It is usually done using the same equipment as tympanometry. A series of loud sounds are played into the ear canal while it's still pressurised to see if the muscles react.

Acoustic reflexes can provide information about the middle ear, inner ear and neural pathways related to hearing.



Some children with deafness also have problems with their balance. Your child may be offered vestibular (balance) function tests. Find out more at ndcs.org.uk/balance.

Hearing tests and children with additional complex needs

If your child has additional complex needs, the tests used will depend on their age as well as their stage of development. It should be possible to test the hearing of any child, whatever their stage of development, but it's more likely that several different tests will need to be done to get a clear picture of your child's hearing level. Objective tests (such as otoacoustic emissions and auditory brainstem response) don't need a child to respond to a sound 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.

Your child may be tested using techniques often used with younger children. If your local audiology service is not confident about testing your child, they will refer you to another hospital, or you can ask to be referred to a centre with more experience of testing children with additional complex needs.



"I became better at saying, "We need a break. Let's rearrange the appointment to a later date." It's so important when you have more than one child with additional needs to look after yourself and recognise that it's OK to take a step back when needed."

- Jenni



If you would like further information on asking for a second opinion, contact our Helpline at ndcs.org.uk/helpline.

Checklist: Making appointments easier for children with additional needs

- ✓ Before an appointment, contact your audiology team to explain your child's needs.
- ✓ Tell the audiology team about any pain issues, health problems, needs and communication difficulties affecting your child. What's the best time of day for your child?
- ✓ Talk about what may help to reduce anxiety. Does the service have a written or digital social story that explains clearly what will happen, including pictures?
- ✓ Is it possible to arrange an introductory visit without examination or treatment to familiarise your child with the environment?
- ✓ Will all staff involved be aware of your child's needs, including reception staff?
- ✓ Is it possible to book a double appointment to allow extra time?
- ✓ Could the service reduce waiting time by booking an appointment at the beginning of the day or at a less busy time?
- ✓ Can the sensory environment be altered? For example, can bright lights be turned down or background noise reduced?
- ✓ How can the test procedure be adapted? For example, if your child has a visual impairment, could bright lights, high contrast or tactile rewards help to get their attention?
- ✓ Can the service delay any procedures that aren't urgent during the first visit?
- ✓ Can you bring any toys from home?



For more information about supporting deaf children with additional needs, visit ndcs.org.uk/additional-needs.

5 Audiograms

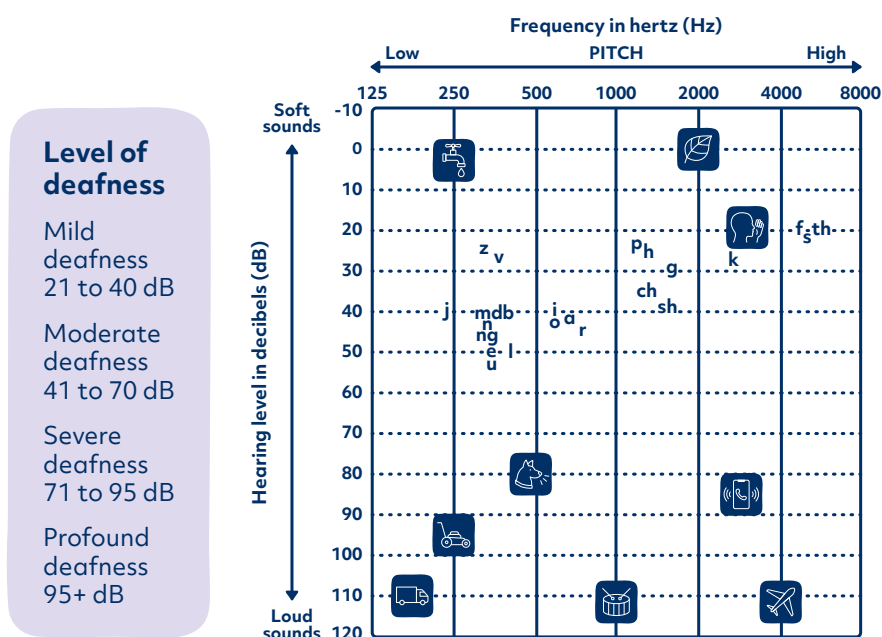
Some of your child's test results will be written on a chart like the one below, known as an audiogram. It shows you how loud a sound has to be, and at what frequency (pitch), before your child can hear it.

Your child's test results may be plotted on one chart (as below) or two separate charts (one for each ear). Crosses always show results for the left ear, and circles for the right ear. Your child may be deaf in one ear (unilateral deafness) or both ears (bilateral deafness). If your child is deaf in both ears, the deafness may be similar in both ears (symmetrical deafness) or different in each ear (asymmetrical deafness).

Soundfield results where either ear could be picking up the sound are usually denoted by a 'B', but sometimes alternative symbols are used. If it isn't clear which ear is being referred to, ask your audiologist to clarify. You can also ask whether the result is 'threshold' (the quietest sound your child can hear) or 'suprathreshold' (likely to be above the level of the quietest sound your child can hear).

Your child's deafness may also be described as high frequency or low frequency, measured in hertz (Hz). We often think of frequency as the pitch of a sound. A piano keyboard runs from low-pitch sounds on the left to high-pitch sounds on the right, and the audiogram is the same.

There are different levels of deafness. These can be described as a decibel hearing level (dBHL) (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. With typical hearing, the quietest sounds people can hear are from 0 to 20dB.



Visual representation of the loudness and pitch of a range of everyday sounds.

On the audiogram on the previous page, there are pictures of common sounds that give us an idea of loudness and frequency. There are also speech sounds from English 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. The vowels also lie in the lower frequency range. Consonants tend to carry the meaning of the words and help with understanding the sound.

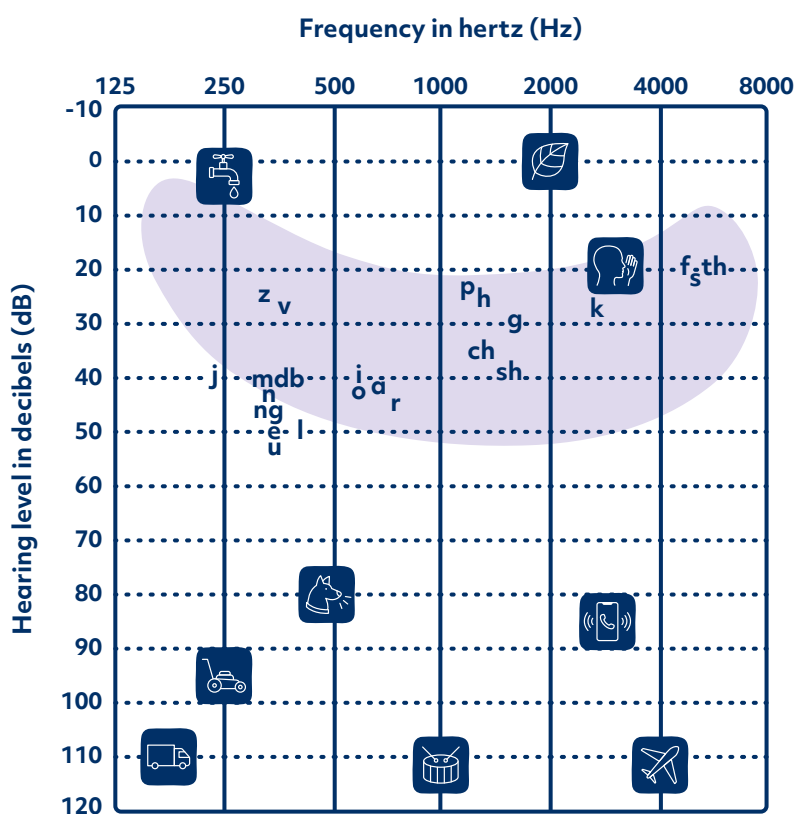
Try writing a short sentence using only the vowels of the words and then repeat the same sentence using consonants only. It's far easier to guess the words with the consonants than the words with only the vowels.

For example, compare:

- _i_ a _ _i_
- f_sh _nd ch_ps.

Which phrase is easier to guess?

Children with a hearing loss in the high frequencies mishear those vital consonants which carry the meaning of the word, making it more difficult to understand, particularly in background noise. It's 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.



Audiogram showing a speech curve or 'speech banana'

When the speech sounds are plotted on an audiogram, they tend to fall within an area of frequencies forming a shape similar to that of a banana, hence the name 'speech banana' (also known as a speech curve). Most of the letters of the alphabet and letter combinations (such as 'ng', 'ch', 'sh' and 'th') lie within the speech banana. By looking at your child's audiogram, you can see which of the speech sounds your child can hear. Not being able to hear in those frequencies can affect a child's spoken language development.

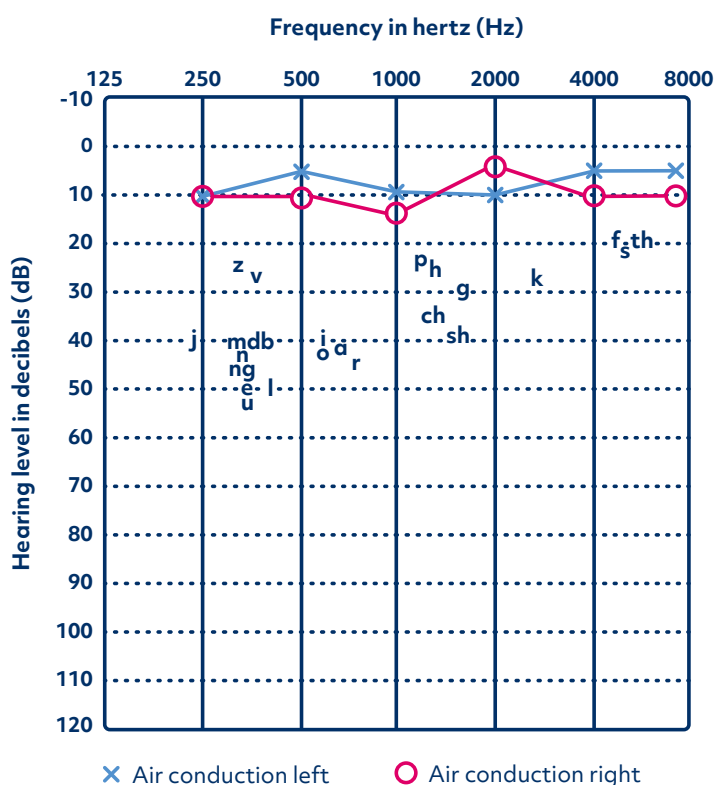
Any sounds below the circles/crosses on the audiogram will be audible to your child. However, the speech banana can move depending on whether the speaker is male or female, or loud or quiet. For example, a child with low-frequency hearing loss may find it easier to hear women than men.

Ask the audiologist to explain your child's hearing test results to you and how they will affect your child's ability to hear speech.

Some examples of different audiogram results

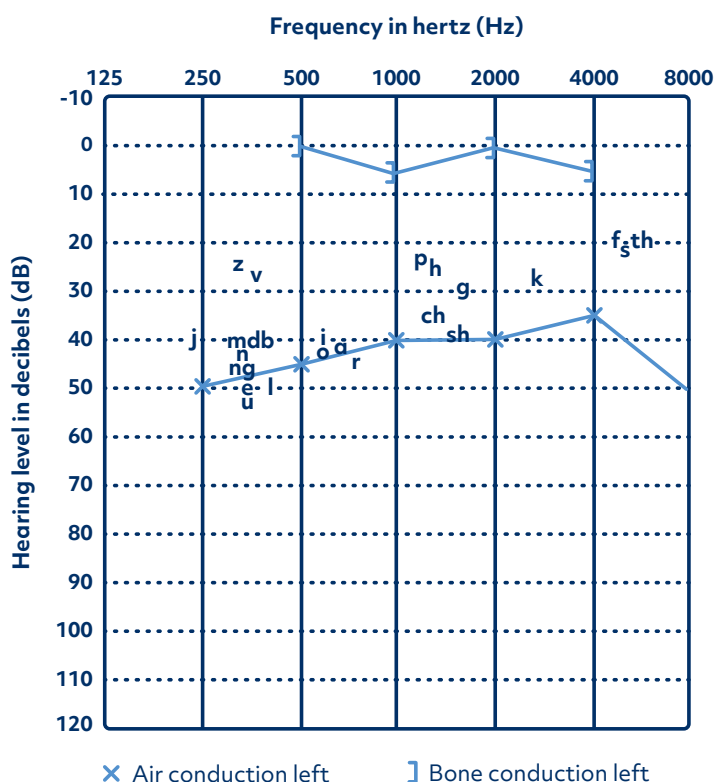
Typical range of hearing

This audiogram shows the level of hearing and range of frequencies for a person with typical hearing levels. The person can hear everything below the circles and crosses. This person can hear all the speech sounds.



Conductive deafness in the left ear

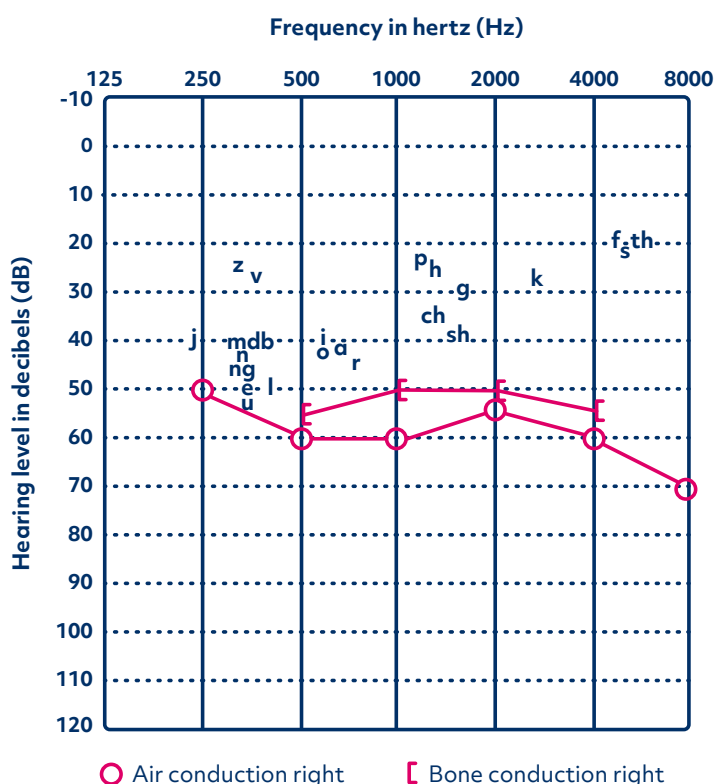
This audiogram shows a typical conductive deafness in a child's left ear. There are two lines – one shows the result of air conduction tests (with headphones or earphones in the ears) marked by crosses, and the other shows bone conduction results marked by square brackets ([]). The bone conduction test shows that the inner ear is receiving the signal clearly, but the air conduction test shows that the sound is being blocked by fluid or another obstruction in the outer or middle ear. This child may have a temporary conductive hearing loss as a result of glue ear or a permanent conductive deafness.



Sensorineural deafness in the right ear

This audiogram shows a sensorineural deafness in the right ear. You can see that both the air and bone conduction tests give similar results.

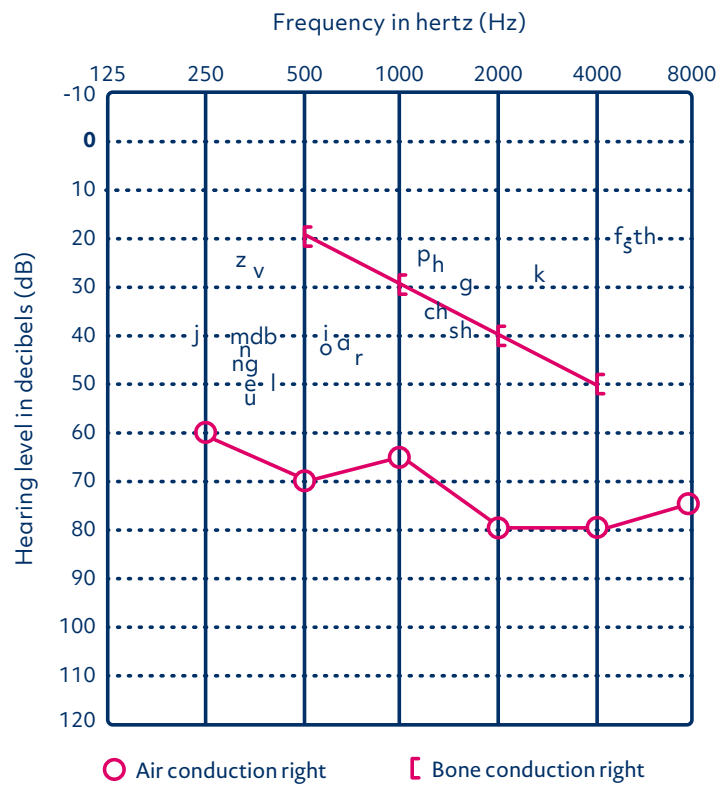
Most speech sounds lie above the air conduction line, suggesting that the child will have significant difficulty hearing speech in their right ear.



Mixed deafness in the right ear

The last audiogram gives an example of mixed deafness in the right ear. Both the bone conduction and air conduction tests show that there is a hearing loss. Because the results are very different, this child has more than one cause of deafness.

This child will hear very little, if any, speech sounds in the right ear.



6 Hearing tests with hearing aids

If your child has a hearing loss, they may be fitted with hearing aids. Visual reinforcement audiometry, soundfield audiometry and speech testing can all be performed while your child wears hearing aids, and the results will 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.

Real ear measurements (REM)

Your child's hearing aids will be programmed for their individual 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 be different, and this can alter the signal (or frequency response) coming from the hearing aid. The audiologist will use a probe 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 conduction hearing aids.

Audiologists use speech signals to check the output from the hearing aid and ensure it matches the child's prescription.

For very young children, real ear measurements are not practical as they require a child to sit still in front of a speaker for around 15 minutes. For babies and young children, audiologists measure a 'real-ear-to-coupler difference' by taking a quick measurement of the frequency response in the ear canal and then comparing that with a response taken in a test cavity/artificial ear canal, so the hearing aid can then be programmed using the test cavity.

Other ways to assess the benefit of hearing aids

Your audiologist or Teacher of the Deaf (ToD) will go through a questionnaire or checklist with you and your child to find out how well they listen in different situations with their hearing aid (for example, how well they can identify different sounds at home, work in groups at school, or use the phone). If your child is very young, your observations using 'Success from the start' may be used. The results of these can be used to fine-tune the settings of their hearing aids if necessary.



Download 'Success from the start' at
ndcs.org.uk/successfromthestart.

7 Causes of permanent deafness

There are many reasons why a child can be born deaf or become deaf early in life, and you or your doctor may feel it's important to know the cause of deafness. It's not always possible to identify the cause, but you may be offered tests to try to find out. It's very unlikely you could have done anything differently to change your child's hearing. Finding out what caused your child's deafness can be an emotional time, and you may want to get support by contacting our Helpline.

Causes before birth (pre-natal causes)

Around half the deaf children born in the UK every year are deaf because of a genetic (inherited) reason. Deafness can be passed down in families, even though there appears to be no family history of deafness. About 70% of these deaf children won't experience any other problems. For the other 30%, the gene involved may cause other disabilities or health problems. Further tests can be done to find out if this is the case.

Deafness can also be caused by complications during pregnancy. Infections such as rubella, cytomegalovirus (CMV), toxoplasmosis and herpes can cause a child to be born deaf.

Causes in early childhood (post-natal causes)

Being born prematurely, having severe jaundice or a lack of oxygen at the time of birth can all increase the risk of a child being born deaf or becoming deaf early on.

Premature babies are often more prone to infections, and some medications used to treat them are known to be ototoxic (damaging to the ear). Infections during early childhood, such as meningitis, measles and mumps, can be responsible for a child becoming deaf.

Occasionally, a head injury or exposure to loud noise can damage the hearing system.



Find out more about causes of deafness at ndcs.org.uk/causes.

8

Medical tests used to help identify the cause of permanent deafness

The process to find out why a child is deaf is sometimes called an 'aetiological investigation'.

The tests listed in this section can find the reason for a child's deafness in 40% to 50% of cases. For the other 50% to 60% of cases it's not possible to find out why a child is deaf. Although this can be frustrating, it may be helpful for you to know what didn't cause it.

Doctors may sometimes suggest tests on other parts of your child's body, like the kidneys or heart, to help find out the cause or rule out conditions that can be associated with deafness. Deafness can be part of a syndrome, meaning a collection of symptoms or signs that commonly appear together. Although medical conditions associated with deafness are quite rare, it's important to get this checked.

As with many services provided by the NHS, you may be asked to give your written permission first for some tests.

What happens when you see the doctor?

The doctor will take details of your child's medical history. This will include questions about the pregnancy and the birth, including any medication that was taken during the pregnancy and the mother's health before, during, and after the birth.

The doctor will ask you about your child's immunisations (routine baby jabs). In toddlers and older children, the doctor will ask about your child's development (including speech and language and milestones such as when your child was sitting or walking). The doctor may also ask whether your child has:

- had meningitis, mumps, measles or other illnesses
- been exposed to loud noises
- taken any prescribed medication
- suffered any head injuries
- had any ear infections
- had sight problems
- had balance problems

All of the above are important for the doctor to know when investigating the cause of deafness.

The doctor will also ask about the hearing of other family members, on both sides of the family.

Physical examination

The doctor will look at your child's head and face and may 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 that may help to identify the cause of your child's deafness. The doctor will assess your child's development in relation to the expected ages and developmental stages. See your baby's personal child health record – the red book – for further information on this.

The doctor may ask close family members to have a hearing test, known as pure tone audiometry.

Imaging

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

The doctor uses an MRI scan or a CT scan to look at the structures of the ear and hearing nerve. Both types of scan are commonly used with children who have a 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 – they are painless and safe. An MRI scan can be carried out on a child from birth, but you should be given a choice of when the scan is done. If it's important for your child to have the scan, the doctor will explain this to you.

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. The scan involves exposing your child to radiation in the form of X-rays. The level of radiation used is kept as low as possible to prevent damage to body cells, and the amount of radiation your child is exposed to depends on the number of images taken. It's generally accepted that there's little risk to health from one scan, but with repeated tests there's a risk that the radiation may damage body cells. The earlier in life your child is exposed to radiation, the greater the risk. You and the doctor may prefer to wait until your child is a little older. Your child's doctor will discuss this with you and answer any questions you may have.

Your child will need to be absolutely still for the time it takes to do a CT or MRI scan. Very young babies may be able to have these scans while they are asleep. Each hospital will have its own procedure, and your doctor will explain this to you. Children aged over three months will normally be given something to help them sleep. This may be a light sedative or a short general anaesthetic.

For children aged over two, you may be offered a stronger sedative or a general anaesthetic. Although modern anaesthetics are very safe, there are small risks associated with having an anaesthetic, and your doctor should explain these to you. The risks of having an anaesthetic reduce as the child gets older. Usually, children over the age of five can lie still for the scan without needing a sedative or anaesthetic.

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's similar to the scans used during pregnancy. It's only likely to be used to rule out a rare syndrome or if there's a family history of kidney problems. There are no risks associated with this type of scan.

Your doctor may advise that one or more of the above scans are done at an early stage, for example, if:

- your child has had meningitis
- their hearing loss is progressing or changing over time
- there are characteristic features that may suggest your child's deafness could be part of a syndrome

In these examples, your doctor will need to look closely at the structure of the cochlea (inner ear), balance organs and hearing nerve to be able to give you advice on possible management or treatment options.

Electrocardiography (ECG)

An ECG is a recording of the rhythm and electrical activity of the heart. There's a very rare syndrome (Jervell and Lange Nielsen syndrome) linking severe and profound hearing loss to a heart problem. If this heart problem is found, it can be treated.

Depending on your child's and your family's medical history, your doctor may advise an ECG for a small number of children with severe to profound deafness to help rule out this condition. There are no risks associated with having this test.



For a list of the most common syndromes associated with hearing loss, visit ndcs.org.uk/causes.

Blood and urine tests

Depending on your child's and your family's medical history, the doctor may ask for one or more routine blood and urine tests. These tests can help doctors to identify the cause of the deafness.

An example of a blood or urine test your child may be offered is an infection screen. An infection screen is used to look for certain infections that sometimes result in deafness. Some of these tests will give useful results only if they're carried out in the weeks or months shortly after your child's birth. When the child is older, an infection screen may give a negative result and rule out an infection being the cause of the deafness. However, a positive result later in life may not give useful information as your doctor couldn't prove that the infection was there at birth and so caused the deafness.



There are several infections that can cause deafness in babies if the mother contracts them when pregnant. The most common infection causing babies to be born deaf is cytomegalovirus (CMV). CMV is very common in the general population and doesn't normally cause any illness. Pregnant women are therefore unlikely to know if they have caught the virus, but it can affect the baby if the mother catches it for the first time at this stage. CMV infection in an unborn baby is called congenital CMV (cCMV). 'Congenital' means from birth.

About one baby in every 150 is born with CMV, and some of these babies can be affected by it. cCMV can cause deafness or, very occasionally, it can affect a child's development. It causes about 10 to 20% of deafness in children in the UK.

Doctors can test for CMV in the first few weeks after birth, or, for older children, look for signs of the CMV infection in the heel-prick blood test cards which are stored after birth. Deafness following infection can sometimes progress over time.

If a young baby is found to have the CMV infection, it may be possible to give treatment that may prevent the deafness from progressing. This must be given in the first four weeks after birth.

Less common infections that can cause deafness include rubella (German measles), toxoplasmosis and syphilis. Your doctor will be able to give you more information about these infections. All the blood tests are usually done using one blood sample.



For more information, visit ndcs.org.uk/cmV.

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. Between 20 to 60% of children with sensorineural deafness also have an eye problem. This may just mean that the child will need to wear glasses when they are older, but an eye test can also help to diagnose a syndrome associated with deafness such as Usher syndrome.

As babies can't tell us what they can see, a developmental assessment can check that the eyes are healthy. It's recommended that all children with hearing loss are referred for an eye test and have regular eye tests throughout their childhood. Sometimes deaf children will have a special eye test. Your ophthalmologist (eye doctor) will discuss this with you if necessary.



9 Understanding genetics

Just as children inherit features such as hair colour or eye colour from their parents, sometimes deafness is inherited. This means that the deafness is genetic.

If your child's deafness could have a genetic cause, you may be offered genetic testing. Genetic testing 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

You may be offered the chance to discuss these things with a trained genetics counsellor.

Some families find it helpful to know whether the deafness and any other medical condition were inherited. Other families prefer to wait until their children are grown up and able to decide for themselves.

Genetic testing

Genetic testing may be able to identify a specific genetic cause for your child's deafness. However, it's important to know that genetic testing only identifies a clear cause of deafness in up to a third of children tested, based on current figures.

If a genetic cause is not identified, this doesn't necessarily mean that your child's deafness is not genetic. They may have a gene variant that causes deafness, but the variant has not yet been identified or it's not tested on the current standard test. Alternatively, there may be a non-genetic cause that cannot be identified.

Many families go through the process of aetiological investigations (which is any investigation looking for the cause of deafness, including genetic testing), without realising that there's a good chance they won't find out the cause of their child's deafness by the end of it.

Where genetic testing can identify a cause, it can help with the following situations.

Related health conditions

For 30% of children whose deafness is genetic, their deafness is part of a syndrome. This means that they may have, or be at risk of, associated health conditions. Knowing this diagnosis can identify specific future screening or monitoring to be offered. On the other hand, identifying a non-syndromic genetic cause means there are unlikely to be any associated health implications.

Whether hearing will change or stay the same

Understanding the cause of deafness can give professionals more information about which part of the ear isn't working. Sometimes this knowledge can help predict whether your child's level of deafness is likely to stay the same or decline over time.

Knowing more about your child's type of deafness can help inform decisions about technology options (such as cochlear implants or hearing aids), communication choices and education.

Chances of passing on a particular condition

Some parents want to know more about the chances of other children or grandchildren they may have being deaf or hearing.

Implications for other family members

Some families want to know whether other family members may have also inherited a health condition related to their child's deafness.

What are the different types of genetic testing?

Until recently, genetic testing could only be offered routinely for the Connexin 26 gene. Following developments in genetic testing technologies, wider NHS genetic testing into causes of deafness is now available across the UK.

These developments mean that your child's paediatrician, other audiology doctor or audiologist can now discuss and arrange tests for both genetic and non-genetic causes of deafness. Previously, almost all genetic testing was arranged only once you'd been seen in a genetics service.

There are different genetic tests that you may be offered. Some families have been through genetic testing but are not clear which tests were done. Be sure to ask your clinician exactly what types of tests they're doing and what each will involve. Knowing which types of tests were performed will help you better understand the results and may also be helpful for your child if they want to do their own research when they're older.

Most genetic tests are carried out using a blood sample from the child. Occasionally, it may be possible to use an alternative sample such as saliva. Sometimes samples from parents are needed alongside their child's to help with interpreting the results.



Questions for your audiologist

- Which genetic tests have been performed?
- What does this test involve?
- If results from these tests are inconclusive (don't show a cause of deafness), are there any other tests we could try?

Is genetic testing right for us?

Deciding whether to have genetic testing is a personal choice, and there's no right or wrong answer. To make the most informed decision for your child and family, you should make sure you understand the full implications of having a genetic test and what it might mean for your family. The results may show what's caused your child's deafness, but they may also reveal new information about other family members. For example, you may find out that your child's parent or grandparent is a carrier for a genetic condition.

The health professional discussing genetic testing with you should explain the possible outcomes and implications. You and your family may want to consider in advance how you feel about these. Don't be afraid to ask questions during your appointment and remember you can always say 'no' if you don't want to proceed with genetic testing.

Genetic testing may not be appropriate or recommended for all families. Some types of deafness may be more likely to have a genetic cause than others. For example, deafness in both ears (bilateral deafness) is more likely to have a genetic cause than if it's one ear only (unilateral deafness). There's also a higher chance genetic testing could find a cause of deafness if there's a family history of deafness or signs that could fit with a syndromic type of deafness.

While some families may find it helpful or reassuring to know the cause of their child's deafness, others may not want to know for a variety of reasons. They may also prefer to wait until their child is grown up and able to decide for themselves.



"We were advised throughout the process that it was likely we may never know why our son is deaf and that a cause may not be found. I found this really helpful as it meant my expectations were always fairly realistic. I did still feel a little disappointed when we got the initial genetics letter advising no genetic cause had been found." - **Gill**

"We are extremely glad we opted to go ahead with genetic testing for Joshua as this uncovered the cause of our son's hearing loss. It's helped us to develop a more appropriate support package for our son and be aware of risks for our future children and grandchildren." - **Julie**

"We're anxious yet excited to start this process as we hope to get some answers, but we're not going to get our hopes up as we have been made aware we may never have an answer as to why Freddie's deaf, and that's OK." - **Beth**



For more information on the genetics of deafness visit
ndcs.org.uk/genetics.

10 Audiology appointments questions list

To help you understand the hearing tests that your child may have, and what the results mean, you may want to ask some of the questions below at your child's audiology or ear, nose and throat (ENT) appointment.

1. Could I have a copy of my child's most recent hearing test and the report?
2. What tests have been carried out? Will there be a need for further testing?
3. How can I prepare my child for their next appointment?
4. What is the purpose of each test?
5. What type of hearing loss does my child have (sensorineural, conductive or mixed)? Please explain what this means.
6. Will my child need to be referred to another health professional? For example, an eye doctor or a geneticist?
7. Do you think our family should have genetic counselling?
8. Does my child need other tests, such as CT or MRI scans, heart, blood, urine or eye tests? What will these tests tell you about my child's hearing loss?
9. Is my child's hearing likely to change in the future?
10. What caused my child's hearing loss?
11. Do we and other members of our family need to have our hearing tested?
12. What options are there to support my child's hearing, such as grommets, hearing aids or implants?
13. Will a hearing aid help my child to hear?
14. When will my child be fitted with hearing aids?
15. Is a cochlear implant an option for my child? Where can I get more information?
16. Will the hearing loss affect my child's development?
17. How can I best ensure my child can hear me?
18. What can I do to support my child's hearing and communication skills?
19. How often will my child come back for a follow-up appointment?
20. What do I need to tell my child's school or nursery to make sure they're supported?
21. How do I get a Teacher of the Deaf (ToD) for my child?
22. Are there any local support services or resources you can recommend?

11

Tess's tips for audiology appointments



We never expected the first audiology appointment we had to be the first of many. After getting referred following a failed newborn screening test when Lucas was a day old, we honestly expected to be told he had a load of mucus in his ears and for him to be given a clean bill of health. Turns out we were wrong, and after a diagnosis of severe to profound permanent hearing loss at the audiology appointment, it became apparent that we would be regular visitors to our local hospital for the foreseeable future.

As a new mum of an 8 week old, I definitely hadn't mastered the art of getting out of the house on time for anything and found car travel with a baby and a pram a real faff. I wasn't confident feeding Lucas in public yet and was not looking forward to long waits in hot hospital waiting rooms with a grizzly baby. He's nearly six months old now, and we've had about eight different hospital appointments so far. I'm learning from every appointment, but I thought these tips might be of use to those of you who are also new to this.

Tess is mum to Lucas, who has a severe-to-profound sensorineural hearing loss and wears cochlear implants. Read Tess's tips for audiology appointments.





Get there on time

Most NHS appointments only give you a 10-minute grace period, so giving yourself loads of time is essential. Work out how long it takes to drive to your appointment and add another half hour on top to account for traffic, parking, fiddling with the pram, walking from the carpark etc. This extra time helps you to avoid feeling rushed and means you can arrive in a more positive mindset.



Scope out parking ahead of time

Suss out where the parent and child parking is so you don't get stuck not being able to get the car seat back in the car. We have a big multi-storey carpark at our hospital, and the parent and child spaces are right on the top floor. It's worth calling your local hospital to find out where your parking is and then make a beeline for it.



Brings layers

Hospitals are hot. Dress your baby in layers that are easy to remove. Lucas had a big meltdown in the ENT consultant's office after two hours of appointments a few months ago, and it was because he was overheating and overtired. I now always dress him in cardigan layers so I can strip him down to a vest once we get there.



Find out if there's a quiet room

Most hospital departments have one of these, and I've found them very handy when Lucas has been grizzly. You can switch the lights off to get your little one to sleep, or feed them in a more private environment. I always pack a dummy in the change bag now too so that he can soothe himself to sleep if he's too distracted to feed.



Don't fit too much in one day

Things will overrun. If you can, try not to schedule anything else in that day. Invariably, appointments run late, and you don't want the added pressure of having to rush back for anything. And appointments tend to be exhausting for you too, so don't try and fit too much in. Also, take some entertainment with you for baby - a favourite toy or a teether.

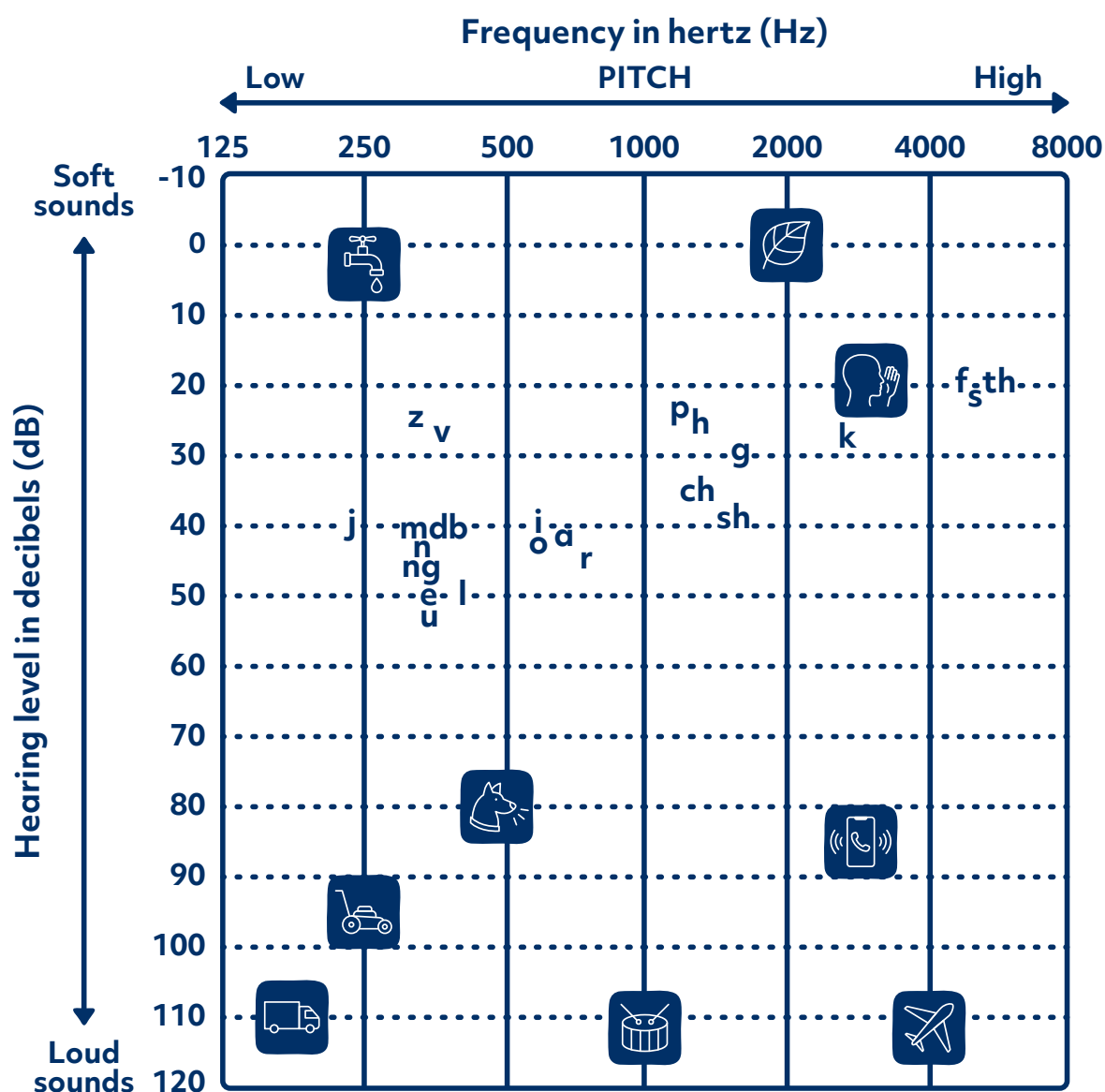


Take notes

If you're attending appointments alone with your little one, take a notebook so you can jot important information down which is easy to forget. Always ask for results or appointment information to be printed out for you too.

12 Audiogram for your child's test results

Ask your audiologist to print off a copy of your child's hearing test results for you or plot their results on this chart. This can help you explain what your child can and can't hear, with and without their hearing aids.



13

Our information and support

We're here for every deaf child who needs us - no matter what their level or type of deafness or how they communicate. As your child grows, we'll be here to support your family every step of the way.

The right information at the right time

We offer free, balanced information about all aspects of childhood deafness, both on our website ndcs.org.uk and in our publications. Our online content and information booklets can help you learn about childhood deafness and make decisions. Our free app, My NDCS, provides information tailored for your child's age and location.

Helping you make informed choices

Every deaf child is different, and families should be able to make decisions that are right for them. That's why we give independent support, setting out all the options, so families can make informed choices about how they want to communicate, or which type of hearing technology is best for them. We want to do all we can to give you the confidence to make the right decisions for you and your family.

Support when you need it

Any questions? We're here to help.

Helpline: **0808 800 8880**

helpline@ndcs.org.uk

ndcs.org.uk/helpline





We're the National Deaf Children's Society, the charity for deaf children with any level of hearing loss.

Helpline

0808 800 8880 (voice and text)
helpline@ndcs.org.uk

ndcs.org.uk



We're always looking for ways to improve our information.

Use your smartphone's camera to scan this QR code and share your feedback on this resource.

You can also give us your feedback by emailing your comments to informationteam@ndcs.org.uk.

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