

## Why does my child have a hearing loss?

### Introduction

This factsheet will tell you about the range of tests that can be carried out to try to find the cause of your child's hearing loss. The process to find out why a child is deaf is sometimes called an 'aetiological investigation'.

The tests listed below can find the reason for a child's deafness 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.

Note: we use the term 'deaf' to mean all levels of permanent deafness, but not temporary deafness such as glue ear.

**The National Deaf  
Children's Society**  
15 Dufferin Street  
London EC1Y 8UR

**NDCS Freephone helpline:**  
0808 800 8880 voice & text  
(Mon – Fri 10am-5pm)  
**Fax:** 020 7251 5020

**Website:** [www.ndcs.org.uk](http://www.ndcs.org.uk)  
**Email:** [helpline@ndcs.org.uk](mailto:helpline@ndcs.org.uk)

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

This factsheet will also tell you about the tests that will be done to make sure your child does not have any other medical condition.

**Please note, throughout this factsheet we give the following words the meanings shown below.**

- We use the term 'your doctor' to mean the doctor in charge of your child's audiological care. This may be an audiological physician, ENT specialist, or community paediatrician in audiology.
- The term 'your family' includes your child's grandparents on both sides.

### **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, 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, language and milestones such as when your child was sitting, walking and so on). The doctor may also ask about 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; or
- had balance problems.

All of these factors are important when investigating the cause of hearing loss.

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 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. The doctor will assess your child's development in relation to the expected ages and 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 an audiogram.

### **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 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. An MRI scan can be carried out on a child from birth, but you should be offered a choice of when the scan is done. If it is 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. A CT 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 is generally accepted that there is little risk to health from one scan, but with repeated tests there is a risk that the radiation may damage body cells. There is only a risk if your child has a high number of x-rays or scans. 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. Generally, 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 extremely 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 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.

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

- your child has had meningitis;
- your child is being referred to be assessed for a cochlear implant;
- the hearing loss is getting worse or changing over time; or
- there are characteristic features that may suggest your child's hearing loss could be part of a 'syndrome'.

In these examples, your doctor will need to look closely at the structure of the cochlea (inner ear) 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 is a very rare syndrome linking severe and profound hearing loss to a heart problem. If this heart problem is detected 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 hearing

loss to help rule out this condition. There are no risks associated with having this test.

## **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 hearing loss.

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 hearing loss. Some of these tests will give useful results only if they are 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 hearing loss. However, a positive result later in life may not give useful information as your doctor could not prove that the infection was present at birth and so caused the hearing loss.

There are several infections that can cause hearing loss in babies if the mother contracts them when pregnant. The most common infection causing babies to be born with hearing loss is CMV (cytomegalovirus). CMV is very common in the general population and does not normally cause any illness. However, it can affect the baby if the mother catches it while pregnant. About one baby in a hundred is born with CMV and some of these babies can be affected by it. CMV can cause deafness or, very occasionally, it can affect a child's development. Doctors can test for CMV in the first few weeks after birth. Hearing loss following infection can sometimes get worse over time. If a young baby is found to have the CMV infection, it may be possible to give treatment that may prevent the hearing loss from increasing.

Less common infections that can cause hearing loss include rubella (German measles), toxoplasma 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.

### **Ophthalmology (eye test)**

All children learn from what they see and hear around them. Children with hearing loss 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, a developmental assessment of the eye is done to make sure the eyes are healthy. Sometimes special eye tests are used. Your ophthalmologist (eye doctor) will discuss this with you if necessary. It is recommended that all children diagnosed with a 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. Most people have 23 pairs of chromosomes – microscopic structures that contain genes – in each of their cells. In each pair of chromosomes, one is inherited from the mother and the other from the father. Sometimes there is a change in a particular gene that results in a different characteristic, for example hearing loss. The change in a gene is known as a 'fault' or 'mutation'.

Genes can work in different ways. Sometimes a child only needs to inherit one gene, from either their mother or their father (dominant inheritance), to have that particular characteristic.

Sometimes the child would need to have the same gene passed on from both parents in order to have a condition (recessive inheritance).

Some people are known as 'carriers'. This is where they have the gene but don't have the characteristic. So a person who is a carrier for hearing loss would have the gene but would not have a hearing loss themselves. However, they can pass it on to their children.

Sometimes a gene mutation is a 'one-off' that simply happened by chance. This means that, although the hearing loss has a genetic cause, it has not been inherited. However, the gene mutation could be passed onto future generations.

It is impossible to summarise in this leaflet all the different areas that could be covered in genetic counselling. If your child's hearing loss could have a genetic cause, you should be given the chance to discuss this with a trained genetics counsellor. 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; and
- how likely you are to have another child with the same condition.

Some families find it helpful to know whether the hearing loss 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

You may be offered a genetic test. This will involve 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. All the genes related to hearing loss have not yet been identified and for most there is no routine test. This means that even if the hearing loss is inherited, it may not be possible to confirm this with a genetic

test. 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. Again, some of these syndromes can be confirmed with a genetic test, but many cannot.

Some children have a rare genetic hearing loss that can get worse if they are given certain medications. It is possible to have a genetic test to identify whether they have this gene. If they do, then this information will help inform you and your doctor if, in the future, your child needs treatment. Your doctor may recommend that this test is done at an early stage depending on your child's history.

Your doctor (audiological physician, paediatrician or ENT specialist) may refer you to a clinical geneticist or a genetic counsellor. Alternatively, they may discuss genetic testing with you themselves. Individual health authorities decide whether to offer routine genetic testing through the audiology or ENT department. Some areas offer genetic testing through this route only if it would provide a direct benefit to the child being tested. For example, if the child has an inherited medical condition it may be of direct benefit to have a genetic test. This is because the results may help to identify treatments that should be made available to the child. Your doctor will discuss the options available in your area with you. If you want to ask more about genetic testing, you can ask to be referred to your local clinical genetics service and your doctor will discuss with you if this is appropriate.

## What next?

Some or all of the tests in this factsheet will be offered to your child. Your doctor will give you more information which will help you decide whether you want your child to have the available tests and the best time for the tests to be carried out.

Test results that may offer immediate benefit to your baby are best done at an early stage, for example when the infection screen is carried out.

Some tests (for example, some routine genetic testing) will not offer an immediate or direct benefit to a baby and may be delayed until a later stage. Your doctor will tell you if particular tests should be done (for example, if your doctor suspects that your child's hearing loss is part of a syndrome). You and your doctor can use this information when planning how to manage the hearing loss and any associated condition in the future.

Most of the tests are not urgent. It is important that you feel comfortable with your child having them. Discuss any worries with your doctor.

### **Further information**

Your doctor should be able to answer any questions you have.

For general information on issues relating to your child's deafness (for example, education, communication and financial support) phone our Freephone helpline on 0808 800 8880 (voice and text) between 10 am and 5pm, Monday to Friday. You can also fax us on 020 7251 5020 or send an email to [helpline@ndcs.org.uk](mailto:helpline@ndcs.org.uk). Our website address is [www.ndcs.org.uk](http://www.ndcs.org.uk).

We would like to thank the members of the Aetiological Investigations Group for the valuable advice they gave to help us produce this factsheet.

**This information is available in large print, in Braille and on audio tape.**

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