Centre for Community Child Health

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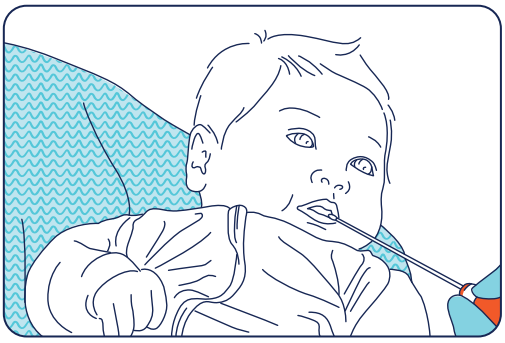
Identifying the cause of congenital hearing loss

What is congenital hearing loss?

Congenital hearing loss is hearing loss that is present at birth. It affects about 2 in 1000 children and has many causes. Medical tests may identify the cause of hearing loss, help families understand what to expect and, sometimes, how to minimise worsening hearing loss. This guide provides information on tests recommended for babies with newly diagnosed hearing loss.

Testing for causes of hearing loss

CMV: Cytomegalovirus testing



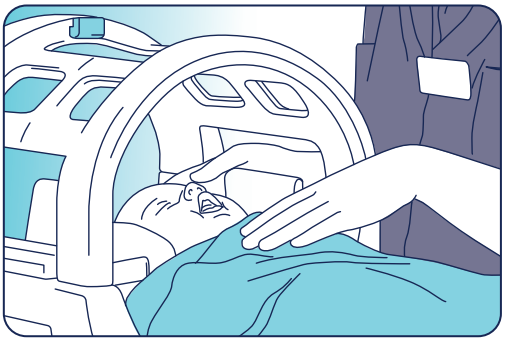
Cytomegalovirus (CMV) is a common virus and the most common infective cause of hearing loss. Several tests can be performed to check if your child may have been infected with CMV during pregnancy.

For babies younger than 21 days, a **saliva swab** from your child's mouth or a **urine test** can be taken to test for CMV. Either test can be done, depending on which test is available in your area.

For babies older than 21 days, we do not recommend a saliva swab or urine test. Instead, your doctor may request to access your child's heel-prick card to test for CMV. Babies born in Australia are routinely offered the **heel-prick test** at birth, which can be used to detect CMV.

The Centre for Community Child Health is a department of The Royal Chidren's Hospital and a research group of Murdoch Children's Research Institute

MRI: Magnetic Resonance Imaging



Magnetic Resonance Imaging (MRI) uses a strong magnetic field and radio waves to take pictures of the inside of the body. It does not involve any radiation. An MRI scan of your child's brain and inner ear may be performed to identify possible structural causes (e.g. abnormalities of the inner ear), of your child's hearing loss. Sometimes, detecting a particular structural cause can help your doctor advise you on ways to avoid further hearing loss. If your child has severe to profound hearing loss, the scan may also inform whether a cochlear implant would be helpful.

An **MRI scan** usually requires a child to remain still for at least 40 minutes. If your child is younger than 4 months, an MRI scan may be done without sedation or anaesthetic. We can feed and settle your baby to sleep for the MRI. This is a process called 'feed and wrap'. The availability of this option may depend on your location.

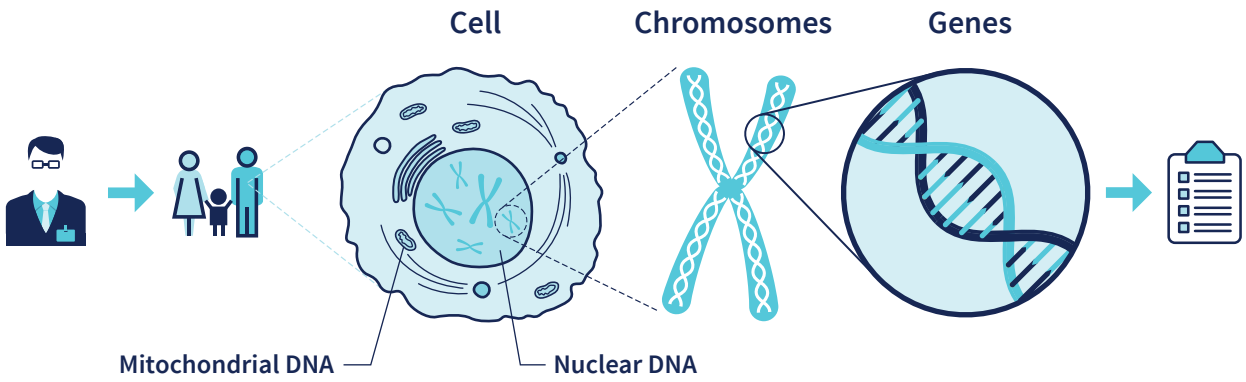
If your child is older than 4 months, discuss the need for an MRI scan with your doctor. This is because older children may need a general anaesthetic to put them to sleep and help them stay still during the MRI. A doctor will need to assess whether an MRI can be done at a later stage when your child is old enough to lie still without an anaesthetic (usually around school age).

Genetic testing

More than 50 per cent of congenital hearing loss is caused by a genetic variation or change. If your child has bilateral hearing loss (hearing loss in both ears), genetic testing and counselling are available to you. Genetic testing will check for any variations or changes in genes that could cause hearing loss. There are different types of genetic tests – some are government funded.

It is your choice to have genetic testing done on your child, as you need to consider the pros and cons of doing the tests for your child and your family. It is important to talk to a genetic counsellor or doctor before considering genetic testing. Reasons for doing genetic testing include:

- the test may identify a genetic change that can cause hearing loss
- if a genetic change is identified, it may help you:
  - understand why your child has hearing loss
  - understand whether your child may have additional medical or developmental needs in the future
  - decide on what to do for family planning.



The first genetic test we recommend is called the **Connexin test**. It is a blood test (or saliva test, depending on availability) that identifies changes to the GJB2 and GJB6 genes, which are responsible for making connexins. Their alterations are the most common genetic cause of hearing loss and they are responsible for 20 per cent of bilateral hearing loss at birth.

If the Connexin test is negative, there may be other genetic tests available to you. Currently, these genetic tests may not be covered by Medicare or may not be available in your local area. Your doctor may refer you to a genetic specialist to discuss available tests.

If your child has unilateral hearing loss (hearing loss in one ear), genetic testing is unlikely to be helpful. However, if there is a family history of hearing loss, discuss the role of genetic testing with your doctor.

Hearing tests for family members

Hearing loss can often be undetected. Therefore, we recommend your child's immediate family (child's siblings and parents) have a hearing test with an audiologist. This can provide more information about the possible cause of your child's hearing loss. You can arrange a hearing test for your child with Hearing Australia (www.hearing.com.au) or the audiologist who assessed your child with hearing loss. For adult tests, contact your doctor for more information.

Vision assessment

Children with hearing loss may also have problems with their eyesight. We recommend that children with hearing loss have their vision tested by an optometrist or ophthalmologist before starting school. Speak to your doctor about the options.

Further information

For more information about congenital hearing loss, testing and CMV, visit the Australasian Newborn Hearing Screening website: www.newbornhearingscreening.com.au

This information is produced by the Childhood Hearing Australasian Medical Professionals (CHAMP) network with the support of Deaf Children Australia and the Australasian Newborn Hearing Screening Committee. Special thanks to Christie Yung.

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Deaf Children Australia