

Childhood hearing loss - an overview

How common is hearing loss in children?

1 in 500 newborns have at least mild/moderate permanent hearing loss, as identified by newborn hearing screening. This number goes up to 1 in 300 by age 9. If you include all types of hearing loss, 1 in 20 teenagers have at least mild hearing loss.

What are the effects of untreated hearing loss in children?

The effects of hearing loss vary depending on the age of the child. In babies, hearing loss can make it harder for them to develop normal speech and language. Older kids can have trouble learning in school, behavioral problems, and attention deficits. Even mild or one-sided hearing loss has significant effects on quality of life.

How do we test and diagnose a hearing loss?

Initial testing and diagnosis of a hearing loss are performed by an audiologist and an otolaryngologist/ENT.

An **audiologist** is a health-care professional who evaluates, diagnoses, treats, and manages hearing loss, tinnitus, and balance disorders in newborn, children, and adults.

An **otolaryngologist**, or **ENT**, is a physician trained in the medical and surgical management and treatment of patients with diseases and disorders of the ear, nose, throat (ENT), and related structures of the head and neck.

Different tests can be done to figure out why your child has hearing loss. It is helpful to find out the cause of hearing loss. Sometimes it can help us treat the hearing loss better, prevent further hearing loss, or be able to better predict what to expect in the future. Other times it can help us know whether there are or are not other associated health issues that need to be looked into. Finally, genetic testing can help you and your child with family planning now and in the future.

Audiologic Testing

The audiogram, or hearing test, tells us whether your child has *conductive* or *sensorineural* hearing loss. In *conductive* hearing loss, sound cannot get from the outside of your body to the inside of your ear. We can usually figure out the most likely cause of most conductive hearing losses by examining the ear in the clinic. In *sensorineural* hearing loss, the inner ear organ (cochlea) that converts sounds into signals sent to your brain is not functioning correctly. Testing for this is more complicated, and may include genetic testing, imaging, or CMV testing.

Genetic testing

What is this? Genes are the molecular instructions that make our bodies ourselves. We get some genes from our mom, and others from our dad. Sometimes, genes have errors, called mutations, that can lead to hearing loss when present in certain combinations. The most common gene involved in hearing loss is Connexin 26.

How is it done? A blood test checks for mutations in Connexin 26, which cause 15-20% of all childhood hearing loss. Additional testing can be done to look for mutations in over 70 other genes, or even new hearing-loss genes.

What happens if we find something? The results may give some hints as to how the hearing loss will change in the future, and plays a role in family planning.

Imaging

What is this? A CT or MRI scan, which can detect structural abnormalities in the inner ear or cochlea. About 15-20% of childhood hearing loss is caused by structural abnormalities.

How is it done? The type and timing of imaging depends on a number of factors, including the exact information that we are looking for, need for anesthesia, radiation risk, and impact on subsequent treatment. CT is generally faster and looks at fine bony anatomy well, but does carry a small risk associated with radiation exposure. MRI is a more involved exam and is best at looking at nerves, but does not require radiation.

What happens if we find something? Results can help determine potential surgical management and/or cochlear implant candidacy or give some hints as to how the hearing loss will change in the future.

CMV testing

What is this? Infection with cytomegalovirus (CMV) during pregnancy ("congenital CMV") causes about 15-20% of all childhood hearing loss. This infection is very common in pregnant women and usually does not cause any identifiable symptoms.

How is it done? Testing for congenital CMV infection can only be done in babies up to 3 weeks of age, or by retrieving the newborn blood spot that was acquired shortly after birth, and testing it.

What happens if we find something? The results may give some hints as to how the hearing loss will change in the future, and in very select instances could be considered for treatment.