



Fact Sheet for Parents:

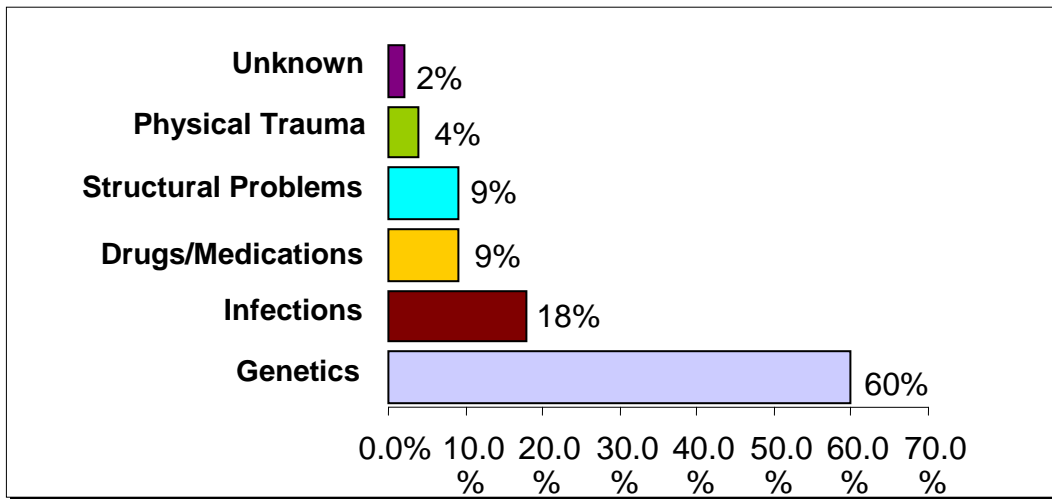


Why Does My Child Have a Hearing Loss?

This fact sheet gives general information about genetic evaluations and the causes of hearing loss. It is not intended to replace the clinical judgment of a healthcare provider.

What causes permanent childhood hearing loss?

Permanent childhood hearing loss can have many causes. The most common are:



Over half of permanent childhood hearing loss has a genetic cause.

What do you mean by a genetic cause?

Genes are the instructions that tell our bodies how to grow and develop. Some genes are responsible for hearing. A change in one or more of these genes may cause a person to have hearing loss.

Genetic causes of hearing loss are mainly divided into two types: syndromic and nonsyndromic.

1. **Syndromic hearing loss** means that the gene change that causes hearing loss also causes other health problems. About 30% of people who have genetic hearing loss are found to have a syndrome. Examples of

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syndromes with hearing loss include:

NAME OF SYNDROME	OTHER FEATURES (besides hearing loss)
Alport	Kidney problems
Pendred	Thyroid gland enlargement
Usher	Vision impairment
Jervell and Lange-Nielsen	Heart problems

2. **Nonsyndromic hearing loss** means that the gene change that causes hearing loss does not cause other health problems. This is also called “isolated hearing loss”. Most people with genetic hearing loss have no other health problems.

About half of **nonsyndromic hearing loss** is caused by a change in a gene called *Connexin 26*. Three of the most common *Connexin 26* changes linked to hearing loss are:

- 35delG, found mostly in Northern European families
- 167delG, found mostly in Ashkenazi Jewish families
- 235delC, found mostly in Japanese and Chinese families

How can I find out what caused my child’s hearing loss?

One option is to have an evaluation by a genetics team. This will likely consist of:

- a physical exam
- providing a medical and family history
- education about genetics and hearing loss
- discussion of options such as clinical evaluation and genetic testing

Sometimes the cause of hearing loss can be identified without genetic testing. Genetic testing is not necessary for every child. It may not be offered as part of the genetics evaluation. If genetic testing is offered, the genetics team can discuss the possible benefits and limitations. They can provide you with information to help you make an informed decision. Genetic testing is a personal decision for each family.

What are some possible benefits of genetic testing?

Genetic testing can sometimes help confirm the cause of hearing loss. Finding out if your child has syndromic or non-syndromic hearing loss may have an impact on your child’s health care. Your child may be able to avoid the need for further diagnostic tests if a genetic cause is found. Finally, genetic testing may help you

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and other relatives find out the chances of having more children with hearing loss.

Will genetic testing tell me why my child has a hearing loss?

Genetic testing is only one part of the evaluation process. There is a chance that genetic testing will not identify the cause of your child's hearing loss. Over 400 genes that are linked to hearing have been identified. However, not all gene changes that cause childhood hearing loss are known.

Sometimes we receive a genetic test result that has not been reported before. Everyone has some natural differences in their genes. We may not be able to determine whether a specific gene change causes hearing loss or is just a natural difference.

Current testing technology may not be able to detect all gene changes. This means that we cannot be absolutely sure there isn't a genetic cause even if the test results do not show a gene change.

Will non-genetic causes be considered during a genetics evaluation?

Yes, a genetics team also looks for other causes of hearing loss during a genetics evaluation. They consider possible causes such as infections before or after birth, exposure to certain drugs or medications, physical trauma, and structural differences inside or outside the ear.

Some factors seem to increase a child's chances for hearing loss. For example, children seem more likely to have a hearing loss if they have had:

- Conditions requiring more than 5 days in a neonatal intensive care unit
- Low oxygen levels
- High bilirubin levels
- Very premature birth

Hearing can change over time. You and your child's healthcare provider should pay extra attention to your child's hearing and speech-language development and watch for changes in how your child responds to sound.

What else can I do to help my child while I wait for the genetic evaluation to be completed?

Early Intervention Services

If your child is less than three years old, he or she may qualify to receive early intervention (EI) services. Children with hearing loss do best if they begin EI

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services before six months of age. The lead agency for EI in Hawai`i is the Department of Health.

Services such as audiology, speech therapy, family support, and information sharing can begin even before all the exams are completed and before hearing aids are obtained. EI services are available at no cost to families, although use of insurance is encouraged.

To contact the Hawaii Keiki Information Services System (H-KISS) for more information about enrolling your child for EI services, call 594-0066 from Oahu or toll-free from Neighbor Islands at 1-800-235-5477.

Hearing Aids

You can decide if you want hearing aids for your child. Hearing aids can be fitted as early as one month of age. They help get sound to the brain. Your child can obtain hearing aids as soon as possible if the audiologist recommends them.

Babies are born with brains that are ready to hear. The ears are a way to get sound to the brain. If sound is not reaching the brain, children begin using the part of the brain that usually hears for other things. Obtaining hearing aids early and helping your child learn to wear them can protect your child's long-term hearing potential.

Overall Health

You can work closely with your child's doctor to monitor you're his or her health and obtain medical treatment when needed. Ear infections can flare up and affect hearing loss. It's important to contact your child's doctor as soon as possible if you think he or she is having more trouble hearing than usual or isn't feeling well.

Sometimes the cause of your child's hearing loss will remain unknown after all the exams are completed. Not knowing why your child has a hearing loss doesn't mean you can't get help. Beginning intervention services as early as possible, helping your child learn to wear hearing aids if recommended, and taking your child to see a healthcare provider and audiologist regularly will help your child stay healthy and build strong language, communication, and listening skills that will last a lifetime.

Who can I contact for a genetics evaluation?

Hawaii Community Genetics provides pediatric and adult genetics clinical services across the state of Hawai'i.

Hawai'i Community Genetics
1441 Kapi'olani Blvd, #1800
Honolulu, HI 96814
Tel: 808-973-3403
Fax: 808-973-3401

Your medical care provider at Kaiser Permanente or Tripler Army Medical Center may also provide genetic evaluations for hearing loss.

Where can I find more information about hearing loss?

Here is a list of websites you may find helpful:

National Center for Hearing Assessment and Management
<http://www.infanthearing.org/>

My Baby's Hearing
<http://www.babyhearing.org/>

Hands and Voices
<http://www.handsandvoices.org/>

U.S. National Library of Medicine Genetics Home Reference on non-syndromic deafness
<http://ghr.nlm.nih.gov/condition=nonsyndromicdeafness>

Center for Disease Control and Prevention on the genetics of hearing loss
<http://www.cdc.gov/ncbddd/hearingloss/genetics.html>

National Institute on Deafness and Other Communication Disorders
<http://www.nidcd.nih.gov/health/hearing/>

Orphanet is a database dedicated to information on rare genetic diseases. Use the search function to read more about deafness.
<http://www.orpha.net/consor/cgi-bin/index.php>

Hawaii Genetics Program
<http://health.hawaii.gov/genetics/programs/genetics/>

Hawai'i Newborn Hearing Screening Program
<http://health.hawaii.gov/genetics/programs/nhsp/>

Hawaii Department of Health
Updated: August 2015