COMMON CAUSES OF HEARING LOSS

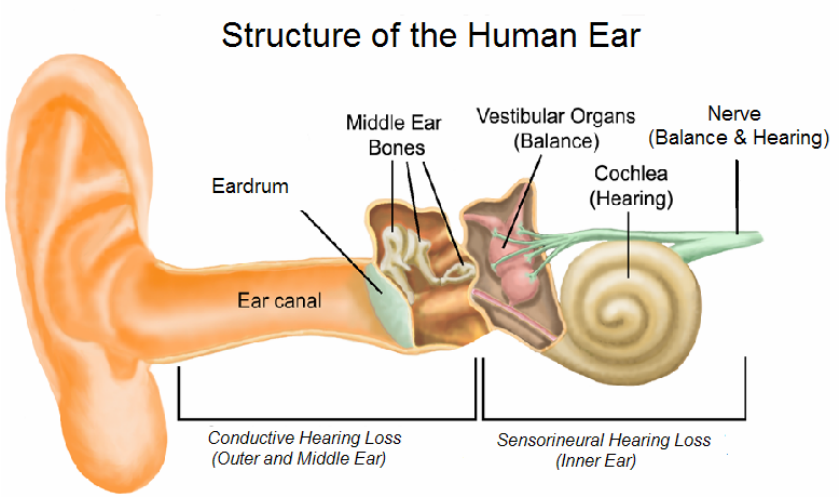
FOR PARENTS & FAMILIES

HARVARD MEDICAL SCHOOL
CENTER FOR HEREDITARY DEAFNESS

When your child is diagnosed with a hearing loss, it may be very overwhelming. This may be a difficult time for you and your family. However, gaining a greater knowledge in this area is crucial in helping with your child's needs. This handbook is aimed at helping parents and families to understand better various types and causes of hearing loss.

About Hearing Loss

Our ears contain many parts, all of which need to work properly for us to hear sounds.



The type of hearing loss that occurs depends on what part of the ear is not working properly. For example, if there is something not working in the

- ear canal
- eardrum
- middle ear bones *or*
- middle ear space (e.g. fluid)

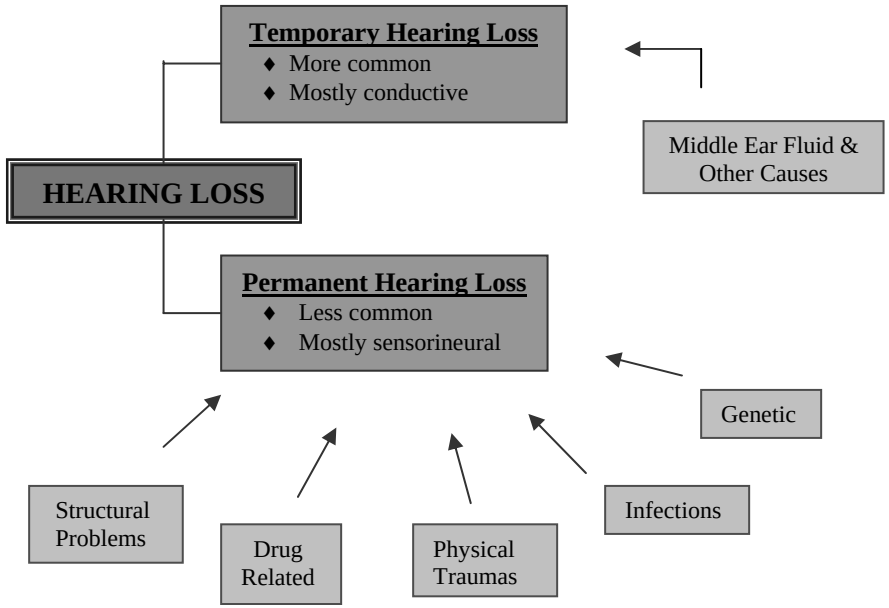
it may result in a ***conductive hearing loss*** (sometimes called "mechanical hearing loss"). If, on the other hand, something is not working in the

- cochlea
- auditory nerve *or*
- brain

it more commonly results in a ***sensorineural hearing loss*** (also called "nerve hearing loss"). When someone has both sensorineural and conductive hearing losses, it is called a ***mixed hearing loss***.

Duration of Hearing Loss

Depending on the cause, hearing loss may be temporary or permanent. Temporary hearing losses are almost always conductive and are far more common than permanent hearing losses. Permanent hearing losses are usually sensorineural, but may also be conductive. A chart below demonstrates this relationship and outlines the causes.



WHAT CAUSES TEMPORARY HEARING LOSS?

There are several causes of temporary hearing loss. The most common cause is fluid in the middle ear (i.e. due to ear infections). Other causes may include:

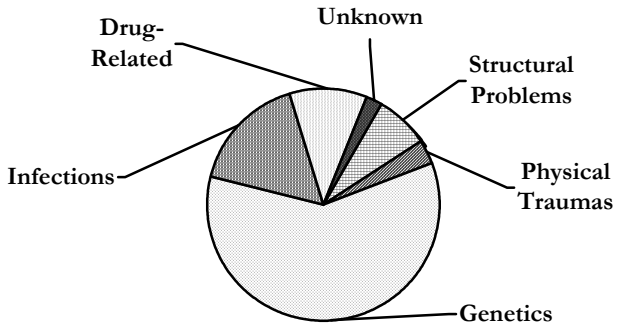
- Eardrum perforation (when there is a hole in the eardrum from a bad ear infection, or ear trauma)
- Narrowing of the ear canal due to surgery or disease
- Excessive ear wax that plugs the ear canal

DID YOU KNOW?

Poking objects too deeply into the ear canal (Q-tips, pencils, food, buttons, pins, etc) can cause hearing loss. It is the most common ear injury in toddlers.

WHAT CAUSES PERMANENT HEARING LOSS?

Permanent hearing losses are less common than temporary hearing losses. There are many causes of permanent hearing losses. These include genetic, infectious, drug-related, physical trauma and structural causes. A rough distribution of these is seen below:



Genetics

Although there are many reasons why permanent hearing loss may occur, genetics is the most common. It accounts for over half of all cases of hearing loss in infants. Saying that hearing loss is "genetic" means that there is a change in a gene.

- We have the same set of genes in every cell of our body
- Genes tell our cells how to function
- When a change in a gene occurs it is called a "mutation"
- A mutation can stop the gene from working properly

DID YOU KNOW?

While there are some families that have hearing loss in many generations, most people with genetic hearing loss do not have family members with hearing loss.

Syndromic vs. Non-syndromic Hearing Loss

There are two types of hearing loss caused by genetics. About 30% of people with a genetic type of hearing loss have what is called a ***syndromic hearing loss.*** Syndromic hearing loss means that there are other health problems in addition to hearing loss. Some examples 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

The other 70% of genetic hearing loss is called ***non syndromic hearing loss***. In this case, the person only has hearing loss.

Genetic testing may help determine if the hearing loss is genetic. This information may allow better treatment and management of the hearing loss, and predict the chances that future children will have hearing loss. To learn more about genetic causes of hearing loss, you may download a free booklet at http://hearing.harvard.edu.

Infections

New vaccines have prevented some hearing loss caused by infections. However, other infectious causes still exist. Some examples include:

During Pregnancy

- Infections caused by viruses (Cytomegalovirus [CMV], herpes, rubella)
- Syphilis
- Toxoplasmosis (may be associated with consumption of food or water contaminated with animal feces, or of raw or undercooked meat)

After Birth

- Infections caused by bacteria (bacterial meningitis, syphilis)
- Infections caused by viruses (measles, mumps)

Drugs

Drugs and other chemicals can also cause hearing loss. Some drugs can affect the fetus, while others are harmful after birth. Some of these include:

During Pregnancy

- Alcohol
- Medications (e.g. Accutane®, Dilantin®, chemotherapy)

After Birth

- Ototoxic drugs (e.g. aminoglycoside antibiotics, diuretics, cisplatin)

Physical Trauma

The ear is a delicate structure and can be easily damaged in a number of ways, such as:

- Head injuries (skull fractures or concussions)
- Exposure to loud noise (either a single blast or many exposures to loud noises)

Structural Problems of the Ear

A person can be born with an ear that is not formed normally. This could affect the outer, middle or the inner ear. Examples include:

- Stenosis (narrowing of the ear canal)
- Absence or malformation of the outer ear, ear canal, ear drum, ear bones
- Inner ear malformations (e.g. enlarged vestibular aqueduct)

Unknown Causes

Sometimes the cause of hearing loss cannot be identified. This may be because there are no available tests to find the cause. For instance, there are no tests for many genetic causes of hearing loss.

In addition, there are other causes of hearing loss that we do not yet understand. For example, a higher incidence of hearing loss has been seen in newborn babies with many different risk factors such as.:

- Lengthy stay in the Neonatal Intensive Care Unit (NICU)
- Very premature birth
- High levels of bilirubin
- Low oxygen levels

The relationship between many of these risk factors and hearing loss is unclear.

IN SUMMARY

Finding the cause of hearing loss is often not an easy task. Making an accurate diagnosis and finding a way to manage the hearing loss requires a combination of

- a physical exam
- family history
- clinical tests (such as genetic tests, MRI, EKG, CT scans, etc.)
- ongoing hearing tests with an audiologist

Sometimes, even with all exams, the cause of hearing loss may remain unknown. However, it is best if hearing loss is identified early. It is also important that help for the hearing loss begin as early as possible. Help can include communication therapy, parent support and hearing devices such as hearing aids and cochlear implants. For help your child should be enrolled in an early intervention program. This is true even if the cause is still being examined. This gives a child with hearing loss the best chance at language development, social interactions and education.

ADDITIONAL RESOURCES

- **American Academy of Otolaryngology Head and Neck Surgery**
  Click on Public and Parents, and then on ENT Health Info to view information on ear infections, ear function, plastic surgery of the ear, cochlear implants and other links.
  www.entnet.org

- **Boys Town National Research Hospital**
  Information on newborn hearing screening, hearing testing, high risk factors for hearing loss, genetic information on hearing loss and hearing aids. Includes www.babyhearing.org.
  www.boystownhospital.org

- **Harvard Medical School Center for Hereditary Deafness**
  Information on genetic forms of hearing loss, genetic testing, and a gene mutation database.
  http://hearing.harvard.edu

- **Laboratory for Molecular Medicine at HPCGG**
  A clinical laboratory offering the most comprehensive genetic testing for hearing loss. The lab is located within the Harvard Medical School-Partners Healthcare System Center for Genetics and Genomics.
  http://www.hpcgg.org/lmm

- **National Institute on Deafness and Other Communication Disorders**
  Information on disorders associated with hearing loss, cochlear implants, ear infections, hearing aids, American Sign Language, noise-induced hearing loss, tinnitus, and resources for educators.
  http://www.nidcd.nih.gov

- **Parents Guide to Hearing Loss**
  A booklet with information on communication with your child, hearing tests, hearing devices, as well as programs for children with hearing loss.
  http://medhome.med.utah.edu/file.cfm?file_id=190&

- **Raising Deaf Kids**
  Information on learning and communicating with hearing loss, general information on hearing loss and other links.
  www.raisingdeafkids.org

FOR ADDITIONAL RESOURCES PLEASE VISIT OUR WEBSITE:
http://hearing.harvard.edu/resources.htm

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