



**DIAGNOSIS –  
Causes/Etiology of Hearing Loss**

# EARLY HEARING DETECTION AND INTERVENTION (EHDI) EARLY INTERVENTION SERVICES (Birth-School Years)

## Medical

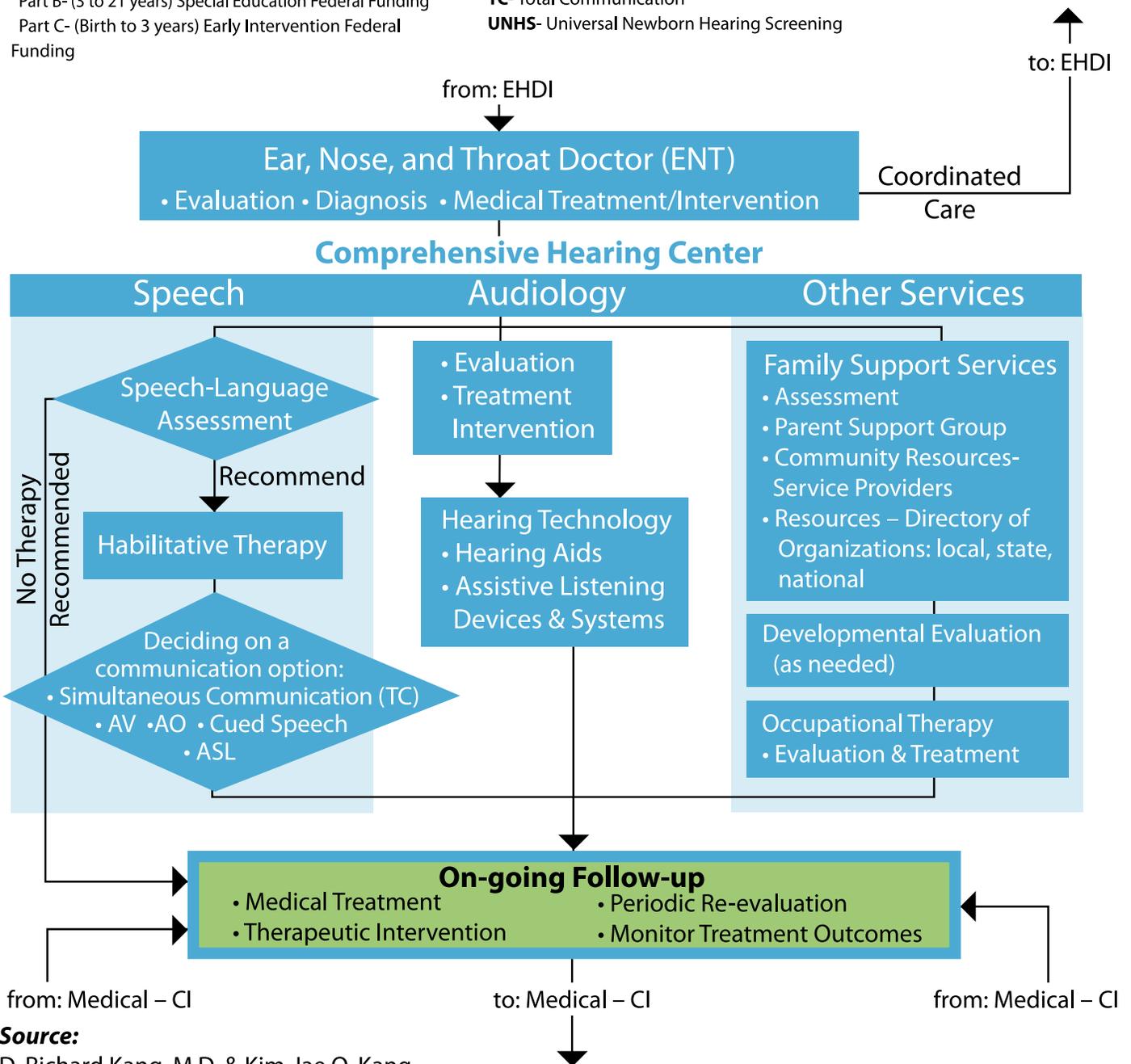
### Acronym Key:

**ALDS**- Assistive Listening Devices and Systems  
**AO**- Auditory Oral (Communication Modality)  
**ASL**- American Sign Language  
**AV**- Auditory Verbal (Communication Modality)  
**BEIS**- Bureau of Early Intervention Services- under ODH  
**CI**- Cochlear Implant  
**EI**- Early Intervention  
**ENT**- Ear, Nose, and Throat Doctor (Otolaryngology)  
**HMG**- Help Me Grow- under ODH  
**IDEA**- Individuals with Disabilities Education Act  
 Part B- (3 to 21 years) Special Education Federal Funding  
 Part C- (Birth to 3 years) Early Intervention Federal Funding

**IEP**- Individualized Educational Plan  
**IFSP**- Individualized Family Service Plan  
**LEA**- Local Education Agency  
**ODE**- Ohio Department of Education  
**ODH**- Ohio Department of Health  
**RIHP**- Regional Infant Hearing Program- under ODH  
**Section 504 Plan**- of Rehabilitation Act of 1973- Civil Rights Law, protects children with disabilities from discrimination  
**TC**- Total Communication  
**UNHS**- Universal Newborn Hearing Screening

### Color Key

 **ODH**  
 **Medical**  
 **Medical – CI**  
 **Education – ODH & ODE**



### Source:

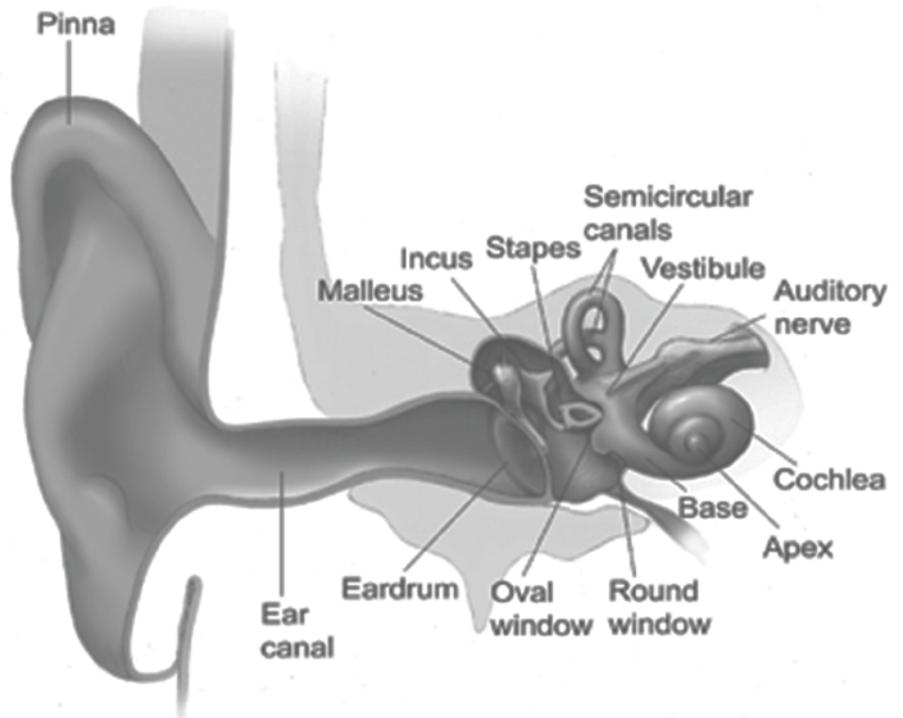
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## How Do We Hear?

Sound waves enter our ears and are amplified by the ear drum and middle ear bones (ossicles), allowing it to create a traveling wave in the fluid within the inner ear (cochlea). The waves bend microscopic hair cells in the cochlea which stimulate the hearing nerve, sending signals to the brain. The brain processes these signal into “sounds” that we learn to recognize and understand.



*Source: NIDCD NIH Medical Arts. Reprinted with permission.*

## The Anatomy of Hearing

Our ear is made up of three sections that make up our hearing system: the outer ear, the middle ear, and the inner ear.

## Parts of the Ear Structure

**The Outer Ear** – is the only part of our ear that we can see.

It is made of two parts:

- the ear flap, called the *pinna*, and
- the *ear canal*

Sound travels from this part of the ear into the middle and inner ear. The outer ear also protects the internal structures of the middle and inner ear.

**The Middle Ear** – sits between the ear drum and the inner ear. It includes these structures:

- *ear drum* also called *tympanic membrane*
- *ossicles* – one chain of three small bones
- *eustachian tube* – connects the middle ear to the upper part of the throat to allow drainage and air exchange. Its function is to keep the pressure of the middle ear equal to the pressure of the outside air.

Sound waves that enter from the outer ear, funnel through the ear canal and enter the middle ear, where it strikes the ear drum and causes it to vibrate. Those vibrations from the ear drum cause the ossicles to move. The middle ear structures amplify the sound vibration energy so that it has enough energy to move the fluid in the inner ear (cochlea).

**The Inner Ear** – consists of three parts:

- *vestibule* – the central inner ear cavity
- *cochlea* – looks like a snail shell and is considered the sensory organ of our hearing system
- *semicircular canals* – three attached loops, which are referred as the vestibular system or balance system

The cochlear fluid in motion causes the movement of the *basilar membrane*, which is in contact with thousands of nerve cells called *hair cells*. The bending of these hair cells generate electrical signals that activate nerve impulses that travel through the auditory nerve up to the brain. Thousands of these hair cells are moved either by, a low, middle, or high frequency sound.

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## Types of Hearing Loss

There are three main types of hearing loss in children. These may occur bilaterally (in both ears) or unilaterally (in one ear).

- 1. Conductive** – hearing loss occurs in the outer or middle ear due to a disruption or mechanical blockage of conduction of sound waves before it reaches the inner ear. In most cases, this type of hearing can be corrected and is considered reversible.
  - **Causes** – blockage of ear canal, punctured ear drum, fluid in the middle ear (otitis media), fixed or discontinuous ossicles from infection or tumor, congenital malformation of the outer or middle ear and ossicles.
  - **Treatments** – in most cases it can be corrected medically or surgically. In those rare cases where this is not possible, hearing aids are very effective in correcting conductive loss.
- 2. Sensorineural** – hearing loss occurs in the inner ear (cochlea), the auditory (hearing) nerve or the brain. This type of hearing loss is generally permanent, since nerve cells do not regenerate (grow back).
  - **Causes** – either environmental or genetic. (See section on Causes/Etiology of Hearing Loss)
  - **Treatments** – use of amplification devices, such as hearing aids, Baha® device, Assistive Listening Devices and Systems (ALDS) such as FM and sound systems, and sometimes cochlear implants.
- 3. Mixed** – both conductive (middle ear pathology) and sensorineural (cochlear or auditory nerve) hearing losses are present together in the same ear. In this case, when the conductive loss is corrected, there is still some hearing loss. It is important that after a child has their conductive loss corrected (such as insertion of tubes), a hearing test is performed.
- 4. Other Conditions of Hearing Loss**

**Auditory Neuropathy Spectrum Disorder (ANSD)** also known as **Auditory Neuropathy/Auditory Dysynchrony (ANAD)** – Part of the cochlea is functioning normally, but some hair cells or the auditory nerve is not working properly. It results in a confused nerve impulse signal being sent to the hearing centers of the brain. The mechanism of this disorder is not fully understood and the treatment varies depending on the degree of functional hearing.

**Central Auditory Processing Disorder (CAPD)** – this is not a hearing loss as the inner ear works normally, rather the brain seems to have difficulty interpreting and processing hearing input. A child with CAPD has problems localizing and discriminating sounds, and has difficulty understanding speech and following spoken instructions.

### **Degrees of Hearing Loss**

The degrees of hearing loss are defined as the severity of hearing loss. In hearing test, it is the softest sound at which the test sound is heard.

<b>Normal</b>	0-20 dB
<b>Mild</b>	21-40 dB
<b>Moderate</b>	41-55 dB
<b>Moderate-to-Severe</b>	56-70 dB
<b>Severe</b>	71-90 dB
<b>Profound</b>	91+ dB

*Note: These are standards used by Nationwide Children’s Hospital – Department of Audiology.*

These thresholds vary (5 to 10 dB) in different parts of the country.

To see a graphic representation of degrees of hearing loss, refer back to the *Section V. Audiological Testing*, under *Audiogram*

### **Causes/Etiology of Hearing Loss**

Causes of hearing loss are divided into two general categories: environmental and genetic. In some cases, the exact cause is not known.

#### **1. Environmental**

- Viral Infections (e.g. Rubella measles or cytomegalovirus (CMV))
- Certain medication that can be toxic to ear
- Injuries caused by lack of oxygen or inadequate blood flow

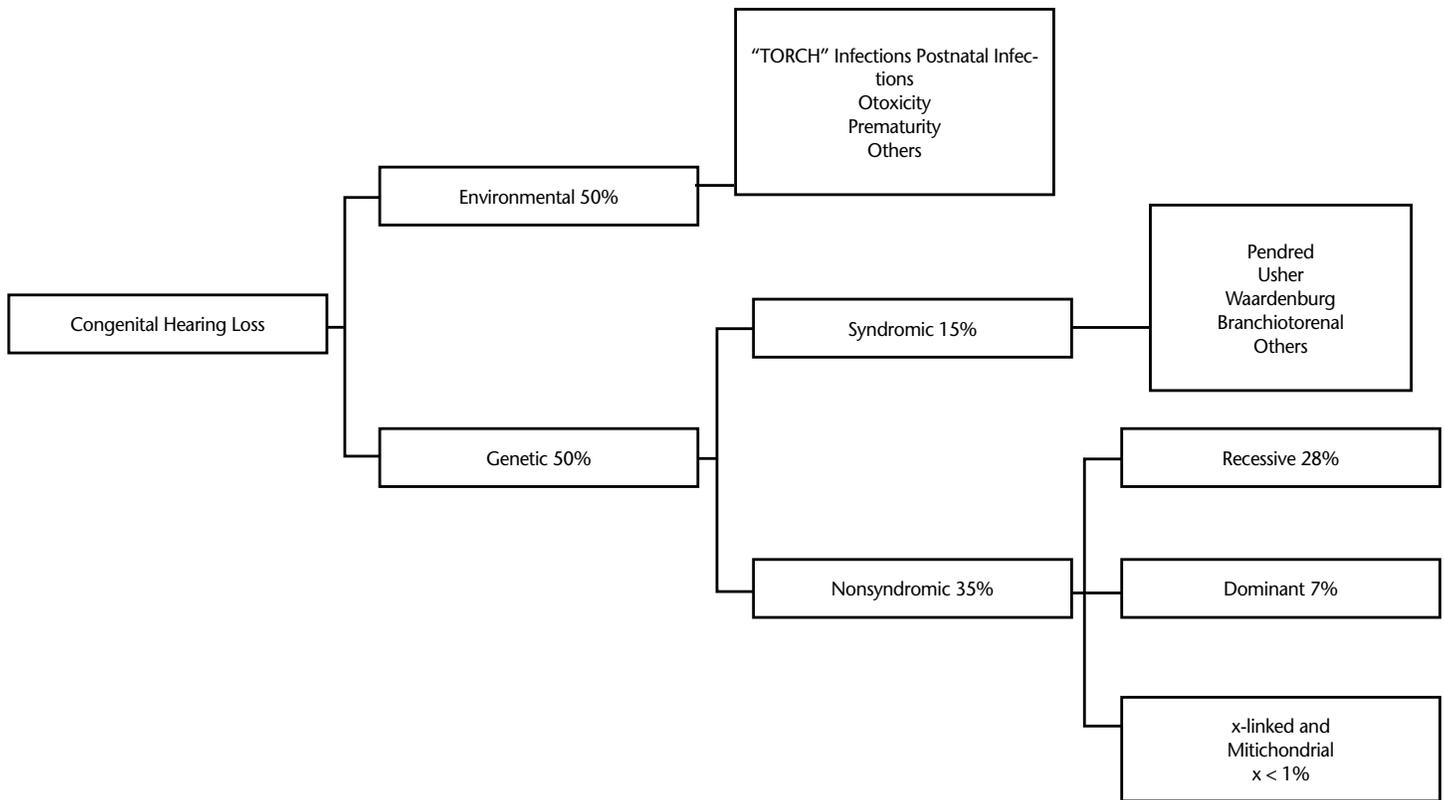
#### **2. Genetic** – numerous genes have been found to be associated with hearing loss. Genetic causes can be divided into two types:

- **Syndromic** – those associated with other organ abnormalities
- **Nonsyndromic** – Connexin 26 is the most common genetic cause and it is a nonsyndromic autosomal recessive genetic abnormality.

Very few of the gene abnormalities are readily available for routine testing. Connexin 26 testing is one of these available tests.

Genetics and heredity are very complex. If your child is diagnosed with genetic hearing loss, ask your child's doctor to have you talk with a geneticist or genetic counselor.

Below is a diagram on the causes/etiology of hearing loss.



For more information on the genetics of hearing loss, see CDC website: "Parent's Guide to Genetics and Hearing Loss"  
<http://www.cdc.gov/ncbddd/ehdi/documents/Edmaterials/ParentsGuidetoGeneticsEN.pdf>



# NOTES