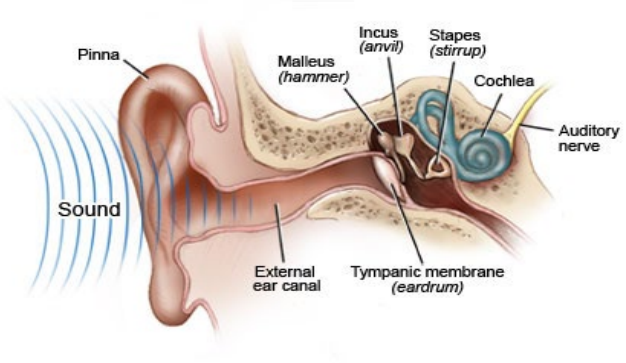
Auditory Neuropathy Spectrum Disorder (ANSD)

PENTA Pediatric ENT of Atlanta
Where Children Are First

What is ANSD?

ANSD is a condition in which the ear can detect sound normally, however, there is a breakdown in the signal getting to the brain. The disruption of the sound getting to the brain could be caused by structures in the inner ear (cochlea) not processing the sound in the synchronous way the brain needs. This is referred to Auditory Dysynchrony and is part of the "spectrum" of ANSD. On the other hand, the disruption of the sound getting to the brain could be caused by the auditory nerve itself.



What causes ANSD?

ANSD can be inherited genetically or it can be caused by trauma or disease. Risk factors for ANSD include, but are not limited to:

- Lack of oxygen
- Severe jaundice
- Infectious diseases
- Immune disorders
- Neurological disorders

How is ANSD diagnosed?

An ANSD diagnoses is made by a comprehensive evaluation that includes a variety of tests. Other tests may be included in the evaluation that is not listed below, however these are the most ordered tests.

Primary Tests:

- Auditory Brainstem Response (ABR) - This test determines they synchrony of the auditory nerve and is completed by an Audiologist.
- Otoacoustic Emissions (OAE) – This test determines if the inner ear (cochlea) is detecting the sound normally and is completed by an Audiologist.

Additional Tests:

- Magnetic Resonance Imaging (MRI) – imaging used to view the auditory nerve.

Common Features of ANSD

ASND can present itself in many ways. These are the most common presentations amongst children with ANSD:

- Inconsistent responses to speech
- Difficulty understanding speech in background noise or noisy environments
- Hearing appearing to change
- Appearing to have hearing loss

Treatment for ANSD:

There is currently no medical treatment for ANSD. However, the following treatment options are available.

- Traditional Hearing Aids (non-surgical) – some children with ANSD receive benefit from hearing aids, however this is not the case for every child with ANSD. A trial with amplification may be recommended.

- FM Systems (non-surgical) – ear-level FM systems look like hearing aids, however instead of amplifying sound, they improve the signal-to-noise ratio by having the speaker wear a microphone (transmitter) and child wear the receiver. This system streams the voice of the speaker directly into the child's ears making the speaker louder than the noise around them.

- Cochlear Implants (surgical) – Not all children with ANSD are a candidate for cochlear implants. The child will go through an extensive evaluation to determine candidacy.

- Watch and Wait Approach (monitoring) – This method is used when no reliable, behavioral audiological information can be obtained for the child. This means a hearing test was not able to determine a hearing level for the child.

Sources:
https://www.cincinnatichildrens.org/health/a/auditory-neuropathy
https://www.hopkinsallchildrens.org/Patients-Families/Health-Library/HealthDocNew/Auditory-Neuropathy-Spectrum-Disorder-(ANSD)