

# Auditory Neuropathy Spectrum Disorder

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## What is auditory neuropathy spectrum disorder (ANSD)?

**Auditory neuropathy spectrum disorder (ANSD)** is a type of hearing loss that happens when the inner ear has problems sending sound to the brain. It can happen at any age, and we don't know how many people are affected by ANSD.

On hearing tests, some people with ANSD will detect (be able to hear) sounds in the normal hearing range, while others can have mild to profound hearing loss. Even if the ear detects sound and send sound signals to the brain, the brain may not process the signals in an organized way. This can affect speech and language development (the ability to talk) and speech perception abilities (the ability to understand what others are saying).

## How is ANSD diagnosed?

An **audiologist** (a doctor specializing in diagnosing and treating hearing loss) will complete different hearing tests with you. This will help them understand how well you can hear and how much hearing loss you may have. These tests include otoacoustic emissions (OAE) and auditory brainstem response (ABR) evaluations.

- An **otoacoustic emission (OAE) evaluation** tests the function of the hair cells in your inner ear (**cochlea**). This is important to understand if sound is reaching the inner ear in the right way.
- An **auditory brainstem response (ABR) evaluation** measures brain waves to understand how your brain responds to sound at different sounds.
- Because ANSD affects how sound travels from the inner ear to the brain, the OAE test typically comes back normal, meaning the hair cells in the

inner ear are functioning normally. However, if you have ANSD, the ABR shows that your brain is not responding to sounds as it should.

- The audiologist will also do a hearing test in a sound booth, because it can help us understand how a person with ANSD detects sounds. However, it cannot give us information on how the detected sound is or isn't processed by the brain.
- A speech and language evaluation is important for understanding how ANSD is affecting language development in children. When a child is diagnosed with ANSD, they will receive a speech and language evaluation performed by a speech therapist. This will give us information on **receptive language** (how a child understands language) and **expressive language** (how a child uses words to express themselves).

## **What are other conditions that someone might have if they have ANSD?**

ANSD can commonly occur with genetic disorders, such as Charcot-Marie-Tooth disease, so genetic testing can be helpful. Some other conditions that happen with ANSD include:



- Friedreich's ataxia
- Cerebral palsy
- Kernicterus

## **How is ANSD treated?**

ANSD can affect people in different ways. Some people with ANSD will develop normal speech and language, and others cannot. That is why a team approach — including an ear, nose, and throat doctor (ENT), an audiologist, a speech-language pathologist, and the patient and their family and friends — is important in deciding how to treat ANSD. Cochlear implants have been very

successful treatments for children with ANSD who are having trouble with speech and language development.

### What are some other resources to learn more about ANSD?

<p><b>National Organization for Rare Disorders</b></p> <ul style="list-style-type: none"><li>• Overview of ANSD: <a href="https://rarediseases.org/rare-diseases/auditory-neuropathy-spectrum-disorder">rarediseases.org/rare-diseases/auditory-neuropathy-spectrum-disorder</a></li></ul>	
<p><b>National Institute on Deafness and Other Communication Disorders</b></p> <ul style="list-style-type: none"><li>• Fact sheet on auditory neuropathy: <a href="https://www.nidcd.nih.gov/sites/default/files/Documents/health/hearing/AuditoryNeuropathy-508.pdf">www.nidcd.nih.gov/sites/default/files/Documents/health/hearing/AuditoryNeuropathy-508.pdf</a></li></ul>	

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