Auditory Neuropathy Spectrum Disorder: What is it?

Auditory Neuropathy Spectrum Disorder (ANSD) is a type of hearing loss that ranges from approximately normal access to sounds to not being able to hear at all. The particularity of this type of hearing loss is that most affected individuals can hear sounds but cannot understand what is being said.

The auditory system is composed of many parts that allow us to hear a variety of sounds (from soft to loud, from low to high pitches) and make sense of them (is it speech, music, noise?).

In the inner ear, we have small cells (outer hair cells) that amplify sounds and help us detect the different pitches. Those cells are usually working normally in ANSD.

Other cells (inner hair cells) convert the sound into a message that is delivered to the auditory nerve, which in turns sends that information to the brain. In some individuals with ANSD, some or all of the inner hair cells do not function properly. In other individuals, the nerve itself is not functioning properly.

The severity of ANSD ranges from mild to profound. Typically, children will hear some sounds but will have trouble understanding speech, particularly in noise environments. Many children will not be able to develop language without intervention.

Working closely with your audiologist and members of your child's team is necessary in order to find the best management strategy so that your child can learn language and thrive in life.

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AUDITORY NEUROPATHY SPECTRUM DISORDER

What is it and how do we manage it?

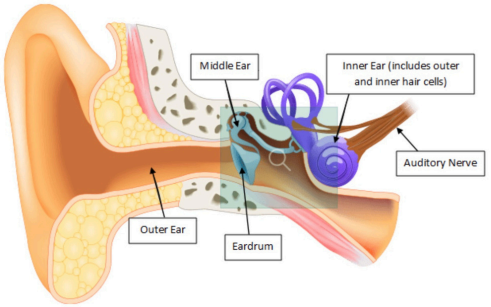
How is ANSD Diagnosed?

The diagnosis of ANSD is based on a series of tests sensitive to both cochlear and auditory nerve function. Some of these tests include:

- Tympanometry: measures the flexibility of the eardrum.
- Middle ear muscle reflexes: evaluates the muscles within the middle ear which protect the ear from loud sounds. The middle ear muscle reflexes are typically abnormal or absent in ANSD.
- Otoacoustic emissions: assess the function and of the outer hair cells. Otoacoustic emissions are usually normal in ANSD.
- Auditory brainstem responses: evaluates the inner hair cells and the auditory nerve functions. The ABR of individuals with ANSD is characteristic and unlike the ABR seen with other types of hearing problems.

Following the diagnosis of ANSD, other testing should be done to help specify the site of lesion of ANSD and if other body functions are affected:

- Genetic testing: ANSD could be caused by a genetic mutation. A genetic mutation exists when there is something abnormal with a gene. Several genetic mutations are known to cause ANSD and finding the right one can help with forming the correct management plan.
- MRI: An MRI will also be important to evaluate if the auditory nerve is present and of normal size.
- Cortical Auditory Evoked Potentials: The test can help understand how well your child is able to process sounds in their brain.
- Vestibular and balance testing: Many disorders of the ear can also affect balance and an early diagnosis is likely to lead to positive outcomes.
- Speech language evaluation: Regular speech and language evaluations are essential to evaluate the degree of severity of ANSD.



How Do We Manage ANSD?

Most children with ANSD need intervention in order to develop speech and language through hearing without any delay. There is a critical period for language development between the ages of 0 and 3 years. It is important to pay close attention to your child's progress, or lack of progress, during this time since their auditory system needs the correct stimulation in order to develop spoken language.

The team may recommend a hearing aid trial for your child. However, ANSD is usually a problem of clarity of speech, not access to sounds. In other words, hearing aids cannot really help with speech clarity.

While we know some children may benefit from amplification to learn language, we also know that this is the case for less than 10% of children. It may soon become clear that a combination of listening therapy with hearing aids will not be enough to help your child learn to hear and speak. At that time, the team will discuss the option of cochlear implantation with you. Independently of the assistive devices chosen, reducing noise in the listening environment of your child will be beneficial.

Questions?

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