



ANSD

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What is Auditory Neuropathy Spectrum Disorder (ANSD)?

- Auditory Neuropathy Spectrum Disorder (ANSD) is when there is damage to the inner ear nerve connection that carries the electrical message from a sound does not do so properly, causing the messages to be jumbled or lost.
- Some reasons this could happen include: family history of ANSD, premature birth or problems during birth such as low oxygen, jaundice, medications that may have been needed to save a baby's life after birth, and/or syndromes or neurological disorders.
- A child diagnosed with ANSD may not understand speech clearly, may seem to hear better at some times more than others, and may have trouble telling one sound from another.
- Symptoms of ANSD may not always be easily seen in a newborn and may appear as the child starts to develop.
- Children with ANSD may have normal hearing but still have difficulty understanding and speaking clearly. Other children may have hearing loss and ANSD.
- Other signs of ANSD may include limited or low language and speech, no response to sounds around them, fluctuating hearing, difficulty understanding in noise, poor speech articulation, poor attention and inability to follow directions, and difficulty learning.
- ANSD is diagnosed by an audiologist, using a combination of tests including Otoacoustic Emissions (OAE's), Auditory Brainstem Response (ABR) testing, and Middle Ear Reflexes. ANSD is diagnosed when OAE's are normal, but middle ear reflexes are absent and the ABR looks abnormal or is absent.
- A child diagnosed with ANSD will be referred to an Ear, Nose, and Throat (ENT) doctor who specializes in pediatrics. They will discuss the baby's health and family history and may order more testing to investigate the cause of the child's ANSD, including:
 - Imaging testing or an MRI to look for inner ear or nerve problems/malformations
 - Genetic testing
 - Neurologic testing to look for any other nerve problems, which can occur in approximately 10% of babies with ANSD
 - Vision testing, as sometimes vision and hearing issues occur together.
- Most children with ANSD will benefit from using an assistive listening device/remote microphone in school.
- Enrolling in early intervention (EI) and finding the language options that are the best fit for the family will help ensure good language and speech development and positive educational, social, and life outcomes for the child.