

# Auditory Neuropathy/ Dyssynchrony Spectrum Disorder (ANSD)

## What is ANSD?

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In ANSD, children have normal hair cells in the cochlea, but the hearing nerve is dyssynchronous (not synchronous). This means that instead of a smooth flow of information from the ear to the brain, the signals are not synchronized - and information will not be relayed to the brain in a consistent way. The amount of dyssynchrony can vary from person to person and can fluctuate over time.

## An Analogy

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You are driving between two cities a considerable distance apart. You have a talk radio station playing on the FM radio. As you leave the range of the radio station behind, the signal develops static. You can still hear the speech, but it is much harder to understand. Turning up the radio volume does little to improve your understanding of the speech presented on the talk radio station. For children with ANSD, the amount of dyssynchrony, can be relatively stable (about the same at all times) or can vary greatly from day to day.

## How is ANSD Diagnosed?

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In ANSD, the otoacoustic emissions (OAEs) test are usually normal. OAEs are sounds given off by the inner ear when the cochlea is stimulated by a sound. This test looks for a response to sound by the outer hair cells of the cochlea. See the section *Evaluating Hearing* in this toolkit for more information on OAEs.

In contrast, even a mild hearing loss would result in an absent OAE. A child may have ANSD and hearing loss.

However, children with ANSD exhibit no Auditory Brainstem Response (ABR). The ABR is performed by pasting electrodes on the head - similar to electrodes placed around the heart when an electrocardiogram is run - and recording brain wave activity in response to sound.

**To summarize: in ANSD, a normal OAEs are usually present but the ABR will be abnormal.**

### **Are there Risk Factors or any History Information that can Help in Identifying Children Who Potentially have ANSD?**

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Yes, but many children who have ANSD do not have any risk factors. The following risk factors are associated with ANSD: (1) hyperbilirubinemia (high level of bilirubin in the blood), (2) prematurity (25 – 36 weeks gestational age), and (3) perinatal asphyxia (baby's brain and other organs do not get enough oxygen during birth). ANSD can also run in families.

ANSD may be suspected when a child has high-quality hearing aids and still does not progress in speech and language development, despite wearing the hearing aids consistently.

### **How can Children with ANSD Vary in their Ability to use Hearing?**

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If you've seen one child with ANSD, you've seen one child with ANSD. There is wide variation in how well children with ANSD hear and understand. Most people with ANSD have both ears affected; however, some people have ANSD only on one side. Hearing levels vary from typical hearing to profound hearing loss.

Auditory abilities can be unpredictable and variable over time. **All children with ANSD have difficulty if any background noise is present.**

Some children with ANSD do not progress in speech and language development despite good intervention and wearing their hearing aids consistently. In this case, they may have better results with cochlear implants. Children who are not progressing and who do not receive cochlear implants may need to learn language through visual means (i.e., sign language, cued speech). Some children actually show a worsening of dyssynchrony symptoms over time. Other children seem to improve in their awareness of sound, but continue to have difficulty in noise and delayed language development.

**Although hearing aids have been found to be helpful in quiet environments, it is apparent that even if a little background noise is present, children with ANSD have great difficulty understanding.** Assistive technology, such as FM or DM technology, is *essential* in the classroom.

### **How can Cochlear Implants Benefit People with ANSD?**

Research has found that cochlear implantation is a viable treatment option for children who are not making progress with hearing aids. Researchers believe that it is possible that electrical stimulation, like that from a cochlear implant, can help to synchronize activity in the brain. Even for children who receive implants, it is important to allow them to gradually move from a visual language system to an auditory language. Abrupt removal of the communication system that a child has depended upon prior to receiving a cochlear implant is not recommended and could interrupt language development.

## **Are there any Communication Methods that Facilitate Development of Speech and Language in Children with ANSD?**

Learning speech and language through the auditory channel exclusively is very difficult for some children with ANSD. This is because it is difficult or impossible to achieve a clear and consistent auditory signal (unless the person has a cochlear implant) in a dyssynchronous auditory system. Research shows that the use of a visual communication system, such as sign language or cued speech, is recommended to develop language. Auditory-Verbal therapy by itself, before cochlear implantation, has not been observed to work as the sole method of teaching language to ANSD children.

*Information was adapted from Karen Anderson's excerpt of Hood, L. J. (2002). Auditory neuropathy/auditory dyssynchrony: New insights. The Hearing Journal (March). Posted to Supporting Success for Children with Hearing Loss Jan 2015. <http://successforkidswithhearingloss.com/ansd>*

## **References**

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Additional articles also available from the Alberta Hands & Voices lending library:

Hands & Voices: Auditory Neuropathy

[http://www.handsandvoices.org/comcon/articles/aud\\_neuropathy.htm](http://www.handsandvoices.org/comcon/articles/aud_neuropathy.htm)

National Institute on Deafness and Other Communication Disorders (NIDCD)

<http://www.nidcd.nih.gov/health/hearing/pages/neuropathy.aspx>

Yahoo Groups: Support to Parents of Children Diagnosed with ANSD

<https://groups.yahoo.com/neo/groups/AuditoryNeuropathy/info>