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AUDITORY NEUROPATHYSYNDROME DISORDER (ANSD)

What is auditory neuropathy syndrome disorder?

ANSD is a disorder which affects the normal synchronous activity in the auditory nerve but does not affect the ability of the inner ear to amplify sounds. Patients with ANSD frequently report that they can hear sounds but cannot understand what is being said. ANAD can be present in some infants, children and adults.

The typical audiologic presentation of ANSD is normal otoacoustic emissions (which measures outer hair cell function in the cochlear) and abnormal neural functioning at the level of the VIIIth nerve through ABR testing. ABR results for patients with ANSD may demonstrate either absent, severely abnormal wave forms, and/or demonstrate a flip of the waveform when polarity of the test signal is reversed. A certain percent of people (about 20%) do not have otoacoustic emissions so the absence of OAE's does not rule out the diagnosis of auditory neuropathy. Abnormal ABR testing is the most critical diagnostic indicator for a diagnosis of ANSD. Middle ear muscle acoustic reflexes are typically absent.

Neither ABR nor OAE's test hearing directly. The OAE measures the response of the outer hair cells of the cochlear to sound and the ABR is a test of neural synchrony. The use of the ABR as a test of hearing depends on the ability of the auditory neurons to respond to precisely to the timing of the stimulus and to respond synchronously to the auditory stimulus. When the auditory system is intact, these tests can predict hearing levels. When it is not intact, they cannot. However, they are very important in determining if the auditory system is intact.

Hearing testing using behavioral evaluations is critical in determining how a person with ANSD will perform in daily listening situations. Test results can be very variable. Some patients with ANSD have normal hearing thresholds and others have severe to profound hearing losses. Speech perception abilities can also vary significantly from mild problems in which the person hears well in many situations, to very significant problems in which the patient cannot understand speech. The ability to understand speech when there is competing noise is frequently poor.

Auditory neuropathy syndrome disorder is a relatively newly identified disorder. It has only been known for about the past 15 years. Prior to the identification of the disorder some patients with ANSD were identified with auditory processing disorders and others were diagnosed with hearing loss.

What causes ANSD?

Some infants with ANSD have had significant neonatal illness such as hyperbilirubinemia. It has also been related to other non-auditory neuropathies. There have been families in which siblings have been identified with ANSD indicating that there may be a genetic factor for some patients. As with many other disorders, it is not unusual for parents to be unaffected but to have a child with the disorder. Since ANAD can be very variable, it is not unusual for siblings to function differently, with one sibling having a more mild form of the disorder than another.

Is there more than one kind of auditory neuropathy syndrome disorder?

Most likely ANSD reflects more than a single disorder and research is currently being conducted to attempt to identify different varieties of ANSD but it has not yet possible to do so.

What other problems to patients with auditory neuropathy syndrome disorder?

Some patients may have other mild neuropathies outside of the auditory system but many only have auditory problems. Patients identified with ANSD should have a neurological evaluation to identify any problems that may exist.

Does the hearing loss caused by ANSD ever change?

Some patients with ANSD have fluctuations in their hearing. Occasionally the hearing loss may progress and become worse. It is not possible to predict the changes in hearing so every child needs to have hearing monitored carefully on a regular basis.

How is ANSD managed?

Management is different for every patient and is determined primarily by the behavioral hearing testing. If the child is hearing at normal hearing levels, we usually adopt a “watchful waiting” attitude and monitor the child’s hearing as well as speech and language development. If necessary, we may recommend an FM system for use in school and other situations in which there is competing noise.

If the behavioral testing indicates a hearing loss, we initially begin by fitting the child with hearing aids in the same way we would if the child had a hearing loss that was not caused by ANSD. If the child is doing well with hearing aids than, again, we adopt a “watchful waiting” attitude and monitor the child’s hearing and arrange for speech-language therapy. If we determine that the child is not doing well with hearing aids, then we may consider cochlear implants.

Many children with ANSD, like their peers with hearing loss of other causes, go to mainstream schools and receive on-going speech-language therapy and some educational assistance. Every child is different and every child needs to be managed individually.

REFERENCES

www.nidcd.nih.gov/health/hearing/neuropathy.asp