JOHN TRACY CENTER

Ideas & Advice
for Parents of Children with Hearing Loss

Addressing ANSD (Auditory Neuropathy Spectrum Disorder)

Early hearing loss is unfamiliar to most parents when their child is identified. As families look for answers they realize there are different types and levels of hearing loss. When told that their child has Auditory Neuropathy (AN) or Auditory Neuropathy Spectrum Disorder (ANSD) parents can begin discussing this term and obtaining specific advice. Exploring ANSD includes learning what it is, why it happens, how to manage it, which devices are considered, when communication approaches are suggested and where families can assist. Gradually parents can increase their understanding and become active partners in addressing their child's ANSD.

ANSD refers to a problem in the hearing nerve, or in the connection between the hearing nerve and the cochlea (the sense organ for sound). A child's hearing with ANSD can vary from normal to severe, occurring in one ear or both. Many parents describe inconsistent responses because their child sometimes appears to hear and sometimes does not. Listening in noisy environments can be especially difficult for a child with ANSD. Responses may also decrease when a child is very tired or not feeling well. When families share observations of a child's listening and language they assist in identifying how he hears and what supports his learning.

Causes of ANSD

Parents often wish to know why their child has ANSD. There can be many reasons for ANSD but sometimes a cause is not identified. The outer, middle and inner ear form during the first 20 weeks of pregnancy and then the hearing system begins to function so babies hear parents' voices even before birth. While the ears and neurological system are forming, a baby's hearing development is impacted by factors including genetics, oxygen, nutrition and the mother's health. After birth the hearing nerve may be damaged by illness or injury. Possible causes for ANSD include:

- Premature birth (25 to 36 weeks of pregnancy)
- Low birth weight
- Anoxia and hypoxia (lack of oxygen)
- Neurological conditions which might include cerebral palsy, microcephaly or hydrocephalus
- Syndromes such as Friedrich ataxia, Stevens-Johnson, Ehlers-Danlos, and Charcot-Marie-Tooth
- Hyperbilininemia (severe jaundice)

- Illnesses such as mumps and meningitis and some viruses
- Head injuries

Diagnosing ANSD

ANSD can be confirmed at any age and is determined by the results of tests that check how the cochlea and hearing nerve are working. When one test indicates the possibility of ANSD, at least one other test must be done to rule out or confirm a diagnosis. The tests used and the results indicating ANSD are:

Screening or diagnostic Otoacoustic Emissions Test (OAE) evaluates the response of the outer hair cells of the cochlea. When OAE testing confirms that these cells amplify sound, ANSD might be identified. Diagnostic OAE assesses more frequencies (tones) for additional information about hair cell function. In some cases and over time, OAE responses might decrease or not be obtainable.

Screening or diagnostic Auditory Brainstem Response Test (ABR) evaluates the response of the hearing nerve. Absent or abnormal response indicates the possibility of ANSD especially when normal results are obtained from OAE testing. Diagnostic ABR testing also might show a cochlear microphonic which means the cochlea is functioning and the hearing nerve is not.

Middle ear muscle reflex (acoustic reflex) checks the response of the hearing nerve to loud sounds. The reflex is absent or elevated when the nerve is unable to stimulate the muscle. Used with ABR and OAE, these results help identify ANSD.

Identifying Hearing Levels

Audiology evaluations are similar for any type of hearing loss but the results for a young child with ANSD may identify broad, not exact, levels of hearing. Hearing may also decrease or remain the same over time. Even when there is no change in hearing, testing should be ongoing. If there appears to be a significant difference in a child's responses to sounds, immediate re-testing can be done.

For some babies who have been diagnosed with ANSD, hearing might improve or become normal within the first 8 – 12 months. This is called Transient Auditory Neuropathy. If a child has Transient Auditory Neuropathy, assessments conducted regularly until the third birthday can document listening and spoken language progress.

Clearer responses might be obtained from Visual Reinforcement and Play Audiometry evaluations done when a child is able to participate by looking or doing an action when a sound is heard. Results can vary from test to test and identifying the hearing of a child with ANSD may require multiple evaluations.

Parents can report when their child responds to speech, what environmental sounds he hears, and if he seems not to hear as well during certain situations. Together parents and audiologists can define the child's broad hearing levels. What a child hears will become clearer as parents continue to communicate with the audiologists and combine home observations with hearing evaluation results.

Using Listening Devices (hearing aids, cochlear implants)

To help a child with ANSD develop listening and speech, audiologists may suggest listening devices. The cause of ANSD, other health conditions, hearing levels and speech recognition skills are considered in a device recommendation. The benefit of using a hearing aid is not certain since ANSD is a neural condition. During a trial period with hearing aids families can watch closely for reactions to sound, especially speech. If a hearing aid does not help significantly then a cochlear implant (CI) might be considered. The benefit from a CI is also not certain but tends to be more effective since it stimulates the hearing nerve. The results from a CI vary but families can share observations and ask about ongoing expectations as they work with the implant team. There are some children with ANSD who use both a hearing aid and a CI but some will use neither.

Considering Communication

There is not one specific communication approach for ANSD. Listening, language and speech

milestones can be assessed starting from infancy to determine a child's communication strengths. The approach initially used by the family might change as the child gains language and shows how he learns best.

If a child is responding to speech, families are encouraged to use spoken language. When a child seems to hear but not understand speech easily, parents might add strategies such as getting down on his level and using natural gestures to help him comprehend. More visual or auditory strategies might be suggested to enhance interactions.

If a child seems not to hear speech well, families can explore with service providers other communication approaches. They might consider sign language or total communication (a combination of speech and signs). After benefitting from a combined approach, a child may show a preference to use only speech or only sign.

Partnering with Service Providers

Auditory Neuropathy Spectrum Disorder involves a range of responses and skills. Parents can learn much about their child's hearing by working closely with audiologists and other service providers. Continuing research is providing information and insight for professionals studying ANSD. Together with families they can problem solve and devise plans. Listening devices, strategies and services can be identified as parents address a child's ANSD to help him acquire the language and skills needed for school!