

Hearing & Amplification

Causes of Hearing Loss

Auditory Neuropathy

Sensorineural hearing loss is the most common type of permanent hearing loss. A less common type of hearing loss is Auditory Neuropathy or Auditory Dysynchrony. With this type of hearing disorder, the inner ear or cochlea seems to receive sounds normally. However, signals leaving the cochlea may be disorganized or the hearing nerve itself may not process sound normally.

Auditory Neuropathy was first identified in the 1980s when advanced testing procedures became available to measure the action of the cells in the cochlea. There are some common features that are usually seen with auditory neuropathy, including:

- Hearing loss of some degree.
 - ~ The degree of hearing loss can vary widely from mild to profound loss.
 - ~ The degree of hearing loss is not predictable.
- Difficulty understanding speech, especially in noise.
- Speech understanding difficulties that are worse than can be predicted from other tests of hearing function.
- Hearing that appears to fluctuate from day-to-day or even from hour-to-hour.
- Other neuropathies that may affect coordination for activities like writing, running or talking.

To listen to a simulation of Auditory Neuropathy, use the following link and scroll to #2. The link is provided courtesy of Fan-Gang Zeng, Ph.D. University of California - Irvine: <http://www.ucihs.uci.edu/hesp/Simulations/simulationsmain.htm>.

What Causes Auditory Neuropathy?

There appear to be multiple causes for auditory neuropathy. It can be inherited genetically or caused by trauma or disease. The most common causes of auditory neuropathy that are known at this time are:

- Lack of oxygen (anoxia) at birth
- Hyperbilirubinemia requiring blood transfusion, associated with severe jaundice during the newborn period
- Infectious Disease such as mumps
- Immune Disorders
- Nonsyndromic Recessive Auditory Neuropathy (see Genetics Section for a further explanation of these terms)
- Neurological disorders such as Charcot-Marie-Tooth syndrome or Friedreich's Ataxia.

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See the following sites for more information about these disorders:

http://www.ninds.nih.gov/disorders/charcot_marie_tooth/charcot_marie_tooth.htm;

http://www.ninds.nih.gov/disorders/friedreichs_ataxia/friedreichs_ataxia.htm

There are large individual differences among children with this hearing disorder. For some, hearing may improve over time. This is most common when the cause of the disorder is hyperbilirubinemia.

How Is Auditory Neuropathy Diagnosed?

The diagnosis of auditory neuropathy is based on a certain pattern of results from a number of different hearing tests. Auditory Brainstem Response (ABR) and Otoacoustic Emission (OAE) results are used to find this condition.

An ABR test uses small bandaid-type electrodes placed behind each ear and the head. Sounds are presented to the ears using miniature earphones. The electrodes pick up responses from the hearing nerve and estimate how the cochlea and hearing nerve are responding to sound. The OAE test uses a miniature earphone to present a series of tones to the ear. A sensitive microphone measures an echo response from the inner ear. This information estimates how the inner ear responds to sound. The most striking finding with auditory neuropathy is that Otoacoustic Emissions (OAEs) are normal. This means that hair cells in the inner ear are working normally. At the same time, ABR responses are absent or very abnormal. This shows poor responses from the hearing nerve. It appears that only a small percentage of people with hearing loss have auditory neuropathy.

Depending upon what method is used for newborn hearing screening (OAEs or ABRs), some children with auditory neuropathy may pass an initial hearing screening conducted in the hospital. This may delay the identification of auditory neuropathy. Parents and physicians may not suspect hearing loss right away if an infant's response to sound changes from day to day.

As children get older, more hearing information becomes available. Ongoing testing may show poorer than expected speech understanding and great difficulty hearing in noisy backgrounds. A wide variety of hearing loss degrees and hearing loss shapes can be seen. Children with auditory neuropathy may also have hearing responses that fluctuate or worsen over time.

What Are The Treatment and Management Options for Auditory Neuropathy?

The location of the problem that causes auditory neuropathy is not known at this time. It also may be different from person to person. Therefore, medical treatment of auditory neuropathy is not currently available. Management typically involves either the use of hearing aids or cochlear implants.

Audiologists vary in their opinions about using hearing aids for a child with auditory neuropathy. Some children benefit from hearing aids. However, many children get limited hearing aid benefit or no benefit at all. At this time, there is no reliable way to predict who will and will not benefit. Hearing aids must be set carefully to prevent damage to the parts of the ear that receive sounds normally. Some professionals have recommended the use of Frequency Modulated (FM) systems. FM systems can improve listening in noisy backgrounds.

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Previously, it was thought that children with auditory neuropathy would not be good cochlear implant candidates. Now we know that some children benefit more from a cochlear implant than from hearing aids. Children with some types of genetically inherited auditory neuropathy may benefit more from cochlear implants than children with other causes of the disorder. We will know more as future genetic and hearing science research becomes available.

It is safe to say that no single teaching approach fits all children with auditory neuropathy. It is helpful to work closely with a team of professionals. A team can help parents observe their child and determine what techniques bring about the best responses. Some children benefit from teaching that focuses only on learning to listen and speak. Sometimes, a child with auditory neuropathy has great difficulty understanding what is heard. These children benefit from visual communication approaches (i.e., sign language or Cued Speech).

It is difficult to predict how any one child will do. It is important to be flexible. With guidance, families can try an approach and evaluate how well it is working over time. Approaches can be modified to bring about success. Teachers and other professionals can help families to decide whether or not an approach is working. If it works, the child should make steady progress. Communication and understanding should improve over time. Frustration around communication should lessen. This gives families increasing confidence and success in communicating with their child.

FOR MORE INFORMATION:

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

http://www.medschool.lsuhs.edu/otorhinolaryngology/deafness_article1.asp

<http://auditoryneuropathy.tripod.com/ANindex.html>

<http://www.raisingdeafkids.org/hearingloss/types/an/>