Auditory Neuropathy

What is auditory neuropathy?

Auditory neuropathy is a hearing disorder in which sound enters the inner ear normally but the transmission of signals from the inner ear to the brain is impaired. It can affect people of all ages, from infancy through adulthood. The number of people affected by auditory neuropathy is not known, but the condition affects a relatively small percentage of people who are deaf or hearing-impaired.

People with auditory neuropathy may have normal hearing, or hearing loss ranging from mild to severe; they always have poor speech-perception abilities, meaning they have trouble understanding speech clearly. Often, speech perception is worse than would be predicted by the degree of hearing loss. For example, a person with auditory neuropathy may be able to hear sounds, but would still have difficulty recognizing spoken words. Sounds may fade in and out for these individuals and seem out of sync.

What causes auditory neuropathy?

Although auditory neuropathy is not yet fully understood, scientists believe the condition probably has more than one cause. In some cases, it may involve damage to the inner hair cells—specialized sensory cells in the inner ear that transmit information about sounds through the nervous system to the brain. Other causes may include faulty connections between the inner hair cells and the nerve leading from the inner ear to the brain, or damage to the nerve itself. A combination of these problems may occur in some cases. Although outer hair cells—hair cells adjacent to and more numerous than the inner hair cells—are generally more prone to damage than inner hair cells, outer hair cells seem to function normally in people with auditory neuropathy.

What are the roles of the outer and inner hair cells?

Outer hair cells help amplify sound vibrations entering the inner ear from the middle ear. When hearing is working normally, the inner hair cells convert these vibrations into electrical signals that travel as nerve impulses to the brain, where the impulses are interpreted as sound.

Are there risk factors for auditory neuropathy?

Several factors have been linked to auditory neuropathy in children. However, a clear cause and effect relationship has not been proven. Some children who have been diagnosed with auditory neuropathy experienced certain health problems as newborns, or during or shortly before birth. These problems include jaundice, premature birth, low birth weight, and an inadequate supply of oxygen to the unborn baby. In addition, some drugs that have been used to treat medical complications in pregnant women or newborns may damage the inner hair cells in the baby's ears, causing auditory neuropathy.

Auditory neuropathy runs in some families, which suggests that genetic factors may be involved in some cases. Some people with auditory neuropathy have neurological disorders that also cause problems outside of the hearing system. Examples of such disorders are Charcot-Marie-Tooth syndrome and Friedreich's ataxia.

How is auditory neuropathy diagnosed?

Health professionals, including otolaryngologists (ear, nose, and throat doctors), pediatricians, and audiologists, use a combination of methods to diagnose auditory neuropathy. These include tests of auditory brainstem response (ABR) and otoacoustic emissions (OAE). The hallmark of auditory neuropathy is a negligible or very abnormal ABR reading together with a normal OAE reading. A normal OAE reading is a sign that the outer hair cells are working normally.

An ABR test monitors brain wave activity in response to sound using electrodes that are placed on the person's head and ears. An OAE test uses a small, very sensitive microphone inserted into the ear canal to monitor the faint sounds produced by the outer hair cells in response to stimulation by a series of clicks. ABR and OAE testing are painless and can be used for newborn babies and infants as well as older children and adults. Other tests may also be used as part of a more comprehensive evaluation of an individual's hearing and speech-perception abilities.

Does auditory neuropathy ever get better or worse?

Some newborn babies who have been diagnosed with auditory neuropathy improve and start to hear and speak within a year or two. Other infants stay the same, while some get worse and show signs that the outer hair cells no longer function (otoacoustic emissions). In adults with auditory neuropathy, hearing can remain stable, fluctuate up and down, or progressively worsen, depending on the underlying cause.

What treatments, devices, and other approaches can help people with auditory neuropathy to communicate?

Researchers are still seeking effective treatments for people with auditory neuropathy. Meanwhile, professionals in the hearing field differ in their opinions about the potential benefits of hearing aids, cochlear implants, and other technologies for people with auditory neuropathy. Some professionals report that hearing aids and personal listening devices such as frequency modulation (FM) systems are helpful for some children and adults with auditory neuropathy. Cochlear implants (electronic devices that compensate for damaged or nonworking parts of the inner ear) may also help some people with auditory neuropathy. However, no tests are currently available to determine whether an individual with auditory neuropathy might benefit from a hearing aid or cochlear implant.

Debate also continues about the best ways to educate and provide communication skills for children who have hearing impairments such as auditory neuropathy. However, most hearing health experts agree that parents should work with a team of professionals who considers the situation and options for each child as well as the child's family members and caregivers. Most also agree that parents and caregivers should interact often with infants who have auditory neuropathy by holding, facing, smiling at, and responding to the child.

There are two main philosophies of how to teach infants and children with auditory neuropathy how to communicate. One philosophy favors using sign language as the child's first language. The second philosophy encourages the use of listening skills and skills in spoken English together with technologies such as hearing aids and cochlear implants. A combination of these two approaches can also be used. Some health professionals believe it may be especially difficult for children with auditory neuropathy to learn to communicate only through spoken language because their ability to understand speech is often greatly impaired. Adults with

auditory neuropathy and older children who have already developed spoken language may benefit from learning how to speech read (also known as lip reading).

What research is being done for auditory neuropathy?

Scientists are working to understand the causes of auditory neuropathy, and are searching for genes that may be involved in causing this condition. Researchers are also continuing to investigate the potential benefits of cochlear implants for children with auditory neuropathy, and are examining why cochlear implants may benefit some people with the condition but not others.

Where can I get more information?

NIDCD maintains a directory of organizations that can answer questions and provide printed or electronic information on auditory neuropathy. Please see the list of organizations at www.nidcd.nih.gov/directory.