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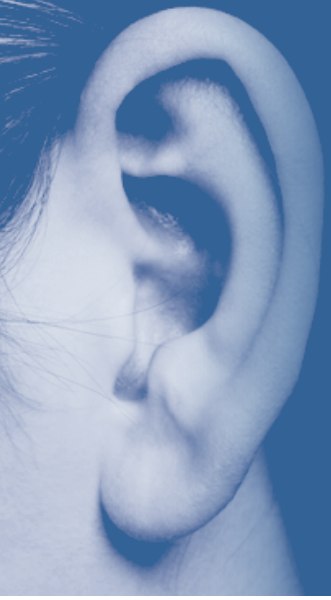

National Institute on Deafness and Other Communication Disorders

NIDCD Fact Sheet | **Hearing and Balance**

Auditory Neuropathy

What is auditory neuropathy?

Auditory neuropathy is a hearing disorder in which the inner ear successfully detects sound, but has a problem with sending sound from the ear to the brain. It can affect people of all ages, from infancy through adulthood. The number of people affected by auditory neuropathy is not known, but current information suggests that auditory neuropathies play a substantial role in hearing impairments and deafness.

When their hearing sensitivity is tested, people with auditory neuropathy may have normal hearing or hearing loss ranging from mild to severe. They always have poor speech-perception abilities, meaning that they have trouble understanding speech clearly. People with auditory neuropathy have greater impairment in speech perception than hearing health experts would predict based upon their degree of hearing loss on a hearing test. 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 or seem out of sync for these individuals.

What causes auditory neuropathy?

Researchers report several causes of auditory neuropathy. In some cases, the cause 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. In other cases, the cause may involve damage to the auditory neurons that transmit sound information from the inner hair cells to the brain. Other possible causes may include inheriting genes with mutations or suffering damage to the auditory system, either of which may result in faulty connections between the inner hair cells and the auditory nerve (the nerve leading from the inner ear to the brain), or damage to the auditory nerve itself. A combination of these problems may occur in some cases.

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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 brain interprets the impulses as sound.

Although outer hair cells—hair cells next to and more numerous than 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.

Are there risk factors for auditory neuropathy?

There are several ways that children may acquire auditory neuropathy. Some children diagnosed with auditory neuropathy experienced particular health problems before or during birth or as newborns. These problems include inadequate oxygen supply during or prior to birth, premature birth, jaundice, low birth weight, and dietary thiamine deficiency. In addition, some drugs used to treat pregnant women or newborns may damage the baby's inner hair cells, causing auditory neuropathy. Adults may also develop auditory neuropathy along with age-related hearing loss.

Auditory neuropathy runs in some families, and in some cases, scientists have identified genes with mutations that compromise the ear's ability to transmit sound information to the brain. Thus, inheritance of mutated genes is also a risk factor for auditory neuropathy.

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.

Health professionals—including otolaryngologists (ear, nose, and throat doctors), pediatricians, and audiologists—use a combination of methods to diagnose auditory neuropathy.

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 an absent 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 uses electrodes placed on a person's head and ears to monitor brain wave activity in response to sound. 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 auditory stimulation. 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 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 (abnormal otoacoustic emissions). In people with auditory neuropathy, hearing sensitivity can remain stable, get better or worse, or gradually 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. No tests are currently available, however, 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 improve communication skills in infants and children who have hearing impairments such as auditory neuropathy. One approach favors sign language as the child's first language. A second approach encourages the use of listening skills—together with technologies such as hearing aids and cochlear implants—and spoken language. A combination of these two approaches may 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 severely impaired. Adults with auditory neuropathy and older children who have already developed spoken language may benefit from learning how to speechread (also known as lip reading).

What research is being done on auditory neuropathy?

Scientists have identified genes involved in causing some cases of auditory neuropathy, and are working to identify what goes wrong in the auditory system when a person inherits a mutant gene. 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.

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Where can I find additional information about auditory neuropathy?

The NIDCD maintains a directory of organizations that provide information on the normal and disordered processes of hearing, balance, taste, smell, voice, speech, and language. Visit the NIDCD website at *https://www.nidcd.nih.gov/directory* to search the directory.

More NIDCD fact sheets on Hearing and Balance:

- American Sign Language
- Assistive Devices for People with Hearing, Voice, Speech, or Language Disorders
- Cochlear Implants
- Telecommunication Relay Services
- Usher Syndrome
- Your Baby's Hearing Screening

Visit the NIDCD website at *https://www.nidcd.nih.gov* to read, print, or download fact sheets.

For more information, contact us at:

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