

Hearing loss - auditory neuropathy

Auditory neuropathy is a rare type of hearing loss that is caused by an abnormality in the transmission of nerve impulses travelling from the inner ear to the brain. Both ears are usually affected, and the hearing loss ranges from mild to severe. In some cases, the affected person can hear, but has difficulty in understanding spoken words, particularly in noisy environments. Hearing and understanding speech may be improved by the use of hearing aids or cochlear implants.

Symptoms can occasionally fluctuate; for example, a person may experience both improvements and deteriorations in their hearing. The long-term outlook varies too. For example, the hearing of a person diagnosed with auditory neuropathy may improve, deteriorate or remain unchanged, and there is no way to predict the outcome. The cause is unknown and there is no cure. Auditory neuropathy is sometimes referred to as a form of neural or nerve deafness.

This is a rare form of hearing loss and many of the symptoms can also be caused by more common types of hearing loss.

Symptoms

The symptoms of auditory neuropathy vary from one individual to the next, but can include:

- Mild to severe hearing loss
- Sounds tend to fade in and out
- Understanding spoken words (speech perception) may be difficult
- Hearing may be normal, but with poor speech perception
- Speech perception may worsen in noisy environments
- Symptoms may fluctuate.

How sounds are experienced

The different structures involved in hearing include:

- **Outer ear** – the part you can see. Sounds are funnelled into the middle ear.
- **Middle ear** – separated from the outer ear by the eardrum. The eardrum moves in response to sound waves. The movement is amplified by tiny bones within the middle ear.
- **Inner ear** – the amplified sound waves are picked up by a little spiral-shaped organ called the cochlea. Tiny hair-like nerve endings within the cochlea sense the vibration and pass the message – interpreted into electrical impulses – to the cochlear nerve.
- **Brain** – the impulse is sent by the cochlear nerve along the eighth cranial nerve and finally to the auditory pathways in the brain stem. The brain stem is located at the base of the skull, and is considered the 'bridge' between the brain and the spinal cord. The electrical impulses are interpreted by the brain and the sound is 'heard'.

The cause is unknown

In auditory neuropathy, some of the structures within the ear may be working perfectly, but the information from the ear may be distorted on its way to the brain. The cause of auditory neuropathy isn't known, but researchers suspect that a number of factors may be responsible, including:

- Damaged hair cells within the inner ear
- Damage to the connections between the hair cells and the cochlear nerve
- Damage to the cochlear nerve
- Damage to the eighth cranial nerve (the combination of the cochlear and vestibular nerves)

- Damage to auditory pathways in the brain stem.

Risk factors

People of any age and either gender can be affected, but known risk factors for children include:

- **Health problems in utero** – such as reduced oxygen supply to the fetus.
- **Particular drugs** – certain drugs that are used to treat obstetric complications are thought to affect the inner hair cells in susceptible babies.
- **Health problems at birth** – such as premature birth, jaundice or low birth weight.
- **Other disorders** – in rare cases, auditory neuropathy is associated with other neurological disorders such as Charcot-Marie-Tooth disorder and Friedrich's ataxia.
- **Other affected family members** – some children with auditory neuropathy have similarly affected family members. This suggests a genetic component.

Diagnosis methods

Auditory neuropathy is diagnosed using a number of tests including:

- **Auditory brainstem response (ABR)** – electrodes on the ears and head detect brain wave patterns when the person is exposed to a series of sounds. Typically, a person with auditory neuropathy has little or no response.
- **Otoacoustic emissions (OAE)** – a tiny microphone is placed inside the ear canal to check whether the cochlear hair cells are working. If so, the microphone will pick up the faint sounds made by the hairs as they respond to noise. Typically, a person with auditory neuropathy has some properly functioning hair cells.
- **Other hearing tests** – such as speech recognition. Generally speaking, a person with auditory neuropathy performs poorly when the test is given with background noise.
- **Other tests** – to rule out other conditions that may present similar symptoms, such as multiple sclerosis.

Treatment options

There is no cure for auditory neuropathy. The variable nature of this hearing disorder means that some treatments work in some cases and not others, which may explain why the medical field is currently divided on best treatment practice. Unfortunately, there are no tests to predict whether a particular treatment will work or not – the patient may have to experiment to find out. Options include:

- **Hearing aids** – to amplify all sounds. Generally, adults with auditory neuropathy don't find hearing aids particularly helpful.
- **Frequency modulation (FM) systems** – a portable receiver and headset that amplifies sounds without the need for wiring.
- **Cochlear implant** – a surgically implanted device that stimulates the nerves of the inner ear. These implants have been shown to help many people with this condition improve their ability to understand speech.

Communication skills

The medical field is also divided on how to teach children with auditory neuropathy to communicate. One school of thought advocates teaching the child sign language; the other suggests that the child should be equipped with devices (such as hearing aids) and given support to learn to listen and speak. Some medical professionals believe that a combination of the two approaches is best, because there is no way of knowing whether the child's hearing will improve, stay the same or deteriorate.

Exposure to spoken language is important in case the child's hearing improves. For adults who already have spoken language skills, learning to use supplemental cues such as lip reading may help.

Where to get help

- Your doctor
- Ear, nose and throat specialist (your doctor can refer you)
- Audiologist (hearing scientist)
- Otolaryngologist (ear, nose and throat specialist)
- The Royal Victorian Eye and Ear Hospital Tel. (03) 9929 8666

Things to remember

- This is a rare form of hearing loss and many of the symptoms can also be caused by more common types of hearing loss.
- Auditory neuropathy is a type of hearing loss that is caused by a disruption of nerve impulses travelling from the inner ear to the brain.
- In some cases, the affected person can hear, but has difficulty in understanding spoken words, particularly in noisy environments.
- The variable nature of this hearing disorder means that some treatments work in some cases and not others.

This page has been produced in consultation with, and approved by:

Royal Victorian Eye and Ear Hospital (RVEEH)

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