

Fact Sheet

Auditory Neuropathy

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How Does Our Hearing Work? To understand auditory neuropathy it is critical to understand how we hear. In simple terms the ear is shaped to “catch” sound. The outer ear directs the sound down the ear canal to the ear drum. As sound moves through the ear drum into the middle ear, the tiny bones of the middle ear amplify the sound. The sound is “pushed” into the cochlea of the inner ear.

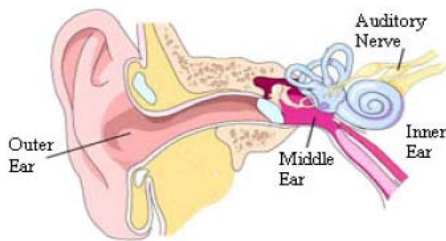


Image retrieved on March 23, 2007 from <http://www.workplacegroup.net/article-ear-anatomy.htm>

The outer hair cells of the cochlea serve as an amplifier while the inner hair cells turn the sounds waves into electrical impulses. The impulses are transmitted to the auditory nerve, which carries the sound to the brain, where it is interpreted. Something can go wrong anywhere in the process.

What Is Auditory Neuropathy? How Does It Relate To The Hearing Process? Auditory neuropathy refers to a condition in which it

appears that sound enters the ear normally, is transmitted through the middle ear and enters the cochlea. However, abnormal responses in the area of the inner hair cells, auditory neurons, or the 8th cranial nerve occur and so “sound” is not appropriately transmitted to the brain for interpretation. Given questions about whether this is a true neuropathy has given rise to the term “auditory dys-synchrony.” “Auditory dys-synchrony and “auditory neuropathy” have come to be synonymous.

Potential Risk Factors: It is important to note that some who are diagnosed with auditory neuropathy might not have any of these risk factors in their histories. Nor does the presence of one of the factors automatically mean the person will have the condition. Risk factors may include: (a) extreme prematurity, (b) neonatal hyperbilirubinemia; (c) anoxia; (d) Infections / high fever; (e) mitochondrial and genetic disorders; (f) congenital brain abnormalities; and (g) genetics or family history

What Are Characteristics Of Auditory Neuropathy?

1. Hearing can range from normal to a profound loss as measured by pure-tone thresholds

2. Difficulty understanding speech/language, especially in noise
 3. Difficulty understanding speech/language is disproportionate to the identified degree of hearing loss
 4. Child might respond to sound, but sound may be distorted and difficult for the brain to interpret
 5. May lead to failure to develop normal auditory behaviors and oral language
 6. Hearing may sometimes seem to change with the child sometimes appearing to be hear better than at other times
 7. Some children may get worse over time: other may improve
- Conventional hearing aids have limited success
 - Communication partners should be instructed to speak clearly to produce more intelligible speech than typically occurs in casual conversation.
 - Use of visual communication methods (e.g., Cued speech, sign language, signed English) can aid in language development
 - Improve listening environment to aid in making the most of speech heard (e.g., use wall and floor coverings, reduce background noise, use personal FM system, use a sound-field system)
 - Cochlear Implants are a management option

Implications for Management:

- Management is difficult due to no standard treatment

How is Auditory Neuropathy Diagnosed?	
There is a combination of normal otoacoustic emissions with absent or severely abnormal auditory brainstem responses. Tests that might be used as part of the diagnostic process are:	
Acoustic Reflex Threshold Testing (ARTs)	Measures middle ear muscle reflexes. Deviation from normal thresholds can indicate possible abnormalities of the auditory nerve.
Otoacoustic Emissions (OAEs)	Tests the outer hair cells of the cochlea. Measured by presenting a series of clicks to the ear thru a probe inserted in the ear canal.
The absence of acoustic reflexes with normal OAEs would warrant referral for further evaluation, we could include an automated brainstem response	
Auditory Brainstem Response (ABR)	Abnormal results indicate that the auditory nerve and/or brainstem might not correctly process sounds. Electrodes are attached to the scalp to measure electrical activity in response to sound clicks

Selected Resources

Auditory Neuropathy: <https://www.nidcd.nih.gov/health/hearing/auditory-neuropathy>

Auditory Neuropathy and auditory dys-synchrony:

https://medschool.lsuhsu.edu/otorhinolaryngology/kresge_about.aspx

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Fact Sheets from the Colorado Services for Children and Youth with Combined Vision and Hearing Loss Project are to be used by both families and professionals serving individuals with vision and hearing loss. This information applies to children and youth, birth through 21 years of age. The purpose of the Fact Sheet is to give general information on a specific topic. More specific information for an individual student can be provided through personalized technical assistance available from the Colorado Project. For more information call (303) 866-6681 or (303) 866-6605. Updated: 1/09