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PATH MEDICAL Academy

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## Auditory Neuropathy (AN)/Auditory Neuropathy Spectrum Disorder (ANSD)

**Auditory neuropathy** is a disorder characterized by having normal outer hair cell function and abnormal function of the auditory nerve. It can be found in individuals of all ages, from infants to adults. It is typically bilateral but can occur unilaterally as well.

It probably is not a new phenomenon as there are descriptions in the literature from the late 1980's e.g., Davis & Hirsh, 1979; Worthington & Peters, 1980; Kraus, Ozdamar, Stein & Reed, 1984; Lenhardt, 1981. These descriptions include reports of inconsistent pure tone thresholds, abnormal ABRs, extreme difficulty understanding speech, word recognition scores much worse than expected from the degree of hearing loss and no help from hearing aids. It was not until Otoacoustic Emissions (OAEs) came into the clinics that enabled differentiation of sensory versus neural hearing loss. In1996, the term auditory neuropathy was introduced by A. Starr and colleagues who described ten individuals with progressive hearing losses, attributable to disease of the VIIIth nerve, showing symptoms in childhood or young adults and had present OAEs, absent or grossly abnormal ABRs, absent acoustic reflexes and speech intelligibility worse than predicted from hearing levels.

The prevalence is estimated to be from 1-10% of individuals with hearing loss. There is a range because of the various types of hearing impairments and the different clinical criteria used across studies (Moser & Starr, 2016). The different clinical criteria led many to use the term anytime there was present otoacoustic emissions (OAEs) and/or present cochlear microphonic (CM) and absent or abnormal auditory brainstem responses (ABRs). Most often, there was not neurologic corroboration that the VIIIth nerve was involved. Rapin & Gravel (2003, 2006) cautioned about the indiscriminate use of the term neuropathy without knowing if there was VIIIth nerve involvement. That is, hearing test results alone are not

sufficient to label an auditory disorder. They went on to say that the term auditory neuropathy should not be used when the main site of pathology is in the brainstem or more centrally in the auditory pathway. The term neural hearing loss should be used then. The term auditory neuropathy should be reserved for patients in whom there is "evidence that the pathology involves the spiral ganglion cells or their axons (8th nerve)". This advice has not been heeded over the years and in 2008, a panel coined the term Auditory Neuropathy Spectrum Disorder (ANSD) because of the wide range of etiologies associated with similar presentations of test results.

**The audiologic evaluation** should encompass a test battery that includes OAEs for assessment of outer hair cell function, ABR to assess VIIIth nerve dysfunction and look for the CM, acoustic reflex testing, and pure tone audiometry at minimum. Depending on the patient's age and abilities other tests should be included such as word recognition in quiet and with ipsilateral noise, TEOAE suppression, masking level difference (MLD), tone decay, gap detection, sound localization, and frequency discrimination or the ability to detect a change in frequency.

Note: for the ABR, stimulus polarity should be both rarefaction and condensation to look for the CM inversion with changes in the polarity of the stimulus. If using insert earphones, care must be taken to ensure that the response is the CM and not stimulus artifact. This is accomplished by clamping the tubing of the insert earphone and repeating the testing. If the response does not disappear with clamping, the response is due to stimulus artifact.

The following are the expected outcomes for those with AN:

| TEST | Expected Outcome |
|---|---|
| OAEs | Normal generally, although the OAEs can be absent or can disappear over time |
| ABR | Abnormal or absent ABR; Cochlear Microphonic present |
| Tympanometry and middle ear muscle reflexes | Normal tympanogram with absent ipsilateral and contralateral reflexes but non-acoustic reflexes are present |
| MLD | 0 dB HL |
| Pure tone thresholds | Normal, mild to moderate as described by Starr, but now includes severe/profound hearing losses of all configurations. Hearing loss is usually stable but there are reports of fluctuations and progression. |

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|---|---|
| Speech recognition in quiet | Variable; slightly reduced to greatly reduced |
| Speech recognition in noise | Generally poor |
| TEOAE suppression | No suppression |
| Tone decay, gap detection, sound localization, frequency discrimination | Typically, all have abnormal findings |

**Management** usually starts with mild gain hearing aids and often progresses to cochlear implant when there is evidence that the hearing aids are not of benefit. However, given the inclusion of a broad range of pathologies e.g. inner hair cell disorder, VIIIth nerve aplasia, and hyperbilirubinemia that are now called ANSD it presents challenges for management. For example, hearing can improve in cases of hyperbilirubinemia and cochlear implants may not be successful when there is aplasia of the VIIIth nerve. Therefore, in addition to the audiologic evaluation, ensuring that there is a multidisciplinary approach including developmental and communication assessments and otologic, neurologic, genetic, ophthalmologic and neurologic evaluations to determine the etiology can help to determine the most appropriate management.

## References


Davis H, Hirsh SK. The audiometric utility of brain stem responses to low-frequency sounds. Audiology. 1976 May-Jun;15(3):181-95. doi: 10.3109/00206097609071775. PMID: 938332.

Kraus N, Ozdamar O, Heydemann PT, Stein L, Reed NL. Auditory brain-stem responses in hydrocephalic patients. Electroencephalogr Clin Neurophysiol. 1984 Jul;59(4):310-7. doi: 10.1016/0168-5597(84)90048-0. PMID: 6203720.

Lenhardt ML. Childhood central auditory processing disorder with brainstem evoked response verification. Arch Otolaryngol. 1981 Oct;107(10):623-5. doi: 10.1001/archotol.1981.00790460035011. PMID: 7283826.

Moser T, Starr A. Auditory neuropathy--neural and synaptic mechanisms. Nat Rev Neurol. 2016 Mar;12(3):135-49. doi: 10.1038/nrneurol.2016.10. Epub 2016 Feb 19. PMID: 26891769.

Rapin I, Gravel J. "Auditory neuropathy": physiologic and pathologic evidence calls for more diagnostic specificity. Int J Pediatr Otorhinolaryngol. 2003 Jul;67(7):707-28. doi: 10.1016/s0165-5876(03)00103-4. PMID: 12791445.

Rapin I, Gravel JS. Auditory neuropathy: a biologically inappropriate label unless acoustic nerve involvement is documented. J Am Acad Audiol. 2006 Feb;17(2):147-50. doi: 10.3766/jaaa.17.2.7. PMID: 16640067.

Starr A, Picton TW, Sininger Y, Hood LJ, Berlin CI. Auditory neuropathy. Brain. 1996 Jun;119 ( Pt 3):741-53. doi: 10.1093/brain/119.3.741. PMID: 8673487.

Worthington DW, Peters JF. Electrophysiologic audiometry. Ann Otol Rhinol Laryngol Suppl. 1980 Sep-Oct;89(5 Pt 2):59-62. doi: 10.1177/00034894800890s517. PMID: 6786193.IHC