

## DIAGNOSIS AND MANAGEMENT OF PEDIATRIC AUDITORY NEUROPATHY

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Auditory neuropathy (AN) is a complex auditory disorder, which results in a distinct pattern of audiometric test results and a wide range of symptoms and auditory complaints. The range of functional auditory abilities in individuals with AN is vast and may include normal hearing sensitivity and limited auditory complaints, fluctuating hearing sensitivity and difficulty understanding speech, especially in noise, or profound hearing loss and inability to understand even amplified speech in quiet despite evidence of normal cochlear outer hair cell function.

Early reports of auditory neuropathy described the paradox of individuals with normal pure-tone audiograms but absent or severely abnormal auditory brainstem responses (ABR). In some cases, individuals demonstrated pure-tone audiograms with mild to moderately-elevated detection thresholds, absent/abnormal ABRs, and absent middle ear muscle (acoustic) reflexes. The addition of otoacoustic emissions (OAEs) to the audiometric test battery as well as improved techniques for recording the inner-ear pre-neural cochlear microphonic (CM) from the ABR helped clarify the puzzling physiology of this disorder. Individuals with auditory neuropathy commonly demonstrate presence of OAEs and/or CM ~ responses generated by the cochlear outer hair cells. Given normal cochlear outer hair cell function and absent or abnormal auditory nerve conduction, the site of dysfunction could be localized to inner hair cells, or inner hair cells-auditory nerve fiber synapse, or the auditory nerve itself. There is only limited evidence of inner hair cell abnormality in humans, but compelling genetic evidence of inner hair cell-auditory nerve fiber synaptic dysfunction, and unequivocal magnetic

resonance imaging (MRI) evidence of auditory nerve abnormality. For an excellent review of auditory neuropathy, see Yvonne Sininger and Arnold Starr (editors): *Auditory Neuropathy A New Perspective on Hearing Disorders*.

Although the scientific and professional community have made significant progress in understanding the nature and variable consequences of AN, optimal clinical management options for individuals with this disorder remain elusive, especially for infants and very young children. For infants with "typical" sensory hearing loss who are identified through newborn hearing screening, excellent and detailed guidelines for identification, diagnosis, and early intervention are widely accepted (Joint Committee on Infant Hearing, 2007). These guidelines do not address, however, some of the specific challenges in providing families with appropriate management options for infants with AN. For example, early amplification fitting is a recommended option for infants with "typical" sensory hearing loss, and fitting protocols can be based on the infant's frequency-specific hearing thresholds as estimated by electrophysiological measures such as ABR to frequency-specific signals or auditory steady state response (ASSR). In contrast, neither ABR nor ASSR provide accurate predictions of frequency-specific hearing thresholds in individuals with AN. In newborns and very young infants with AN, there are currently no audiometric measures, either behavioral or physiologic, which predict audibility or degree of auditory deficit the baby will experience. The uncertainty and ambiguity about the developmental impact of AN for an infant is frustrating and anxiety-provoking for parents and professionals alike.

To address the unique needs of infants and young children with AN, an international panel of experts in audiology, hearing science, medical genetics, neonatology, and neurology gathered in Como, Italy, in June 2008 to discuss and debate issues relevant to identification and management of infants and young children with AN. The panel developed a set of guidelines addressing terminology, diagnostic criteria, recommended comprehensive assessments, recommended audiological test battery, recommended amplification strategies, special considerations for cochlear implantation, recommended habilitation for communication development, screening newborns for auditory neuropathy, monitoring infants with “transient” auditory neuropathy, and counseling families of infants with this disorder. Some of the more challenging issues that the panel addressed are summarized below.

### **Terminology.**

The term “auditory neuropathy” was coined by Starr et al (1996) to describe perplexing auditory symptoms exhibited by a group of 10 patients from various clinical sites; seven of these patients exhibited generalized peripheral neuropathy. Other terms for this disorder have been proposed including auditory neuropathy/auditory dys-synchrony, auditory neuropathy type I (pre-synaptic) and type II (post-synaptic), and auditory nerve disorder. The panel considered these multiple designations as well as those proposed at the conference and concluded that “auditory neuropathy spectrum disorder” was the preferred usage. Three principles drove this consensus. First, despite potentially inexact usage, the term “auditory neuropathy” has gained widespread acceptance and usage, both in the professional literature and among parent/consumer organizations. Renaming the disorder could lead to confusion for patients and other professionals whereas retaining current terminology would provide continuity for the scientific community. Second, the expression of this disorder in everyday listening and communication behaviors encompasses a spectrum ranging from

limited or mild effects (complaints of difficulty “hearing” in noisy listening conditions) to profound effects (inability to “hear” in any listening condition, functionally “deaf”). Finally, the term “spectrum” was felt to expand the concept of this disorder to include sites of lesion other than the auditory nerve. In practice, especially when communicating with families, using the shortened term, “auditory neuropathy” would appear appropriate to avoid confusion with other childhood conditions that present on a spectrum.

### **Diagnostic Criteria.**

Auditory neuropathy is characterized by evidence of cochlear hair cell (sensory) function and absence/ abnormality of auditory nerve function. Therefore, the (minimum) test battery needed to diagnose AN requires tests of cochlear hair cell (sensory) function and auditory nerve function is as follows:

1. Tests of cochlear hair cell (sensory) function:
  - a. Otoacoustic emissions: Standard screening or diagnostic protocol using Transient-evoked OAEs (TEOAEs) or Distortion Product OAEs (DPOAEs), and/or
  - b. Cochlear microphonics: Click-evoked ABR using insert earphones and separate polarity recordings to responses of positive vs. negative stimulus.
2. Test of auditory nerve function:
  - a. Click-evoked ABR to high-level stimuli (80-90 dB nHL).

### **Special Considerations in Diagnosing Infants with Auditory Neuropathy**

When using these test procedures in newborns and very young infants, recording conditions must be optimum to obtain valid, artifact-free, unambiguous test results. Infants should be quietly sleeping in either natural or sedated sleep to avoid movement artifact or “noisy” recordings. Caution should be used in interpretation of results when these tests are used in infants below 36 weeks gestational age. Repeated measures, over several weeks or months, are recommended to determine the reliability of test results. Because “transient” AN has been

reported in a percentage of infants, particularly those with hyperbilirubinemia, hypoxia, ischemia, and central nervous system immaturity and low birth weight, frequent monitoring by the AN test battery is recommended to establish the stability of test results.

**Recommended Audiological Test Battery.** Because the developmental effects of AN cannot be predicted by electrophysiological audiologic tests, behavioral evaluation of infants with this diagnosis is of paramount importance. The recommended test battery permits assessment of middle ear status as well as functional hearing abilities and auditory development using behavioral response to pure tones, speech reception, and speech recognition. In concert with JCIH recommendations (2007), the test battery includes:

1. Otoscopic examination and acoustic immittance measures of middle ear function; as with any infant, infants with AN may develop middle ear dysfunction and otitis media with effusion resulting in mild conductive hearing loss. Because acoustic (stapedial) reflexes are absent in individuals with AN, otoscopy and tympanometry will be most useful for identifying infants with middle ear dysfunction.
2. Behavioral assessment of pure-tone thresholds using developmentally-appropriate, conditioned test procedures (visual reinforcement audiometry {VRA}, or conditioned orientation reflex {COR} audiometry). For very young or developmentally-delayed infants, behavioral observation audiometry (BOA) may be used to observe the infant's reflexive response to sound; however, results should not be interpreted as representing behavioral threshold or minimal response levels.
3. Speech reception and speech recognition measures. For very young infants, response threshold to repetitive consonant-vowel combinations (e.g., ba-ba, ga-ga) is appropriate; for toddlers, pointing to body parts may yield acceptable speech threshold results. As children's vocabulary develops, speech recognition measures using standardized picture-pointing

(e.g., Word Intelligibility by Picture Identification, WIPI) or open-set tests should be employed. Standardized taped materials are preferable to live-voice presentation to obtain consistency of stimuli across test sessions.

4. Otoacoustic emissions utilizing either TEOAEs and/or DPOAEs. Although initially present, OAEs may disappear in individuals with AN.
5. Cortical evoked potentials to speech or speech-like signals are not yet a standard clinical measure for infants or toddlers. However, these measures show promise as objective clinical tools for predicting speech-recognition performance in young children with AN.

#### **Recommended Amplification Strategies.**

For infants with "typical" sensory hearing loss, hearing aid fitting can proceed in the earliest months of life based on electrophysiological estimates (e.g., ABR, ASSR) of hearing sensitivity. For infants with AN, however, electrophysiological methods do not predict auditory detection thresholds. Clinicians and parents must rely upon the infant's behavioral response to sound to guide the hearing aid fitting decision. If an infant or young child with AN demonstrates elevated pure-tone and speech detection thresholds with consistent test-retest reliability, hearing aid fitting should be considered and a trial use of amplification should be offered to families.

Hearing aid fitting strategies for children with AN should follow established guidelines for the fitting of amplification in infants and toddlers (American Academy of Audiology Pediatric Amplification Protocol, 2003).

#### **Special Considerations for Cochlear Implantation.**

In addition to standard cochlear implant criteria for children, special consideration for cochlear implantation for children with AN include:

1. Cochlear implantation should not be considered until auditory test results are stable and demonstrate unequivocal evidence of permanent AN.

2. Evidence of auditory nerve sufficiency should be obtained by appropriate imaging techniques prior to implantation.
3. For children with AN who do not demonstrate good progress in speech understanding ability and aural/auditory language development, cochlear implantation should be considered, regardless of behavioral audiometric thresholds.

**Recommended Habilitation for Communication Development.**

Families of infants with AN should be informed that their baby’s auditory capacity or speech, language, and communication development cannot be predicted on the basis of the initial evaluation. Ongoing monitoring of their infant’s auditory, speech, language, communication, and general development is essential. Families should be made aware of all communication options presented in an unbiased manner. Infants with this diagnosis should receive referral to early intervention programs that assess language, cognitive skills, auditory skills, speech, vocabulary, and social-emotional development of children at six month intervals during the early years of life.

**Counseling Families of Infants with AN.** Counseling families of infants and young children with AN is one of the greatest challenges associated with this disorder. Because the developmental effects of AN cannot be predicted from test results obtained in the earliest months or even years of life, families struggle with the uncertainty of what this diagnosis means relative to their infant’s growth and development. Strong support systems, including parents of children with similar diagnoses and professionals with expertise in clinical social work and family counseling, should be available to meet the ongoing and changing needs of families.

Summary papers by each panel member as well as the guidelines were published by the Bill Daniels Center for Children’s Hearing, The Children’s Hospital – Colorado (to request a copy, e-mail [billdanielscenter@tchden.org](mailto:billdanielscenter@tchden.org)). Appreciation is extended to the panel who contributed their expertise to these guidelines: Yvonne Sininger, Ph.D.,

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## Vision

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