Auditory Neuropathy/Dys-Synchrony Disorder

Diagnosis and Management

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KEYWORDS

• Auditory • Neuropathy • Dys-synchrony • Diagnosis • Management
• Cochlear implant • Characteristics

KEY POINTS

- Effect, directly or indirectly, is on neural processing of auditory stimuli.
  - Physiologic measures are needed to accurately characterize patients with auditory neuropathy spectrum disorder (ANSD).
  - Clinical behavioral responses vary greatly and are not useful diagnostic measures.
- Patients with ANSD have greater difficulty listening in noise than those with other types of hearing disorders.
  - Separating detection ability from discrimination ability is critical in considering various management approaches.
- Without clear auditory input, visual information is needed for auditory communication and speech/language development.
- Many patients benefit from cochlear implants; fewer benefit from hearing aids.
- Patients should be followed closely, as changes in auditory function may occur over time.

Auditory neuropathy (AN),[1] auditory neuropathy/dys-synchrony (AN/AD),[2] and, more recently, auditory neuropathy spectrum disorder (ANSD) are variable terms used to describe an auditory disorder seen in patients ranging in age from infants to adults. With knowledge of the inherent problems presented by each term, ANSD

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Abbreviations

ABR	Auditory brainstem response
AN	Auditory neuropathy
AN/AD	Auditory neuropathy/auditory dys-synchrony
ANSD	Auditory neuropathy spectrum disorder
APD	Auditory processing disorder
CM	Cochlear microphonic
EVA	Enlarged vestibular aqueduct
FM	Frequency modulation
HMSN	Hereditary motor sensory neuropathy
IHC	Inner hair cells
MEMR	Middle-ear muscle reflex
MOCR	Medial olivocochlear reflex
NICU	Neonatal intensive care unit
OAE	Otoacoustic emission
OHC	Outer hair cells
OTOF	Otoferlin
SNHL	Sensorineural hearing loss

is used here with the understanding that no one term is completely definitive or descriptive.

OVERVIEW OF CHARACTERISTICS

ANSD is characterized by evidence of intact outer hair cell (OHC) function, shown by the presence of otoacoustic emissions (OAEs) and/or cochlear microphonics (CMs), accompanied by poor eighth nerve–brainstem responses, demonstrated by absent or highly abnormal auditory brainstem responses (ABRs).[3] Further evidence of effects on neural function is demonstrated by absent or elevated middle-ear muscle reflexes (MEMRs)[4] and abnormal medial olivocochlear reflexes (MOCRs).[5] Although understanding of speech in noise is poorer than that observed in sensorineural hearing loss (SNHL), word recognition in quiet is highly variable, and thresholds for pure tones range from normal to profound losses. Most ANSD patients show bilateral characteristics, although function may be asymmetric between ears, and patients with unilateral ANSD have been documented.

Despite fairly similar findings on auditory physiologic measures, patients vary considerably in functional communication abilities.[6,7] Clinical presentation typically includes difficulty listening in noise, may include fluctuating hearing ability, and, in the case of infants and children, most often involves delays in speech and language development. Patients with ANSD typically demonstrate poor temporal resolution[8] and may have neural deficits in other systems.

INCIDENCE

ANSD occurs in about 10% of individuals who have a dys-synchronous ABR, or an ABR consistent with an estimate of severe or profound hearing loss. This estimate is based on data from several sources that include screening of more than 1000 children enrolled in schools for the d/Deaf in North America,[9] a similar smaller-scale study in Hong Kong,[10] a hospital-based study of children in Australia,[11] and a multicenter newborn screening study in the United States.[12] A higher incidence of 17.3% and 15.4%, respectively, was reported among children identified with hearing loss following newborn hearing screening.[13,14] In the neonatal intensive care unit (NICU),