

Auditory Neuropathy: Diagnosis and Case Management

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Historically, a diagnosis of sensorineural hearing loss was attributed to cochlear dysfunction; primarily of the outer hair cell cells. The addition of new diagnostic tools to the audiological test battery assisted the definition and understanding of auditory neuropathy (AN). Auditory neuropathy is term used to classify patients with abnormal auditory neural responses in the presence of normal cochlear function. This article will review the diagnosis and demographics of AN reported in the literature as well as discuss the clinical management of this challenging hearing disorder.

Clinical characteristics of AN:

Persons diagnosed with AN present with varying degrees of hearing loss from mild to profound with extremely poor speech discrimination regardless of degree of hearing loss. This inconsistency between speech discrimination and degree of hearing serves as the first indication that a patient may have AN. In order to confirm a diagnosis of AN, the following three characteristics should be evident; extreme difficulty “hearing” in various situations regardless of hearing level, absent or abnormal auditory neural function and normal cochlear function. The test battery recommended to confirm each of these characteristics includes auditory brainstem response (ABR) test, stapedial reflex test, air and bone conduction audiometry, otoacoustic emissions test and speech discrimination testing. One must present with an abnormal ABR, indicating abnormal neural function, and present CM or OAE, indicating normal cochlear function, at a minimum for a diagnosis of AN. The test battery and results consistent with a diagnosis of AN are summarized in the table below.

Audiometric Test	Clinical Finding in Cases of AN
Auditory Brainstem Response (ABR)	Absent or severely abnormal morphology regardless of audiometric thresholds
Cochlear Microphonic (CM)	Present
Otoacoustic Emissions (OAE)	Typically Present
Stapedial Reflexes	Absent
Audiometric thresholds (air and bone)	Variable, normal to profound hearing loss
Speech discrimination	Poor regardless of hearing thresholds

Demographics of AN:

AN has a variable age of onset, with reports from birth to over age 60 years (Sininger and Oba, 2001). The majority of reported cases show an onset before the age of ten years. Case distribution appears equal among males and females. Associated risk factors include; genetic hearing disorders, prematurity, hyperbillirubinemia, and hereditary sensory-motor neuropathies. Approximately 25% of patients have no associated risk factors (Sininger and Oba, 2001). The degree of audiometric hearing loss is variable with the majority in the moderate to severe range and typically bilateral in nature. The overall incidence of AN has been elusive. The trend to incorporate OAEs into standard clinical test batteries and infant screening in conjunction with ABR should assist in estimating an appropriate incidence. To date, reports have suggested that 10% of patients with “deafness” my potentially have AN (Rance et al., 1999).

Potentials Site of Dysfunction:

There are several potential sites of dysfunction for AN, yet none are unanimously agreed upon. It could be an isolated inner hair cell dysfunction (Harrison, R. 1998; Gibson, 2002) or a synaptic dysfunction between the nerve and hair cell. The peripheral portion of auditory nerve could be demyelinating or have an axonal neuropathy. Multiple sites of dysfunction cannot be ruled out either. In evaluating AN, it is important to implement a through test battery. In addition to the audiological work up, a neurological consult should be included for evaluating the presence of other system neuropathies (Starr, A., 2001).

Cases of AN diagnosed at House Ear Institute:

Sixteen pediatric cases of AN were diagnosed at the House Ear Institute's Children's Center between 1996 to the present. Several other cases were referred to center for research purposes following a diagnosis of AN at another clinic, but were not included in this assessment. The degree of hearing loss for the sixteen cases are as follows; 5 were mild to moderate, 2 were moderate to severe, 7 were severe to profound and 2 were fluctuating in nature. Several of the cases had multiple associated risk factors. Hyperbillirubinemia and prematurity were suggested in 24% and 19% of the cases respectively. Other risk factors identified included genetic syndrome (10%), low birth weight (5%) and hydrocephalus (5%). No associated risk factors were identified in 37% of the cases.

In addition to the case demographics, the educational placements of the children diagnosed with AN were also analyzed. Nine of the children (56%) utilized an Oral Mode of communication while seven children (44%) utilized Total Communication. Five of the children (29%) were enrolled in mainstream academic settings with normal hearing, age matched peers. Five children (29%) were enrolled in an oral education class for the hearing impaired (no sign language) while seven children (42%) were enrolled in a Total Communication class in which American Sign Language is consistently used. Although none of the children from this center used Cued Speech (most likely due to a lack of availability in the area at the time), there are case reports of success with this rehabilitation approach and children with AN (Shallop et al., 2001).

All of the sixteen cases were fitted with conventional hearing aids on a trial basis. Six children continue to use conventional amplification, 2 now use a cochlear implant, 5 have discontinued use of amplification and 3 are unknown and have electively not returned to the center for follow up appointments.

Case Management of AN:

A trial period with conventional amplification, either hearing aids or assistive listening device, is typically recommended for patients with AN. Reports have suggested adequate benefit in some cases (Cone-Wesson et al., 2001, Rance et al., 1999). For many patients however, conventional amplification is unsuccessful. Therefore, physicians and clinicians began investigating the possible benefit of cochlear implantation in cases of AN. At first, many were suspect of the efficacy of implanting a neural stimulator in a case of suspected neural dysfunction. However, research suggested that electrical stimulation can actually restore neural synchrony in cases of demyelination (Zhou et al., 1995), promote neural survival (Araki et al., 1998) and restore temporal coding (Shannon, 1993). Children with severe to profound sensorineural hearing loss are routinely implanted. In many of these cases, the status of the auditory nerve is unknown at the time of implantation. It is not unlikely that

some of the children implanted over the past 20 years, had undiagnosed AN (given the lack of diagnostic tools at that time of surgery). Unfortunately, attempts to identify these cases post implant to evaluate outcomes are difficult. Approximately one third of AN cases meet the candidacy criteria for cochlear implantation. More recently, implant centers have been implanting children and adults with AN (Miyamoto et al., 1999, Trautwein et al., 2000 and Shallop et al., 2001). The outcomes in these cases have been consistent with the outcomes of patients with sensorineural hearing losses routinely recommended for implantation (Trautwein et al., 2000; Trautwein et al., 2001). Post-operative case studies of AN and cochlear implants report positive outcomes. Many of the children in these studies demonstrate improved listening and communications skills enabling them to take advantage of multiple communication options (Shallop, et al., 2001). In addition, the neural responses that were absent or severely abnormal pre-operatively, are obtainable post-operatively in response to electrical stimulation (Trautwein et al., 2001; Shallop et al., 2001). Further, restoration of the neural synchrony has been demonstrated beyond the peripheral nerve, throughout the auditory pathway (Shallop et al., 2002). Therefore, cochlear implantation is now considered a feasible option for patients with AN.

Summary:

AN is a relatively new classification of a hearing disorder characterized by extremely poor speech discrimination regardless of the audiometric thresholds. Patients typically present with an absent ABR but present OAES and CM. Case management typically includes a trial with conventional forms of amplification and assistive listening devices. In some cases, a cochlear implant may be an option. Many of the patients are enrolled in rehabilitation programs with an emphasis on lip reading or visual modes of communication (sign language, cued speech). Close monitoring of pediatric cases is recommended to ensure appropriate language skills are developed.

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