Cochlear Implantation of Auditory Neuropathy

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Abstract

Auditory neuropathy (AN) is a hearing disorder that presents with a grossly abnormal or absent neural response as measured by evoked potentials in the presence of normal outer hair cell function evidenced by present otoacoustic emissions or cochlear microphonics. Rehabilitation for patients with AN is challenging due to abnormal temporal encoding at the auditory nerve leading to severely impaired speech perception. Although patients with AN may demonstrate improvement in thresholds with amplification, temporal encoding dysfunction, and consequently speech perception degradation, is not alleviated by amplification. Another issue is the heterogeneity of the AN population in terms of audiologic and neurologic findings, in addition to uncertain etiology and pathophysiology. For children with prelingual onset of AN, development of auditory and oral communication skills is particularly compromised. All children with hearing loss in the severe-to-profound range who do not benefit from conventional amplification can be considered candidates for a cochlear implant (CI). This paper presents a case study of a child with AN who received a CI. Whereas no synchronous neural response auditory brainstem response could be elicited to acoustic stimuli, an electrically evoked auditory nerve action potential was evident following implantation, suggesting restoration to some degree of neural synchrony. Significant improvement in speech perception was found post-CI. Recommendation to implant all patients with AN would be premature, but these findings suggest that electrical stimulation in some cases of auditory neuropathy can be a viable option.

Key Words: Auditory neuropathy, cochlear implant, electrically evoked compound action potential, speech perception

Abbreviations: AN = auditory neuropathy, CI = cochlear implant, CM = cochlear microphonic, DSL = desired sensation level, EAP = electrically evoked compound action potential, ESP = Early Speech Perception test, IHC = inner hair cells, LTASS = long-term average speech spectrum, NRT = neural response telemetry, OAE = otoacoustic emission, TAC = Test of Auditory Comprehension, TEOAE = transient evoked otoacoustic emission

Auditory neuropathy (AN) is a term used to describe a recently recognized type of hearing disorder (Berlin et al, 1993; Sininger et al, 1995; Starr et al, 1996; Deltenre et al, 1997; Doyle et al, 1998; Rance et al, 1999). Symptoms of AN include (1) hearing loss, usually bilateral of any degree; (2) normal outer hair cell function evidenced by present otoacoustic emissions (OAEs) and/or cochlear microphonic (CM); (3) abnormal evoked potentials beginning with wave I of the auditory brainstem response (ABR); and (4) poor speech perception. The site of lesion for patients with AN remains unknown. The possibilities include the inner hair cells (IHC), synaptic junction between the IHC and auditory nerve, and the auditory nerve itself.

Rehabilitation for patients with AN is challenging due to the heterogeneity of the population in terms of audiologic and neurologic findings, in addition to the uncertain etiology and pathophysiology. One pervasive observation with AN is poor temporal encoding (Starr et al, 1996; Zeng et al, in press) and, consequently, degraded speech perception ranging from limited to no open-set discrimination. Since
conventional amplification cannot address temporal function, trials with hearing aids, FM systems, or tactile aids have demonstrated only limited success (Rance et al., 1999). Data from our laboratory indicate that approximately one-third of patients with AN have audiometric thresholds in the severe-to-profound range and, therefore, could be considered candidates for cochlear implantation.

Cochlear implantation is routinely performed in children and adults with hearing losses in which the cochlea is the primary site of dysfunction and the status of the auditory nerve remains elusive. However, for patients with AN, the efficacy of implantation could be questioned because of the suspected disease of the auditory nerve, which could be axonal degeneration, demyelination, or both (Starr et al., 1996). If the nerve is the site of dysfunction, electrical stimulation provided by the cochlear implant (CI) may promote neural survival (Araki et al., 1998; Mitchell et al., 1997) and restore temporal encoding (Shannon, 1993). Even more promising, Zhou et al. (1995a, b) report that electrical stimulation produces synchronous ABRs in the presence of peripheral auditory nerve demyelination. This suggests that the stimulation provided by a CI may, in fact, provide reliable, consistent neural firing even in the presence of a diseased nerve. If the site of dysfunction is cochlear, that is, IHC or the synapse (Harrison, 1998), a CI may be a viable compensation as it is in patients not diagnosed with AN who are routinely implanted as treatment for cochlear hearing losses.

Although the potential benefits of cochlear implantation for patients with auditory neuropathy are exciting, drawbacks do exist. The standard risk of the surgical procedure and anesthesia obviously remain. However, these risks are no greater for children with auditory neuropathy than children with cochlear hearing loss routinely being implanted. Studies have suggested that the insertion of the electrode array may damage cochlear structures and destroy residual hearing (O'Leary et al., 1991; Gstoettner et al., 1997). Therefore, the implantation procedure may damage structures in the cochlea, that is, the outer hair cells, which are functioning normally.

The following case presentation details the cochlear implantation of a child with AN. The case study serves as one example of the potential benefits a child with AN may experience from cochlear implantation.

**CASE REPORT**

An 18-month-old boy was referred to the House Ear Institute CARE Center for the dispensing of hearing aids following a non-response ABR at another local center. The parents reported a healthy, full-term pregnancy. Developmental milestones with the exception of speech were reached at age-appropriate times. The patient is an only child, and a family history of hearing loss was denied. Behavioral audiologic assessment revealed audiometric thresholds in the severe-to-profound range (Fig. 1). Immittance

![Figure 1](image)
testing revealed normal tympanograms and absent ipsilateral acoustic reflexes. Before this point in time, OAE testing had not been attempted; therefore, a diagnosis of AN was not yet known. The child was immediately fit with conventional, power amplification using the Desired Sensation Level (DSL) fitting algorithm (Seewald et al., 1985; Cornelisse et al., 1995). Aided audiometric thresholds for both ears were within the long-term average speech spectrum (LTASS) (Ling, 1978) through 1000 Hz. The child was monitored at regular intervals every 3 months, and hearing sensitivity was stable throughout repeat evaluations. The child enrolled in an oral preschool for hearing-impaired children. Despite the consistent use of amplification and intensive auditory training for 16 months, the child’s auditory-oral language skills did not improve. Therefore, the child was referred for a CI pre-evaluation.

The child was 2 years, 11 months of age at the time of the pre-evaluation. As part of the evaluation, transient evoked otoacoustic emission (TEOAE) testing was performed for the first time. TEOAEs were clearly absent for the left ear (Fig. 2). However, despite the high noise rejection level and low waveform reproducibility, an emission appeared to be emerging in the right ear (see Fig. 2) and warranted further testing. Therefore, a repeat ABR was obtained using condensation and rarefaction clicks to possibly elicit a CM. A CM is a cochlear potential generated by the hair cells (Tasaki et al., 1954). Figure 3 shows the ABR recording in which a neural response is absent while CM is present for both ears. Clearly, the CM is more robust in the right ear, which had the emission, than the left ear, which did not have a TEOAE.

However, as the incidence of unilateral AN is very rare based on the data from our laboratory, these test results were taken as sufficient evidence to support a diagnosis of bilateral AN. Speech perception testing in the binaurally aided condition revealed no closed- or open-set speech discrimination. The CI team reviewed the case and discussed it at length with the child’s parents. The family chose to proceed with the CI with guarded expectations.

The child was implanted at 3 years, 3 months of age with the Cochlear Corporation’s (Denver, CO) Nucleus CI24M CI. The left ear was implanted based on the lack of measurable OAEs. The surgeon reported no complications with a full insertion of the device. The child was fit with the SPEAK (McKay et al., 1992; Skinner et al., 1994) processing strategy and monitored closely at 3-month intervals. Amplification of the right ear (opposite CI) was discontinued. Figure 4 plots functional gain with the CI at 1-year postimplantation when the child was 4 years, 4 months of age. These responses are within the LTASS.
In addition, TEOAE testing was repeated for the right ear opposite the CI (Fig. 5), at this time revealing a robust OAE most likely due to the discontinued use of a hearing aid.

It was hypothesized that electrical stimulation provided by an implant may enhance temporal encoding and neural synchrony for this patient. One way to assess the enhancement of temporal coding and neural synchrony is through evoked potential recordings. The most direct measure of auditory nerve activity in CI recipients is the electrically evoked compound action potential (EAP). Until recently, these measurements were limited to intraoperative recordings or in patients with percutaneous plugs (Gantz et al, 1994). The new Nucleus CI24M CI incorporates telemetry, known as neural response telemetry (NRT), used to measure the EAP in CI24M recipients. The reader is referred to Abbas et al (1999) for detailed explanation. Briefly, NRT allows the voltage across a pair of electrodes along the array to be sampled following stimulation of another electrode along the array. A subtraction technique is used to extract the EAP from stimulus artifact by recording two tracings, one with and one without the removal of the neural potential using a forward masking paradigm. The EAP is typically recorded as a negative peak (N1) followed by a positive peak (P1) with amplitude measured in μV. Figure 6 represents an NRT recording of the EAP for this child. The response waveforms are presented as a function of current level. Although a neural response could not be elicited prior to implantation (see Fig. 3), a robust EAP is recorded in response to electrical stimulation. The response amplitude increases with increasing current level as would be expected of a functioning neural system. Figure 7 plots the EAP as a function of increasing stimulus rate from 10 to 80 Hz. The amplitude of the response was not altered at faster presentation rates, suggesting proper neural integrity. Rates above 80 Hz were not assessed due to equipment limitations. The fact that an EAP can be recorded in this patient even at faster stimulus rates indicates that electrical stimulation has restored some degree of neural synchrony and temporal encoding at the level of the auditory nerve.

Restoration of neural synchrony and temporal encoding to some degree may enhance the development of speech and language skills often compromised in children with AN. The speech perception skills of children with cochlear hearing losses who receive CIs are closely monitored. A wide range of performance among implant users is well documented (O'Donoghue et al, 1998; Cohen et al, 1999; Miyamoto et al, 1999). However, it is helpful to compare the performance of the child with AN to children with cochlear hearing loss as implantation for the latter is routinely recommended. Therefore, speech perception data on three measures were
compared for the child with AN and a set of 10 children with cochlear hearing loss (Figs. 8, 9, 10). The children were selected from the patient database at the House Ear Institute CARE Center. The children with cochlear hearing loss were similar to the AN patient in that all were implanted between the ages of 2.8 and 4.0 years with the Nucleus 22 or CI24M device and use the SPEAK processing strategy. Speech perception testing data at 1 year postimplantation for the child with AN were compared to the speech perception scores of the 10 children with cochlear hearing loss.

Figure 8 plots the preimplantation and 1 year postimplantation score for each child on the Ling Six Sounds Test (Ling, 1978). This test assesses a patient's ability to either detect or discriminate the phonemes /a, u, i, s, sh, m/ when presented auditory only (i.e., no visual cues). Each phoneme is presented five times in a random order for a maximum score of 30. Prior to receiving the CI, the child with AN actually had better detection of the sounds than the children with cochlear hearing loss. However, the child with AN could only detect these sounds and demonstrated no discrimination of the sounds without the visual cue of lip reading. One-year postimplantation, the child with AN could discriminate the sounds with accuracy as did most of the children with cochlear hearing loss.

The Early Speech Perception (ESP) test (Moog and Geers, 1990) is designed to be used with children with limited vocabulary and language skills in order to assess speech perception ability. The test consists of three sets of test stimuli and each is administered as a closed set. The results
Cochlear Hearing Loss © Auditory Neuropathy

Subjects

Figure 9 ESP test results for the child with AN and 10 children with cochlear hearing loss 1-year post-CI. Prior to implantation, all children were in category 1. ESP = closed-set task with four-category scoring; category 1: no pattern perception; category 2: pattern perception; category 3: some word identification; category 4: consistent word identification.

Figure 10 TAC results for the child with AN and 10 children with cochlear hearing loss. This test was not administered to any of the children prior to implantation. TAC: closed-set task of 10 subtests increasing in degree of difficulty. Subtest 1: linguistic/non-linguistic; 2: linguistic/human non-linguistic/environmental; 3: stereotypic messages; 4: single element core noun vocabulary; 5: recall two critical elements; 6: recall four critical elements; 7: sequence three events; 8: recall five details; 9: sequence three events with competing messages; 10: recall five details with competing messages.

DISCUSSION

This case illustrates one example in which electrical stimulation provided by a CI in a child with AN produced electrically evoked neural responses and significant improvement in speech perception ability. Although the results obtained in this case appear promising, the decision to implant should still be approached cautiously. The diagnosis of AN should not be an immediate referral for a CI. Some infants diagnosed with AN, who initially presented behaviorally as deaf, show improved or fluctuating auditory responses with increasing age (Berlin et al, 1999). Although few, other patients have experienced benefit with conventional amplification (Rance et al, 1999). Since no clear relationship between audiometric threshold and performance with conventional amplification has been established, a trial period with amplification is the only way to determine an individual's benefit. Studying the electrically evoked neural responses and their relation to speech perception in patients with AN may offer insight into the pathophysiology of this disease. Continued research in the areas of diagnosis, etiology, and pathophysiology of AN is needed prior to addressing the possibility of implanting patients with better audiometric thresholds who obtain no benefit from amplification.
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REFERENCES


