The Role of FM Technology in the Management of Patients with Auditory Neuropathy/Dys-synchrony

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Introduction

Persons with auditory neuropathy/dys-synchrony display dys-synchronous peripheral neural function in the presence of intact outer hair cell function (e.g., Berlin et al., 1993; Starr et al., 1996). The majority of patients also report hearing difficulty characterized by difficulty understanding speech, particularly in background noise. Patients with AN/AD show variation along several dimensions, including sound awareness, the presence of other non-auditory sensory or motor neuropathies, age of onset, and residual ability to utilize auditory information. Utilizing auditory information alone for learning language is, in our experience, unsuccessful without cochlear implantation. Thus, we recommend use of visually based communication such as Cued Speech or some form of sign language. In those patients where some residual speech understanding ability remains, it is generally very poor in noise. Thus, there is a subgroup of patients with AN/AD who are candidates for use of FM technology either with or without cochlear implants.

Identification of AN/AD patients

Dys-synchronous peripheral neural function is revealed by absent or highly abnormal auditory brainstem responses (ABR) and absent, or in a few patients elevated, middle-ear muscle reflexes (MEMR). Intact outer hair cell function is documented by the presence of otoacoustic emissions (OAE) and/or cochlear microphonics (CM). The CM is distinguished from the ABR by obtaining separate responses using condensation and rarefaction clicks (Berlin et al., 1998). Cochlear microphonics will invert with polarity while neural responses to clicks generally show only small latency shifts. Furthermore, CM does not shift in latency as intensity changes, while neural responses do shift. The test results found in patients with AN/AD is shown in table 1.

The difficulties that AN/AD patients experience can be further probed through studies of processing using psychophysical measures. Sound contains three dimensions: frequency, intensity, and time. Studies from Zeng et al. (1999, 2001) indicate that AN/AD patients have particular difficulty with the timing (temporal) characteristics of sound. Intensity processing is comparable to normal individuals and frequency discrimination is normal for higher frequency stimuli, but not in the lower frequencies. Measures of temporal processing, specifically temporal gap detection and temporal modulation, show that AN/AD patients require substantially longer amounts of time to make these discriminations.

Temporal processing is particularly important in understanding speech. While normal individuals can detect temporal changes of less than 10 msec, patients with AN/AD may not be able to discriminate changes of less than 100 msec. Speech changes rapidly and identification of attributes such as voicing and formant transitions, necessary to distinguish among speech sounds, requires the ability to make 30 to 50 msec discriminations. This, coupled with the role of the nervous system in marking the onset of stimuli, following changes in signals, and preserving timing information, underscores the difficulties that patients with desynchronized neural responses experience.

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Table 1. Test results in auditory neuropathy/dys-synchrony patients.

Test	**Outcome**
Otoacoustic emissions (OAE)	Normal
Cochlear microphonic (CM)	Present (inverts with stimulus polarity reversal)
Auditory Brainstem Response (ABR)	Absent (or severely abnormal)
Middle ear muscle reflexes (MEMR) Ipsilateral and contralateral	Absent (in rare cases, present but elevated)
Pure tone thresholds	Normal to severe/profound hearing loss (any configuration, can be asymmetric)
Speech recognition in quiet	Generally poorer than expected; slightly reduced to greatly reduced
Speech recognition in noise	Generally very poor
Masking Level Difference (MLD)	No MLD (i.e., 0 dB)
Olivocochlear reflex: TEOAE suppression	No suppression

Potential Underlying Mechanisms and Incidence

There is considerable variation among AN/AD patients which may be explained in part by the fact that several underlying mechanisms can result in the audiologic test profile seen in these patients. Normal outer hair cell function, reflected by present OAEs and CMs, and dys-synchronous neural responses can be due to an absence of inner hair cells, compromised synaptic connections between the inner hair cells and auditory nerve, or axonal loss or demyelination of the auditory nerve itself. Changes in neural firing patterns as a result of abnormalities at any of these sites could account for reduced or dys-synchronous responses (Starr, 2001). Preservation of outer hair cell function suggests that the biological cochlear amplifier is intact.

Estimates of the incidence of AN/AD suggest that it occurs at a rate of about 10% in those individuals who have a dys-synchronous ABR. This is based on evidence from several sources. Berlin et al. (2000) screened over 1000 hearing-impaired children in schools for the Deaf in the United States and found that 1–2% had robust OAEs and 10–12% had evidence of residual outer hair cell function shown by presence of OAEs in limited frequency bands. A similar smaller scale study in schools for hearing-impaired children in Hong Kong found similar results (Lee et al., 2001). Rance et al. (1999) reported that 1 in 9 infants with severe or profound permanent hearing loss shown by ABR had cochlear microphonics. Finally, Sininger et al. (2002) reported that approximately 10% of infants enrolled in the NIDCD Newborn Hearing Screening Study had OAEs and no ABR, consistent with AN/AD.

Problems in Understanding Speech in Patients with AN/AD

A key in analyzing the potential success that patients might gain from FM technology relates to speech recognition ability. Understanding speech is the primary auditory difficulty that AN/AD patients report. Test results show that word and sentence recognition in quiet is most often poorer than audiometric thresholds would predict (Starr et al., 1996). Listening ability in noise is universally reduced.

We evaluated a subset of 62 patients aged 4 years and older from our AN/AD database of 225 patients (Berlin et al., 2003b) where speech recognition testing was attempted. Younger patients were excluded due to differences in procedures and materials. With this evaluation, we wished to determine the number of patients who have some measurable speech recognition ability in quiet and in noise.

Measurable word recognition, defined as greater than 0%, was obtained in 26 patients (42%). It should be noted, however, that in the majority of these patients, the word recognition scores were much poorer than audiometric thresholds would predict, based on the norms of Yellin et al. (1989). Furthermore, most of the test results in children were obtained using closed set stimuli. The mean word recognition scores in quiet were 44.7% and 48.9% correct for the right and left ears, respectively. Measurable word recognition in noise (at a +10 signal-to-noise level) was obtained in only 4 patients (6%). The mean word recognition scores for these four patients were also severely below normal with mean scores of 17% and 29% for the right and left ears, respectively.

Considerations in the Management of AN/AD

Several factors define the important considerations in the management of AN/AD. Patients are often able to detect sound, but unable to discriminate among sounds. This is an important distinction in understanding responses from patients, and children in particular, to various management strategies. The critical element in the management of infants and children is language development. We work closely with speech/language pathologists, early interventionists, and educators in planning management. Our experience has taught us that visual communication methods (e.g., Cued speech, sign language, signed English) are necessary for language development. Auditory/Verbal therapy by itself, before cochlear implantation, has not worked in our practice as the sole method of teaching language.

Overall performance with hearing aids has been poor, though there are reports of improvement in a limited number of patients (e.g., Deltenre et al., 1997; Rance et al., 1999). In our patient database, a survey of 24 patients who assessed overall outcome with hearing aids, we found 3 patients who reported some functional interaction ability, 5 who reported primarily detection ability, and 16 who reported that they were only able to hear environmental sounds (Berlin et al., 2003a).

Cochlear implants are an important management option for patients with AN/AD. Post-implant neural response telemetry, electrical ABR, and MEMRs demonstrate restoration of neural synchrony and are comparable to responses in usual cochlear implant patients (Shallop et al., 2001). Comparisons of performance in AN/AD and non-AN/AD cochlear implant children has demonstrated comparable performance (e.g., Trautwein et al., 2000; Peterson et al., 2003).

Why Might FM System Use Be Helpful?

There are several reasons why FM systems may be helpful in some AN/AD patients. First, AN/AD patients with some residual speech recognition ability in quiet generally show very poor or no ability to understand speech in background noise. Thus, removing interference from noise would be beneficial in allowing those patients to maximize ability to use their residual speech recognition ability. Second, efferent feedback function, demonstrated by the middle-ear muscle (MEMR) and olivocochlear (OCR) reflexes, is disabled in AN/AD patients due to apparent compromise of afferent function (Hood et al., 2003). According to an anti-masking model proposed by Liberman and Guinan (1998), both of these reflexes are thought to assist in listening in noise. Finally, some AN/AD patients with residual speech recognition ability in quiet report that FM systems are helpful when listening in background noise.

A recently completed follow-up survey of 72 patients from our Kresge Lab AN/AD database indicates that 25% of the patients presently utilize FM systems with or without cochlear implants, in school, and in other situations. There are consistent reports of improved comprehension in these patients when they use FM systems, particularly in conjunction with their cochlear implants.

A Continuum of AN/AD

Patients with AN/AD are quite variable. Berlin et al. (2001, 2003b) describe a continuum along which these patients distribute in relation to their functional communication ability. There are a few patients at one extreme who exhibit no overt delays or auditory complaints until adulthood or until they are first evaluated with MEMRs or ABR. They are the patients who generally demonstrate the greatest residual speech recognition ability in quiet, though they report difficulty in noise. These patients would most likely be good candidates for FM technology since they generally report a considerable increase in listening difficulty in noisy situations.

At the other extreme are the AN/AD patients who exhibit a total lack of sound awareness. Between these two groups is the largest group of patients we have seen. These patients demonstrate inconsistent auditory responses, manage the best in quiet, and the poorest in noise. Their audiograms are inconsistent with other test results. The ABR is always desynchronized and middle-ear muscle reflexes are absent. Visual phonetic language usually works best until cochlear implantation, unless the family prefers cultural Deafness.

Conclusions

AN/AD is a clearly identifiable clinical entity, though there is considerable patient variation. While outer hair cells appear functional, neural processing is de-synchronized and severely compromised. Physiologic measures are needed to correctly identify auditory neuropathy/dys-synchrony. AN/AD infants and children have extreme difficulty learning language by auditory means alone and are rarely successful. Thus, visual communication methods are recommended prior to cochlear implantation, or if a cochlear implant is not chosen as a management option. It is important to distinguish detection of sound (sensitivity) from discrimination (especially in noise) when evaluating auditory function and hearing aid benefit in AN/AD patients. Some patients have residual speech recognition ability and may benefit from FM system use.

Further information on Auditory Neuropathy/ Dys-synchrony can be found at the Kresge Hearing Research Laboratory website (*www.kresgelab.org*) in the section titled “Information on Deafness”. This website contains articles and other information about AN/AD, links to other websites and resources, and a link to a listserve for parents and patients with AN/ AD.

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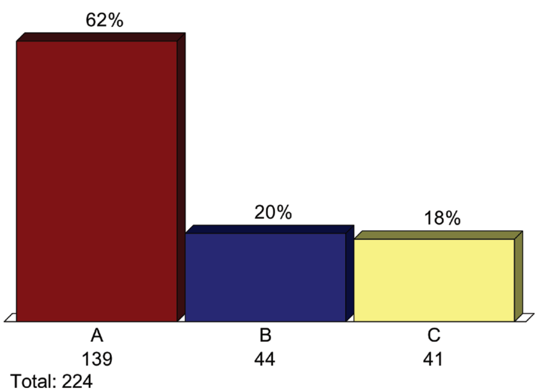
Participant Surveys

Have you seen any patients with auditory neuropathy/dys-synchrony?

A. Yes
B. No
C. Not sure



How many patients with dys-synchronous ABRs will have OHC responses and be classified as AN/AD?

A. 1 in 10,000
B. 1 in 1,000
C. 1 in 100
D. 1 in 10