J Am Acad Audiol 18:380–390 (2007)

Cortical Auditory Evoked Potentials in the Assessment of Auditory Neuropathy: Two Case Studies

Wendy Pearce*†
Maryanne Golding*†
Harvey Dillon*†

Abstract

Infants with auditory neuropathy and possible hearing impairment are being identified at very young ages through the implementation of hearing screening programs. The diagnosis is commonly based on evidence of normal cochlear function but abnormal brainstem function. This lack of normal brainstem function is highly problematic when prescribing amplification in young infants because prescriptive formulae require the input of hearing thresholds that are normally estimated from auditory brainstem responses to tonal stimuli. Without this information, there is great uncertainty surrounding the final fitting. Cortical auditory evoked potentials may, however, still be evident and reliably recorded to speech stimuli presented at conversational levels. The case studies of two infants are presented that demonstrate how these higher order electrophysiological responses may be utilized in the audiological management of some infants with auditory neuropathy.

Key Words: Auditory neuropathy, cortical auditory evoked potentials, hearing aids

Abbreviations: ABR = auditory brainstem response; AN = auditory neuropathy; CAEPs = cortical auditory evoked potentials; CMs = cochlear microphonics; DPOAEs = distortion product otoacoustic emissions; ECochG = electrocochleography; IHC = inner hair cell; NAL-NL1 = National Acoustic Laboratories Non-linear 1; OAEs = otoacoustic emissions; PBK = phonetically balanced kindergarten

Sumario

Los niños con neuropatía auditiva y posibles posibles trastornos auditivos están siendo identificados a edades tempranas con la implementación de programas de tamizaje auditivo. El diagnóstico se basa en la evidencia de una función coclear normal pero de una función anormal del tallo cerebral. Esta falta de función normal de tallo cerebral es muy problemática cuando se trata de

*National Acoustic Laboratories, Chatswood, New South Wales, Australia; †Cooperative Research Centre for Cochlear Implants and Hearing Aid Innovation, Melbourne, Australia

Ms. W. Pearce, National Acoustic Laboratories, 126 Greville St., Chatswood, NSW 2067 Australian Hearing; Phone +61 9412 6842; Fax: +61 9411 8273; E-mail: wendy.pearce@hearing.com.au

prescribir amplificación en niños pequeños, porque las fórmulas de prescripción requieren el insumo de los umbrales auditivos que se estiman normalmente a partir de las respuestas del tallo cerebral ante estímulos tonales. Sin esta información, existe una gran incertidumbre en cuanto a la adaptación final. Los potenciales evocados auditivos corticales pueden, sin embargo, registrarse con confiabilidad a partir de estímulos de lenguaje presentados a niveles de intensidad para la conversación. Se presenta el estudio de dos casos de niños, que demuestran como estas respuestas electrofisiológicas de orden mayor pueden utilizarse en el manejo audiológico de algunos niños con neuropatía auditiva.

Palabras Clave: Neuropatía auditiva, potenciales evocados auditivos corticales, auxiliares auditivos

Abreviaturas: ABR = respuestas auditivas del tallo cerebral; AN = neuropatía auditiva; CAEPs = potenciales evocados auditivos corticales; CMs = microfónica coclear; DPOAEs = emisiones otoacústicas por productos de distorsión; ECochG = electrococleografía; IHC = células ciliadas internas, NAL-NL1 = Laboratorios Nacionales de Acústicas No-lineal 1; OAEs = emisiones otoacústicas; PBK = palabras fonéticamente balanceadas para niños preescolares

The term "auditory neuropathy" (AN) first appeared in audiological literature in the mid-1990s (Sininger et al, 1995) although findings consistent with AN had been reported for some years before this time (Worthington and Peters, 1980; Kraus et al, 1984; Widen et al, 1995). One of these early studies reported cases where the auditory brainstem response (ABR) was absent but behavioral thresholds had been obtained in the absence of other neuropathology (Kraus et al, 1984), and the other study reported children with speech delay who were found to have ABR results that did not correlate with behavioral audiometric data (Worthington and Peters, 1980). In more recent times it has become clear that the term "AN" is too broad, or possibly inappropriate in some cases, and so other terms such as "auditory dyssynchrony" (Berlin et al, 2002), "auditory de-synchrony" (Ray et al, 2006) or "neural hearing loss" (Rapin and Gravel, 2003) have also come into use.

The diagnosis of AN is commonly made when normal cochlear function (i.e., otoacoustic emissions [OAEs] and/or a cochlear microphonic [CM] are present) but abnormal brainstem function is evident (i.e., absent, elevated, or grossly abnormal ABR results) (Starr et al, 1996; Berlin et al, 2003). Despite the disruption to the ABR, it has been reported that behavioral thresholds measured in people with AN range from normal (Kraus et al, 2000) to profound and any degree of behavioral hearing loss in between (Rance et al, 1999). In addition, when speech perception results from open set speech tests are compared between people with AN and those with sensorineural hearing loss measured behaviorally, of similar degrees, performance outcomes may be equal, or results for those with AN may be much poorer (Rance et al, 2002).

While the term "neuropathy" refers to pathology of peripheral nerve fibers, AN may result from any one of a number of disorders or combination of them. It has been suggested that the site of lesion may be at the inner hair cells (IHC) and/or the synapse with Type 1 auditory nerve fibers (Foerst et al, 2006). The disorder may also result from a breakdown of the simultaneous transmitter release from vesicles

attached to the synaptic ribbon of the IHCs, which results in impaired timing of afferent neuron firing (Fuchs et al, 2003; Khimich et al, 2005). AN may also arise if demyelization of Schwann cells around the auditory nerve fibers and/or alterations to nerve axons impair the normal synchrony of axonal conduction velocity (Starr et al, 1996; Rapin and Gravel, 2003). This disruption to peripheral function, which often leads to poor ABRs, does not necessarily affect cortical auditory evoked potentials (CAEPs) as these later responses are not as reliant on timing as the earlier evoked responses (Hood, 1998; Rapin and Gravel, 2003). Rance et al (2002) reported that in a sample of 18 children diagnosed with AN, CAEPs were present in 50% of cases. The mere fact that some children with AN had CAEPs and some did not lends increased support to the hypothesis that AN describes a number of auditory dysfunctions and underlying conditions and should not be thought of as a single disorder.

CAEPs have been recorded since the 1960s although its popularity as a clinical tool waned with the advent of ABR techniques. Davis (1966) reported that CAEP thresholds for tonal stimuli are within 10 dB of behavioral thresholds in 90% of normal-hearing and hearing-impaired adults and children with a sensory loss. CAEPs to a variety of speech elements are also robust in normal-hearing infants, at least at conversational levels, but using this technique to estimate threshold in infants is problematic as keeping them awake, but quiet enough to elicit a threshold response, is difficult (Cone-Wesson and Wunderlich, 2003). They have also been recorded in children and infants with hearing impairment, to demonstrate the detection of speech stimuli at the cortical level after hearing aid fitting (Rapin and Graziani, 1967; Gravel et al, 1989), but this is by no means a routine clinical application.

Hearing aid fitting in infants that is based on a prescriptive fitting formula requires hearing thresholds to be estimated and entered into the formula. Tone-burst ABR thresholds are typically used as the estimates, but when a diagnosis of AN has been made, there is great uncertainty about the appropriateness of using these values. More generally, there is uncertainty about the application of a prescription rule that has been derived based on the characteristics of people with sensorineural hearing loss to people with AN. Hearing aid fitting for these children should therefore also be dependent on all available information including behavioral test results and the family's perspective on when the habilitation process should commence (King et al, 2005).

Since the implementation of newborn hearing screening in the state of New South Wales, there has been an increase in the number of infants diagnosed with AN and subsequently referred to Australian Hearing for hearing habilitation. This report presents the case studies of two infants who failed their newborn hearing screening and were referred to experienced pediatric audiologists for diagnostic audiological assessments at state hospitals and test facilities. These assessments consisted of high-frequency probe-tone tympanometry, tone burst ABR, and OAEs, and on the basis of these results, the diagnosis of AN was made. On referral to Australian Hearing, they were assessed with CAEPs to provide additional guidance in the habilitation process.

METHOD

Procedure

The test stimuli were /m/ (duration 78 msec), /g/ (duration 31 msec), and /t/ (duration 78 msec), which were presented using alternating onset polarity at typical conversational levels (i.e., 65 dB SPL or 75 dB SPL), and an interstimulus interval of 1125 msec. These stimuli were generated from natural speech tokens consisting of an initial consonant followed by the vowel /ae/, which was extracted from a recording of running speech that was spoken by an average male Australian. The frequency response of the final test stimuli is shown in Figure 1. They included very little of the vowel transition and were recorded with digitization rates of 40 kHz. They were gated off at a zero crossing to minimize audible clicks, and no further modifications of the onset or offset characteristics were made. These consonants were chosen because they had a spectral emphasis in the low, mid-, and high-frequency regions,

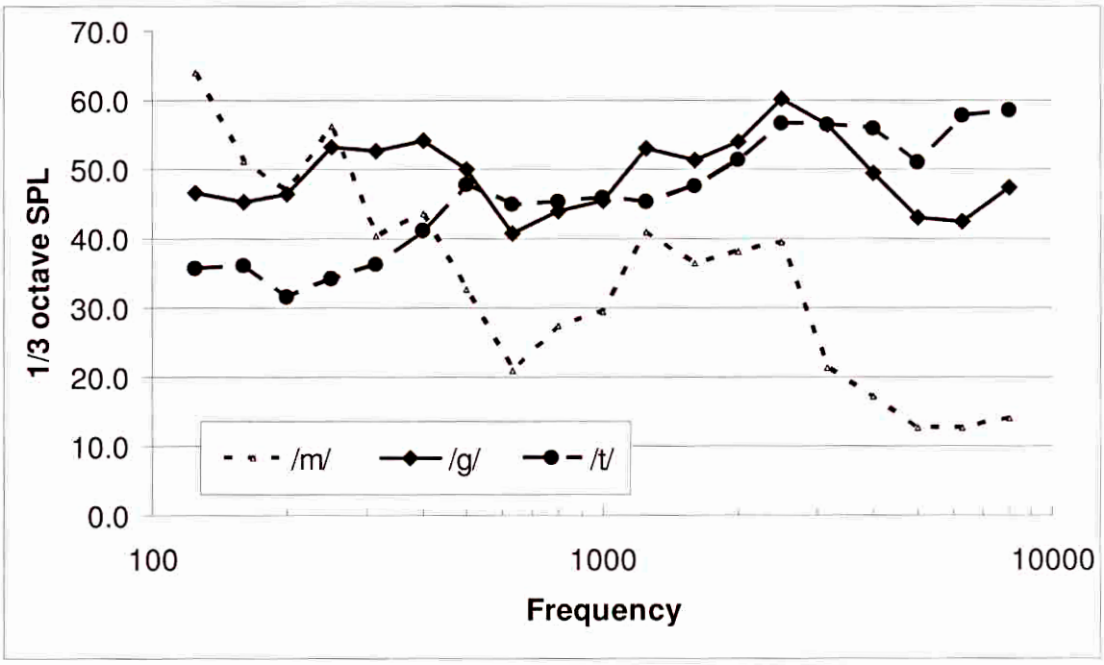

Figure 1. Spectral analysis of the three stimuli /g/, /t/, and /m/ showing the primary energy of /m/ below 400 Hz, /g/ between 1200 and 4000 Hz, and /t/ above 3000 Hz.

respectively, and thus had the potential to give diagnostic information about the perception of speech sounds in different frequency regions. Prior to testing, the stimuli outputs at the sound-field test position were measured as 65 dB SPL and 75 dB SPL (impulse time constant) using a microphone suspended from the ceiling and connected to a measuring amplifier in the observation room. The microphone was then retracted to a point above head height for continuous monitoring of the signal.

Brain electrical activity was recorded using the Neuroscan™ system with electrodes positioned at Cz, C3, and C4 referenced to one mastoid with forehead as ground. During cortical testing, infants were awake and seated on their mother's lap or in a baby chair, distracted by another adult if required. Stimuli were delivered via a loudspeaker positioned at 45 degrees azimuth on either side of the subject. If unaided, stimuli were presented through the speaker to the left side as the default setting. If aided, the speaker nearest the test ear was used while the opposite ear was occluded by the child's own earmold and hearing aid in the off position. Individual sweeps of the electroencephalic activity were amplified and analog filtered online at 0.1–100 Hz using a 24 dB/octave slope and subsequently filtered off line at 1–30 Hz. The recording window consisted of a 100 msec pre-stimulus baseline and a further 600 msec post-stimulus. Artifact reject was set at ±150 μV.

In keeping with our standard protocols, each stimulus was presented in blocks until 100 artifact-free EEG samples were acquired and, where possible, each block of stimuli was presented on two occasions with a randomized stimulus order. If the block was not repeated because the infant grew tired of testing, responses in the single stimulus block were separately averaged for the odd and even stimulus presentations. Response detection was based on the two replicated waveforms being overlaid and inspected for repeatability by an examiner who was experienced in identifying infant CAEPs. For the purposes of these case studies, no attempt was made to mark peak latency or amplitude.

CASE STUDIES

Infant 1

Infant 1 was born at 28 weeks gestation with low birth weight, a poor Apgar score, jaundice, and respiratory distress. He was seen for diagnostic audiology at 12 weeks of

age (i.e., 40 weeks gestational age) by experienced pediatric audiologists at a large teaching hospital. The clinicians reported that there were no responses to tone-burst ABR at 500 Hz and 2000 Hz at the limits of the equipment (i.e., 85 dB nHL for 500 Hz and 100 dB nHL for 2000 Hz), but distortion product OAEs (DPOAEs) were present across all frequencies bilaterally. Although the infant was referred to Australian Hearing for habilitation on the basis of these results, the family reported that the infant was responsive to sounds at home such as familiar voice and the stereo, and so a repeat ABR was performed approximately six weeks later at the hospital. At this second visit, click stimuli were presented with a possible threshold of 95 dB nHL achieved in both ears.

Unaided CAEP testing was performed one week later (i.e., at seven weeks corrected age). Repeatable responses for all three speech stimuli (i.e., /m/, /g/, /t/) were observed with presentation levels of 65 dB SPL as shown in Figure 2. This result shows that responses to CAEPs were

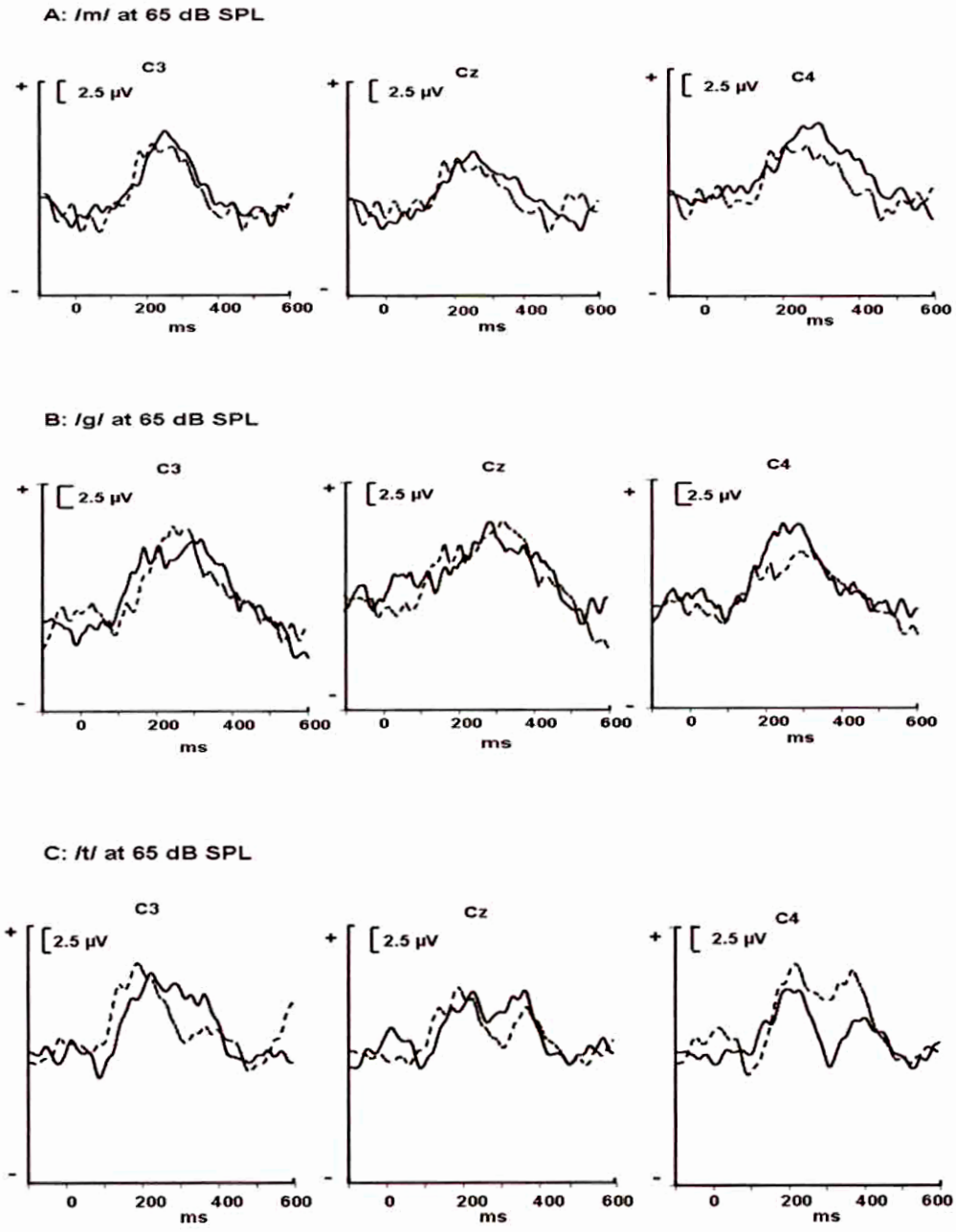

Figure 2. Repeatable CAEP responses for infant 1 are shown in response to the three speech stimuli: (*A*) stimulus /m/, (*B*) stimulus /g/, (*C*) stimulus /t/.

obtained at significantly lower stimulus levels than for ABR. A severe hearing loss could be excluded as behavioral responses and cortical responses were seen at moderate levels but no attempt was made to establish threshold using CAEP testing. A mild-to-moderate loss could not, however, be excluded, and therefore, hearing aids were fitted using Australian Hearing recommendations (King et al, 2005). In brief, these recommendations advise that all available measures of auditory function, including electrophysiological and behavioral test results, be considered along with parental desire to proceed with the fitting. On the basis of the CAEP results and this infant's parental observations, and to avoid potential overprescription of the hearing aid gain, the audiogram was estimated at 30 dB at 500 Hz, 40 dB at 1000 Hz, 50 dB at 2000 Hz, and 50 dB at 4000 Hz. This audiogram was entered to NAL-NL1 (National Acoustic Laboratories Non-linear 1) prescription formula, and he was fitted with digital behind-the-ear hearing aids using wide dynamic range compression and monitored regularly by his clinician. With this degree of amplification, there was no possibility of damage to the cochlea even if the auditory thresholds proved to be normal.

Given the ongoing uncertainties regarding the infant's final thresholds, electrocochleography (ECochG) was carried out five months later by the infant's physician. There were no responses at 110 dB nHL to tonal stimuli, but CAEP testing two weeks later again showed repeatable responses for all three speech stimuli (i.e., /m/, /g/, /t/) with presentation levels of 65 dB SPL

Regular adjustments were made to the fitting based on parental report, the outcomes of behavioral tests, and another unaided CAEP test, which continued to demonstrate repeatable responses at 65 dB SPL. At the age of two-and-a-half years, behavioral responses to visual reinforcement orientation audiometry were obtained at 30 dB SPL at 500 Hz, 25 dB SPL at 1000 Hz, 30 dB SPL at 2000 Hz, and 30 dB SPL at 4000 Hz. The child was no longer using hearing aids and was enrolled in an early learning program due to delay in his speech development.

Infant 2

This infant was born at full term with the only risk factor being a family history of two paternal relatives who used cochlear implants. Their etiology is unknown. He was diagnosed with AN at ten weeks of age by experienced pediatric audiologists at a specialist diagnostic facility. Initial ABR tests, using click stimuli, showed no response at the maximum limit of 100 dB nHL for the left ear and a repeatable wave V response at 100 dB nHL for the right ear as shown in Figure 3. Clear CMs and robust DPOAEs were, however, seen for both ears. In keeping with this clinic's policy for infants with AN, a repeat ABR was ordered with consultations for habilitation at Australian Hearing and medical review organized in the interim. He was fitted with digital hearing aids at four months of age using Australian Hearing protocols (King et

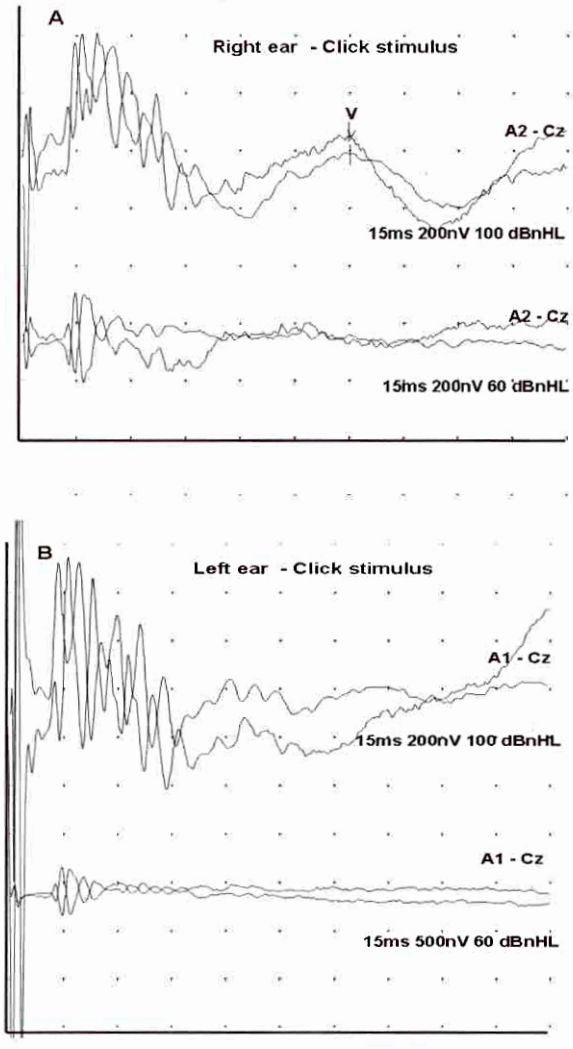

Figure 3. ABR s to click stimuli are shown for infant 2: (*A*) right ear, (*B*) left ear.

al, 2005). A conservative audiogram that predicted a moderate loss was entered into the NAL-NL1 prescription in the first instance as there were no clear behavioral responses to auditory stimuli and limited electrophysiological information.

A click and tone burst ABR was performed three months after the first test and showed no response to click stimuli in the left ear at maximum output, although cochlear microphonic activity was still reported to be evident. Testing for the right ear was limited to two frequencies as the infant did not sleep well. No response to 4000 Hz at 105 dB nHL or 2000 Hz at 90 dB nHL was observed, which was consistent with the results from the first test.

Aided CAEP testing was performed two weeks after the second ABR test. Parents felt that their baby was a little more responsive with his hearing aids on than without them, but there were very few clear examples of auditory behavior. The aided CAEP test results for 65 dB SPL presentation levels are shown in Figures 4 and 5.

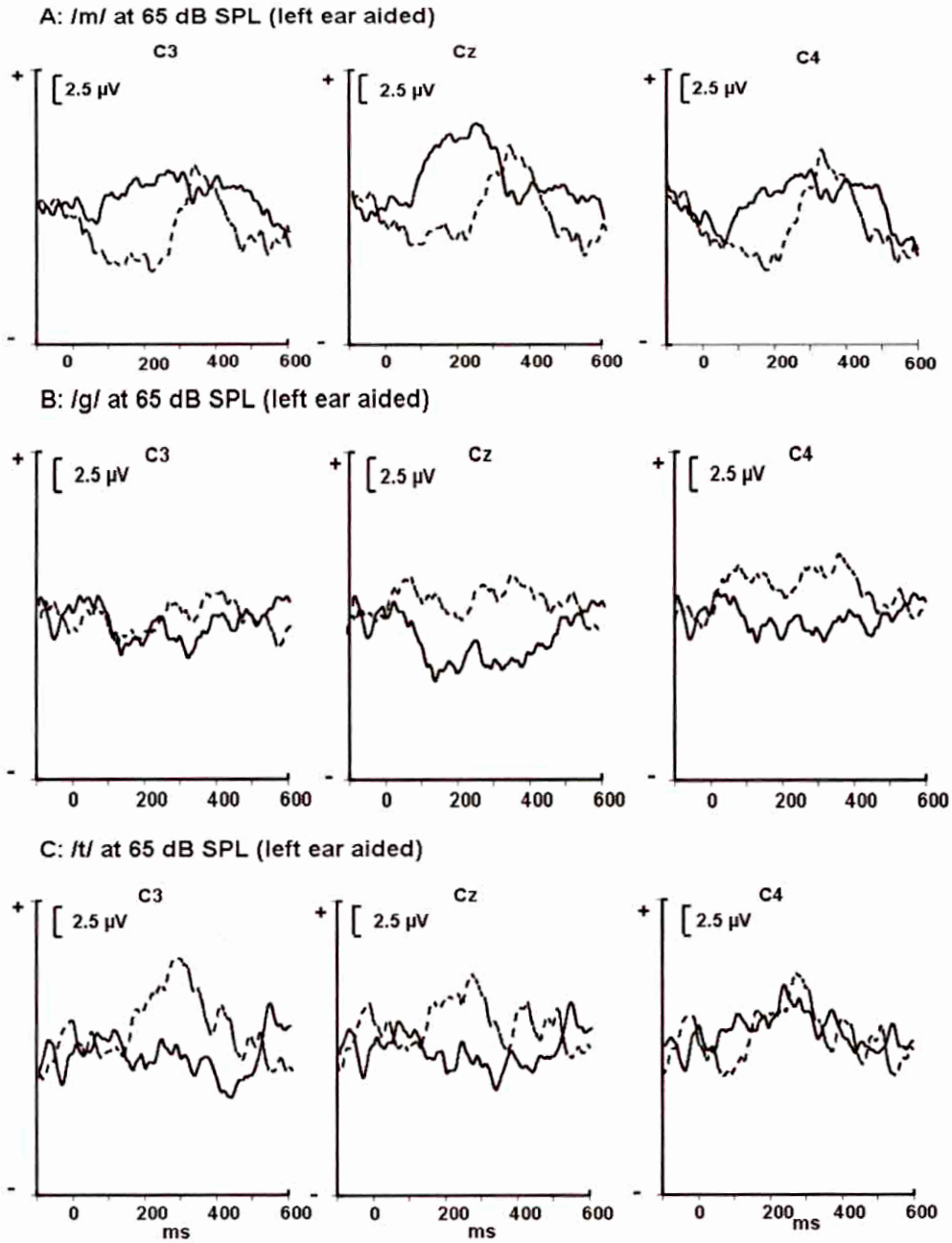

Figure 4. Nonrepeatable CAEP responses for infant 2 (left ear aided) are shown in response to the three speech stimuli: (*A*) stimulus /m/, (*B*) stimulus /g/, (*C*) stimulus /t/.

There were no repeatable cortical responses to any of the three speech stimuli presented at 65 dB SPL to the left or the right ear when aided. Limited testing was performed at 75 dB SPL as the infant became restless, with no response observed in either ear to /t/ at 75 dB SPL. The infant's hearing aids were subsequently adjusted to provide greater gain by re-estimating the degree of hearing loss and recalculating the NAL-NL1 prescription. Aided CAEP testing was performed a second time after the adjustment, but no response to stimulation at 75 dB SPL could be observed on this second occasion, and therefore, the hearing aids were further adjusted to provide their maximum gain. CAEP testing was repeated within a few days of this final adjustment, but still no response could be observed.

The child underwent a series of tests for cochlear implantation. He exhibited a severe hearing loss using ECochG, and there was evidence of auditory nerve function using electrically evoked ABR testing.

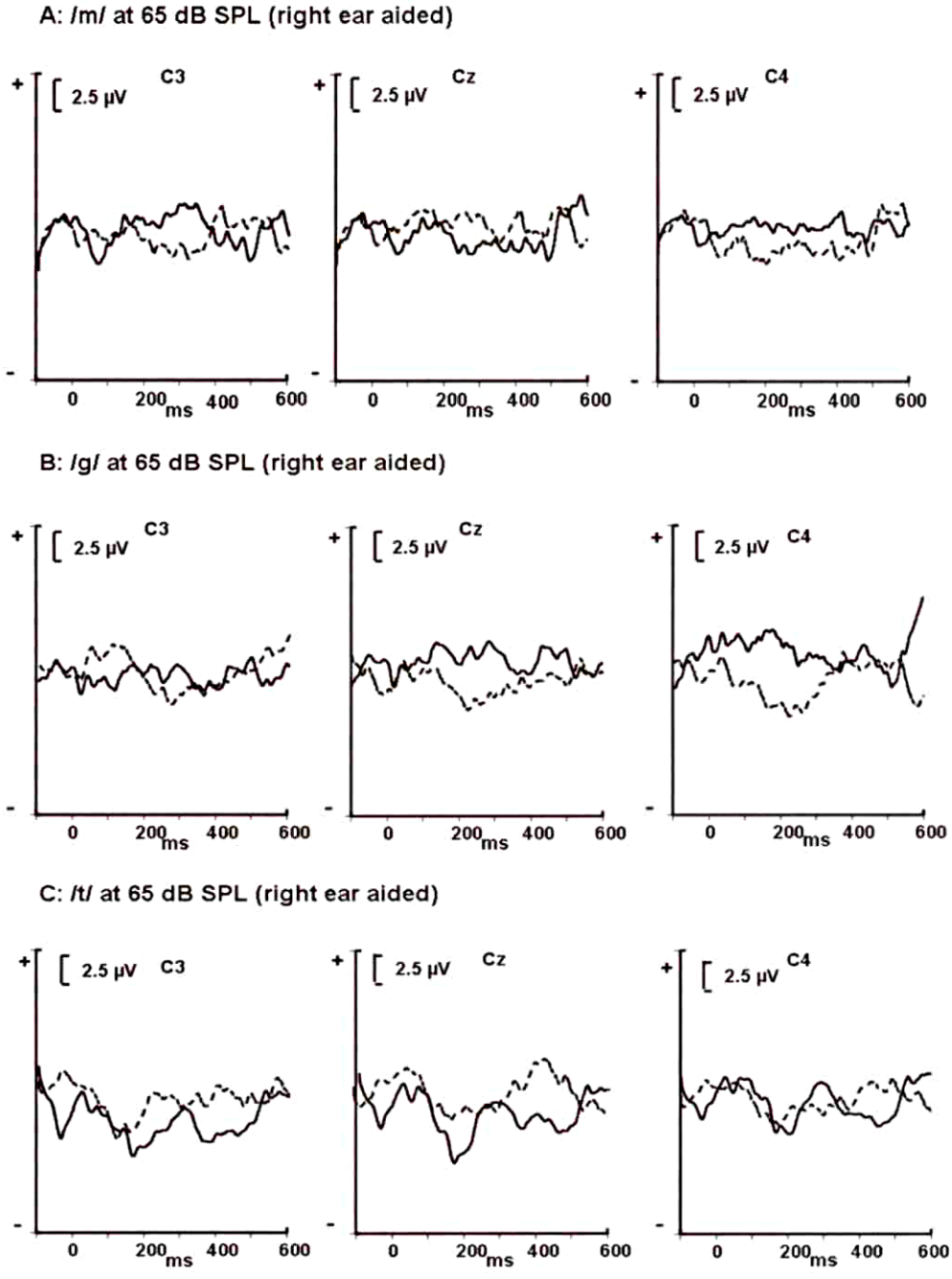

Figure 5. Nonrepeatable CAEP responses for infant 2 (right ear aided) are shown in response to the three speech stimuli: (*A*) stimulus /m/, (*B*) stimulus /g/, (*C*) stimulus /t/.

There were no inner ear abnormalities found on computerized tomography and magnetic resonance imaging studies. After extensive family counseling to ensure that the results were understood and that a positive commitment to the process existed, he received a cochlear implant at one year and four months of age.

DISCUSSION

Hearing aid prescription for young infants is often based on thresholds that have been estimated from frequency-specific ABR recordings (Dillon, 2001). In cases where AN is present, this approach is highly problematic as the ABR response is often poorly formed or absent and potentially misleading with behavioral hearing thresholds much better than those estimated by the ABR results. The possibility of overprescribing the gain requirements therefore exists if decisions are based on ABR thresholds alone. Although no attempt was made to find CAEP threshold in either case presented in this report, the detection or lack of CAEP responses to speech stimuli presented at conversational levels assisted in clinical decision making for these two children.

In the case of our first infant, it was clear that the three speech stimuli could be detected at the level of the cortex at conversational level without the assistance of amplification. Although it was not possible to infer that hearing thresholds were normal in one or both ears, these results certainly suggested that the degree of hearing loss was much less than that suggested by the ABR results. It has been reported (Berlin et al, 2002) that premature infants, or those suffering with hyperbilirubinemia, may have an initial presentation normally associated with AN, but they are more likely to recover with time. Our first infant was very premature when born at 28 weeks and had poor ABR test outcomes on two occasions over the first 18 weeks of life. It is quite possible that the ABR may have improved over time, and it would have been valuable to observe this improvement, but relying on ABR results alone in these early months would have prolonged the period of anxiety for parents and clinicians and would have resulted in a fitting inconsistent with the child's ability to detect sounds. This child was fitted with hearing aids set for a mild-to-moderate hearing loss as suggested by the CAEP results, and the ensuing clinical decisions and modifications to the fit were based on monitoring the child's auditory behavior and parental report, as well as CAEP testing. Sometime later, behavioral testing was able to confirm that hearing thresholds, in the sound field, were normal. This outcome did not, however, guarantee that the child had good speech perception skills. Kurtzberg (1989) reported that infants with normal CAEP responses were more likely to show normal receptive language at the age of one year. This child's parents reported informally that he had good receptive skills at two-and-one-half years of age (e.g., he could follow instructions well), but speech production was very poor.

Rance et al (2002) found that in children with AN, the presence or absence of CAEPs at presentation levels of 20 to 40 dB SL was positively correlated with aided phonemic scores on the phonetically balanced kindergarten (PBK) word tests. Children with CAEPs had an average aided PBK score of 60%, and those without CAEPs showed an average score of 6%. The authors concluded that the recording of CAEPs might therefore be a means of predicting speech perception skills. Lee et al (2001), however, described two children who had OAEs and attended a school for hearing-impaired children. They had poor ABRs but clear CAEPs. Both these children, who had moderate degrees of hearing loss behaviorally, had rejected their hearing aids and had poor speech-discrimination scores. Hood (1999) also reported the case of an adult diagnosed with Charcot-Marie-Tooth syndrome who similarly had clear CAEPs, robust OAEs, and poor ABR responses. This adult, who had a moderate-to-severe hearing loss behaviorally, did not find hearing aids helpful and was reliant on lip reading and other visual cues for communication. It appears, then, that in cases of AN, the presence of a clear cortical response at suprathreshold presentation levels may not always be predictive of good speech perception skills. Children with AN should, therefore, be regularly monitored for speech-language delay and intervention organized as appropriate.

Our second infant had absent ABRs and

absent aided CAEPs at conversational levels. The infant was therefore fitted using infant protocols for severe-to-profound hearing loss, and regular monitoring of the child's performance both behaviorally and with CAEPs was undertaken. No aided CAEP could be recorded at conversational levels, even after several incremental gain adjustments were made, and improvements to auditory behavior could not be observed. As a result he was evaluated for cochlear implantation at one year of age.

There are of course many children with AN who have some ABR response albeit distorted and potentially well above hearing threshold. In such cases, combined information from CAEP and ABR testing may be valuable in clinical decision making. It is also acknowledged that establishing CAEP thresholds may provide a useful additional perspective on the child's hearing, but it remains unclear whether auditory thresholds can be estimated reliably using this technique in all but the calmest of infants (Cone-Wesson and Wunderlich, 2003). Even without this extra information, the mere presence/absence of a CAEP response to speech stimuli at conversational level can provide useful clinical information. It is our contention that, for infants with AN, hearing aids might be fitted based on an assumed mild-to-moderate hearing threshold when unaided CAEPs are evident in response to speech stimuli presented at conversational levels. It is important, however, to arrange regular formal monitoring of performance using CAEPs, parental questionnaire, and behavioral testing. If ABRs and aided CAEPs are absent, however, a more aggressive approach to intervention may be warranted.

Acknowledgments. We thank colleagues at the Jim Patrick Audiology Centre at the Royal Institute for Deaf and Blind Children, and the New Children's Hospital Westmead for access to the diagnostic test results for these two infants.

REFERENCES

Berlin C, Li L, Hood LJ, Morlet T, Rose K, Brashears M. (2002) Auditory neuropathy/dys-synchrony: after the diagnosis, then what? *Semin Hear* 23(3):29–214.

Berlin C, Morlet T, Hood L. (2003) Auditory neuropathy/dys-synchrony. Its diagnosis and management. *Pediatr Clin North Am* 50(2):331–340.

Cone-Wesson B, Wunderlich J. (2003) Auditory evoked potentials from the cortex: audiology applications. *Curr Opin Otolaryngol Head Neck Surg* 11:372–377.

Davis H. (1966) Validation of evoked-response audiometry (ERA) in deaf children. *Int Audiol* 5(2):77–81.

Dillon H. (2001) *Hearing Aids.* New York: Thieme Publishers.

Foerst A, Beutner D, Lang-Roth R, Huttenbrink K-B, Von Wedel H, Walger M. (2006) Prevalence of auditory neuropathy/synaptopathy in a population of children with profound hearing loss. *Int J Pediatr Otorhinolaryngol* 70:1415–1422.

Fuchs P, Glowatzki E, Moser T. (2003) The afferent synapse of cochlear hair cells. *Curr Opin Neurobiol* 13:452–458.

Gravel JS, Kurtsberg D, Stapells DR, Vaughan HG, Wallace IF. (1989) Case studies. *Semin Hear* 10(3):272–287.

Hood L. (1998) Auditory neuropathy: what is it and what can we do about it? *Hear J* 51:10–18.

Hood L. (1999) A review of objective methods of evaluating auditory neural pathways. *Laryngoscope* 109:1745–1748.

Khimich D, Nouvian R, Pujol R, Tom Dieck S, Egner A. (2005) Hair cell synaptic ribbons are essential for synchronous auditory signaling. *Nature* 434(7035):889–894.

King A, Purdy S, Dillon H, Sharma M, Pearce W. (2005) Australian Hearing protocols for the audiological management of infants who have auditory neuropathy. *Aust N Z J Audiol* 27(1):69–77.

Kraus N, Azdamar A, Stein L, Reed N. (1984) Absent auditory brainstem response: peripheral hearing loss or brainstem dysfunction? *Laryngoscope* 94:400–406.

Kraus N, Bradlow A, Cheatham M, Cunningham J, King C, Koch D, Nichol T, Mc Gee T, Stein L, Wright B. (2000). Consequences of neural asynchrony: a case of auditory neuropathy. *J Assoc Res Otolaryngol* 1:33–45.

Kurtzberg D. (1989) Cortical event-related potential assessment of auditory system function. *Semin Hear* 10(3):252–262.

Lee JS, McPherson B, Yuen KC, Wong LL. (2001) Screening for auditory neuropathy in a school of hearing impaired children. *Int J Pediatr Otorhinolaryngol* 61:39–46.

Rance G, Beer D, Cone-Wesson B, Shepherd R, Dowell R, King A, Rickards F, Clark G. (1999) Clinical findings for a group of infants and young children with auditory neuropathy. *Ear Hear* 20:238–252.

Rance G, Cone-Wesson B, Wunderlich J, Dowell R. (2002) Speech perception and cortical event related potentials in children with auditory neuropathy. *Ear Hear* 23:239–253.

Rapin I, Gravel J. (2003) "Auditory neuropathy": physiologic and pathologic evidence calls for more diagnostic specificity. *Int J Pediatr Otorhinolaryngol* 67:707–728.

Rapin I, Graziani L. (1967) Auditory-evoked responses in normal, brain damaged and deaf infants. *Neurology* 17:881–894.

Ray J, Gibson W, Sanli H, Haddon A. (2006) Brainstem auditory neuropathy, hair cell desynchrony and cochlear implantation.

Sininger Y, Hood L, Starr A, Berlin C, Picton T. (1995) Hearing loss due to auditory neuropathy. *Audiol Today* 7:10–13.

Starr A, Picton T, Sininger Y, Hood L, Berlin C. (1996) Auditory neuropathy. *Brain* 119:741–753.

Widen J, Ferraro J, Trouba S. (1995) Progressive neural hearing impairment: case report. *J Am Acad Audiol* 6:217–224.

Worthington D, Peters J. (1980) Quantifiable hearing and no ABR: paradox or error? *Ear Hear* 1:281–285.