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Objective Hearing Threshold Estimation in Children with Auditory Neuropathy Spectrum Disorder

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Keywords

Auditory neuropathy spectrum disorder; hearing thresholds; cortical auditory evoked potentials

INTRODUCTION

Auditory neuropathy spectrum disorder (ANSD) is a clinical syndrome characterized by evidence of cochlear function in conjunction with an aberrant auditory neural system. The auditory brainstem response (ABR) cannot be recorded or is grossly abnormal in ANSD patients. Consequently, the fitting of amplification is delayed until behavioral testing is possible. For some ANSD children, behavioral testing is infeasible despite advanced age due to other comorbidities. Therefore, it is crucial to identify a reliable measure of auditory thresholds for ANSD children in order to provide appropriate and timely intervention. Despite an absent ABR, the cortical auditory evoked potential (CAEP) can be recorded from ANSD patients[1-4]. Previous studies have also shown that hearing thresholds can be reliably measured with the CAEP response in SNHL children[5] and its accuracy is as good as or better than that of the ABR[6]. This report demonstrates the feasibility of using the CAEP to estimate hearing thresholds in ANSD children.

MATRERIALS AND METHODS

Patients

Five pediatric ANSD patients (S1 – S5) participated in this study. None had any anatomic malformations or a mixed hearing loss. Detailed demographic information for these patients is shown in Table 1. All patients and/or their legal guardians provided written informed consent to the procedures as approved by the local Institutional Review Board.

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Conflict of Interest: None

Procedures

The study protocol included behavioral threshold assessment using pure-tone audiometry and CAEP measures that were undertaken by different researchers. Frequencies tested in this study included octave frequencies between 250 and 4000 Hz.

Pure-Tone Tests—Behavioral audiometric thresholds were obtained in each ANSD subject in a sound-attenuating chamber. Pure tone thresholds were determined using ER-3A insert earphones using standard behavioral techniques.

CAEP Measures

Stimuli: Stimuli were pure-tone bursts with durations of 100 ms. All stimuli were gated on and off with a 20-ms cosine window. All stimuli were presented through an ER-3A insert earphone to the tested ear at a stimulation rate of 1.1 Hz. Stimulation levels were adjusted adaptively for each subject and frequency.

CAEP recordings: Electroencephalographic (EEG) activity was recorded using a Neuroscan system (version 4.4) and a SynAmp 2 amplifier. The EEG was recorded differentially between surface recording electrodes placed at the high forehead (Fz) and the contralateral mastoid. An electrode placed on the low forehead (Fpz) served as the ground. Eye movements were monitored using a pair of recording electrodes placed above and below the eye that was contralateral to the test ear. Electrode impedances were maintained below 5000 Ohms with an inter-electrode impedance difference of less than 2000 Ohms. The recording window included a 100-ms pre-stimulus baseline and a 700-ms post-stimulus time. During acquisition, the EEG was digitized at a sampling rate of 1000 Hz, amplified with a gain of X10, and band-pass filtered on-line between 0.1-100 Hz with a12 dB/octave slope. Epochs with EEG amplitudes greater than ±100 μV were rejected from averaging. Each response represented the average of 100 artifact-free epochs. For each patient, a minimum of two replicates were recorded for each condition. Responses were then baseline corrected, digitally filtered between 1-30 Hz (12 dB/octave) offline before response analysis. For each subject, replicates recorded for the same stimulation conditions were superimposed. *Two* experienced researchers who were *blind to* stimulus condition, patient identity, and results of behavioral audiometric threshold measures independently determined the presence/absence of the CAEP response based on peak latency, waveform morphology, and the replicable property of neural responses. The inter-judge reliability was 96%. The CAEP threshold was defined as the lowest stimulation level that could reliably evoke the CAEP response.

RESULTS

The CAEP responses were recorded across five frequencies from all subjects tested. These responses consisted of a P1 peak followed by a N2 peak occurring approximately 100 ms later. Figure 1 shows a CAEP level series recorded from subjects S4 (upper) and S2 (lower) at 500 Hz (left) and 2000 Hz (right), respectively. P1 peaks and stimulation levels are labeled for the traces. It is apparent that there is a tendency for P1 latencies to increase and amplitudes to decrease as stimulation level decreases. Figure 2 shows the corresponding air

conduction thresholds measured in the five subjects using both techniques. Generally, CAEP and behavioral thresholds are within 5-10 dBs between 500 and 2000 Hz. The discrepancy is larger for 250 and 4000 Hz although consistent. For 250 Hz, CAEP thresholds are 20-25 dB higher than behavioral responses while at 4000 Hz; the discrepancy is 10-15 dB.

DISCUSSIONS

These preliminary results suggest that the CAEP holds great promise for determining auditory thresholds in children with ANSD. While further subjects are needed to confirm these findings and to better estimate the systematic error at 250 and 4000 Hz, it is apparent that this approach potentially represents a breakthrough for the determination of auditory thresholds in children with ANSD. Although auditory thresholds do not correlate well with speech perception ability in ANSD children[7] this approach will allow early fitting of amplification and, if appropriate, subsequent progression to cochlear implantation at an earlier age.

CONCLUSIONS

The CAEP is sensitive to hearing loss in ANSD patients. It can potentially be used to estimate hearing thresholds in ANSD children.

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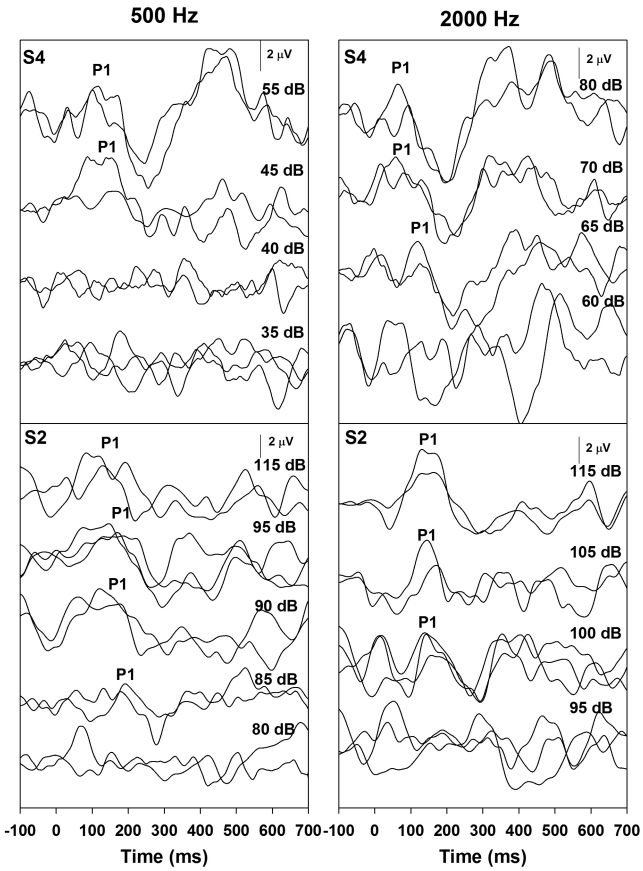

Figure 1.

CAEPs recorded from two ANSD patients at 500 and 2000 Hz. Each trace represents averaged response of 100 artifact free epochs.

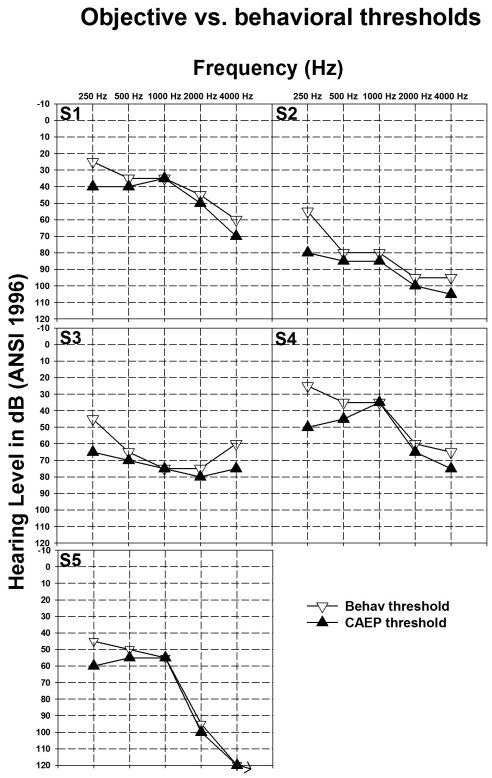

Figure 2.

Hearing thresholds measured with behavioral techniques (unfilled triangles) and the CAEP recordings (filled triangles) in five ANSD patients.

Table 1

Demographic information for implanted children with auditory brainstem implants in this study.

Subject number	Gender	Etiology	Ear tested	Age at implantation (yr)	Age at testing (yr)	Number of active electrodes in the programming MAP
S1	M	Goldenhar Syndrome	R	3.3	10.5	17
S2	M	CHARGE syndrome	L	0.8	6.8	85
S3	F	Premature	L	1.3	8.9	71.7
S4	M	CHARGE syndrome	L	2.4	6.5	66.7