PRELIMINARY REPORT: EARLY LATENCY AUDITORY EVOKED POTENTIALS IN INFANTS WITH DOWN SYNDROME

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ABSTRACT - In individuals with Down syndrome, it has been speculated that compromise of the neural synapses of the auditory brainstem pathway may result in an auditory brainstem response (ABR) in which the evoked potential components exhibit temporal variability. This paper presents our findings on 14 infants with Trisomy 21. Using a mode of stimulation to directly innervate the cochlea, ABR thresholds were obtained which suggest that synaptic abnormality may be present in some cases.

INTRODUCTION

Electric response audiometry is a procedure that involves the far-field measurement of the electroencephalic potentials that occur in response to auditory stimuli using scalp electrodes (Jewett & Williston, 1971). Early latency auditory evoked potentials or the auditory brainstem response (ABR) is a sequence of potentials that occurs within the first ten milliseconds post-stimulus onset (Borg, 1981). Audiologically, the recording of early latency auditory evoked potentials is well established as an accurate procedure for determination of auditory acuity at any age (Hecox & Galambos, 1974; Rowe, 1990; Hyde, Malizia, Ritco & Alberti, 1991). The ABR 'hearing' threshold to a transient acoustic stimulus delivered to the ear via an earphone coupled to the pinna is not significantly different between adults and neonates (Stuart, Yang, Stenstrom & Reindorp, 1993).

The early latency auditory evoked potentials in infants with Down syndrome have previously been found to be similar in waveform morphology to the norm, but with significant differences in the latency-intensity function for wave V at twelve months of age (Folsom, Widen & Wilson, 1983). The amplitude of wave V has also been reported to be smaller (Jiang, Wu & Liu, 1990).

Dendritic abnormalities of the cerebral cortex have been reported in individuals with Down syndrome at all ages, being more pronounced with increasing age, that is, there

is deficient synaptogenesis and dendritic atrophy (Takashima, Iida, Mito & Arima, 1994). Such synaptic abnormality or 'synaptic insecurity' would presumably affect the timing of the single event related potentials, resulting in a greater variance of latencies for these single potentials, with the subsequent average potential reflecting this greater variability by being reduced in amplitude.

"One of the assumptions of the averaging technique [that improves the signal to noise ratio for the ABR] is that the evoked potential waveform remains constant with each presentation of the stimulus" (Picton, Linden, Hamel & Maru, 1983, p.332). Galbraith (1986) suggests that the assumption of response constancy is not valid for individuals with Down syndrome, due to synaptic insecurity of the neurons of the brainstem.

Brainstem electric response audiometry was performed on 14 infants with Trisomy 21, the ABR recorded to a transient stimulus delivered via a bone conduction transducer. A bone conduction ABR provides a direct measure of cochlear reserve. Bone conduction ABR threshold should be depressed in those infants where synaptic insecurity of the brainstem auditory pathway exists, presuming no cochlear dysfunction.

METHOD

Electrophysiological assessment of each infant was performed using a Medilec Audiostar AEP System, with most assessments conducted at the infant's place of residence. Infants were assessed in a natural sleep to minimize electromyogenic activity. All infants were in the age range 4 to 57 weeks at the time of assessment. Auditory brainstem responses were generated by a 100 microsecond electrical pulse delivered to a Radioear B71 bone conduction transducer.

The electroencephalic activity recorded at the scalp electrodes was bandpass filtered (30Hz to 3000Hz) and amplified upto 250 000 times. Analysis time was 10 milliseconds post-stimulus onset. The ADC sampling rate was 31.25 kHz.

The auditory stimulus was presented at one of two stimulus repetition rates, 10Hz or 40Hz. The object was to identify, using a decreasing stimulus intensity series, the stimulus intensity level at which no ABR was identifiable. The presence of an ABR was predicated on the identification of the first replicable evoked potential with a latency > 5.5ms (Edwards, Durieux-Smith & Picton, 1985). The lowest stimulus intensity level at which a replicable evoked potential was obtained was defined as ABR threshold (Stuart, Yang, Stenstrom & Reindorp, 1993).

RESULTS

Results are presented combining the ABR threshold obtained for each stimulus repetition rate. Table one gives the ABR threshold of the 14 subjects, stimulus intensity level referenced to 1 microvolt.

Stimulus Intensity Level	Number of infants
46 dB	5
56 dB	5
66 dB	2
76 dB	2

Table 1. ABR thresholds of the 14 subjects

DISCUSSION

Stuart, Yang, Stenstrom & Reindorp (1993) found for a group of neonates that the range of normal ABR thresholds was 25 to 45 dB (re: 1 microvolt) using technical specifications similar to this study. Cornacchia, Martini & Morra (1983) reported for a group of infants 16 - 20 months of age, using much narrower bandpass filtering than the present study, that ABR thresholds were less than or equal to 15 dBnHL (thought to be approximately 50 dB (re: 1 microvolt)). It is clear from these two studies that the ABR thresholds of 66 dB and 76 dB (re: 1 microvolt) are elevated.

Depressed bone conduction ABR thresholds are highly supportive of Galbraith's hypothesis of synaptic insecurity, assuming no cochlear dysfunction. Why only some of the infants with Down syndrome have depressed bone conduction ABR thresholds is open to conjecture. Further analysis of these findings is obviously necessary. One caveat to the conclusion of synaptic insecurity is in relation to significant, prolongued conductive hearing loss, present from birth, which may affect the development of the auditory central nervous system (Sohmer & Friedman, 1992). Research into ABR bone conduction threshold levels in infants with and without Down syndrome with otitis media present at, or soon after, birth resulting in hearing impairment of moderate degree or greater is necessary to examine further the effect of auditory deprivation on the development of the auditory central nervous system and the subsequently obtained ABR.

Of prime importance from an audiological standpoint is the extension of these findings to interpretation of elevated or absent air conducted ABR's in infants with Down syndrome, and the need to assess cochlear reserve in this population electrophysiologically.

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