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Short communication

CEREBRAL CORTICAL CONTRIBUTIONS TO SENSORY EVOKED POTENTIALS: HYDRANENCEPHALY [1]

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Summary The contribution of the cerebral cortex to the generation of sensory evoked potentials was studied in an infant with hydranencephaly. On CT scan no tissue above the thalamus was noted. Long-latency potentials to auditory stimuli were absent whereas the short-latency or brain-stem auditory evoked potentials and some of the components of the middle latency auditory evoked potentials (No and Po) were present. To visual stimulation only the electroretinogram was detected. To somatosensory stimulation only the spinal cord potentials could be detected. The absence of long-latency components in each of the sensory modalities supports the concept that these potentials require intact cerebral hemispheres in man.

Keywords: *hydranencephaly – sensory evoked potentials – ABR – VER – SSEP*

Sensory stimulation of the auditory, visual and somatosensory systems evokes a series of neural events that may be recorded non-invasively from the skin (Starr 1978). The precise sites of generation of many of the evoked potential components are not known. Patients with hydranencephaly have their cerebral hemispheres replaced by cerebrospinal fluid with or without a significant change in the size of the cranial vault (Halsey et al. 1971). The brain-stem, midbrain and thalamus are usually of normal configuration. We have studied a patient with hydranencephaly to help determine the effects of cerebral hemisphere loss on the various components of sensory evoked potentials (auditory, visual and somatosensory).

Methods

Auditory evoked potentials were recorded from electrodes placed according to the 10-20 international method at Cz and referenced to the mastoid ipsilateral to the ear being stimulated. Upper extremity somatosensory evoked potentials were obtained by placing electrodes at C4′ (3 cm lateral of C4) for right median nerve stimulation or C3′ (3 cm lateral of C3) for left median nerve stimulation and referenced to Fpz. Peripheral nerve activity was measured between Erb's point and Fpz. Spinal cord potentials were recorded between electrodes placed over L1 and L5 of the spinous process. Visual evoked potentials were recorded between an electrode at Oz referenced to Fpz. The electroretinogram was recorded from an electrode placed just below the eye and referenced to Fpz. The ongoing electrical activity was amplified and filtered (3 dB down) from 30 to 3000 Hz for the auditory brain-stem potentials; from 5 to 1500 Hz for both the middle-latency auditory and the visual evoked potentials (including the electroretinogram); from 1 to 500 Hz for the long-latency auditory evoked potentials; and from 1 to 3000 Hz for the somatosensory evoked potentials.

Auditory brain-stem potentials were evoked by monaural acoustic stimulation to clicks (100 μsec duration) presented at 11.1/sec for a series of 2048 trials at an intensity of 75 dB nHL. In addition, acoustic clicks were presented at 41/sec for intensities of 65 dB nHL, 40 dB nHL, and 20 dB nHL to establish estimated hearing levels. Middle-latency auditory evoked potentials were stimulated by tone pips presented at 9.1/sec with a frequency of 1000 Hz for 1024 trials at 85 dB nHL. The same tone burst was used to evoke long-latency auditory evoked potentials but presented at a rate of 0.7/sec for 512 trials. Somatosensory evoked potentials were obtained to stimulation of the median nerve at the wrist or the posterior tibial nerve at the ankle at a rate of 0.9 pulses/sec. The current was adjusted to the threshold for evoking a contraction of the thenar muscles. A series of 256 trials were acquired. Monocular stroboscopic flash stimulation was achieved by placing a patch over the non-test eye and stimulating the test eye at a rate 1.2/sec for a series of 1024 trials. The patient was tested while lying in a crib in a state of sleep or resting quietly and tested several times throughout the first year of life.

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Results

Case report

This 8-month-old male infant was born to an 18-year-old primagravida at term following an uncomplicated pregnancy. An emergency Caesarian section was occasioned by signs of fetal distress. The birth weight was 3345 g and the Apgar scores were 7 at 1 min and 9 at 5 min. Pathologic examination of the placenta showed focal angiomatosis and infarction involving at least 15% of the surface.

At birth, the head circumference was 41.0 cm (greater than the 97th percentile). A precipitous increase occurred over the next 3 weeks to 45.0 cm. At 3 weeks, the cranial-facial ratio was abnormally increased, the cranial sutures were widely split, MacEwen's sign was positive, and transillumination with a standard cuffed flashlight produced a generalized increase in light transmission. Additional positive findings included a high pitched cry, generalized increase in muscle tone, and bilateral extensor plantar responses. A 'setting sun' sign was absent and the eyes remained in the horizontal meridian. No systemic abnormalities were observed. At a chronological age of 3.5 weeks, a ventricular-peritoneal shunt was placed for ease of nursing management. Cerebral spinal fluid protein concentration was 1019 mg/100 ml.

Computed cranial tomography

The CT scan in Fig. 1 shows normal appearing cerebellum, pons, brain-stem and thalamus (Fig. 1A). There were large hypodense areas in the anterior and lateral planes representing fluid which had replaced both cerebral hemispheres. Above this level (Fig. 1B and C), no appreciable cortical tissue could be identified except for a small amount of posterior temporal lobe. The apparent increase in cerebrospinal fluid density is attributed to proteinaceous debris resulting from destruction of the cerebral hemispheres.

Sensory evoked potentials

Sensory evoked potentials for each of the modalities evaluated are seen in Fig. 1 and include 'typical' tracings from a normal-term infant. Table I gives the means and standard deviations for normal latencies in a term infant of sensory evoked potentials for our laboratory. We define abnormal values as latencies that are greater than 2.5 S.D.s beyond the mean. Also, Table I lists the values obtained in this patient.

(1) Auditory evoked potentials

(a) Auditory brain-stem potentials. Auditory brain-stem potentials were recorded 1 week after birth to monaural stimulation at 85 dB nHL and 11.1/sec stimulation rate. Although differences in wave form morphology are seen between the normal infant tracings and between the two tracings in this patient, normal absolute latencies and inter-component conduction times were observed. Wave V was identified as the component that persisted both at the fast stimulus rates (40/sec) and at the lowest sound intensities (40–60 dB nHL). Decreasing the stimulus intensity at fast rates of stimulation (41 clicks/sec) did not result in a reproducible wave V below

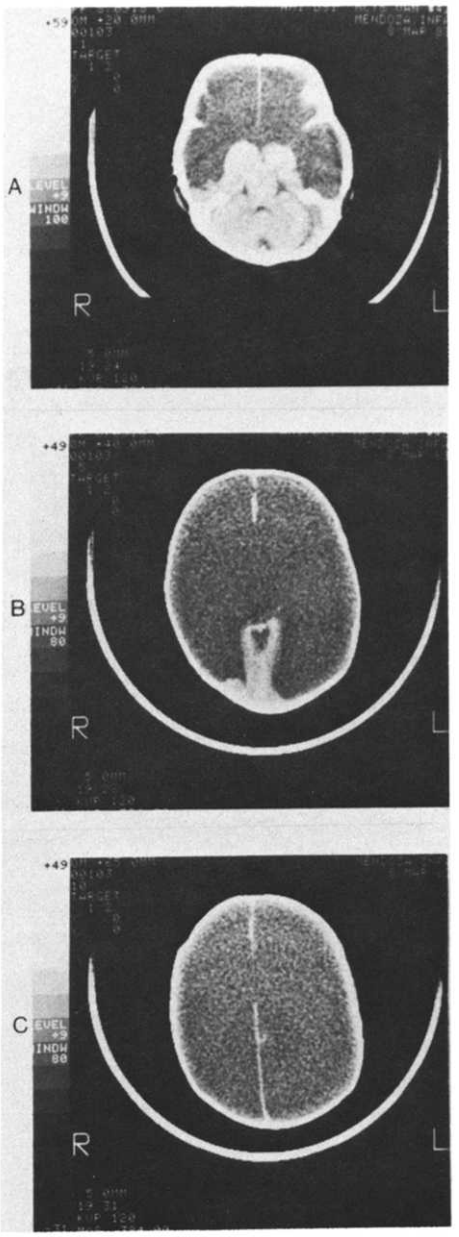

Fig. 1. CT scans showing (A) normal cerebellum, pons, and thalamus with large hypodense areas in the anterior and lateral planes representing the replacement of cerebral hemisphere with fluid; (B) the replacement of the cerebral hemisphere with CSF except for a small amount of the temporal lobe; and (C) a complete absence of cerebral hemisphere at this level.

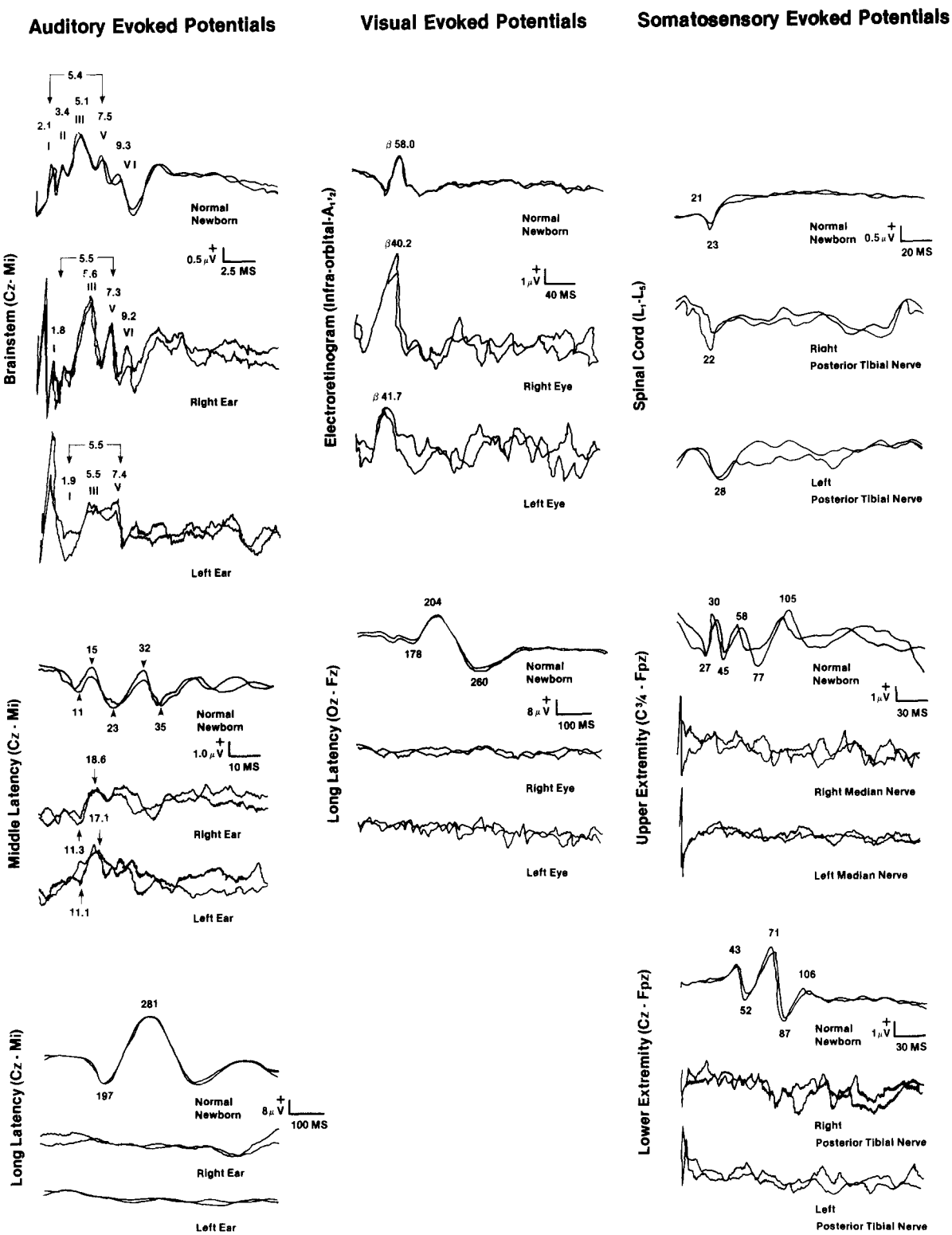

Fig. 2. Sensory evoked potentials in the patients with hydranencephaly and a normal subject for the various modalities are shown. In response to auditory stimulation there are auditory brain-stem, middle-latency and long-latency potentials. Visual stimulation includes the electroretinogram and long-latency potentials. Somatosensory stimulation included spinal cord potentials (upper panel) and cerebral potentials to posterior tibial nerve stimulation (lower panel) and median nerve stimulation (middle panel).

40 dB nHL for right ear stimulation, and 60 dB nHL for left ear stimulation.

(b) Middle-latency auditory evoked potential. Middle-latency potentials were normal for the first negative peak occurring at 11.3 msec for right ear stimulation and 11.1 msec for left ear stimulation, and for the first positive peak occurring at 18.6 msec for right ear stimulation and 17.1 msec for left ear stimulation. The later components (see the normal tracing on

TABLE I

95% confidence intervals for latencies of the sensory evoked potentials in term infants and latencies of the sensory evoked potential in a 40 week gestational age (41 week conceptual age) hydranencephalic infant.

Modality	Wave component latency (msec)			
Auditory evoked potential	I	III	V	I–V
Brain-stem				
95% interval	2.2	5.6	7.8	5.6
Normal infant	2.1	5.1	7.5	5.4
Patients: right ear	1.8	5.6	7.3	5.5
left ear	1.9	5.5	7.4	5.5
Middle latency	No	Po	Na	Pa
95% interval	12.7	18.8	28.4	41.2
Normal infant	11.0	15.0	23.0	32.0
Patient: right ear	11.3	18.6	–	–
left ear	11.1	17.1	–	–
Long latency	N1	P2		
95% interval	No actual norms established			
Normal infant	197	281		
Patient: right ear	–	–		
left ear	–	–		
Visual evoked potential	ERG	N1	P1	N2
95% interval	61	205	276	338
Normal infant	58.0	178	204	260
Patient: right eye	40.2	–	–	–
left eye	41.7	–	–	–

Somatosensory evoked potentials

Spinal cord response		N22					
95% interval		29					
Normal infant		23					
Patient: right nerve		22					
left nerve		28					
Upper extremity *	P/N14	N20	P27	N30	P45	N80	P100
95% interval	30	44	Identified as absent/present				
Normal infant		27	30	45	58	77	105
Patient: right nerve		No consistent responses					
left nerve		No consistent responses					
Lower extremity		N30	P40	N50	P60	N75	P100
95% interval		Identified as absent/present					
Normal infant		–	43	52	71	87	106
Patient: right nerve		No consistent responses					
left nerve		No consistent responses					

Note: labels used are for adult responses and may not be appropriate for an infant population; used for illustrative purposes only.
* Labeling after Mauguière et al. (1983).

Fig. 2 for the middle-latency components) were absent following stimulation of either ear (Fig. 2).

(c) Long-latency auditory evoked potentials. Long-latency auditory evoked potentials occurring between 80 and 200 msec were absent (Fig. 2).

(II) Visual evoked potentials

A well-defined electroretinogram (ERG) for monocular stimulation was observed (B wave latency 41.7 and 40.2 msec for left and right eye, respectively). The long-latency scalp derived cortical components were absent (Fig. 2).

(III) Somatosensory evoked potential

(a) Short-latency upper extremity potentials. Stimulation of the left and right median nerves at 1 week of age showed no cortical components (Fig. 2, middle panel) as well as no peripheral nerve components from Erb's point. However, in our experience approximately 40% of normal newborns do not show such a peripheral nerve component even though the stimulus at the wrist causes the thenar muscles to contract and cortical components can be recorded from the scalp.

(b) Lower extremity somatosensory evoked potentials. Lower extremity somatosensory evoked potentials could not be successfully obtained at age 1 week due to movement artifacts. However, at 8 months of age a clear spinal cord potential was noted for both left and right posterior tibial nerve stimulation. No long-latency cortical potentials over the scalp were present (Fig. 2, upper and lower panels).

Electroencephalography (EEG)

The EEG at 4 days of age was markedly abnormal with severe suppression of all activity.

Discussion

In this patient with hydranencephaly, no long-latency potentials were elicited to auditory, visual, or somatosensory stimulation. The short-latency auditory evoked potentials were successfully recorded and reflect the sequential activation of the auditory pathway along the brain-stem (Buchwald and Huang 1975; McPherson et al. 1985). The earliest components of the middle-latency auditory potentials (MLRs) were also observed (No and Po) but the later waves (Na, Pa, and Nb) in this complex were absent. The MLR is believed to represent activity of the medial geniculate and polysensory nuclei of the thalamus (Picton et al. 1974; Polich and Starr 1983).

In both the somatosensory and visual systems there was evidence of activity in peripheral parts of these sensory systems. Nerve root and spinal cord activity as recorded over the lumbar region to stimulation of the lower extremity and a normal ERG to light flash were detected.

As in most cases of hydranencephaly (Halsey et al. 1971), liquefaction of the cortex appeared to be complete except for a small remnant of the posterior temporal lobe. Norman (1958) has suggested that the proteinaceous debris in the CSF may be related to prenatal occlusion or agenesis of the carotid arteries.

The findings in this case indicate that all of the so-called 'long-latency' sensory evoked potentials in man, i.e., auditory, visual and somatosensory, require the presence of intact cerebral hemispheres. This result is consistent with other observations of lesions in man from a variety of causes (Spehlmann et al. 1977; Knight et al. 1980; Phelps et al. 1981; Chiappa 1983; Mauguière et al. 1983). Of particular interest is the finding that in infants for the first negative response (N27, Table I) of the upper extremity SSEP was absent. In adults, Chiappa (1983) reports that the first negative response (N20) is thalamic. It appears that this may not be the case in infants and that the response is generated in the cortex. The results from the present case suggest that the Na, Pa and Nb waves of the middle-latency auditory evoked potentials would appear to require the preservation of cortical structures as well. Kraus et al. (1982) found that in unilateral lesions of the temporal lobe, waves Na are present and unaffected; in contrast, the amplitude of Pa was asymmetrical demonstrating a reduced amplitude over the affected hemisphere. Kraus et al. (1982) concluded that Na and Pa are generated separately, Na deriving from subcortical structures and Pa from auditory cortex. Both these components were absent in our recordings from this patient, whereas the earlier No and Po components were present. The precise location of the generators for No and Po are not known but their origin in thalamus or midbrain seems likely. The observation in this patient of a normal ABR and radiographic evidence of intact brain-stem and thalamic structures with small amounts of posterior temporal lobe lends support to the concept that both the ABR waves and the earliest components of the middle-latency response (No and Po) depend on intact structures up to and including the thalamus.

Résumé

Contributions du cortex cérébral à des potentiels évoqués sensoriels: hydranencéphale

La contribution du cortex cérébral à la production de potentiels évoqués sensoriels a été étudiée chez un nouveau-né hydranencéphale. Au scanner, aucun tissu n'avait été observé au-dessus du thalamus. Les potentiels de longue latence à des stimulus auditifs étaient absents alors que les potentiels à courte latence ou les potentiels évoqués du tronc cérébral ainsi que certaines des composantes du potentiel évoqué auditif à moyenne latence (No et Po) étaient présentes. Pour des stimulations visuelles, seul l'électrorétinogramme était détecté. Pour des stimulations somesthésiques, seuls les potentiels spinaux étaient obtenus. L'absence de composantes à longue latence pour les 3 modalités sensorielles confirme que l'existence de ces composantes nécessitent l'intégrité des hémisphères cérébraux chez l'homme.

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