EVOKED POTENTIALS AND DOPPLER VASCULAR STUDIES IN HYDRANENCEPHALY

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SUMMARY

A study of brainstem auditory evoked potentials, electro-encephalography and doppler vascular scans were conducted in an infant with hydranencephaly. The abnormalities detected were consistent with the gross cerebral cortical anomaly in hydranencephaly.

Key Words: Hydranencephaly , Brain stem auditory evoked potentials (BAEP) , Somatosensory evoked potentials (SSEP), Doppler vascular scan, Electro encephalography (EEG)

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INTRODUCTION

Hydranencephaly is a clinically recognizable syndrome of increased cranial transilluminability in a macro or normocephalic newborn or infant. Pathologically it is characterised by marked reduction in the total brain substance which is largely replaced by fluid within the cranium (1). Electrophysiological studies, including brain stem auditory evoked potentials (BAEP), somatosensory evoked potentials (SSEP) and electroencephalography depend upon the contribution from the various components of the central nervous system; these will show abnormalities in the presence of major defects in the brain in hydranencephaly. In addition, doppler vascular scan which can demonstrate decreased velocity of blood flow in the carotid arteries or its major branches can provide diagnostic clues regarding the vascularity of the brain. The present paper reports the results of these tests in an infant with hydranencephaly.

METHODS

BAEPs were recorded from electrodes placed at central midline (Cz) and referenced to the ipsilateral mastoid, with frontal midline (FPz) as common electrode (10 — 20 system of EEG electrode placement). Electrode resistance was kept below 2 k ohms. Mono-aural accoustic stimuli rarefaction clicks of 100m sec duration and 90 dB nHL intensity were delivered through ear phones at 11.4/sec from a series of 2000 clicks. Sampling time was 10 msec. The signals were led to a Nicolet C4 evoked potentials system with filter band pass 150-3000 Hz. The BAEPs were recorded twice from each ear for consistency of results.

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The SSEP was done by stimulating the left median nerve at the wrist and recording at the Erb's point (spinous process of the seventh and first cervical vertebrae) and from the contralateral scalp overlying the somatosensory cortical area using measurements described by Giblin (2). The reference electrode was placed on the left mastoid and the grounding electrode on the left arm. Electrode impedances were kept below 5 k ohms. Rectangular pulses of current of 0.2 msec duration and intensity twice that required to produce a twitch of the thumb were used. The signals to 500 stimuli were averaged in a Nicolet C4 system with filter band pass 2 — 2000 Hz at a stimulus rate of 2/sec and with sampling time of 100 msec.

By means of a Vasoscan bidirectional continous wave doppler system, with 4 and 8 MHz probes, the common, internal and external carotid and vertebral arteries were insonated in the neck and the doppler flow velocity spectra recorded. The flow velocities in the superficial temporal, supratrochlear, subclavian and ophthalmic arteries were also studied. An attempt was also made to detect flow signals in the anterior, middle and posterior cerebral arteries from appropriate anatomic positions over the scalp of the hydranencephalic child.

RESULTS

Case Report

T.S., a 2 months old female baby was born to a 27 year old healthy woman at home following an uncomplicated pregnancy at term. The birth weight was 3080g and the newborn was reported to have cried immediately after birth. Head circumference was not noted at birth. The baby was reported to be ' normal ' by the mother until 4 weeks of age when refusal of feeds was first observed. At about this time, involuntary jerky movements were observed by the mother. These were brief and were localised to the lower limbs, they were unassociated with eye movements, salivation, loss of consciousness or urinary incontinence, and used to occur 2 — 3 times daily. The mother observed a progressive enlargement of the head at 6 weeks after birth and occasional vomiting after feeds and decided to seek medical treatment.

On examination, the body weight was between the 10th and 50th percentiles; the height was on the 50th centile and the head circumference above the 90th centile. The infant was conscious and alert but was unable to follow objects or smile at the mother; menace reflex was absent. Eye movements were full, using doll's eye man-

ouver; the pupils were 3mm in diameter and were reactive to light. Fundus examination revealed bilateral atrophy and hypoplasia of the optic discs. On palpation of the head, sagittal and coronal sutural separation was evident. The anterior and posterior fontanellae were full. There was a uniform hypertonia in all four limbs, more in the arm flexors and extensors of the lower limbs. The infant was able to move all four limbs. Occasional decorticate posturing was observed during examination. The plantar reflexes were extensor and the deep tendon reflexes were exaggerated. Crossed extensor and adductor reflexes were elicitable. The primitive reflexes including snout, palmomental, sucking, rooting moro and grasp were demonstrable. Cardiovascular, respiratory and abdominal examination was normal. Transillumination of the head using a standard cuffed transilluminator produced a brilliant transillumination. A CT Scan revealed normal appearing cerebellum, brainstem, thalamus and large hypodense areas were seen in the region of cerebral hemispheres. Appreciable cerebral cortical tissue could not be identified (Fig. 1).

Somatosensory evoked potentials (SSEP) revealed an attenuated brachial plexus potential. The spinal and cortical components were absent (Fig 2). The early components of BAEP were absent (Fig 3). By doppler imaging, normal blood flow velocities were seen in the subclavian, common and external carotid arteries in the neck. In comparison, flow velocities were reduced in the internal carotid in the neck and the opthalmic arteries. Electroencephalogram showed an isoelectric tracing except for slow wave activity in the temporal regions. Focal spikes in phase reversal across T2 were seen in the temporal region indicating right temporal lobe epilepsy (Fig 4).

DISCUSSION

Hydranencephaly is a condition which is characterised by the absence of all the areas of the telencephalon and variable portions of the brainstem and cerebellum. The presumed time of onset, pathogenesis, regions of involvement of the central nervous system and anatomical organization of the brain remnants vary from case to case (1). Clinically the affected infant may have severe neurological abnormality at birth, or the neurological development may be arrested beyond the newborn period. Rarely, however, the development may appear to be normal in a few of the chronic survivors. In the present case, the infant was stated to be apparently normal at birth although a neurological assessment was not done since she was delivered without medical supervision at home. At 2 months, the development was delayed as evidenced by lack of fixation, absence of social smile, and lack of visual following. The primitive reflexes were however, demonstrable. Preservation of these reflexes has been a feature of most of the chronic survivors in hydranencephaly, possibly indicating the presence of brainstem and diencephalon. Larber (3) described two cases of children who were subjected to ventriculography because of slightly enlarged heads. No brain substance could be identified. Remarkably, the development of one of these children was normal. But this is an unusual finding in chronic survivors. Optic atrophy has been a frequent finding in most of the cases of hydrencephaly as in our case. Hydrocephalus is frequently present and probably often plays an accessory role in the morphogenesis (1). However, clinically the infant may have normocephaly. External examination may reveal no other obvious facial or limb malformations.

The occurrence of seizures in hydrancephaly is an interesting but not an unusual event. Various types of generalised or partial seizures have been described (3). Infantile spasms and myoclonic jerks have also been documented (1). However, temporal lobe seizures, as documented by electroencephalography, as in our case, has not been documented to our knowledge. This possibly suggests the presence of remnants of cortex in the temporal lobe. Generally, however an isoelectric EEG activity in an otherwise normal infant is suggestive of hydranencephaly.

The contribution of the cerebral cortex to sensory evoked potentials is highlighted by the fact that the cortical components of auditory and visual evoked potentials are markedly absent in hydranencephaly. Lott et al (4) studied the cerebral cortical contributions to auditory, visual and somatosensory evoked potentials and reached the same conclusion. In brain stem auditory evoked potentials (BAEP), the early components are believed to arise from the brainstem; they were absent in our study. The finding is in contrast to that reported by Lott et al who could record the early components of the BAEP. The absent BAEP in our case signifies additional brainstem anomaly. The somatosensory evoked potentials (SSEP) showed absent cortical and spinal elements; this result is similar to the one obtained by Lott et al (4). In early infancy, over 40% of normal infants have absent brachial plexus and spinal potentials; however their cortical components are preserved (5). In the present case, the results probably denote the atrophy of the posterior columns in the spinal cord. Doppler vascular study shows a decreased flow in the internal carotid and ophthalmic arteries. This probably represents the changes secondary to occlusion of the intracranial portion of the internal carotid arteries. In conclusion, studies such as BAEP, SSEP and EEG are useful tools to demonstrate the extent of the cortical loss in hydranencephaly. Doppler vascular study is a non-invasive investigation which holds promise in the study of this condition.

REFERENCES

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Legends For Photographs

Fig. 1. Unenhanced CT Scan shows absence of cerebral cortex and fluid filled cavities replacing the cerebral hemispheres. (C). The diencephalic and cerebellar structures are well-developed. (a)

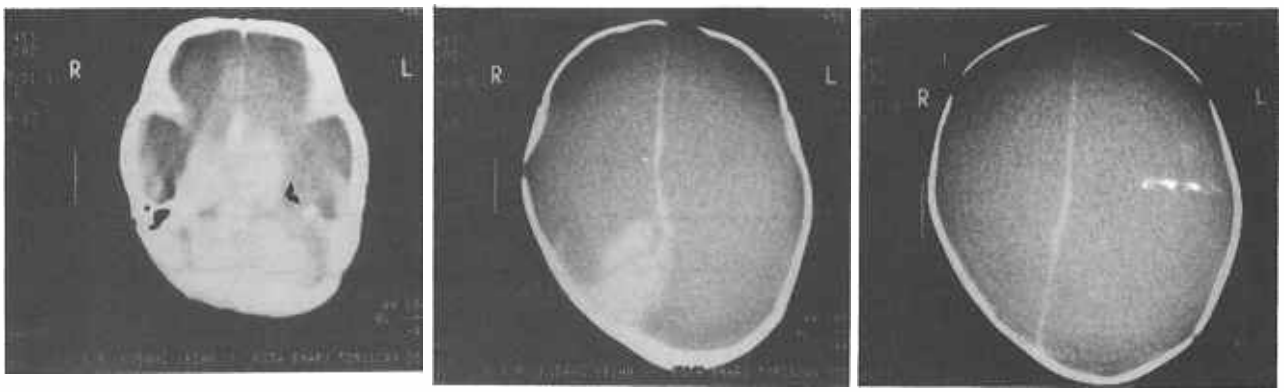

Fig. 2. Somatosensory evoked potential. M1, M2, M3 and M4 are the tracings from the brachial plexus, C7 spinal, C1 spinal and scalp overlying the somatosensory cortex, respectively. M1 shows a poorly defined response from the brachial plexus. The spinal and cortical responses are absent.

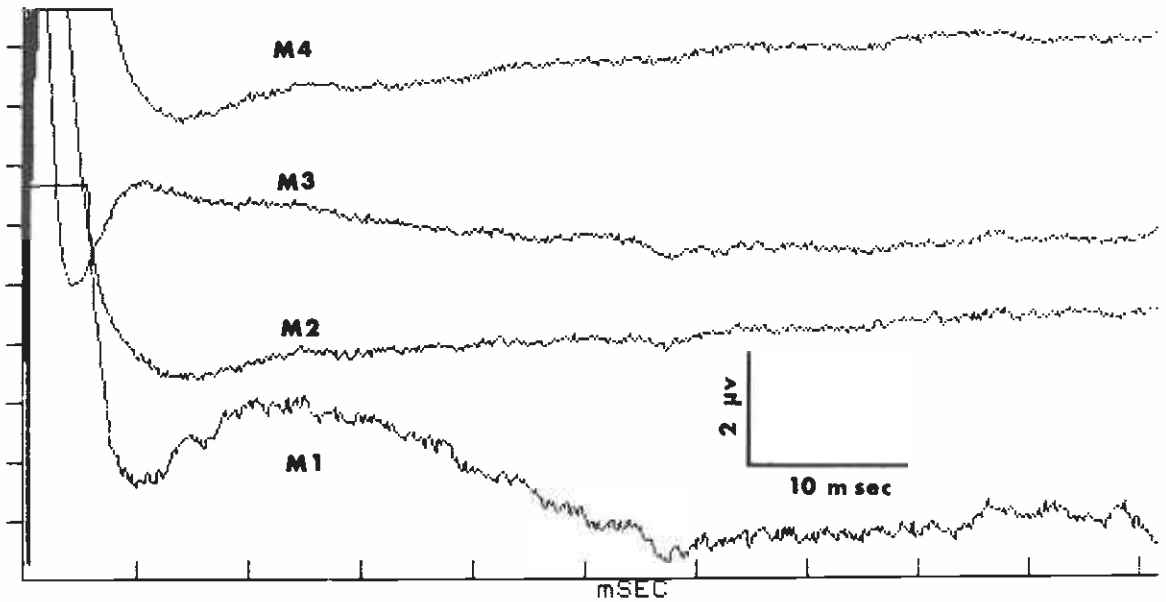

Fig. 3. M1 and M3 are the brainstem auditory evoked potential tracings from left and right ear stimulation respectively. The potentials are absent.

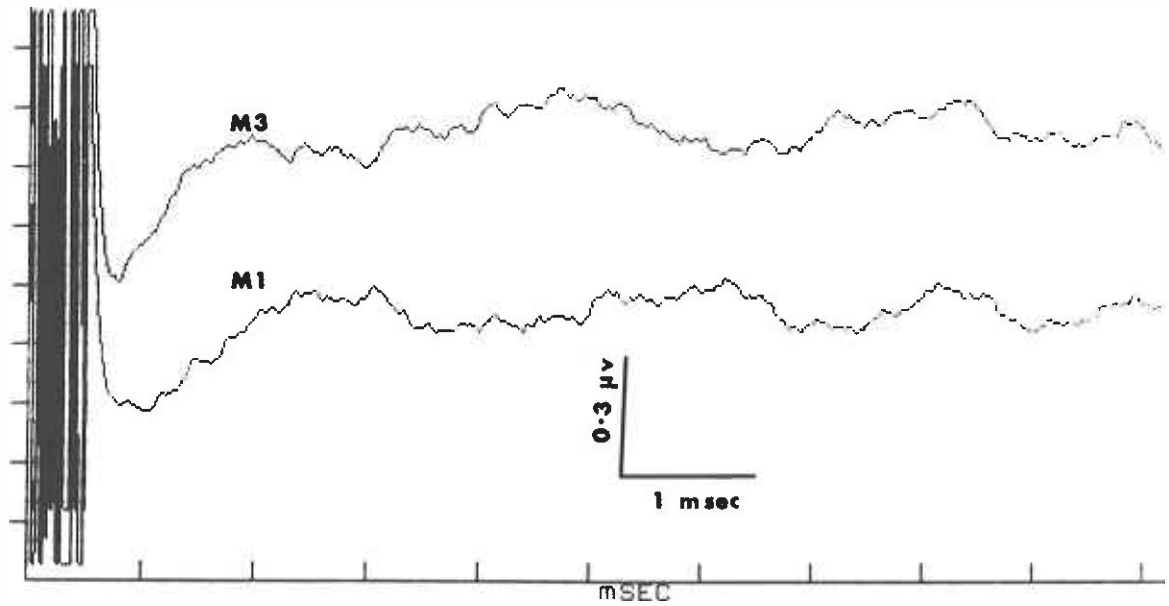

Fig. 4. EEG showing an isoelectric trace except for slow waves in the temporal regions. Focal spikes and slow waves are seen in phase reversal across T2.