The Prognosis of Encephaloceles

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The term "encephalocele" generally denotes a cephalic hernia through a congenital defect in the skull (cranium bifidum). The protrusions contain meninges and cerebrospinal fluid (cranial meningocele) and may also contain some portion of the brain (meningoencephalocele or encephalomeningocele). They may be partially or completely epithelialized. The great majority of encephaloceles are located in the occipital area. Anterior encephaloceles are uncommon and have been classified into sinucipital varieties which visibly involve the nasofrontal region or the extremely rare basal ones which are hidden in the nasopharynx or orbit. The incidence of this malformation has been estimated at one in every 3000 to 10,000 live births. The clinical manifestations and surgical management of encephaloceles have been described in reports dealing for the most part with the larger related problems of spina bifida and myelomeningocele. There are no comprehensive surveys on the morbidity and the quality of survival in individuals born with different types of encephaloceles. Lorber, however, has recently reviewed the prognosis of occipital encephaloceles in a series of infants. There is no comparison of the long-term outlook with this condition before and after ventriculottrial shunt operations were widely adopted for controlling the hydrocephalus associated with many encephaloceles.

This report analyzes the morbidity of encephaloceles and the results of treatment over a 20-year span through 1967 at the Indiana University Medical Center. The study interval includes the decade immediately before and that just after the first ventriculottrial shunting operations were done at this institution. The follow-up has been long enough to define the frequency and magnitude of various physical handicaps and mental sequelae associated with encephalocele and to see how these residuals affect the lives of the patients reaching childhood and adolescence.

Case Material

During a 20-year interval from January 1, 1948, through December 31, 1967, there were 76,287 admissions to the Childrens Hospital at Indiana University Medical Center, with approximately 3700 of these directly to the neurosurgical service. These included 559 infants with myelomeningoceles, 60 with encephalolocles, and four with both myelomeningoceles and encephaloceles. Of the 64 infants with encephaloceles, 23 were seen as newborns and 41 within the first week of life. A 65th case of encephalocele was seen when the patient was 3 years old. Myelomeningoceles and encephaloceles combined accounted for 0.82% of all pediatric admissions, and one of every six pediatric neurosurgical admissions. Encephaloceles constituted 10.3% of this group of cranial and spinal malformations. The 65 patients had 67 encephaloceles, 55 being occipital, three interparietal, eight nasofrontal, and one nasopharyngeal. Forty-four of these 65 patients were females and 21 were males. The sex predilection was limited to occipital encephaloceles where 40 of the 55 cases were females (significance p < .01). In contrast, 6 of 10 encephaloceles in other locations occurred in males.

Results: Non-Surgical

There were 18 patients who were not operated on; 12 of these had occipital encephaloceles with maximum external diameters ranging from 1 to 10 cm, averaging 6.4 cm. The size of the encephaloceles per se was not the reason for withholding treatment (Fig. 1). Surgery was withheld in most cases

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Fig. 1. **A.** Occipital lesion obscured by hair and mistaken for an encephalocele. **B.** Unoperated occipital encephalocele in an infant who lived only 8 days. **C.** Large occipital encephalocele in a young infant who postoperatively is grossly retarded at 6 years of age and has hydrocephalus. **D.** Transillumination of a huge occipital encephalocele in a newborn infant who postoperatively is essentially normal at 17 years of age.

because of poor general condition. Of the 18 patients, 15 are dead. Two of the dead infants had large frontal defects, and another had separate parietal and occipital encephaloceles. Each of these three infants also had a myelomeningocele and was paraplegic. Nine died 4 hours to 23 days after birth, six others died when 2 to 10 months old from inanition, pneumonia, and meningitis. In only one death was an untreated hydrocephalus believed to be an important contributory cause. The incidence of other major congenital problems was high in these 15 infants and included myelomeningocele (3), microcephaly (2), seizures (3), cleft lip and palate (4), dysplastic extremities (2), congenital heart disease (2), and laryngomalacia (1).

Three patients whose cephalic lesions were never operated on are living. One is the adult who came for treatment of an encephalocele when 33 years old; she is now 45 years and has been institutionalized for custodial care. She has always been retarded and childlike but is capable of self-care under supervision. She has had occasional generalized seizures but there are no physical handicaps. The second living patient was admitted on the day of birth with a midfrontal cranium bifidum and a large soft tissue protrusion. An associated lumbosacral myelomeningocele was repaired immediately, and 3 weeks later a ventriculoatrial shunting procedure was necessary for progressive hydrocephalus. Thereafter, with the hydrocephalus controlled, the cranial defect closed spontaneously by the time the patient was 3 years old. This boy is now 5 years old, mildly retarded, paretic below the knees, but able to walk with crutches and braces. His head is of normal shape and size.

The third non-surgical survivor in this series was admitted at 2 weeks of age with what was believed to be an occipital encephalocele (Fig. 1A). Operation was deferred at that time, and when the infant was