Institutional experience with cranial vault encephaloceles

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Object. There is scant literature regarding the long-term outcome in patients with cranial vault encephaloceles, and what literature there is may underestimate long-term deficits. The goal of this study was to address this lack of information.

Methods. The authors performed a retrospective chart review of cranial vault encephaloceles performed at our institution between 1989 and 2003. Fifty-two total patients were identified and 44 of these cases were reviewed. Additionally, 34 of the 44 patients were contacted and given an outcome survey (Hydrocephalus Outcome Questionnaire [HOQ]) to evaluate physical, emotional, cognitive, and overall health outcomes.

Results. The mean age for patients in this cohort was 9.6 years (range 4–17 years) and the mean follow-up time was 9.2 years. There was an equal sex distribution and there were no deaths. Hydrocephalus was found in 60% of occipital and 14% of frontal encephaloceles, and epilepsy was confirmed in 17% of occipital and 7% of frontal lesions. Outcome assessments performed using the HOQ showed that 50% of the patients with occipital encephaloceles had overall HOQ health scores of 0.5 or less and 55% had HOQ cognitive scores of 0.3 or less, compared with 0% of patients in both categories who had frontal encephaloceles. It was also found that the presence of hydrocephalus and epilepsy independently and significantly lowered the overall health scores.

Conclusions. Occipital encephaloceles carry a worse prognosis than frontal encephaloceles, with higher rates of hydrocephalus and seizure. Based on this study, the presence of hydrocephalus and epilepsy are significant additive adverse prognostic factors. Approximately half of the patients with occipital encephaloceles will be severely debilitated and will probably be unable to live and function independently in society. These data may be useful to clinicians in counseling patients and predicting long-term outcome following repair of cranial vault encephaloceles.

(DOI: 10.3171/PED-07/07/022)

KEY WORDS • brain • cranial vault • encephalocele • pediatric neurosurgery

Cranial vault encephaloceles are relatively rare but challenging congenital neural tube defects presenting in the neonatal period with herniation of neural elements through a congenital skull defect. Historically, these entities have carried a poor prognosis, particularly the posterior/occipital encephaloceles.3 For example, in reports from more than 20 years ago, Lorber and Schofield4 found a 57% mortality rate for occipital encephaloceles, and Tsuchida et al.5 observed a 41% rate of mortality within 2 years. However, improved preoperative imaging, surgical techniques and postoperative neonatal care have decreased the morbidity and mortality rates of encephaloceles, particularly for the occipital type, resulting in increased long-term survival of these patients. In the series published by Docherty et al.,6 of 52 patients with occipital encephaloceles treated between 1971 and 1990, this group reported 12 deaths, but only one of these occurred in the last decade. With longer survival, it is uncertain how well these children do as they age. Overall, there is paucity of literature regarding the long-term outcome for these patients and this may perhaps result in underestimates of such deficits. The current study was conducted to observe the long-term outcome in patients with cranial vault encephaloceles to help improve prognostication for this patient population.

Clinical Material and Methods

We conducted a retrospective chart review of cranial vault encephaloceles seen at the Children’s Hospital, Birmingham, Alabama between 1989 and 2003. We did not include parietal cephaloceles in this study because they are not considered to be neural tube defects by many authorities and made up only a small minority of the total cases. Fifty-two total patients with cranial vault encephaloceles were identified and we were able to perform an in-depth review of 44 patients (68% with occipital and 32% with frontal lesions). The 52 patients represented all patients with encephaloceles seen and treated at Children’s Hospital during the noted time period. Care was not withheld from any patients.

Abbreviations used in this paper: HOQ = Hydrocephalus Outcome Questionnaire; SD = standard deviation.
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There were no known early pregnancy terminations due to the prenatal diagnosis of encephalocele and there were no deaths recorded within the first days of life. Six neurosurgeons with comparable treatment philosophies treated this patient cohort. Eight charts were either not available for detailed review or were incomplete, so we were unable to obtain information regarding their medium to long-term outcome data.

Additionally, 34 of the 44 patients (65% with occipital and 35% with frontal lesions) were contacted and the HOQ was used to assess their health status. This questionnaire is a validated outcome measure developed at Toronto’s Hospital for Sick Children and is used to assess the physical, emotional, cognitive, and overall health of patients. The questionnaire is composed of 51 items and is scored based on a parent’s or primary caregiver’s answer regarding how true or not true each statement is as it applies to their child’s health status over the last 4 weeks. Each answer is scored on a 5-point scale. Each section’s score is added and then divided by a simple quotient to provide a score that is a metric between 0.0 (representing worse health) to 1.0 (better health). The questionnaire takes approximately 10 to 15 minutes to complete. The HOQ has been validated and is reliable. The parent test–retest reliability, parent interrater reliability, and internal consistency have been established. The interpretability of the scores has also been reported.

Based on past studies, we used a score of 0.5 or less to represent severe impairment and/or disability. We used SPSS version 11.0 (SPSS, Inc.) and Excel’s Analyze It software for statistical analyses to 1) compare health status scores within each disease subgroup and 2) compare health status scores among disease subgroups. Assuming a normal distribution, parametric statistics were performed, including the paired two-sample t-test, the z-test, and the Fisher exact test, to compare discrete outcomes. The SD was also calculated to determine the amount of variation between variables. A probability value of less than 0.05 was considered to indicate statistical significance.

Results

The mean age for this cohort was 9.6 years (range 4–17 years) and the mean follow-up period was 9.2 years, with a minimum follow-up duration of 3 years. There was an equal sex distribution and there were no deaths in the 44 patients reviewed. There were three patients with postoperative cerebrospinal fluid leakage who were treated without infection or subsequent problems. Hydrocephalus was found in 60% of occipital and 14% of frontal encephaloceles, and epilepsy was confirmed in 17% of occipital and 7% of frontal encephaloceles (Table 1). Interestingly, we found concomitant cerebral arachnoid cysts in 11% of the cohort. There were no deaths at long-term follow up in any of the patients who were surveyed.

The results of the questionnaire identified the following aspects: 1) an overall score of 0.80 for frontal encephaloceles compared with a score of 0.65 for occipital encephaloceles (p = 0.037; Fig. 1); 2) 50% of patients with occipital encephaloceles who were surveyed had overall scores of 0.5 or less compared with 0% of patients who had frontal encephaloceles (Table 1), with the cognitive component having the most significant difference between the two groups (Figs. 2 and 3); 3) 55% of patients with occipital encephaloceles had cognitive scores of 0.3 or less, compared with 0% of patients with frontal encephaloceles (Table 1); and 4) the presence of hydrocephalus and/or seizure significantly lowered the scores of the cohort (Figs. 4 and 5). The scores of 0.5 and 0.3 were arbitrarily chosen as points of comparison to show the significant differences seen between the two groups (see Kulkarni et al. for a more in-depth interpretation of the scores). All patients with an overall score of 0.5 or less were severely disabled and extremely dependent on their caregivers.

Discussion

Our data correlate with previous suggestions that morbidity and mortality rates for cranial vault encephaloceles today are significantly lower than in older reports (prior to 1990). Docherty et al. reported that hydrocephalus developed in 57% of 51 patients with occipital encephaloceles. The rate of hydrocephalus reported in our study is similar to this and confirms that occipital encephaloceles carry a much higher rate of hydrocephalus than frontal encephaloceles. Tsuchida et al., Docherty et al., Date et al., Macfarlane et al., and Martinez-Lage et al. reported that 16 to 31% of their patients with encephaloceles were “physically and/or mentally disabled.” The majority of the assessments were done simply based on the physician’s follow-up examina-

![Fig. 1. Bar graph showing overall HOQ scores comparing frontal and occipital encephaloceles. The whiskers represent the SDs.](image-url)
tion or interpretation of follow-up questions to the patient’s family. There are no previous reports of outcome for cranial vault encephaloceles in which a validated outcome tool was used.

We found a significantly higher percentage of disability in our group. We found that occipital encephaloceles carry a worse prognosis than frontal encephaloceles and have higher rates of hydrocephalus and seizure. Contrary to the findings of Tsuchida et al.,9 we found that the presence of hydrocephalus does have significant adverse effects on patients with encephaloceles—lowering all scores in both frontal and occipital groups. Moreover, our results demonstrate that the presence of epilepsy and hydrocephalus are significant additive adverse prognostic factors for patients with encephaloceles. Regarding patients with occipital encephaloceles, approximately 50% of our patients were severely debilitated at their long-term follow-up evaluations, and would be unlikely to live and function independently in society. This is significantly higher than in previous reports. One factor that may account for this difference is that this is the first study in which cognitive and emotional impairments were assessed along with physical disabilities. When one examines the outcome scores closely, it is the significantly low scores in cognitive functions of many of these patients that make them dependent on their caregivers rather than just purely physical limitations.

Although we did not perform anatomical correlations (for example, the presence or amount of brain tissue within the encephalocele, the size of the defect, or the size of the cranium), Lorber and Schofield6 concluded that cerebral tissue in the encephalocele and associated microcephaly are bad prognostic features. Date et al.1 also found that the presence of gross brain tissue within the encephalocele and the size of the sac are unfavorable factors for prognosis.

The limitations of this study include the following: 1) the heterogeneity of medical/operative records; 2) availability of long-term outcome follow-up data in only 77% of our patients, and the possibility that all of the patients who were not contacted were more debilitated; and 3) lack of anatomical correlation to outcome. Additionally, six neurosurgeons treated this patient cohort. There might be some concern...
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about the use of an outcome questionnaire specifically designed for patients with hydrocephalus to assess general outcomes in a patient population with encephaloceles. Nevertheless, we believe that the HOQ is applicable to this patient population because many of our patients have hydrocephalus, and furthermore may be more sensitive to subtle deficits like cognitive impairment, whereas more general neurological outcome measures like the Functional Independence Measure for Children are much more physically weighted.

Conclusions

Occipital encephaloceles carry a worse prognosis than frontal encephaloceles, with higher rates of hydrocephalus and seizure. Based on our study, the presence of hydrocephalus and epilepsy are significant additive adverse prognostic factors. At least half of our patients with occipital encephaloceles were found to be severely debilitated, and we argue that they will probably be unable to live and function independently in society. These data may be useful to clinicians in counseling patients and predicting long-term outcome following repair of cranial vault encephaloceles.

References

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