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 sin- cipital 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.3,11,12

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.5,7,10,12 There are no comprehensive surveys on the morbidity and the quality of survival in individuals born with different types of encephaloceles. Lorber,11 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 ventriculoatrial 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 ventriculoatrial 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 encephalohaloceles, 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 33 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
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
The Prognosis of Encephaloceles

3½ months old, the lesion was no longer apparent. When she was 10 months old, skull x-rays were repeated and found normal. The patient has developed normally and at present is 12 years old, doing satisfactory school work in the sixth grade. The probability of this lesion having been an occipital cephalhematoma rather than an encephalocele must be strongly considered.

Results: Surgical Treatment of Occipital Encephalocele

Clinical Manifestations. Management and prognosis of patients with occipital encephaloceles differ significantly from those with encephaloceles in other locations, and prompted a separate analysis. Forty patients had occipital encephaloceles excised which ranged in size from 1 to 14 cm in their maximum diameters and averaged 6.4 cm in diameter. The size of the surgically treated lesions were in the same general range as the occipital encephaloceles observed in the unoperated patients.

Operations. Surgery was performed during the first week of life in approximately one third of the patients, by the end of the first month in two thirds, and before 3 months of age in over four fifths (88%). A few infants admitted with collapsed occipital sacs draining cerebrospinal fluid and several cases where rupture of large lesions seemed imminent were operated on as emergencies. Most of the infants, however, had occipital encephaloceles which were fully covered by reasonably healthy skin, and surgery was done electively.

General anesthesia was used for the operative procedure in the majority of patients. Some smaller lesions in young infants were excised under local anesthesia. Operation consisted of opening and exploring the sac of the encephalocele, excision of obviously malformed neural tissue, and tight closure of the basal dural defect. The redundant sac was then trimmed and the skin closed in layers. No attempt was made to fill in the bony defect.

Mortality. Of the 40 patients whose occipital encephaloceles were excised, 15 have died (37.5%). Three died in shock shortly after operation and one 2 weeks postoperatively from a wound breakdown and meningitis. The remaining 11 died between 3 months and 3 years of age; six of these deaths were related to hydrocephalus. Prior to 1958, three grossly retarded and spastic infants died of uncontrolled hydrocephalus, their occipital protrusions having contained gross brain tissue; third ventriculostomies were unsuccessful in two of these cases, and the progressive hydrocephalus in the third was never treated surgically. More recently, three patients who seemed neurologically intact following resection of occipital meningoceles eventually died of acute intracranial hypertension secondary to malfunction of ventriculoatrial shunts.

Five infants who eventually died did not have abnormal enlargement of the head. Four of these patients died between 3 and 11 months of age of pneumonia and inanition, and the fifth patient survived 3 years. Two remained microcephalic, and all showed gross psychomotor damage. Three of these cases had portions of the brain in the occipital hernia, but two of the lesions were cranial meningoceles.

The 25 living patients with surgically treated occipital encephaloceles ranged in age from 18 months to 19 years at the end of 1968. The average follow-up in these survivors has been over 9 years. While 14 are normally intelligent, 11 are disabled mentally and physically.

Disabled Patients. Three retarded patients are educable and enrolled in special school programs; they are able to care for their personal needs. All three are ataxic and one must use a walker; two are handicapped by dysarthric speech. One with a Klippel-Feil deformity frequently has mirror movements with the use of one or both arms, and the other two children have impaired ocular movements. None has had abnormal enlargement of the head; however, a ventriculoatrial shunt was performed in one case when an enlarged ventricular system was demonstrated by pneumoencephalography, but this procedure did not improve the child's mental state nor alter the existing neurological deficit.

Of eight profoundly retarded survivors, five have never walked or talked; three of these are severely spastic, two are blind, and two have recurring seizures. Two developed progressive hydrocephalus, which was never adequately controlled in one despite multiple
shunting procedures from the ventricles to the ureter, salpinx, and parotid duct. Although the hydrocephalus was relieved in early infancy in the second child by a ventriculoatrial shunt, the degree of psychomotor retardation remains extreme at 6 years of age. The three remaining children in the severely retarded category have developed some skills such as feeding themselves. They can walk with help or crawl and communicate on a verbal level. All are badly ataxic and have visual impairments. One is hydrocephalic and has been treated by a ventriculoatrial shunt.

**Competitive Patients (Normal Intelligence).** Of the 14 good recoveries, 13 had surgery for occipital encephalocele in infancy and one in her fifth year. All are considered “competitive” children, have normal intelligence, and four (10% of the operative series) are completely free of any physical defects. Nine of these children, ranging in age from 7 to 17 years, are making satisfactory-to-excellent progress in their school programs. Seven of them are in normal grade levels for their ages, and two are 1 year behind. Mental development in the five preschool children also seems within normal limits.

Ten children with normal intelligence had physical handicaps (Table 1). The stigmata most characteristically associated with occipital encephaloceles were neurological abnormalities, the Klippel-Feil syndrome and hydrocephalus, appearing in multiple combinations or as isolated conditions. These sequelae and other congenital anomalies in three of these cases have not precluded satisfactory growth and development to date.

Five of the six children with demonstrable neurological abnormalities have relatively minor deficits, including incoordination and impairment of ocular motility.

Five intelligent survivors were hydrocephalic with either abnormal enlargement of the head or an acute increase in intracranial pressure. In one case, this condition was very slowly progressive in infancy and early childhood and was apparently arrested spontaneously before puberty. He is 13 years old; his head size has been stable at 2 to 3 cm above the 98th percentile. Three cases had ventriculoatrial shunts in infancy after their occipital encephaloceles were excised and their heads enlarged at abnormal rates. One is obviously shunt-dependent, requiring several surgical revisions during childhood. In the other two cases, the hydrocephalus has been arrested after the shunting procedures and their heads are normal in size. One is now 10 years old and has not required surgical revision of the shunt system. In the other patient, currently 6 years old, who had a surgical revision for a symptomatic relapse in late infancy, the entire shunt assembly has been removed electively without recurrence of increased intracranial pressure over the past 3 years. The fifth competitive survivor with hydrocephalus was normocephalic and had enjoyed normal development for the first 5 years of life except for an untreated occipital encephalocele and associated Klippel-Feil syndrome. When this malformation was excised, however, the patient developed an acute hydrocephalus which required surgical shunting. This patient has since had a number of operative revisions and is considered shunt-dependent.

The six individuals with the Klippel-Feil syndrome show no specific neurologic findings apart from mirror movements of the arms in two cases. Other associated residuals included limitation in the natural movements of the neck and a decrease in physical stature. Several children have been restricted in their range of physical activities, and two in their mid-teens are well under 5 feet tall. Three children had congenital defects in other body areas.

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**Table 1**

<table>
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<tr>
<th>Case No.</th>
<th>Neurological Deficit</th>
<th>Klippel-Feil Deformity</th>
<th>Hydrocephalus</th>
<th>Other Congenital Defect</th>
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*Handicaps in 10 normally intelligent children after surgery for occipital encephalocele*
**Prognosis.** In patients with occipital encephaloceles managed between 1958 and 1967, recovery was superior to that achieved earlier (Table 2). Although fewer patients were admitted in the decade beginning in 1958, the frequency of surgical intervention increased from 66% to 86%. The incidence of satisfying results increased to 15 of 22 cases, compared with 10 of 32 patients managed between 1948 and 1957. The drop in unadjusted mortality associated with occipital encephaloceles from 68% in the first decade to 31% in the following decade reached statistical significance ($p < .05$).

Although the percentage of competent survivors increased from one decade to the next, no numerical or statistically meaningful difference was evident. None of the differences noted above could be correlated with any specific changes in our management of patients with occipital encephaloceles.

**The Time of Surgical Intervention.** For occipital encephalocele, the timing of operation had no clearly discernible influence on the ultimate clinical outcome (Fig. 2). Of the five infants operated on within 24 hours of birth, one premature infant did not survive the procedure. Three other infants also died as a result of operations performed at 1 week, 3 months, and 1 year of age; in each case the sac contained gross brain tissue. There has been no mortality resulting from operations for occipital encephalocele in the last 31 consecutive cases in this series since 1952. Six of the 14 “competitive” survivors were operated on during the first week of life, and three of them are completely normal. No statistically significant differences were found between those having early vs late operations.

![Figure 2](image-url)
TABLE 3

Contents of sac and surgical results in 40 patients with occipital encephaloceles

<table>
<thead>
<tr>
<th>Patient Category</th>
<th>Gross Brain Tissue</th>
<th>Total Cases</th>
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<tr>
<td></td>
<td>Present</td>
<td>Absent*</td>
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<tr>
<td>Competitive</td>
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<tr>
<td>Defective</td>
<td>8</td>
<td>3 (1)</td>
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<tr>
<td>Dead</td>
<td>9</td>
<td>6 (3)</td>
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<tr>
<td>Total</td>
<td>17</td>
<td>23 (8)</td>
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</table>

* Parentheses indicate glial tissue present.

**Size of Lesion.** Although lesion size was not a factor in the selection of patients with occipital encephaloceles for surgical treatment, there was an association of larger lesions with poorer results. The maximum average diameters of the lesions increased from 5.7 cm in the 14 competitive survivors, to 6.1 cm in the retarded children, to 7.3 cm in the patients who died. The validity of this trend cannot be definitely ascertained from these data, and the differences in sizes observed are not statistically significant (t-test). Although the maximum diameters ranged only from 1 to 7 cm (average, 4.3 cm) in the four children who are normal in every respect, the incidence of large lesions (diameter greater than 6 cm) in the competitive group was not significantly different from that found in the patients who died.

**Recognizable Cerebral Tissue.** The presence of cerebellum in the sac was the most important prognostic factor in this series of occipital encephaloceles (Table 3). This finding was not a statistically significant determinant of life or death. However, when brain tissue was encountered grossly, one could confidently predict that either the infant would not survive or that it would be mentally retarded with disabling neurological deficits (chi square, p < .001). Although none of the lesions in the competitive survivors contained frank brain tissue, no corresponding converse relationship was found. Even when the lesion proved to be an occipital meningocele, as in 23 of our cases, the chances of death or mental and physical incapacitation remained high (39%).

The presence of glial tissue *per se* , either histologically in the cyst lining or grossly as a meningogial nubbin at the cranial defect, was not of prognostic significance in our experience.

**Incidence of Hydrocephalus.** Of our patients with occipital encephaloceles who survived the first neonatal month, 36% had hydrocephalus. For our purposes, hydrocephalus was considered of clinical importance when a symptomatic increase in intracranial tension was manifested or when there was abnormal enlargement of the head. As defined, hydrocephalus developed in 1 of 6 patients whose lesions were never excised. Of 38 patients in the surgical group who survived longer than the first month of life, hydrocephalus developed in 15. This complication arose in 11 of 23 patients with occipital meningoceles and in four of 15 infants whose lesions contained gross brain tissue. Although the difference observed in the incidence of hydrocephalus in patients with surgically treated and untreated occipital herniations is non-significant statistically, in several infants and certainly in the child who was nearly 5 years old at operation, excision of the occipital lesion was associated with a precipitous onset of symptoms of increased intracranial pressure.

Hydrocephalus was an unfavorable prognostic factor in our cases, and eight of the 16 patients who developed this complication died as a result of it before they were 21 months old. Three of four hydrocephalic infants whose occipital lesions contained frank brain tissue have died. The morbidity from hydrocephalus was also striking among the 23 patients whose occipital sacs contained only cerebrospinal fluid and meninges, accounting for four of the six deaths in this group.

Survival was possible only if the hydrocephalus became arrested or was alleviated surgically. Two of seven infants with hydrocephalus observed prior to 1958 still survive, and one whose condition arrested spontaneously is competent. In nine cases, the hydrocephalus was treated by ventriculostrial shunting procedures; six of this group are living and four are competitive children. Thus, among 25 long-term survivors after surgical excision of occipital encephaloceles, hydrocephalus was a clinical feature in three of 11 defective children and in five of 14 others who are competent. Although the lowered mortality from hydrocephalus since 1958 is
The Prognosis of Encephaloceles

associated with the use of the ventriculocerebral shunt operation, the differences observed in our case material are not statistically significant. Furthermore, in those patients who lived with the condition arrested or controlled surgically, hydrocephalus had no significant effect on the quality of survival.

Results: Surgical Treatment of Other Encephaloceles

Eight encephaloceles in the following regions were repaired surgically: interparietal (2), nasal and frontal (5), and nasopharyngeal (1).

Interparietal Encephaloceles (Two Cases). Both lesions were considered meningoceles, although glial fibers were described histologically in one of the surgical specimens. One infant had separate meningoceles in the interparietal and occipital regions which were excised at 3 and 5 months of age respectively. Pneumoencephalography was essentially normal, except for a slight deformity of the fourth ventricle. This child, who is now 3 years old, shows normal mental and physical development, but is handicapped by concomitant congenital anomalies. The other patient who had a $3 \times 3$ cm interparietal lesion repaired 16 days after birth now shows normal psychomotor development at 5 years of age.

Nasal and Frontal Encephaloceles (Five Cases). Four of five infants with protruding anterior encephaloceles have survived and are normally intelligent. The patient who died had a large ($8 \times 10$ cm) unilateral cranial defect which contained much of the malformed cerebral hemisphere, including a dilated porencephalic lateral ventricle. Multiple anomalies of the extremities and a cleft palate were also present. The encephalocele was partially excised at 10 weeks of age. The patient had severe psychomotor retardation, frequent seizures, and died when 2 years old with an enlarged head and a persisting craniocerebral deformity.

Although two patients in this group survive as intelligent children, they are handicapped by facial deformities. In one, the right frontal region and orbit were involved. The lesion contained gliotic brain when a transfrontal repair was done at 1 month of age. The patient had four subsequent operations for cranial restoration and cosmetic improvement. However, now in his 16th year he has lost all vision in the involved eye and has a persisting facial deformity. He has not done well in school, having only reached the eighth grade. The other child was born with a grotesque rostral defect with marked hypertelorism, a cleft lip and palate, and bifid nose in addition to the obvious cranium bifidum (Fig. 3). A ventriculogram was normal, and a bifrontal craniotomy and dura-plasty when he was 4 years old ruled out the presence of anomalous brain tissue in the frontal protrusion but did not produce much cosmetic improvement. Apart from the craniofacial deformity, the patient's growth and development up to age 5 years have seemed normal. Further reconstructive facial surgery and cranioplasty are planned.

In two other infants, the anterior defects were localized nasal protrusions and posed fewer problems in surgical management. One patient when 2 years old had a trancranial excision of ectopic brain, repair of the dural defect, and amputation of the pedunculated external mass. Her subsequent development has been normal; her school record is excellent and at 12 years a mild degree of hypertelorism is the only residual abnormality. The other patient with a localized nasal encephalocele had a similar intracranial and extracranial operative procedure when 3 months old. However, the intradural closure of the cranial defect was imperfect, and subsequently the patient has had three plastic nasal operations for a bulge at the root of the nose due to a small persisting meningocele. Now 16 years old he is normal except for a splayed nose and concomitant hypertelorism.

Nasopharyngeal Encephalocele (One Case). This lesion presented as an obstructive mass which was biopsied transnasally at 2 weeks of age. No neural tissue was identified in the biopsy specimen, which showed a cyst compatible with the diagnosis of meningocele. Postoperative cerebrospinal fluid rhinorrhea was successfully treated by nasal packing and has never recurred. Although another more extensive transnasal excision of this lesion was performed subsequently, this otherwise normally-developing four-year-old child has a persisting nasopharyngeal mass which involves the left infratemporal fossa and is causing mechanical obstruction to the upper airway.
Discussion

The reported incidence of encephaloceles in the over-all problem of spina bifida and cranium bifidum ranges from 8% to 16%. The percentage of encephaloceles in the occipital area in our series is slightly higher than that reported previously. However, some of our larger occipital lesions also involved the parietal region so that classifications of this sort are fairly arbitrary. Occipital and parietal encephaloceles together accounted for 87% of the lesions in our series, which is comparable to the combined incidence of 89% of 187 cases reported by Ingraham and Matson. Nasofrontal encephaloceles made up 11% of their series and 12% of ours, with only one basal lesion involving the nasopharynx in each of these reviews. However, among 28 cases recently reported from Thailand, 25 (89%) of the encephaloceles involved the frontoethmoidal region.

Generally, the diagnosis of occipital encephalocele is obvious at birth. The best treatment for occipital encephaloceles in the majority of cases is surgical excision as soon as feasible in the neonatal period, provided the infant's progress appears satisfactory. The technical aspects of the occipital operation ordinarily are not formidable, and there should be little or no surgical morbidity. Our results since 1957 indicate that two-thirds of the infants born with occipital encephaloceles may survive after operation and roughly half of these will be normally intelligent children. These findings are more favorable than those reported recently by Lorber in 55 infants who were treated between 1959 and 1962 and followed 2 to 7 years. Fourteen of his cases died before, during, or soon after operation for occipital encephalocele, and 11 died later. Of the 30 survivors in his series, 10 were considered to be normal.

The prognosis for patients with surgically treated occipital encephaloceles depended largely on whether the lesion contained a portion of the brain. At operation 17 (42%) of our patients had this abnormality, and they either died or were defective. Preoperative ventriculography or angiography seems advantageous in the management of
infants with the larger lesions. In one recent case (Fig. 1 B), the dysplastic cerebellum and most of the brain stem had herniated into the occipital sac. In another patient, who died within several months, a comparable situation was indicated by open brachial angiography, and operation was never recommended. In other reports, brain tissue has been described in 25% to 80% of the lesions in surgically-treated infants; only occasionally was this type of occipital encephalocele compatible with useful survival.\textsuperscript{1,11} Manifest hydrocephalus had an adverse effect on the life expectancy of infants with occipital encephaloceles. This complication, however, was clearly secondary in importance to the nature of the malformation itself. Hydrocephalus developed in 36% of all the cases we surveyed who lived longer than 1 month, but at present in surgically treated patients the subsequent risk of hydrocephalus is probably nearer 50%. Nine of 19 patients in our series who were operated on after 1957 developed hydrocephalus and were treated by ventriculoatrial shunts, as were 22 of Lorber’s 41 cases.\textsuperscript{11} In Lorber’s series and in our cases of occipital meningocele, the incidence of hydrocephalus was virtually the same irrespective of the contents of the malformations. Although the use of ventriculoatrial shunts has simplified the operative treatment of hydrocephalus and probably helped reduce the over-all mortality associated with occipital encephaloceles, several deaths could be attributed to malfunction of the shunts themselves.

The majority of nasofrontal encephaloceles, in our experience, were isolated malformations and did not preclude normal mental and physical development. Occasionally, however, associated malformations can be extreme, including microcephaly and hydrocephalus (Fig. 4). Anterior encephaloceles may be more difficult to recognize when

![Fig. 4. A-B. Newborn microcephalic infant with a large, ulcerated frontal encephalocele. C-D. Same patient after one-stage bifrontal craniotomy, intracranial repair of dural defect, and extracranial excision of malformation. (This case is not included in this series ending December 31, 1967, since it is a more recent case.)](image-url)
they are occult and involve the nose or orbit. Intranasal gliomas and teratomas must also be considered in the differential diagnosis.\textsuperscript{2,3}

Nasofrontal encephaloceles should be treated by frontal craniotomy with intracranial repair of the dural defect and direct excision of any mass present, plus an attempt to repair the bony dehiscence at the same time. Operation may be indicated in early infancy in some cases to prevent infection (Fig. 4). In patients with complex frontal defects, where the risk of infection does not arise, a definitive intracranial operation may be deferred beyond infancy to facilitate simultaneous attempts at craniofacial reconstruction. However, multiple operative procedures may be anticipated in patients whose nasofrontal encephaloceles are associated with marked facial deformities. Preoperative air studies are worthwhile in the majority of cases. When anterior defects are extreme, a ventricular shunting operation in infancy may be preferable to an attempted direct repair of the lesion.

**Summary**

Encephaloceles were diagnosed in 65 patients over a 20-year interval through 1967. Of 18 unoperated cases, 15 have died. Fifty-five had occipital encephaloceles, which predominated in females in a ratio of nearly 3:1. In 15 patients, the lesions were not removed, and 13 of these have died. In 40 patients the occipital encephalocele was excised, in 88% before the age of 3 months, and 25 in this operated group have survived 18 months to 19 years. Four are normal in every respect. Ten others are normally intelligent children, but handicapped to some extent by residuals associated with the occipital encephalocele, including cerebellar ataxia, Klippel Feil syndrome, and hydrocephalus.

The most important adverse prognostic factor was the presence of brain tissue within the sac of the lesion. If this finding was noted grossly at operation, the patient either died or became defective mentally and physically.

Hydrocephalus was an important cause of death before and after ventriculoatrial shunts were used for its treatment, especially in patients with occipital meningoceles, but did not have a significant effect on the quality of survival of our patients.

Encephaloceles in interparietal, nasopharyngeal, and nasofrontal regions were excised in eight patients, and in six of these the surgical results are satisfactory. Nasofrontal protrusions were difficult to repair surgically, especially when associated with extensive facial deformities. Unlike the findings in patients with occipital encephaloceles, there were no characteristic neurological sequelae in patients with anterior malformations.

**References**