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A RETROSPECTIVE STUDY OF ENCEPHALOCELE MANAGEMENT: INSTITUTIONAL EXPERIENCE

Neurosurgery

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ABSTRACT

Goal: To provide an overview of encephalocele care and assess the outcomes in our institutions over a three-year period. **Methods:** A retrospective analysis was conducted on all newborns hospitalised and operated on for encephalocele at the Department of Neurosurgery of UPUMS Saifai hospitals between June 2020 and June 2023. **Results** This research included 55 babies with encephalocele ranging in age from one day to three years, with a mean age SD of 247±22 days. According to location and morphology, 6 types of encephaloceles were treated: 21 (38.1%) occipital, 11 (20%) atretic, 10 (18.18%) vault, 7 (12.7%) occipital-cervical, 4 (7.2%) ethmoidal, and 2 (3.6%) double encephaloceles. We divided encephaloceles into three sizes: small, medium, and giant. We discovered that 18.18% of newborns had hydrocephalus. Only seven deaths were documented, and five of these had nothing to do with encephalocele or its care. **Conclusions:** Encephalocele management entails thorough investigations and accurate diagnosis in order to devise the best surgical approach. A favourable outcome requires meticulous patient preparation, surgery, and thorough postoperative care and follow up. Associated hydrocephalus, which was not significantly associated to encephalocele type or size, as well as neurological and non-neurological diseases, are prevalent and should be considered.

KEYWORDS

INTRODUCTION:

Encephaloceles are congenital malformations characterised by brain herniation with or without meninges via a skull defect caused by lack of normal midline fusion of the cranial neural tube (1 & 2). The occipital area is home to 70% to 90% of all encephaloceles (3). This congenital lesion is a prevalent concern in paediatric neurosurgery across the world (4). It is considered that genetic and environmental variables have a role in the development of such congenital disease (5). Many patients have TORCH infections (toxoplasma, rubella, cytomegalovirus, herpes simplex virus) (6). Trauma, tumours, or iatrogenic damage can all be a cause of acquired encephaloceles, with traumatic birth rupture being the most serious (7). Encephaloceles in the occipital area, followed by the fronto-ethmoidal and parietal regions, should be corrected in the first few months of life because the intracranial content of the sac may be easily detected and full repair of the dural and skull defect can be performed. 3-dimensional computed tomography (CT) is favoured for visualising bone deformities (internal or exterior), while magnetic resonance imaging (MRI) is useful for differentiating herniated content and detecting other anomalies (7).

Purpose

The goal of this study is to look at the care of encephalocele and analyse the outcomes in our institutions over a three-year period.

Patients and Procedures: The research's design: this is a three-year retrospective study.

Study Design: From 2020 to 2023, patients admitted to and operated on in the Neurosurgery departments of UPUMS Saifai were studied. All medical records of babies hospitalised with encephalocele were retrieved and analysed for age, sex, encephalocele location and size, and concomitant neurological and systemic problems. Skull radiography, three-dimensional CT scans, and MRI brain scans were performed.

Inclusion Criteria

All patients with encephaloceles who received surgical correction throughout the research period were included.

Exclusion Criteria

The Babies excluded from the study were: all those with encephaloceles who did not undergo surgical repair; and those with encephaloceles but were not operated within the study period.

Surgical Details: Infants with large posterior encephaloceles were placed in prone position or lateral position, vault encephaloceles in supine or lateral position, and anterior skull base encephaloceles in supine position with 30 degrees flexion. We did an ellipse-shaped skin incision over sac followed by skin dissection then the sac was opened, cerebrospinal fluid (CSF) was slowly evacuated, the sac content was visualised and gently dissected and encountered, then repositioned inside through the dural opening under general anaesthesia with special care taken to the endotracheal tube, close monitoring to electrolytes and vital signs as well as the patient's body temperature to avoid hypothermia. The type of the herniated tissue visualised during surgery (gliotic or not) influences how we deal with the neural tissue; in most situations, we remove gliotic neural tissue. To avoid blood loss, step-by-step hemostasis was essential. The dural defect is revealed and closed with a reabsorbable suture; the skull periosteum surrounding the defect might be removed and utilised to heal the defect. Following the watertight closure of the dura, the hole was filled with an absorbable gelatin sponge, and the incision was closed with carefully approximated sutures to avoid postoperative CSF leaking. Finally, skin repair, hemostasis, and layer closure without drain were performed. To avoid infection, ruptured encephaloceles were treated as emergency cases within 24 hours.

Normal herniated frontal brain with intact meninges through basal bony defect was usually seen in anterior skull base encephalocele, so a modified suture incision was done followed by bilateral frontal bone craniotomy, then open the dura and retraction of frontal lobes with gentle retraction of healthy herniated part of frontal lobe after that extradural dissection of sac from defect followed by cauterization and suturing of redundant dura Except for one youngster who required postoperative critical care owing to unrelated surgical conditions, the patients were transferred to a standard paediatric ward or incubator. Additional procedures included ventriculoperitoneal (VP) shunt (for hydrocephalus associated with encephalocele). 10 procedures were done, 4 patients had VP shunt before encephalocele surgery, and 6 patients need VP shunt after it

Follow-up: All Patients were assessed postoperatively with clinical examination and CT brain with a follow-up period ranging from 12-24 months.

Ethical Approval: This research accepted by Institutional Ethics Committee (IEC) . All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. A written informed consent was taken from parents of infants after explaining all steps of this study.

Statistical Analysis: The program used for statistical analysis was SPSS version 20. Quantitative data were analyzed using mean, and standard deviation (SD), while frequency and percentage were used with qualitative data. Fischer exact test was used to compare frequencies. The corresponding distribution tables were consulted to get the "P" (probability value). Statistical significance was accepted at a P-value ≤ 0.05 while a P-value > 0.05 was considered insignificant.

RESULTS

55 infants with encephalocele were treated surgically in this study, 33 males (60%) and 22 females (40%). Their age ranged from 1 day to 3 years with a mean age of 247 ± 22 days. 6 types of encephalocele according to location and shape were treated in this study; we had 21 (38.1%) occipital, 11 (20%) atretic, 10 (18.18%) vault, 7 (12.7%) occipital-cervical, 4 (7.2%) ethmoidal, and 2 (3.6%)are double .

Table 1: comparison between rupture, hydrocephalus and death according to types of encephalocele

Type (No.)	Rupture (%)	Hydrocephalus (%)	Death (%)
Atretic (11)	0(0.0)	1(10%)	0(0.0)
Vault (10)	3(23%)	1(10%)	1(14.2%)
Occipital (21)	6(46.15%)	5(50%)	2(28.5%)
Occipto-cervical (7)	4(30%)	2(20%)	3(42.8%)
Double (2)	0(0.0)	1(10)	0(0.0)
Ethmoidal (4)	0(0.0)	0(0.0)	1(14.2%)
TOTAL 55	13 (23.6%)	10 (18.18%)	7(12.7%)
Statistical test FET= 2.34	P value 0.99		

At presentation and examination 13 (23.6%) infants had ruptured encephalocele; 6 occipital, 4 occipito-cervical, and 3 vault encephaloceles. We had 10 (18.18%) infants that were associated with hydrocephalus; 5 occipital, 2 occipito-cervical, 1 vaults, 1 atretic, and one double encephalocele case. During follow up we had 7deaths (12.7%) ; 3 deaths in occipito-cervical type, 2 in occipital and 1 in each of vault, and ethmoidal types with one case from hypoxia in early postoperative period one due to cerebritis of ruptured sac and three unrelated general causes in late period. There was no significant statistical relating incidence of rupture, association of hydrocephalus, or deaths to encephalocele types

Associated neurological and non-neurological conditions, Table 2, were in patients with encephalocele in this study. We had 7 infants (12.7%) who had meningomyeloceles, 4 (7.2%) had cord syrinx, 3 (5.4%) had Arnold Chiari Malformation, 23 (41.8 %) had non-surgical brain anomalies (as agenesis corpus callosum, dandy walker cyst, brain atrophy, shizencephally and others) and 15 (27.2%) had other congenital extra neural anomalies (Fallot's tetralogy, cardiac septal defect, syndactyly and others).

Table 2: distribution of the studied group according to associated conditions

Associated conditions	No. (%)
Arnold Chiari Malformation	3(5.4%)
Meningomyelocele	7(12.7%)
Syrinx	4 (7.2%)
Brain nonsurgical anomalies	23(41.8%)
Other congenital (extra neural) anomalies	15(27.2%)
Total	55 (100%)

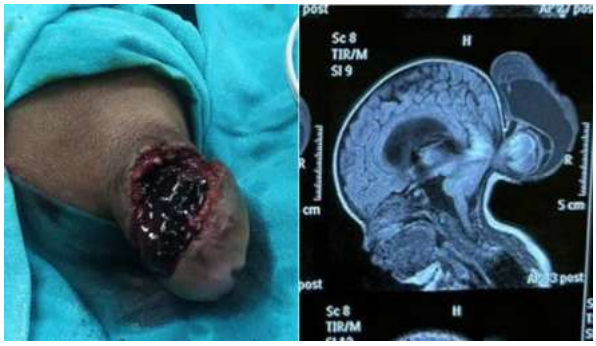
According to size, Table 3, we classified encephaloceles into 3 sizes; small with a volume 4 to 18 cm3, medium with a volume > 18 to 72 cm3 , and large of a volume > 72 to 520 cm3 . We had 18 (32.7%) small, 28 (50.9%) medium and 9(16.3%) large encephaloceles. Hydrocephalus was associated with 3 out 19 of small encephaloceles (16.6%), 5 out of 28 medium sized (17.8%), 2 out of 9 large sized (22.2%) which was statistically insignificant

Table 3: comparison between different size of encephalocele and occurrence of hydrocephalus

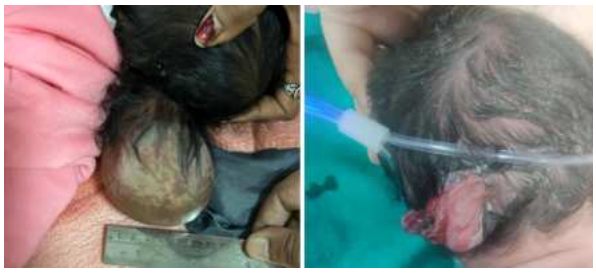
Size (Cm3)	No	Hydrocephalus (%)
Small (4-18)	18	3(16.6%)
Medium (>18-72)	28	5(17.8%)
Large (>72-520)	9	2(22.2%)
Total	55	10 (18.18%)
FET= 0.23	P value 0.89	

DISCUSSION:

In our study, 55 newborns with encephalocele were surgically treated, including 33 boys (60%) and 20 girls (40%). Many neural tube abnormalities have a female predominance, although this is especially noticeable in cases of encephalocele (4.5:1) (8). Female patients are more likely than male patients to have an occipital encephalocele (1.9:1). Females were found to be more impacted than men, which is consistent with recent results by Nath et al(10). In our experience, however, men predominate (60%), which is consistent with another research in the series by Shilpakar et al (11). Age varied from 1 day to 3 years in our research, with a mean age of 247± 22 days. It was previously noted in a prior series that the most common presenting age was 30 days, with only one kid presenting at the age of 6 years (12). Another study of 50 individuals revealed that the average age at presentation was 2.4 months (9). Ravindra et al. (13) found that the age of presentation ranged from a 1-day-old newborn to a 6-year-old child in their research of 19 individuals.



A) Encephalocele containing blood
B) MRI of patient with encephalocele



C) large occipital encephalocele
D) Ruptured Encephalocele

Figure 1 A) Encephalocele containing Haematoma B) MRI of patient with encephalocele C) large occipital encephalocele D) Ruptured Encephalocele

In terms of location and morphology of the lesion, we discovered 21 (38.1%) occipital, 11 (20%) atretic, 10 (18.18%) vault, 7 (12.7%) occipital-cervical, 4 (7.2%) ethmoidal, and 2 (3.6%). According to the Ravindra et al study, occipital encephaloceles accounted for 60%, occipital-cervical encephaloceles 20%, parietal 10%, and 5% had a double encephalocele (one atretic and the other occipital). Three individuals had ruptured encephaloceles at the time of presentation in the study, whereas we found ruptured encephaloceles in 13 newborns. In our study, 12.7% of patients with encephalocele had meningomyeloceles, 7.2% had cord syrinx, 5.4% had Arnold Chiari Malformation, 41.8% had non-surgical brain anomalies (as agenesis corpus callosum, dandy walker cyst, brain atrophy, shizencephally, and others), and 27.2% had other congenital extra neural anomalies. Encephaloceles are frequently accompanied with intracranial abnormalities. The association of some neurological conditions with occipital encephaloceles has been documented to have higher probabilities of posterior fossa malformations such as arachnoid cyst, agyria-pachygyria complex, Dandy-Walker malformations, and callosal anomalies (14). Other conditions associated with Chiari malformation, meningomyeloceles, and syrinx (13)

We divided encephaloceles into three types based on their volume: small (4 to 18 cm3), medium (18 to 72 cm3), and giant (72 to 520 cm3). The size of the sac was estimated to be between 1.5 cm ×2 cm and 25 cm× 12 cm (9). Hydrocephalus was found in 16.6% of small encephaloceles, 17.8% of medium-sized encephaloceles, and 22.2% of large-sized encephaloceles, which was statistically insignificant. We discovered no relationship between sac size and hydrocephalus. In research conducted by Rehman et al (9), it was shown that (32%) of patients had increased head circumference and hydrocephalus. According to a 2007 study by Bui et al (15), hydrocephalus is frequent in 40% to 60% of occipital encephaloceles patients. Postoperative hydrocephalus should be treated with ventriculoperitoneal (VP) shunts

performed in one or two stages (16). Rehman et al (9) reported that 4 (8%) of the patients developed hydrocephalus following sac repair, which was addressed with the implantation of a ventriculoperitoneal shunt. One patient (2%) did not recover from anesthesia. Bui et al (17) previously observed that occipital encephalocele is more usually linked with hydrocephalus than other kinds of encephalocele. Hydrocephalus is a significant predictor of developmental delays in encephalocele patients, and it can occur after surgery in rare situations (18). In these cases, a VP shunt should be implanted before total encephalocele repair (19). In our investigation, VP shunting was performed on ten infants: four before encephalocele surgery and six after surgery. One of the patients developed an encephalocele sac hematoma (Figure 1A). During our study's follow-up period, we experienced 7 fatalities (12.7%); one from hypoxia in the early post-operative time, one from cerebritis of ruptured sac, and five from unrelated general reasons in the late period. Lorber et al found 25 fatalities (14.9%) in a series of 167 individuals. 14 people died before, during, or shortly after the procedure, and 11 people died later.

The presence or absence of brain tissue in the ruptured sac was shown to be the single most relevant prognostic factor in 28 of the survivors (12). big encephaloceles are linked with a high rate of CSF leaking as a result of the big defect, which may predispose to infection (20). Many factors have been mentioned that can affect the disease's prognosis, such as 1) location: location of the sac with its content, which is mainly vermis and rarely cerebellum to varying degrees; however, occipital lobes do herniate on occasion, 2) size of sac and amount of brain tissue, 3) hydrocephalus, 4) associated malformation, and 5) associated infections (21). Early surgery for encephalocele is indicated to prevent sac rupture, and decrease CSF leak, and meningitis, with specific attention during surgery on avoiding hypothermia, minimizing blood loss, and monitoring electrolytes (22).

Giant encephaloceles are uncommon, and undergoing surgery is a difficult undertaking for both neurosurgeons and anesthesiologists. The main reasons for the difficulties are the site's complexity, size, and related bulging contents, which can cause intracranial malformations, intraoperative blood loss, and extended anesthesia (23) Handling airway skills, appropriate intraoperative care, and a search for further congenital anomalies are all necessary in managing such a patient. For a satisfactory outcome, meticulous preparation and perioperative management are necessary.

For these patients, endotracheal intubation is a serious problem. The infant cannot be placed supine and requires lateral positioning for intubation in order to prevent any compression and sac rupture.

It was challenging to place the patient supine for endotracheal intubation because of the extent of the encephalocele. We lifted the torso with the use of a towel drape so that the head ring could easily and without compression accept the encephalocele. Various intubation techniques are employed by other authors, such as the lateral position [24], the child's head being placed beyond the table [25], the infant being lifted on the table [26], and the preoperative aspiration of encephalocele prior to anesthesia in order to obtain correct posture [27].

Worldwide, there are a few standard procedures that are often followed for managing wound dehiscence. Obtaining a wound swab culture should come first, then debridement of non-viable tissue and a lengthy (four to six week) course of systemic broad-spectrum antibiotic treatment [28]. However, long-term usage of broad-spectrum antibiotics has been linked to a risk of acquiring antimicrobial resistance, necrotizing enterocolitis, late-onset sepsis, and mortality [29].

Other treatments for this complication have already been suggested, such as the use of negative pressure wound therapy, which includes vacuum-assisted closure (VAC) devices. These devices help to reduce the need for painful dressing changes, healing times, and an earlier return for the patients to their regular activities by cleaning the wound margins, bringing them together, and encouraging the formation of granulation tissue [28].

Limitations

Our study's sample size and short follow-up time may be limitations that should be addressed before reaching any firm conclusions.

CONCLUSION:

Encephalocele management entails thorough investigations and accurate diagnosis in order to devise the best surgical strategy. For a successful outcome, meticulous patient preparation, surgery, and postoperative care and follow-up are required. Associated hydrocephalus, which was unrelated to encephalocele type or size, as well as neurological and non-neurological problems, are prevalent and should be considered. Proper counselling and education of the parents about the condition (encephaloceles) and its consequences is recommended, as is a long-term follow-up. Folic acid supplementation, which none of the patients in our research had, should be provided before conception to decrease neural tube abnormalities. Counselling should be extensively emphasized in health centers. Encephalocele repair should preferably be done postnatally to allow for acceptable nursing and to reduce the risk of ulceration and stress to the lesion with eventual meningitis. Early healing also allows the face skeleton to develop and reorganize, improving cosmesis.

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Conflict Of Interest: None declared
Ethical Approval: The study was approved by the Institutional Ethics Committee

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