

Therapeutic Outcomes of Encephaloceles at The Bangui Paediatric University Hospital: A Review of 20 Cases

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How to cite this paper: Ouai^{mon}, D.S., Gbelesso, J.S., Sopia, J.I.J., Gbanade-Packo, N., Ngombo-Banguele, C.O., Mboko-Yandombo, J.J., Mbaikoua, T.N.P., Ouaimon, O.D. and Ndoma Ngatchoukpo, V. (2026) Therapeutic Outcomes of Encephaloceles at The Bangui Paediatric University Hospital: A Review of 20 Cases. *Open Journal of Pediatrics*, 16, 425-430.

<https://doi.org/10.4236/ojped.2026.163042>

Received: February 27, 2026

Accepted: April 20, 2026

Published: April 23, 2026

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Abstract

Encephalocele is a rare congenital anomaly characterized by a malformation of the central nervous system consisting of a herniation of the brain through the skull. We report the therapeutic results of 20 cases of encephaloceles in our study at the paediatric surgery department in Bangui. This was a prospective study conducted over a period of five years from January 2020 to December 2025. Males predominated with a sex ratio of 1.5. The average age was 2 months. The occipital location was the most common (70%, n = 14), followed by the frontal location (25%, n = 5), and finally the nasoethmoidal location (5%, n = 1). The majority of parents had a low socioeconomic status (90%, n = 18) and came from the suburbs. Six children (30%) were born to consanguineous parents, and associated malformations were diagnosed at birth. All cases underwent surgical treatment with resection of the non-functional dysgenetic brain and closure of bone defects. The outcome was favorable in most cases (two cases of post-operative hydrocephalus). Encephaloceles are linked to an abnormality in the closure of the neural tube. They usually manifest very early at birth with very suggestive clinical symptoms. Surgical treatment usually results in a permanent cure. Despite all the progress made, the mortality and morbidity rates remain high and the psychomotor prognosis is compromised.

Keywords

Encephalocele, Child, Treatment, Bangui

1. Introduction

Encephaloceles are congenital anomalies that fall within the general category of

neural tube closure defects. They are defined by the protrusion of brain tissue and/or meninges outside the skull through a defect in the skull bone. The overall incidence of encephaloceles is 0.8 to 3 per 10,000 live births [1]. Diagnosis is clinical, with computed tomography (CT) confirming the diagnosis. Treatment is always surgical.

2. Materials and Methods

This is a prospective study of cases of encephalocele hospitalised in the surgery department of the Bangui Paediatric University Hospital Centre (CHUPB) over a period of five years, from January 2020 to December 2025. All patients admitted for encephaloceles were included in the study. Those with incomplete records were excluded. Patient data was collected from patient records and operating theatre registers in the paediatric surgery department. The parameters analysed were age at the time of hospitalisation, gender, consanguinity, reason for consultation, family history, location of lesions and associated malformations. Brain scans and transfontanellar ultrasounds were performed on two patients with large encephaloceles exceeding 32 cm in circumference and whose parents had a high socio-economic status. All patients underwent surgery and were followed up one week, one month and three months after discharge. Epi Info 7.2 software was used for data processing and analysis.

3. Results

During the study period, we identified 20 cases of patients who had undergone surgery for encephalocele, representing an incidence of 0.4%. All patients were admitted during the first few months of life. Males predominated, accounting for 70% (n = 14); most patients came from remote areas (70%), whilst 30% of cases were recruited from the capital. The majority of mothers, 80% (n = 16), were from a low socio-economic background; 85% (n = 17) of pregnancies were poorly monitored, and only 15% (n = 3) of mothers received folic acid supplementation. Occipital location was the most common (70%, n = 14), followed by frontal (25%, n = 5) (see **Figure 1**) and ethmoidal (5%, n = 1) (see **Figure 2**). The average circumference was 36 cm (range 10 - 50 cm). It was voluminous, with a circumference exceeding 32 cm in two patients (10%); a fistula was observed in two cases (10%) and an infection in three cases (15%). The content of the malformation was mixed in 90% (n = 18) (brain + cerebrospinal fluid). Hydrocephalus was associated in 25% (n = 5); other associated malformations included microcephaly in 10% of cases (n = 2), Dandy-Walker syndrome in 10% of cases (n = 2), and umbilical hernia in 40% of cases (n = 8). Transfontanellar ultrasound was performed, as was a brain CT scan, in two patients whose parents had a high socio-economic status and had the means to arrange for the requested investigations. All patients underwent surgery within an average of 7 days; the procedure involved making an incision around the encephalocele and dissecting the adhesions between the skin and the dura mater. All dysplastic tissue was resected, followed by repositioning of the

healthy tissue. A watertight closure of the dura mater was achieved using a suture technique, whilst the subcutaneous layer and skin were closed with simple sutures. A ventriculo-peritoneal shunt was performed in 5 patients (25%). The overall mortality rate was 30% (n = 6). These deaths occurred mainly as a result of post-operative sepsis in four patients and in two patients one week after ventriculo-peritoneal shunting. At 1- and 3-month follow-up, six of our patients presented with lower limb muscle hypotonia.

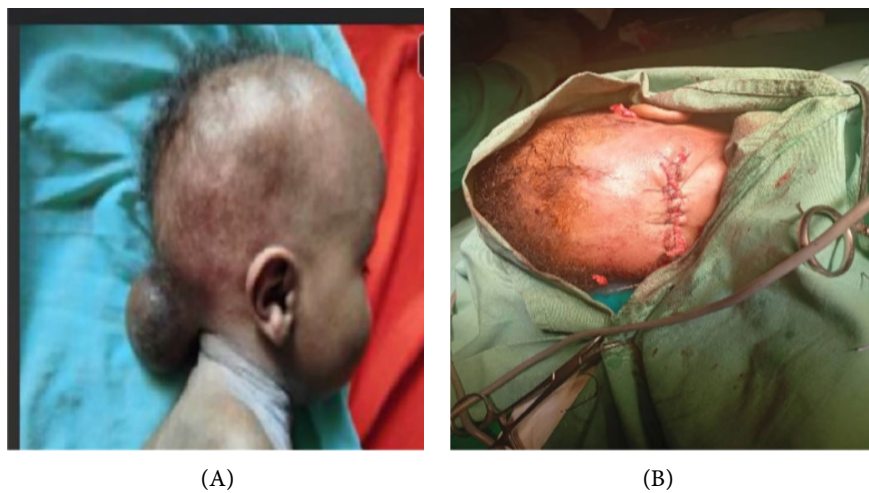


Figure 1. (A) Occipital encephalocele seen in profile [CHUPB]; (B) Post-operative appearance [CHUPB].

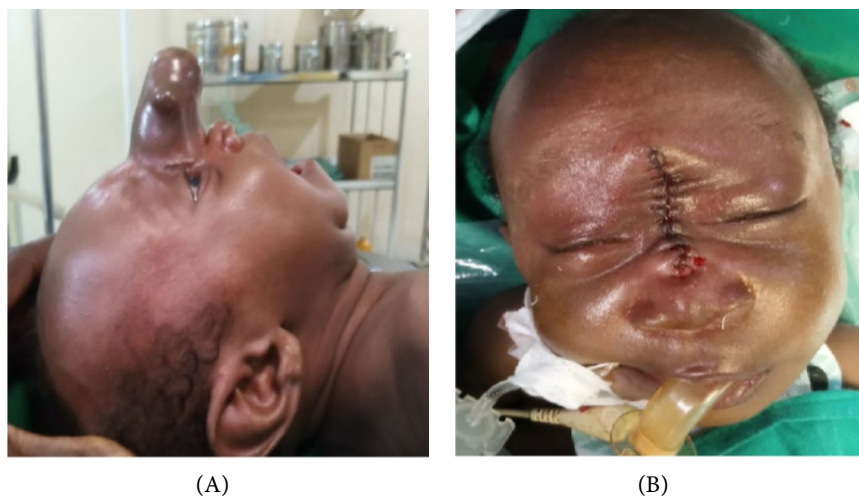


Figure 2. (A) Ethmoidal encephalocele seen in profile [CHUPB]; (B) Post-operative appearance [CHUPB].

4. Discussion

The pathogenesis of encephaloceles is thought to involve a failure of the neural tube to close properly. Encephaloceles, spina bifida and anencephaly are commonly grouped under the term “Congenital Neural Tube Defects” (CNTD) [1]. Their aetiologies are heterogeneous, involving environmental and genetic factors,

and remain controversial. Several theories have been put forward, such as maternal hyperthermia, valproic acid, hypervitaminosis, and vitamin B12 and folic acid deficiency [2] [3]. The incidence of encephaloceles varies according to geographical area and socio-economic status [4] [5]. In the USA and Western Europe, the incidence of encephaloceles is 1 to 3 cases per 10,000 live births [5], whereas other studies conducted in Asia, the Middle East and Africa have reported a prevalence of 23 to 61 cases per 10,000 live births [5] [6]. In our study, we found an annual incidence of 4 cases per year, compared with the study by Ndoma *et al.* [6], which reported 7 cases per year; this could be explained by improved antenatal care and the improved socio-economic status of the parents. The diagnosis was made post-natally in all cases in our study. The occipital form predominated (70%). These findings are consistent with the data published by Nagata, Bhandari, and Radouani [7]-[9]. The majority of studies reported a much higher incidence of occipital forms [9]-[12]. Anterior localisations (frontal, orbital, nasal and ethmoid regions) are less common [13]-[15]. The association of encephaloceles with other malformations such as hydrocephalus, microcephaly, psychomotor retardation, and Dandy-Walker syndrome is common [14] [15]. Antenatal ultrasound performed by trained practitioners is the examination of choice for the antenatal screening of cerebral malformations. It enables the detection of the cranial defect, sometimes accompanied by a brain herniation [15]. Preventive measures such as taking folic acid during the periconceptual period and antenatal diagnosis via imaging (ultrasound or MRI) are factors that help to reduce the prevalence of ACTN [15] [16]. The surgical approach varies, and a common pitfall is insufficient closure of the dura mater, leading to post-operative CSF leakage or the formation of a pseudomeningocele [16]. Early surgical treatment, before significant craniofacial deformities develop, yields satisfactory cosmetic results [13] [14] [16]. We recorded an overall mortality rate of 30%. This rate is similar to that reported by Radiouni *et al.* [3], who reported a mortality rate of 25%. Mortality from encephaloceles is generally very high, particularly in sub-Saharan Africa, where the rate varies from 10 to 33% [17]. The prognosis for children with encephalocele is linked to the timeliness of surgical management, the severity of associated anomalies, the volume of the encephalocele and herniated neural tissue, and the parents' low socio-economic status [17].

5. Conclusion

Encephalocele is a rare but serious congenital condition due to its psychomotor implications. This condition leads to morbidity and mortality in our context; its management requires an upgrade of our technical facilities, raising public awareness to encourage prompt consultation, but above all, strengthening prevention strategies through folic acid supplementation during the periconceptual period.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper

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