



## ENCEPHALOCELE: RETROSPECTIVE ANALYSIS AND OUR CLINICAL EXPERIENCE

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**Abstract:** Encephalocele is defined as extracranial herniation of the CSF, meninges, or cerebral tissue through a midline fusion defect in the cranium. The aim of this article is to present the clinical experience of the authors on encephalocele management. A total of 19 patients who underwent surgery for encephalocele in our hospital between 2015 and 2021 were included in the study. We reached 7 cases who were diagnosed with encephalocele and underwent pregnancy termination between 2018 and 2020 in our hospital. The patients' demographics, neurological examinations, procedure and anaesthesia data, and postoperative follow-up were all evaluated. 15 of 19 patients were female. 2 mothers used folic acid supplementation, but it was not effective. 7 patients were diagnosed prenatally, whereas the others were not followed up during pregnancy. 9 of the patients had parenchyma inside the sac, while the rest had none. 5 patients required shunts. All of the patients were followed up by the departments of neurosurgery, pediatrics, pediatric neurology, neonatal, pediatric gastroenterology, and genetics for their needs. It was demonstrated that folic acid supplementation before conception greatly reduces the incidence of encephalocele. It would be appropriate to inform the families of babies diagnosed with encephalocele in detail at prenatal follow-up about what problems they can expect in the future. Follow-up of encephalocele patients must be done with a multidisciplinary approach to ensure a quality life throughout their life.

**Keywords:** Encephalocele, Hydrocephalus, Seizure, Sac content, Anesthesia

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### 1. Introduction

Encephalocele occurs as a result of a midline fusion defect in the skull bone (Greenberg, 2019). This defect allows the herniation of the cerebral tissue or meninges (Ghatan, 2011). It has a prevalence of 0.8-4 per 1000 births (Ugras et al, 2016). Encephalocele occurs as a result of defective neural tube closure or a defect in post-neurulation (Rolo et al, 2019). It has also been reported that folic acid intake significantly reduces the incidence of all neural tube defects, including encephalocele (Copp et al., 2013). It is frequently located in the midline and occipital region and is accompanied by other malformations and chromosomal anomalies (Chen et al., 2000; Stoll et al., 2007). The presence of parenchyma within the sac, the development of hydrocephalus, and seizures often indicate a poor prognosis, mostly occurring in occipital encephaloceles (Bui et al., 2007). The purpose of encephalocele surgery is to place the herniated functional structures back into the calvarial component (Drake and MacFarlane, 2001). Congenital anomalies that occur with an encephalocele, the size of the sac diameter, hydrocephalus, and hemodynamic disturbances that may develop during surgery complicate anaesthetic management. These patients may have perioperative complications such as electrolyte imbalance (due to CSF drainage), hypothermia, and blood

loss (Mahajan et al., 2011; Singh et al., 2012). Hemodynamic changes caused by blood loss can necessitate a blood transfusion (Mahajan et al., 2011). In addition, during the opening of the encephalocele, rapid drainage of cerebrospinal fluid may lead to hemodynamic disturbances that may cause bradycardia or even cardiac arrest (Mahajan et al., 2011; Singh et al., 2012). Therefore, these patients should be followed up with a multidisciplinary approach.

### 2. Materials and Methods

Encephalocele patients operated between 2015-2021 (2011-KAEK-25 with 2020/12-12 clinical research and ethics committee decision number) were scanned retrospectively. Since the data of 2 of 21 patients who were operated on were not available, they were not included in the study. 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.

In our hospital, 7 cases underwent pregnancy termination upon the diagnosis of encephalocele between 2018 and 2020. We utilized patient files, anaesthesia forms, outpatient follow-up observations,



radiological examinations, and we contacted any families we could contact for the study. We scanned patients' surgical data, length of hospital stay, and other findings that may affect their quality of life. The data we screened were as follows: birth weight, birth head circumference, mother's gravity-parity, gestational week of delivery, prenatal folic acid supplementation, time of prenatal diagnosis, prenatal medication use, type of delivery, infant's APGAR score, microcephaly/ macrocephaly of development, neurological examinations, follow-up period, size of the sac, presence of parenchyma in the sac content, shunt requirement, history of seizure, presence of nutritional problems and growth retardation, operation time, perioperative and postoperative surgery and anaesthesia complications, preoperative and postoperative electrolyte and hemoglobin values, presence of intraoperative blood transfusion, length of ICU and hospital stay, duration of intubation and discharge-ex status. Verbal consent was obtained from all families who could be reached.

### 2.1. Statistical Analysis

Within the scope of the study, 19 patients were compared with 33 different clinical parameters. All analyzes were performed using IBM Statistics SPSS 25 and GraphPad Prizm 8 programs. The quantitative data obtained as a result of the measurements were first subjected to the normality test. Parametric One-Way ANOVA test was applied to the data found to be suitable for normal distribution. The Kruskal-Wallis test was used when comparing clinical data that did not show normal distribution. Chi-square test was used when comparing categorical variables among themselves. In the light of the findings obtained, Spearman r correlation analysis was performed in the analysis of the relationship between clinical data with a statistically significant difference. In the analyses, 95% confidence interval and statistical significance were accepted as  $P < 0.05$  (Önder, 2018).

### 3. Results

The patient data are given in Table 1. Birth weights varied between 2575 and 4000 grams, with no low birth weight infants. The patients' head circumference at birth was between 25 and 36 cm.

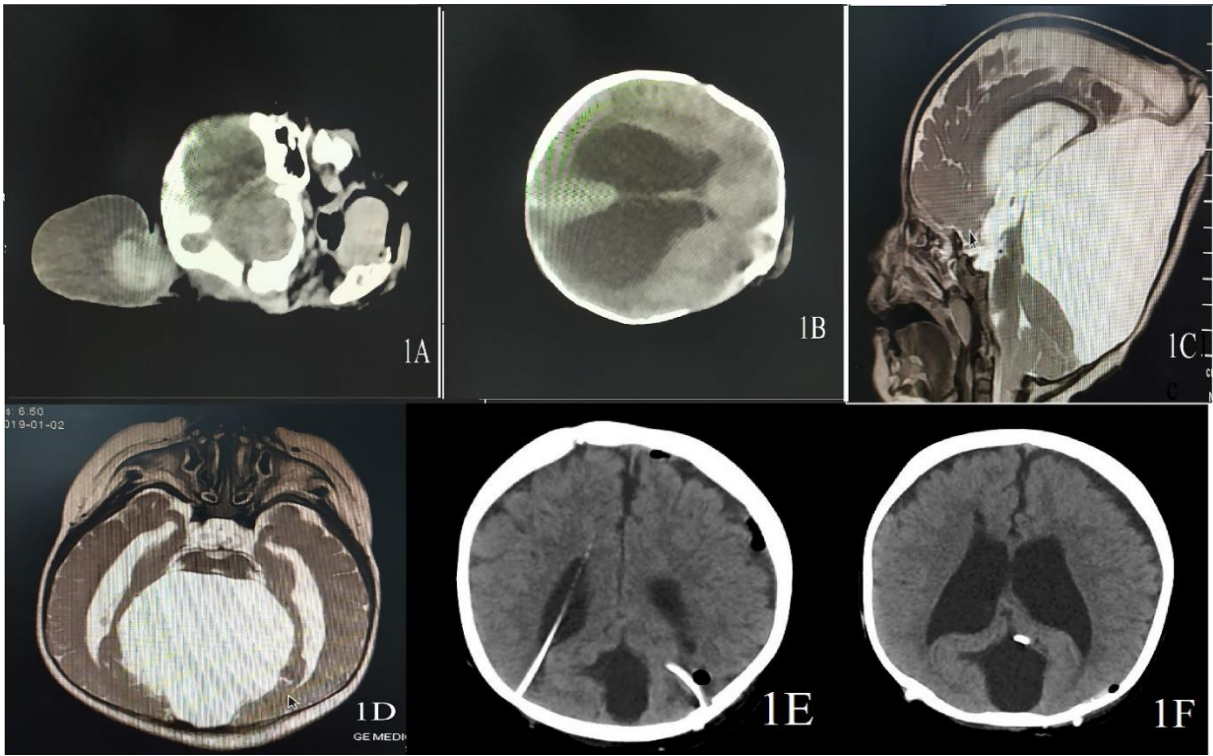
The patients were born at 36 and 39 weeks of gestation. The APGAR scores were reported as 9-10 in 15 patients, 8-9 in 1 patient, and 4-8 in 3 patients. The length of a patients' stay in the newborn intensive care unit is influenced by their Apgar scores at birth. A patient with an Apgar score of 4-8 required 14 days of newborn intensive care unit follow-up. Neurological examinations of 16 of 19 patients were normal, the first of the remaining 3 patients had genu recurvatum deformity, the second was hypoactive and had a dysmorphic facial appearance, with a broad nasal bridge and proptosis-like appearance in the eyes on physical examination, whereas the third patient was hypoactive and had accessory fingers in both hands (Figure 1). One of the patients had

an omphalocele accompanying encephalocele (Figure 2 and Figure 3). 3 patients were found to have microcephaly during follow-up, while the others' head circumferences were within the normal percentile (Figure 4). The size of the sac also varied from 1x1.5 cm to 10x10 cm. Parenchyma was observed within the sac in 9 patients, and only CSF content was present in the sac of 10 patients.

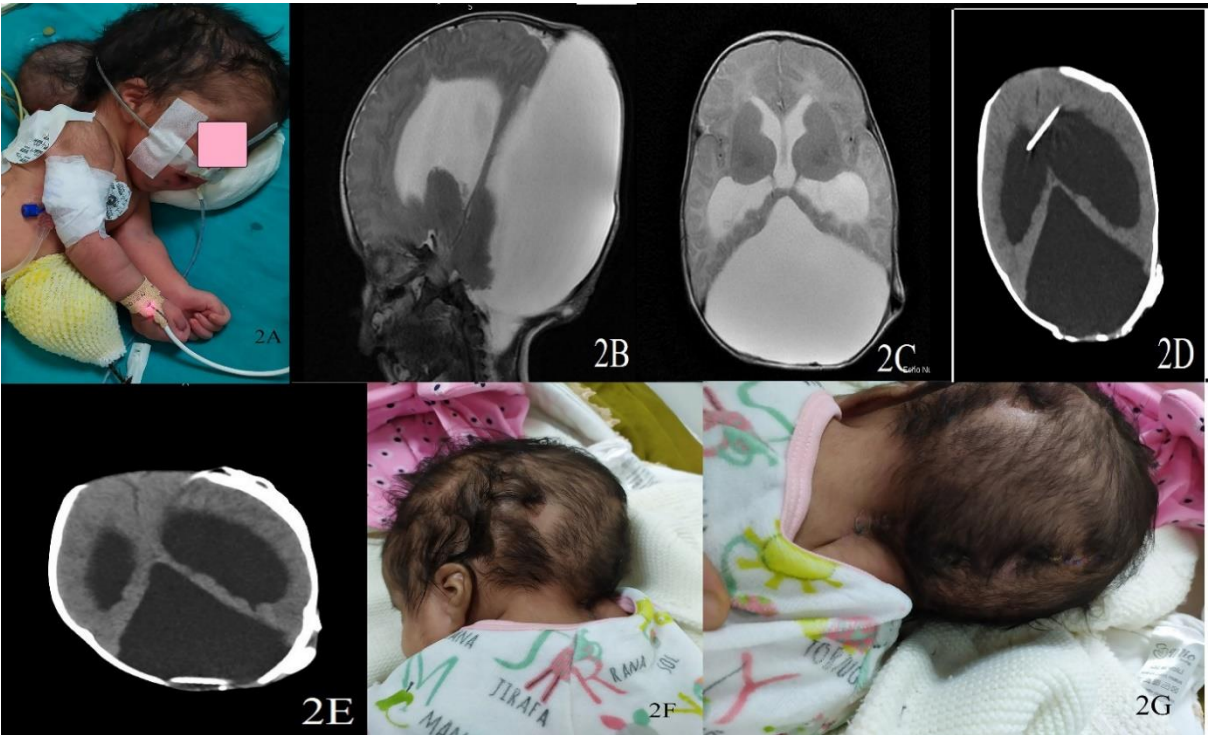
**Table 1.** Descriptive statistics of patients

Gender n (%)	
Female	15 (78.9)
Male	4 (21.1)
Weight (gr); med (min-max)	3128.68 (2575-4000)
Gestational age, week; med (min-max)	38 (36-39)
Apgar 1.minute n/%) $\leq 7$	3 (15.8 %)
Apgar 5.minutes n/%) $\leq 7$	0
Family story n (%)	
No	19 (100)
Head circumference, cm; med (min-max)	34.23 (25-36)
Paranchyma tissue in the sac n (%)	
No	10 (52.6)
Yes	9 (47.4)
Microcephaly vs macrocephaly n (%)	
No	16 (84.2)
Microcephaly	3 (15.8)
Additional anomalies n (%)	
Cardiac	1(33.3)
2 handheld accessory fingers	1 (33.3)
Omphalocele	1 (33.3)

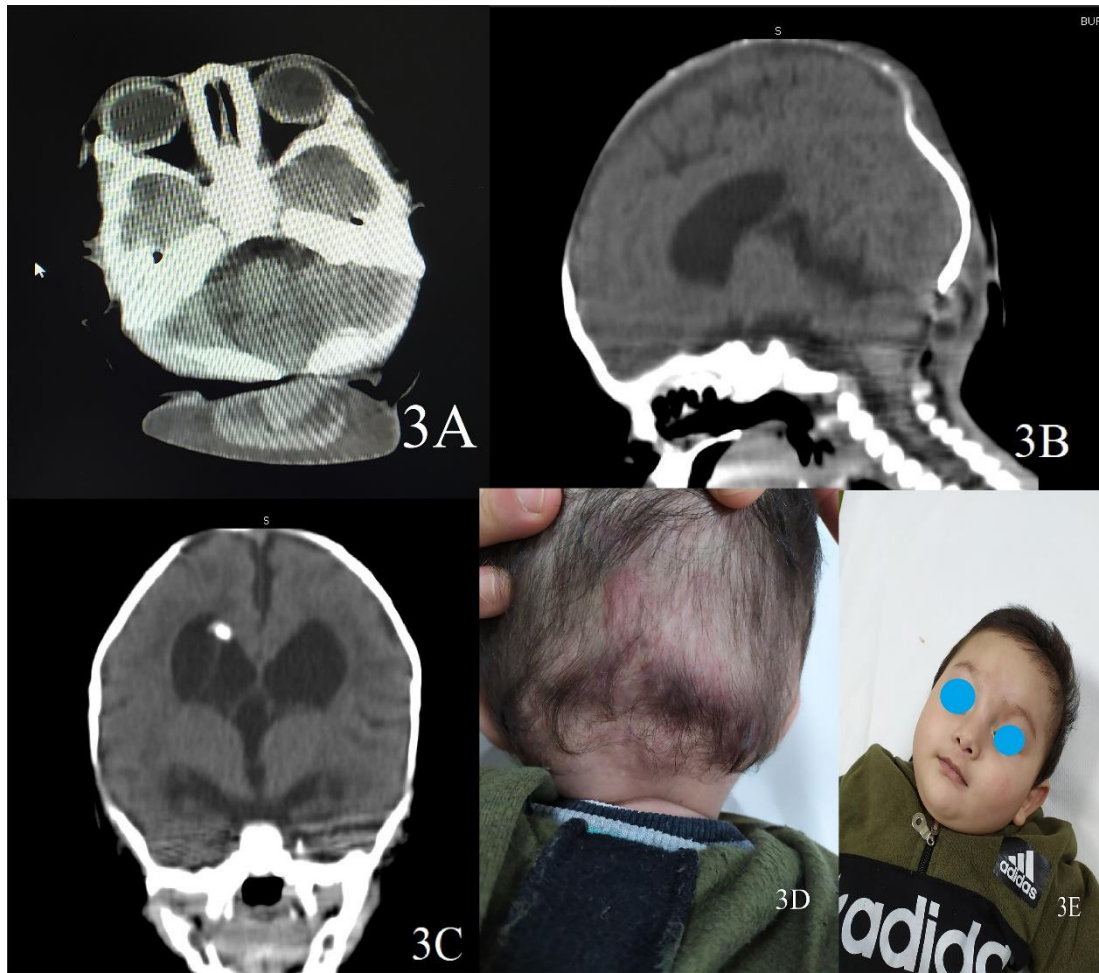
The data of the mothers are given in Table 2. 15 of 19 patients were female and 4 were male. The mean age of the mother was 27 (Range of age group: 21 to 45 years). Two of the mothers had preeclampsia and 1 had DM-HT. No history of prenatal medication use was reported among the mothers. Of 19, 16 mothers never used folic acid. 12 mothers had no pregnancy follow-up. 2 mothers used folic acid supplementation, but the first one started to use it after the 10th week of pregnancy, while the second mother used folic acid in the first 3 months of pregnancy and then stopped. The first was diagnosed at 12 weeks, the second at 16 weeks, the third at 8 months, the fourth at 37 weeks, and the fifth at 12 weeks, according to the statements of five mothers who were followed up by an obstetrician (2 mothers were diagnosed at which week could not be reached). Termination was recommended to those who did not exceed the termination period, but they did not accept it. One patient who was diagnosed at 12 weeks underwent a karyotype analysis and was reported as normal. No diagnosis of midline defect anomaly was present in the family history of any patient.



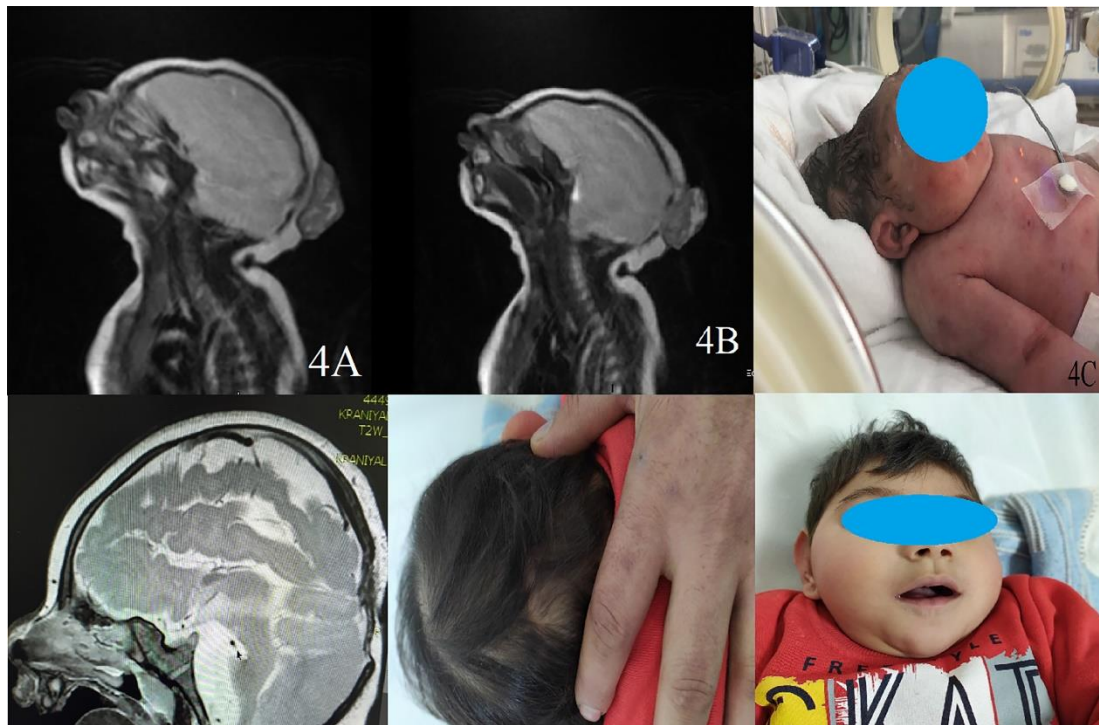
**Figure 1.** 1A: Preoperative cranial CT 1B: Postoperative hydrocephalus developed. 1C-1D: There was a large cystic appearance in the suprasellar area. 1E-1F: VP shunt was inserted in the right occipital and supracerebellar region.



**Figure 2.** 2A: Preoperative view of an encephalocele case with omphalocele association. 2B-2C: Postoperative MR image 2D-E: Control cranial CT after VP shunt from the left frontal and then a VP shunt from the left occipital to the posterior fossa. 2F-G: 5-month outpatient control view.



**Figure 3.** 3A: Preoperative cranial CT view 3B: Postoperative cranial CT image 3C: Control cranial CT after VP shunt surgery. 3D-E: 11-month outpatient control view.



**Figure 4.** 4A-B: Preoperative MR image 4C: Preoperative view of the patient 4D: Postoperative MR image 4E-F: 1 year and 5 months outpatient control view.

**Table 2.** Maternal data

Age, years; med (min-max)	27 (21-45)
Comorbidity of mother n (%)	
DM + HT	1 (5.3)
Preeclampsia	2 (10.5)
No	16 (84.2)
Gravity; med (min-max)	1.89 (1-3)
Parity; med (min-max)	1.73 (1-3)
Use of folic acid during pregnancy n (%)	
No	16 (84.2)
Yes	2 (10.5)
Unfollowed	1 (5.3)
Prenatal diagnosis (%)	
Unfollowed	12 (63.2)
Yes	7 (36.8)
Drug used in pregnancy n (%)	
No	18 (94.7)
Unfollowed	1 (5.3)

DM= Diabetes mellitus, HT= Hypertension.

One of the patients was born by vaginal delivery while the others were delivered by cesarean section under spinal anaesthesia.

Patients' operative and postoperative data are given in Table 3. The operation time varied between 0-720 days after birth. 9 patients were admitted as intubated from the preoperative neonatal unit to the operating room, while the other 10 patients were intubated in the operating room. No difficult intubation was observed in patients operated under general anaesthesia. The duration of operation varied between 45-150 minutes. While 7 patients were taken to the clinic after postoperative extubation (there was no need for newborn intensive care unit), the other 11 patients were taken to the neonatal unit as intubated and 1 patient as extubated. All patients who were extubated were over 1 month old. In this regard, it is important that patients who do not require emergency surgery are included in elective surgery. Our patients did not experience any complications related to perioperative anaesthesia or surgery. None of the patients needed perioperative blood transfusion. 17 patients were extubated on the first postoperative day. One of the other two patients was extubated after 2 days and the other after 4 days. The replacement was not required since there was no serious decrease in the preoperative and postoperative sodium, potassium, and hemoglobin values. Patients admitted to the newborn intensive care unit were admitted to the hospital for 1 to 24 days. 2 patients had prolonged newborn intensive care unit stay. The first patient, who had an Apgar score of 4-8, required 14 days of newborn intensive care unit. The second one was the patient who had omphalocele accompanying encephalocele. The patient who was operated on the same session due to encephalocele and omphalocele developed subsequent hisrodephagia, thereby requiring an EVD replacement. A 3-week EVD follow-up was required for the abdomen to be ready for ventriculoperitoneal shunt placement after

the omphalocele surgery. Therefore, a 24-day newborn intensive care unit stay was required. The patient was taken to the service without any problem following shunt surgery. The length of stay in the hospital also varied between 2-31 days. In the end, all the patients were discharged with full recovery. The follow-up period ranged from 1 month (due to the recently operated patients) to 52 months.

**Table 3.** Operation and after data

Operation time, minute; med (min-max)	81.31 (45-150)
Operation day; med (min-max)	88.73 (0-720)
Intubated when arrives, n (%)	
Yes	9 (47.4)
No	10 (52.6)
Intubated when out of operation n (%)	
Yes	11 (57.9)
No	8 (42.1)
Need for shunt n (%)	
Yes	5 (26.3)
No	14 (73.7)
Facial asymmetry n (%)	
Yes	1 (5.3)
No	18 (94.7)
Seizure n (%)	
Yes	3 (15.8)
No	16 (84.2)
Nutrition and growth retardation n (%)	
Yes	5 (26.3)
No	14 (73.7)
Length of stay in the hospital, day; med (min-max)	12 (2-31)
Follow up time, month; med (min-max)	19 (1-52)
Result n (%)	
Discharge	19 (100)

Only 5 patients required shunts. Parenchyma was present in the sac in 2 of the patients who needed a shunt, whereas the other had no parenchyma. The seizure occurred in 3 patients while there was no occurrence of seizures in the others. 5 of the patients were followed up by the department of pediatric gastroenterology due to malnutrition and growth retardation. One patient was admitted to the clinic due to a wound infection 3 months later and died due to aspiration while receiving antibiotic treatment. The others are still alive.

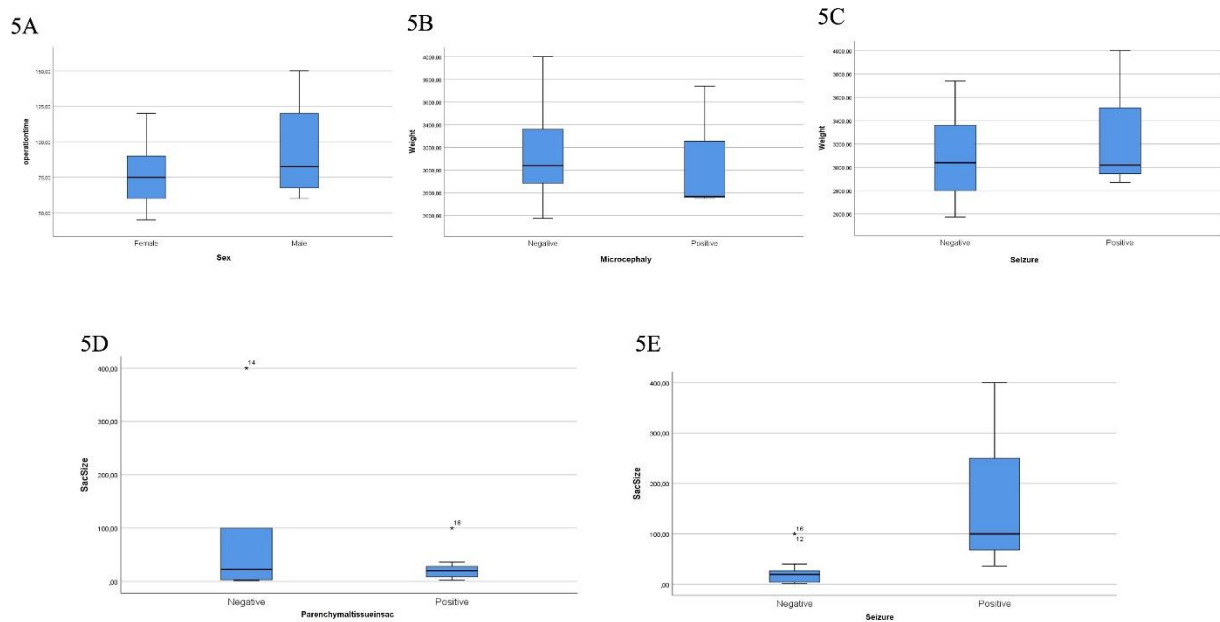
In conclusion, the mean weight of our 19 cases was 3128, the mean age of the mothers was 27, the mean gravity/parity was 1.89/1.73, the mean gestational age was 38 weeks, the infant 1-min/5-min APGAR score was 8/9 mins, taking into account the recently operated cases, the mean follow-up period was 19 months, the mean length of stay in newborn intensive care unit was 8,5 days (7 patients did not need newborn intensive care

unit), the mean head circumference was 34 cm, the duration of operation was minimum 45 and maximum 150 minutes, with a mean of 81 minutes, and the mean operation time was 88 days. According to the statistical comparisons; the operation times were found to be significantly shorter in female patients than in male patients (Figure 5), the incidence of microcephaly increased in low-weight patients (Figure 5), and the chance of seizures increased as the patients' weight increased (Figure 5). Significant changes were found in all parameters with maternal age. As maternal age increases, the frequency of pathological neurological examination increases, pregnancy without follow-up increases, microcephaly increases, the frequency of seizures increases, the parenchyma tissue in the sac increases, and the frequency of nutrition and growth

retardation increases (Table 4).

As the gestational age increased, the duration of postoperative extubation became shorter, and the incidence of pathological neurological examination increased. In addition, we found that patients with pathological neurological examination had lower head circumference and longer operation times.

We found a correlation between the APGAR scores of the patients and some data. We found that APGAR scores were low in microcephalic cases, cases with parenchyma tissue in the sac, cases with seizures, and cases who were intubated on arrival (Table 5). In addition, we found that the size of the sac was higher in those without parenchyma tissue in the sac (Figure 5), and the risk of seizures was higher in those with a larger sac size (Figure 5).



**Figure 5.** The statistical significance of the analyzed parameters. 5A: Operation time and gender, 5B: weight and microcephaly, 5C: weight and seizure status, 1D: The size of the sac diameter and presence of parenchyma in the sac content, 1E: The size of the sac diameter and seizure status.

**Table 4.** The parameters that were found to be statistically significant with maternal age

Maternal age vs. Pathological neurological examination	226.8	Yes	*	0,0121
Maternal age vs. Microcephaly	379.5	Yes	****	<0.0001
Maternal age vs. The parenchyma tissue in the sac	329.3	Yes	****	<0.0001
Maternal age vs. Need for shunt	362.7	Yes	****	<0.0001
Maternal age vs. Seizure status	379.5	Yes	****	<0.0001
Maternal age vs. Nutrition and growth retardation	362.7	Yes	****	<0.0001
Maternal age vs. Additional anomaly	367.8	Yes	****	<0.0001
Maternal age vs. Arrived intubated	329.3	Yes	****	<0.0001
Maternal age vs. Postoperative extubation time	334.6	Yes	****	<0.0001

**Table 5.** The statistical significance of the analyzed parameters

Baby apgar score 1 min vs. Microcephaly	317.9	Yes	****	<0.0001
Baby apgar score 1 min vs. The parenchyma tissue in the sac	267.7	Yes	***	0,0003
Baby apgar score 1 min vs. Need for shunt	301.2	Yes	****	<0.0001
Baby apgar score 1 min vs. Seizure status	317.9	Yes	****	<0.0001
Baby apgar score 1 min vs. Nutrition and growth retardation	301.2	Yes	****	<0.0001
Baby apgar score 1 min vs. Additional anomaly	306.3	Yes	****	<0.0001
Baby apgar score 1 min vs. Arrived intubated	267.7	Yes	***	0,0003
Baby apgar score 1 min vs. Postoperative extubation time	273.1	Yes	***	0,0002
Baby apgar score 5 min vs. Microcephaly	334.7	Yes	****	<0.0001
Baby apgar score 5 min vs. The parenchyma tissue in the sac	284.5	Yes	****	<0.0001
Baby apgar score 5 min vs. Need for shunt	318	Yes	****	<0.0001
Baby apgar score 5 min vs. Seizure status	334.7	Yes	****	<0.0001
Baby apgar score 5 min vs. Nutrition and growth retardation	318	Yes	****	<0.0001
Baby apgar score 5 min vs. Additional anomaly	323.1	Yes	****	<0.0001
Baby apgar score 5 min vs. Arrived intubated	284.5	Yes	****	<0.0001
Baby apgar score 5 min vs. Postoperative extubation time	289.9	Yes	****	<0.0001

#### 4. Discussion

Encephalocele occurs as a result of a midline fusion defect in the skull bone (Greenberg, 2019). This defect allows the herniation of the cerebral tissue or meninges (Ghatan, 2011). The most accepted theory for the origin of a congenital encephalocele is the incomplete separation of the surface ectoderm from the neuroectoderm after the closure of the neural folds (Matos Cruz and De Jesus, 2022). Genetic and environmental factors also play a role in the etiology of encephalocele. Encephalocele has been linked to TORCH diseases, consanguineous relationships, genetic predisposition, and more than 30 syndromes (Yucetas and Uçler, 2017). None of these additional pathologies was encountered in the history of our patients. Although encephalocele is a post neurulation defect, the use of folic acid has been reported to significantly reduce the incidence of all neural tube defects, including encephalocele (Copp et al., 2013). While the mothers of 16 of the 19 patients presented were not taking folic acid, the mother of the other 3 patients was not found to be using it ideally.

The incidence of congenital encephalocele is estimated to be 0.8-2.0 per 10000 live births (Cavalheiro et al., 2020). However, its actual incidence is estimated to be higher as termination is recommended when the diagnosis is made during pregnancy (Matos Cruz and De Jesus, 2022). Encephalocele is diagnosed during pregnancy follow-up in the first trimester (Engels et al., 2016). In our hospital, a total of 42593 deliveries were realized with 18260 C/S and 24333 vaginal deliveries between 2018 and 2020. The obstetrics department conducted 325 terminations in patients who were followed up on during pregnancy, with 7 of them being diagnosed with encephalocele (The data between 2016 and 2018 were not available). We operated 8 encephalocele cases between 2018-2020 and 19 encephalocele cases between 2015-2021 (In our

hospital, a total of 67832 deliveries were realized with 26643 C/S and 41189 vaginal deliveries between 2015-2021. 3.09 out of 10000 live births were operated on with the diagnosis of encephalocele.). 2.1% of the terminations performed between 2018 and 2020 were due to encephalocele, and 1.4 out of 10000 live births were operated on with the diagnosis of encephalocele.

As with all neural tube defects, female gender predominance is seen in encephaloceles. Female patients are more likely to have an occipital encephalocele compared to an anterior encephalocele (1.9/1) (Rehman et al., 2018). Of our 19 cases, 15 were girls and 4 were boys, all of whom were classified as posterior encephalocele, which is frequently located in the midline and occipital region. Approximately, 70-90% of them are located at the occipital region (Matos Cruz and De Jesus, 2022).

The presence of parenchymal tissue within the sac, the development of hydrocephalus, and seizures often indicate a poor prognosis, mostly occurring in occipital encephaloceles (Bui et al., 2007). In our case series, only 6 patients needed a shunt.

In radiological examinations, USG is used to show the content of the sac, CT to expose bone structures, and MRI to reveal the relationship of the sac with the venous sinus and vascular structures (Ozek and Hicdonmez, 2014). Preoperative cranial CT examination was performed in all cases. It provided great guidance for us as it revealed the sac content, any bone defects, and the presence of hydrocephalic appearance (Figure 1). Encephalocele surgery is performed in the prone position under general anaesthesia. The purpose of the operation is to excise the gliotic tissue and the ischemic neural contents in the sac, taking into account the sinus which may be passing through (Murthy et al., 2019). Given that there is only CSF content present in the sac and the sac neck is not wide, then the pedicle of the sac is exposed and the

remaining sac is excised, leaving enough part for dural repair. However, if there is unfunctional neural tissue in the sac, it is coagulated with bipolar and removed, and the dura and skin are closed, and the operation is terminated. If the neural tissue is thought to be functional and cannot be included in the intracalvarial area, the bony exposure is enlarged sufficiently covering the neural tissue with dura and skin, and the operation is terminated (Partington and Petronio, 2001). There was no need for calvarial reshaping in our cases. Additionally, congenital malformations that may accompany encephalocele, hydrocephalus, enlarged sac, and hemodynamic disorders that may develop during the perioperative period make anaesthesia management difficult. These patients may have perioperative complications such as electrolyte imbalance (due to CSF drainage), hypothermia, and blood loss (Mahajan et al., 2011; Singh et al., 2012). A blood transfusion may be required due to blood loss and related hemodynamic changes. In addition, during the opening of the encephalocele, rapid drainage of cerebrospinal fluid may lead to hemodynamic disturbances that may cause bradycardia or even cardiac arrest (Mahajan et al., 2011; Singh et al., 2012).

There was no severe electrolyte imbalance, low hb, and blood transfusion requirement in any of our cases. It is vital for these patients to be followed up with a multidisciplinary approach. Although postoperative newborn intensive care unit follow-up is very important in encephalocele cases, the absence of prolonged intubation and newborn intensive care unit requirement is more ideal for these patients. In this regard, elective surgery eliminates the need for newborn intensive care unit, except for cases with ruptured sac and cases with very thin skin at risk of rupture. 7 of our 19 cases did not need newborn intensive care unit, while 1 case was taken to newborn intensive care unit after extubation. All of these 5 patients were over 1 month old. The other 14 patients were taken into operation at 2-13 days old depending on the patient's condition, due to the need for urgent surgery. Except for the patient with a low Apgar score and the patient with accompanying omphalocele, other patients were extubated on the same day. Since the inclusion of cases in elective surgery eliminates the need for newborn intensive care unit and shortens the duration of intubation, we believe that the cases should be operated on electively unless it is very urgent. (Operated electively except for the cases with a high risk of rupture with ruptured sac or very thin skin). There was no serious electrolyte imbalance and blood transfusion requirement in the perioperative period. Classically, children with encephalocele develop significant rates of spastic paraparesis (11%), moderate to severe developmental delay (7%), and hydrocephalus (20%-65%) (Gandhoke et al., 2017). Posterior encephaloceles exhibit a worse prognosis than anterior encephaloceles due to the higher incidence of hydrocephalus development and seizures and being

neurologically worse in the presence of functional cerebral tissue, and they also lead to greater physical, emotional and cognitive delay (Bui et al., 2007). All of our cases were classified as posterior encephalocele and we did not have any patients with physical and cognitive impairment, except for the case who was born hypoactive with a dysmorphic facial appearance. The seizure occurred in only 3 patients while there was no occurrence of seizures in the others. 5 of the patients were followed up by the department of pediatric gastroenterology due to malnutrition and growth retardation. Parenchyma was observed within the sac in 9 patients, and only CSF content was present in the sac of 10 patients. Meningocele have a better prognosis than encephaloceles. While the mortality due to postoperative infection and shunt complications is 14% in meningoceles, it is 52% in encephaloceles (French et al., 1990; Partington et al., 2001). Motor and mental retardation is found in 83% of all encephaloceles (French, 1990). Of our 19 cases, 9 were classified as encephalocele (48%), and 10 as meningocele (52%).

### 5. Conclusion

Encephalocele cases should be followed up multi disciplinarily before and after surgery because of the risk of concomitant additional malformations, feeding problems, and seizures.

The presence of parenchyma in the sac and its functionality, its relationship with vascular structures, and the presence of hydrocephalus are significant surgical factors, whereas CSF fistula formation, shunt requirement or shunt-related complications, and seizure development are significant postoperative factors.

Additionally, planning elective surgery for encephalocele cases that do not have an indication for emergency surgery also eliminates the need for postoperative newborn intensive care unit. For these reasons, encephalocele cases should be treated with care, taking into account the timing of surgery as well as any potential preoperative and postoperative complications. And most importantly, we hope that the incidence of encephalocele will decrease with greater encouragement of folic acid supplementation and more strict pregnancy follow-up in the future.

### Author Contributions

Concept: E.B.G (%50) and N.K. (50%), Design: E.B.G (%50) and N.K. (50%), Supervision: E.B.G (%50) and N.K. (50%), Data collection and/or processing: E.B.G (%50) and N.K. (50%), Data analysis and/or interpretation: E.B.G (%50) and N.K. (50%), Literature search: E.B.G (%50) and N.K. (50%), Writing: E.B.G (%50) and N.K. (50%), Critical review: E.B.G (%50) and N.K. (50%), Submission and revision E.B.G (%50) and N.K. (50%). All authors reviewed and approved final version of the manuscript.



## Conflict of Interest

The authors declared that there is no conflict of interest.

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## Ethical Approval/Informed Consent

The study was initiated after obtaining approval from the Bursa Health Sciences University ethics committee (Decision number and date: 011-KAEK-25 2020/11-12 / 25.11.2022). 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. Informed consent was obtained from all individual participants in the study.

## References

- Bui CJ, Tubbs RS, Shannon CN, Acakpo-Satchivi L, Wellons JC, Blount JP. 2007. Institutional experience with cranial vault encephaloceles. *J Neurosurg*, 107(1): 22-25.
- Cavalheiro S, Silva da Costa MD, Nicácio JM. 2020. Fetal surgery for occipital encephalocele. *J Neurosurg Pediatr*, 26(6): 1-8.
- Chen CP, Chern SR, Wang W. 2000. Rapid determination of zygosity and common aneuploidies from amniotic fluid cells using quantitative fluorescent polymerase chain reaction following genetic amniocentesis in multiple pregnancies. *Hum Reprod*, 15: 929-934.
- Copp AJ, Stanier P, Greene ND. 2013. Neural tube defects: recent advances, unsolved questions, and controversies. *Lancet Neurol*, 12(8): 799-810.
- Drake JM, MacFarlane R. 2001. Encephalocele repair. In: McLone DG. *Pediatric Neurosurgery*, 4th Ed., WB Saunders Philadelphia, US, pp: 209-213.
- Engels AC, Joyeux L, Brantner C, De Keersmaecker B, De Catte L, Baud D. 2016. Sonographic detection of central nervous system defects in the first trimester of pregnancy. *Prenat Diagn*, 36(3): 266-273.
- French BN. 1990. Midline fusion defects and defects of formation. In: Youmans JR, *Neurological Surgery*, 3rd Ed., WB Saunders, Philadelphia, US, pp: 1081-1235.
- Gandhoke GS, Goldschmidt E, Kellogg R, Greene S. 2017. Encephalocele development from a congenital meningocele: case report. *J Neurosurg Pediatr*, 20(5): 419-422.
- Ghatan S. 2011. Encephalocele: Cranial development Abnormalities: Pediatrics. In: Winn RH, Youmans *Neurological Surgery*, 6th Ed., Elsevier, London, UK, pp: 1898-1905.
- Greenberg MS. 2019. *Handbook of neurosurgery*. 9th Ed., Thieme Medical Publishers Incorporated, London, UK.
- Mahajan C, Rath GP, Dash HH, Bithal PK. 2011. Perioperative management of children with encephalocele: an institutional experience. *J Neurosurg Anesthesiol*, 23(4): 352-356.
- Matos Cruz AJ, De Jesus O. 2022. Encephalocele. URL: <https://www.ncbi.nlm.nih.gov/books/NBK562168/> (March 01, 2022).
- Murthy PS. 2019. Kalinayakanahalli Ramkrishnappa SK. Giant Occipital Encephalocele in an Infant: A surgical challenge. *J Pediatr Neurosci*, 14(4): 218-221.
- Ozek M, Hicdonmez T. 2014. Ensefaloseller. In: Baykaner K, Ersahin Y, Mutluer S, Ozek M. *Pediatric Nöroşürji* (Ed). *Buluş Tasarım ve Matbaacılık Hizmetleri*, Ankara, Türkiye, pp: 365-376.
- Önder H. 2018. Nonparametric statistical methods used in biological experiments. *BSJ Eng Sci*, 1(1): 1-6.
- Partington MD, Petronio JA. 2001. Malformations of the cerebral hemispheres. In: McLone DG. *Pediatric Neurosurgery*, 4th Ed., WB Saunders, Philadelphia, US, pp: 201-208.
- Rehman L, Farooq G, Bukhari I. 2018. Neurosurgical Interventions for Occipital Encephalocele. *Asian J Neurosurg*, 13(2): 233-237.
- Rolo A, Galea GL, Savery D, Greene NDE, Copp AJ. 2019. Novel mouse model of encephalocele: post-neurulation origin and relationship to open neural tube defects. *Dis Model Mech*, 12(11): dmm040683.
- Singh H, Singh D, Sharma DP, Tandon MS, Ganjoo P. 2012. Perioperative challenges in patients with giant occipital encephalocele with microcephaly and micrognathia. *J Neurosci Rural Pract*, 3(1): 68-70.
- Stoll C, Alembik Y, Dott B. 2007. Associated malformations in cases with neural tube defects. *Genet Couns*, 18(2): 209-215.
- Ugras M, Kavak O, Alpay F, Karabekir HS, Biçer S. 2016. New born children with ensefhalocele. *J Neurol Neurosci*, 7(1): 73.
- Yucetas SC, Uçler N. 2017. A retrospective analysis of neonatal encephalocele predisposing factors and outcomes. *Pediatr Neurosurg*, 52(2): 73-76.