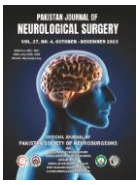

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Original Research

Association of Hydrocephalus in Children with an Encephalocele

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ABSTRACT

Objective: To describe the association between the occipital encephaloceles with hydrocephalus and how to improve the outcome of the patient.

Materials and Methods: This prospective study collected records of fifty patients diagnosed with posterior encephaloceles. Data were collected on the gender, location of the encephalocele, presence of neural tissue, dandy walker, microcephaly, and hydrocephalus.

Results: Twenty-nine females and twenty-one males were present. Over half of the lesions were supratorcular, while the remaining eight (16%) were torcular and the remaining seventeen (34%) were infratorcular. Primary encephalocele repairs were done at an average of 9 days (range 2.5–120 days). In twenty-five of the instances, a diagnosis of hydrocephalus was made. Twenty-four patients had a ventriculo-peritoneal shunt implanted. The average age of implantation of VP shunt was only 1.2 months (range 0.3–9 months). One patient underwent an endoscopic third ventriculostomy with good results. Hydrocephalus was related to Dandy-Walker and ventriculomegaly before encephalocele surgery was performed (*p* values 0.01 and 0.05, respectively). For hydrocephalus therapy, CMIII malformation was detected in two patients, which required the insertion of a VP shunt. In nine cases, microcephaly was found. The death rate was estimated to be 8%, and death was associated with Chiari malformation, microcephaly, and Dandy-walker.

Conclusion: Patients born with encephaloceles frequently suffer from hydrocephalus (50%). This is especially true in cases where the patient also has Dandy-Walker syndrome, CMIII malformation, or pre-existing ventriculomegaly. The severity of large encephaloceles is a limiting factor for the development of hydrocephalus when connected with torcular types and microcephaly.

Keywords: Dandy-Walker malformation, Microcephaly**,** Chiari III malformation, Encephalocele, Hydrocephalus, Ventriculo-peritoneal shunt.

Abbreviations: CM-III: Chiari III malformation. **DWM:** Dandy-Walker malformation. **NTD:** Neural tube defect, CSF Cerebral Spinal Fluid. **CI:** Confidence interval. **OR:** Odds ratio. **VP SHUNT:** Ventriculo-peritoneal shunt.

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INTRODUCTION

Herniation of the intracranial content is the defining feature of encephalocele, which is caused by a defect that is present at birth in the skull. The term "meningocele" refers to a herniation of the meninges, while "meningoencephalocele" refers to a herniation of both the meninges and the brain. "hydro encephalomeningocele" refers to a herniation that involves a portion of the cerebral ventricles.[1] Occipital regions are the most common sites for encephaloceles, accounting for over 75% of all cases. Infratorcular or supratorcular encephaloceles of the occipital region can be distinguished from one another based on their location concerning the torcula.[2] A torcular type is yet another division.[3]

The actual cause of the condition is unclear, and it often arises in the context of other types of brain abnormalities. The primary theory of its etiology proposes that a deficiency in the dura and the calvaria, which might allow the cerebral material to protrude through, results from an aberration in mesodermal development,[4-5] Even though some researchers have hypothesized a link between encephaloceles and spina bifida and anencephaly.[6-7] There is a wide range of encephalocele incidence, ranging from 0.8 to 5.6 per 10,000 live births.[8] Despite the disease's rarity, it is still in high demand in developing and emerging countries, such as Brazil, where abortion is only permitted in extremely rare circumstances. Mortality and morbidity in neonates have decreased as time went on as diagnostic testing, surgical procedures, and Newborn care following surgery have improved throughout the years. Despite this, there are very few investigations on occipital encephaloceles and hydrocephalus that have been done.[9] It is the goal of this study to describe our series, encourage a reassessment of the literature, and examine incidence rates of hydrocephalus in patients with posterior encephaloceles and often encountered abnormalities such as Chiari III malformation, microcephaly, Dandy-Walker malformation, and others.

Surgical morbidity was evaluated through the initial follow-up consultation. A longer follow-up period was not included in this study because the researchers knew that the complication rate would be lower than if a shorter follow-up period were considered. Instead, they wanted to see how common VPS insertion–related problems are in the short term.

Cephaloceles and encephaloceles are both types of neural tube defects that can occur during pregnancy (NTD). As many as 0.8 per 10,000 births are thought to be affected by them.[10] Depending on where the herniation occurs, this deformity might affect the occipital, frontal, temporal, or parietal regions. Cephaloceles can be found in a variety of places around the world. Occipital or posterior cephaloceles account for 66% to 89 percent of all cases in Europe and North America, with the remaining 15% occurring in the frontal or parietal lobes. Frontal (sincipital) vs. occipital placement is more common in Thailand and other southern Asian countries.[11]

MATERIALS AND METHODS

Study Design and Setting

The study was carried out in the Neurosurgery department of Jinnah Postgraduate Medical Center Karachi from September 2022 till May 2023. A total of 59 patients having encephalocele repair were considered in the study.

Inclusion Criteria

Inclusion criteria included all patients having encephalocele with complete radiological evaluation.

Exclusion Criteria

Exclusion criteria comprised patients who did not follow up with radiological imaging and those

who did not consent to be a part of the study. Therefore 9 out of 59 were excluded and the total sample size was 50 patients.

Collection of Data

MRI neuro-spinal axis was done to assess the size of the bone defect, the contents of the encephalocele, the presence or absence of hydrocephalus, and other associated intracranial anomalies. A predesigned proforma was used to document patient demographics as well as the location and size of an encephalocele, amount of neural tissue within the malformation, concordant hydrocephalus, presence of microcephaly, Chiari malformation, and other malformations. All of the patients then subsequently underwent encephalocele repair with or without VP shunt placement. Those lesions with minimum brain tissues within the sac are described as having CSF content as described in the table.

RESULTS

A total of 50 people sought treatment at (ABC) for encephalocele "between" September 2022 to May 2023.

Gender Distribution

We counted 50 people, 29 of whom were females and 21 males, for a female-to-male ratio of 1:1.38.

Age Distribution

A total of fifty children were included in this study. Their age ranged from 0 to 24 months. 30 patients were under 1 month old and had a mean age of 2.3 months.

Patient's Cohort

Patients 21 and 3 underwent surgery at 4 months and 2 months old, respectively, however, the majority of the patients underwent neonatal surgery. Primary encephalocele repair was performed on 46 patients (92%). Four individuals were ineligible for surgery because of the presence of torcula in the severe microcephaly and/or malformation. Table 1 highlights the cohort's characteristics.

Table 1: Patient cohort.

Sr. #	Gender	Contents	Associated malformations	Hydrocephaly	Microcephaly	Follow-up Period (Months)
1.	Female	NT	No	-	-	3
2.	Female	NT and venous sinus	Abnormal sulcation	-	+	4
3.	Female	CSF	-	-	-	2
4.	Female	CSF	Dandy-walker malformation	+	-	4
5.	Male	NT and venous sinus	Abnormal sulcation	-	+	1
6.	Female	CSF	Arachnoid cyst	+	-	1
7.	Female	NTs and ventricle (giant encephalocele)	Ventriculomegaly	+	-	3
8.	Male	CSF	Dysgenesis of the corpus callosum + large ventricles	+	-	4
9.	Female	NT	Ventriculomegaly	+	-	4
10.	Female	CSF	Dysgenesis of the corpus callosum + large ventricles	+	-	1

11.	Female	NT	Ventriculomegaly	+	-	2
12.	Female	NT + CSF	Ventriculomegaly	+	-	2
13.	Female	CSF	Dandy-walker malformation	+	-	3
14.	Female	NT and ventricles (giant encephalocele)	Dysgenesis of the corpus callosum	-	+	4
15.	Female	NT and CSF	Dysgenesis of the corpus callosum + large ventricles	+	-	4
16.	Female	NT	Dysgenesis of the corpus callosum	-	-	3
17.	Male	NT	Dysgenesis of the corpus callosum	-	+	4
18.	Male	NT	-	-	-	
19.	Male	NT	Ventriculomegaly	+	-	1
20.	Female	NT and ventricle (giant encephalocele)	Abnormal sulcation	-	+	2
21.	Male	CSF	-	-	-	3
22.	Female	NT and venous sinus	Abnormal sulcation	-	-	Dead
23.	Male	CSF	Dandy-walker malformation	+	+	3
24.	Male	NT	Abnormal sulcation	-	+	No follow-up
25.	Male	NT + CSF	Dandy-walker malformation	+	-	4
26.	Male	NT + CSF	-	-	-	No follow-up
27.	Female	CSF	Ventriculomegaly	+	-	No follow-up
28.	Female	NT + CSF	Abnormal sulcation	-	+	No follow-up
29.	Female	CSF	-	-	-	2
30.	Male	CSF	-	-	-	2
31.	Male	CSF	-	-	-	1
32.	Male	CSF + cerebellum (Chiari III)	Dysgenesis of the corpus callosum + large ventricles	+	-	2
33.	Male	NT	-	+	-	3
34.	Male	NT	-	+	-	3
35.	Female	NT	Ventriculomegaly	+	-	4
36.	Male	CSF	Ventriculomegaly	+	-	1
37.	Male	NT	Ventriculomegaly	-	-	3
38.	Male	NT	Ventriculomegaly	+	-	2
39.	Female	CSF	-	+	-	2
40.	Female	CSF	-	-	-	3
41.	Male	NT	Ventriculomegaly	-	-	3
42.	Female	NT + ventricle (giant encephalocele)	Dysgenesis of the corpus callosum + abnormal sulcation	-	+	Dead
43.	Female	NT + CSF	Ventriculomegaly	-	-	No follow-up
44.	Female	NTs + CSF + venous sinus (Giant encephalocele)	Abnormal sulcation	-	-	Dead
45.	Female	NT and ventricle	Abnormal sulcation	-	+	Dead
46.	Female	NT	-	+	-	No follow-up
47.	Male	CSF	Dandy-walker malformation	+	-	No follow-up
48.	Female	CSF	Ventriculomegaly	-	-	1

49.	Male	NT+CSF	-	+	-	No follow-up
50.	Female	CSF+cerebellum (Chiari III)	-	+	-	116-days

Key: (-) indicates No results **(+)** indicates presence torcula, **(NT =** Neural tissues**)**

Location of Encephalocele And Its Association with Hydrocephalus

There were 25 (50%) encephaloceles identified as supratorcular, whereas eight (16%) were classed as torcular and 17 (34%) were classified as infratorcular, according to the torcula connection (Table 2). Follow-up lasted anywhere from 0 to 9 months of age, on average. Diagnosed hydrocephalus occurred in 25 of the cases (50 percent). Before encephalocele surgery, 5 (20%) of these patients had DWM, and 13 (52%) had ventriculomegaly. Hydrocephalus was found to be related to DWM and a history of ventriculomegaly in a bivariate analysis, as shown in Table 3 (*p* values 0.01 and 0.05, respectively). We were unable to corroborate this conclusion in the logistic regression model because of the small sample size. Table 2 shows the distribution of cases of hydrocephalus according to the kind of encephalocele, with supratorcular forms being seen more frequently than other types.

Table 2: Encephalocele location x frequency of hydrocephalus.

Location	Hydrocephalus		Frequency
	Yes	No	
Infratorcular	8 (47.1%)	9 (52.9%)	17 (34%)
Torcular	2 (25%)	6 (75%)	8 (16%)
Supratorcular	15 (60%)	10 (40%)	25 (50%)
Total	25 (50%)	25 (50%)	50 (100%)

Treatment for Hydrocephalus

The treatment for hydrocephalus was either an endoscopic third ventriculostomy or a ventriculoperitoneal shunt (also known as VP shunts) (ETV). 23 of the 25 patients who were diagnosed with hydrocephalus had a ventriculoperitoneal (VP) shunt placed as their primary treatment because of their young age and unfavorable anatomy for ETV placement.

Complications of Surgery

All patients were given a medium-sized fixed differential pressure valve. The average age of placement of a VP shunt was 1.3 months (range 0.3–9 months). The proximal obstruction was detected in one patient, while infection was found in the other, both of whom had VP shunt malfunction (8.3%). CSF leaks and surgical wound infections were detected in seven of the patients with huge encephaloceles, which is a typical occurrence. Shunts were placed in four individuals who had previously suffered from hydrocephalus, and wounds healed and Trans fontanelle punctures were made in these cases. In these situations, no additional difficulties were reported. Nine people had microcephaly. Four of them (44 percent) succumbed to other injuries. It was known that 8% of the total were dead. 17 patients had CSF leaking out of their ruptured sacs, 15

Table 3: Bivariate correlation analysis of neurological abnormalities and hydrocephalus.

Variables	Hydrocephalus		Bivariant Analysis	
	Yes(n=25)	No (n= 25)	OR (95% CI)	*p* Values
NT	14 (43%)	18 (56%)	0.4 (0.1-1.3)	0.3
Dandy-walker malformation	5 (100%	0 (0%)	2.3 (1.6-3.1)	0.04*
Ventriculomegaly	13 (76.5%)	4 (23.5%)	5.6 (1.5-2.1)	0.02*
Chiari III malformation	2 (100%)	0 (0%)	2.0 (1.5-2.8)	0.14
microcephaly	0 (0%)	9 (100%)	2.5 (1.7-3.7)	0.02*

*Statistically significant

patients had brain tissue, 9 patients had brain tissue mixed with CSF, 9 patients had brain tissue mixed with cerebral ventricles, and 9 patients had brain tissue mixed with venous sinuses. A bivariate study looked at whether or not the presence of neural tissue was associated with an increased risk of hydrocephalus and found no correlation (Table 3). In six cases, the abnormality contained neural tissue and cerebral ventricles, which were linked to large encephaloceles. 66% of these cases were accompanied by microcephaly, of whom 3 died, 2 had shortened follow-up (1 month), and only 1 developed hydrocephalus needing shunting. VP shunts were used to treat hydrocephalus in two individuals, both of whom had Chiari III malformation, and both were stable in follow-up.

DISCUSSION

There are several rare congenital disorders of the nervous system called encephaloceles. Affected individuals have protruding brain tissue in their skulls as a result of these conditions.[12] As a result of certain disorders or by itself, it can occur.[13] As of now, there is no clear pathophysiology for this damage in the literature. Even though the lesion was initially categorized as a neural tube defect (NTD), the fact that it was epithelialized and did not contain dystrophic brain tissue suggests that the mechanism for the formation could be related to a mesodermal insufficiency, in a manner that differs from other NTDs.[5,14] Some researchers believe folic acid supplements can provide some protection against these conditions as well as encephaloceles, which share similarities with spina bifida and anencephaly.

It remains a very dangerous disease that has a high mortality and morbidity rate, despite advances in surgical techniques and research into its origins. Various literature sources mention a wide range of death rates. Kotil et al, had a mortality rate of 33.3%.[14] A death rate of 8 percent was observed in our cohort. The absence of follow-up may lead to fatalities or serious disabilities. It has been documented that 16 to 31% of patients with encephaloceles have substantial physical and cognitive impairments, despite the differing mortality rates among the studies. However, the research is consistent as far as mental and motor abilities are concerned.[15–17] It has been determined that the prognosis of the patient is impacted by how much neural tissue there is inside the abnormality, as well as any concomitant congenital defects or infections that may be present, presence of vascular structure, size of encephalocele compared to the size of the skull.[14,15. 18,19]

According to the research, people with posterior encephaloceles are more likely to develop hydrocephalus than those with other forms of this deformity.[9,13,20,21] However, Warf et al. showed a comparable incidence of hydrocephalus in patients with encephaloceles in the occipital, sincipital, and parietal regions.[22] A prevalence rate of fifty percent was found in this study for hydrocephalus. A Supratorcular encephalocele occurs 60% of the time, while an Infratorcular encephalocele occurs 47% of the time, and a torcular encephalocele occurs 25% of the time or less frequently. Torcular cases were the most severe in our experience. There have been two fatalities and three cases of microcephaly associated with this form, which usually occurs in pairs with enormous encephaloceles. Considering that torcular encephaloceles frequently result in death, it is reasonable to assume the affected individuals would not survive long enough to develop hydrocephalus. This also explains why there is a lower incidence of hydrocephalus in cases where microcephaly is present, as shown in Table 3. Da Silva et al,[9] recommended that Hydrocephalus has been linked to the presence of neural tissue within the defect, as well as brain malformations, massive encephaloceles, seizures, and microcephaly.

An analysis of multivariate logistic regression

determined there was only one predictive feature with a statistical significance close to significant: the presence of neural tissue in the defect. In 64 percent of the cases we examined, neural tissue was found inside the sacs. The prevalence of hydrocephalus, however, did not reveal any statistical correlation with this discovery. Eight individuals, including four patients who passed away, were lost to follow-up, which may account for the lack of association. Markovic et al,[8] mentioned that we hypothesize that the twisting or stenosis of the Sylvian aqueduct is to blame for the development of hydrocephalus after surgery. We had a few cases involving patients with hydrocephalus who had the Sylvian aqueduct permeable before encephalocele repair. Five patients were diagnosed with DWM, while 13 patients had ventriculomegaly. The correlation was statistically significant (Table 3). It is uncommon for the DWM to be reported in encephalocele series. Bindal et al,[23] concluded that there is a 16% chance of this happening. DWM was discovered in 10% of the cases in our series. Most cases of DWM have hydrocephalus and massive encephaloceles have been linked to increasing ventriculomegaly in the past.[24] The increased intra-cystic pressure may be a compensatory response to the elevated intracranial pressure that occurs throughout pregnancy.[25] There are, however, instances where symptoms of hydrocephalus are not recognized for some time after a corrective operation for an encephalocele has taken place.[22] Considering this, it may be possible that hydrocephalus and encephalocele are separate conditions caused by DWM.[24,26] In this group of patients with DWM associated with occipital encephalocele, ventriculoperitoneal shunting was required in all cases (p-value 0.05). Giant encephaloceles were not found to be associated with any of these cases.

The possibility of Chiari II malformation in infratentorial encephaloceles must be brought to light, as well as high occipital or cervical encephaloceles that exhibit Chiari II malformation symptoms. [27] The development of CMIII has been explained by a few hypotheses. In general, encephaloceles have already been associated with mesodermal insufficiency, but some hypotheses involve CSF leakage or aberrant neuronal development.[27,29] It appears that altered CSF flow dynamics contribute to the onset and progression of hydrocephalus in these patients. There were two cases of CMIII in this group, both of which needed VP shunting to treat their hydrocephalus. Meckel-Gruber syndrome was present in one of them.[12]

In the literature, it has been suggested that patients with respiratory stridor, encephaloceles, hydrocephalus, and neural tissue within the sac have a worse prognosis than those without these problems[28]. Despite significant cognitive delays, both of our patients were able to make it through their treatment. VP shunts are the most common treatment for hydrocephalus in encephaloceles. According to a study by Rehman et al,[13] before encephalocele repair can be completed, a VP shunt must be installed. Kabré et al,[7] suggested that when preoperative hydrocephalus is present, encephalocele should be addressed as well. Surgical wounds from encephaloceles should be treated with antibiotics only after the hydrocephalus has healed to reduce the risk of infection. Medical literature does not readily report shunt dysfunction rates in encephalocele series.[7,13,14] DaSilva et al,[9] published that in 3.7 years of follow-up, found a malfunction rate of 20%. 90% of shunt dysfunction events occurred in the first 24 months of follow-up in our cohort, which had an 8% shunt dysfunction rate, as seen by the Kaplan–Meier survival curve that was shown. Encephalocele patients with endoscopic third ventriculostomy are uncommonly reported in the literature. According to the largest known series, 13 patients under the age of a year have undergone endoscopic third ventriculostomy/ choroid plexus cauterization; two patients under the age of a year have undergone ETV alone.[22]

Approximately 79% of individuals who begin treatment within one year are successful, based on the Kaplan–Meier survival study. Although a few years have passed since the study began, there has been little follow-up. Our series included only two cases where ETV was used. A VP shunt was performed on the first patient after his treatment failed. In the second case, however, there was no shunt and it was followed for 15 years before treatment was initiated. Though most series are based on real-life events.[7,9,13,14] VP shunts are the most common treatment for hydrocephalus, although ETV may be an option when the patient's age and anatomy are appropriate. This study has several drawbacks, including a retrospective nature, a limited sample size, and no follow-up data. A more thorough follow-up study might even categorize non-hydrocephalic patients as having hydrocephalus, especially those with encephaloceles are associated with higher mortality rates.

CONCLUSION

Patients with posterior encephaloceles are more likely to experience hydrocephalus, which is more common in those with the Supratorcular form. DWM, CMIII, and preexisting ventriculomegaly are also more likely to be associated with the connection. Even when combined with torcular types and microcephaly, the severity of gigantic encephaloceles prevents the development of hydrocephalus because of the quick natural history of evolution and structural alterations found in microcephaly.

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Additional Information

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AUTHORS CONTRIBUTION

S. No.	Author's Full Name	Intellectual Contribution to Paper in Terms of:
1.	Sagheer Ahmed	Study design and methodology.
2.	Iram Bokhari	Literature review and referencing.
3.	Tanveer Ahmed	Final review and approval.
4.	Rabail Akbar	Data collection and calculations.
5.	Raheel Gohar	Interpretation of results.
6.	Anas Ahmed	Analysis of data.