OPERATIVE SURVIVAL OF CHILDREN WITH CONGENITAL ENCEPHALOCELE AT ZEWDITU MEMORIAL HOSPITAL, ADDIS ABABA UNIVERSITY, COLLEGE OF HEALTH SCIENCES AND SCHOOL OF MEDICINE, DEPARTMENT OF SURGERY, NEUROSURGERY UNIT.

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Operative survival of Children with Congenital Encephalocele at Zewditu Memorial Hospital, Addis Ababa University, College of health sciences and School of Medicine, Department of Surgery, Neurosurgery Unit, Addis Ababa, Ethiopia.

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Academic Research Submission

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The full title of the research project	Operative survivals of children with congenital Encephalocele; An institutional based a retrospective chart reviews at ZMH Pediatric neurosurgical center, affiliated hospitals under Addis Ababa university, in Addis Ababa, Ethiopia.
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Abstract

Background; Encephalocele, a subtype of neural tube defects, is characterized by the protrusion of cranial contents through a defect in the skull. The goal of surgery for encephalocele involves excision of the sac, followed by watertight dural repair, and often reconstruction of the bony defect. Survival of children born with EC varies widely across the regions.

Objective; the main purpose of the study was to determine the operative survival rates of children with congenital encephalocele and the factors predicting operative survival outcomes. The study also aimed to assess patterns of congenital encephalocele occurrence.
Materials and Methods; we retrospectively reviewed the medical charts of 126 patients who underwent repair of congenital encephalocele at ZMH Pediatric Neurosurgery Center between January 2018 and June 2024. Relevant demographic and clinical data, including age, sex, birth status, location, size and content of encephalocele, neurologic status, associated congenital anomalies, perioperative, and follow-up data were recorded. Survival analyses, including Life Table, Kaplan–Meier, log-rank test, and Cox proportional hazard model, were used to determine the probability of survival at specific times, examine variation in survival of variables by time of surgery, and assess prognostic factors.
Results; Out of a total of 126 cases, 40.5% were males and 59.5% females. The mean age in days at surgery was 130+-50. At delivery, most of the patients (90.5%) were term. Occipital encephalocele was the most common, accounting for 67.5%, followed by occipito-cervical encephalocele (13.5%) and fronto-ethmoidal (9.5%). The sac of EC contained no neuronal tissue in 66.7% of patients and had contained dysplastic/neuronal tissue in 33.3%. Motor strength of the extremities was intact in majority of patients (78.8%) and incomplete in 28 patients (22%). A diagnosis of congenital anomaly was made in 40 patients (31.7%.. The survival probability was 98% at 1 month, 92% at 2 years, 85% at 4 years, and 75% at 6.5 years, with a median follow-up time of 26.86 months (95% CI: 63.102, 73.996). The Kaplan-Meier log-rank test showed that the presence of neuronal tissue within the sac, occipito-cervical location, preterm delivery, presence of preoperative neurologic deficit, and associated congenital anomalies possibly correlated with worse survival outcomes. However, the Cox proportional hazard model demonstrated, presence of neuronal tissue within the sac($p=0.01$) and associated congenital anomalies($p=0.006$) were statistically significant in predicting operative survival outcomes.
Conclusion;
Our study demonstrated that surgery for congenital encephalocele is a common practice in our institution. The presence of neuronal tissue within the sac, and associated congenital malformations, were factors statistically significant correlated with worse survival outcomes. Despite the challenges, our study's overview of the survival outcomes of children operated on for congenital encephalocele has shown almost comparable result to other centers. We believe that this may serve as a foundation for future prospective and long-term studies. It is also important for decision-makers and caregivers who plan for subsequent follow-up and long-term care in patients with congenital Encephalocele.

Keywords: Encephalocele, Operative survival, cranial anomalies

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List of acronyms and abbreviations

AAU Addis Ababa University

ANC Antenatal care

CDC Center for Disease for control
CSF Cerebrospinal fluid
Chiari-II Chiari Type –II Malformation
Chiari-III Chiari Type –III Malformation
CT Computed Tomography
CS Cesarean Section
DWM Dandy walkers' malformation
EC Encephalocele
ETV Endoscopic Third ventriculostomy
HC Head circumference
HCP Hydrocephalus
MRI Magnetic resonance imaging
MMC Meningomyelocele
NTD Neural Tube Defect
SVD Spontaneous Vaginal Delivery
SPSS Statistical Package for the Social Sciences
TASH Tikur Anbessa Specialized Hospital
USG Ultrasonography
VPS Ventriculoperitoneal Shunt
ZMH Zewditu Memorial Hospital

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1. Introduction

1.1 Background

Encephalocele, a subtype of neural tube defects, is characterized by the protrusion of cranial contents through a defect in the skull(1)(2). The composition of the encephalocele can vary, ranging from a sac containing only cerebrospinal fluid (meningocele) to a sac containing cerebrospinal fluid and brain tissue (meningoencephalocele), and even to a sac containing brain tissue and ventricles (cystoencephalocele). Severity of encephalocele correlates with extent and part of the neuronal tissue that has herniated through the defect. In more than half of the cases, encephaloceles are associated with other brain and systemic congenital malformations which influences further neurological morbidity and mortality in children(3,4) .

The estimated overall incidence of encephalocele is between 0.8 and 3.0 per 10,000 births, accounting for 10% of all neural tube defects(5) . The occurrence of encephaloceles varies geographically, with occipital encephaloceles being more common in the USA, Europe, and the Middle East, while sincipital encephaloceles are more prevalent in Southeast Asia (6)(7). Currently, the incidence of encephaloceles is declining, possibly due to the widespread introduction of folic acid fortification and the early detection of cases through prenatal screening with Sonography, which allows for the termination of pregnancies that would result in significant malformations incompatible with life. The precise pathophysiologic mechanism of encephalocele is not fully understood, but it is hypothesized to be the result of a defect in the separation of the ectoderm and neuroectoderm during the embryonic stage of brain development, particularly between the 5th and 9th weeks of gestation(8)(4) .

Encephaloceles can be classified into two groups based on their etiology: primary (congenital) , and secondary encephaloceles which occur as a result of trauma or other pathological processes(1) . Depending on the location of the sac, encephaloceles are classified as occipital, parietal, sincipital (frontoethmoidal), or basal (9)(10). In relation to the coronal suture, EC are further grouped into anterior and posterior encephaloceles [14]. Swelling over the craniofacial region is most common presentation at birth except atretic or basal EC which could present without obvious external swelling(11)(3).

Encephalocele can be detected prenatally by means of maternal alpha-fetoprotein elevation and Ultrasonography since the late stages of first trimester of pregnancy. A postnatal diagnosis of encephalocele is established on the basis of clinical evaluation of the sac and, with greater precision, by employing imaging findings such as US, CT or MRI in order to identify the anatomical location, estimate the size and extent of tissue contained within the sac, evaluate the presence of hydrocephalus, and identify any associated congenital malformations of the cranium, thus facilitating of surgical treatment planning (12)(11)(13).

Management of Encephalocele involves multidisciplinary approach including; Neurosurgeons, pediatricians, anesthesiologist among others. The goal of surgery for encephalocele involves excision of the sac and its dysplastic tissue, followed by watertight dural repair and reconstruction of the bony defect. The optimal timing of surgery is not described yet, however surgical intervention is recommended as soon as possible once diagnosis is mad to minimize further herniation neural tissue, risk of meningitis and neurologic deficit. Although Open Trans cranial surgery is the most widely practiced approach, currently minimally invasive surgery with endoscopic approach is evolving and fetal surgery for intrauterine repair of encephalocele has also been reported. Outcomes of EC varies widely, but with advanced neurosurgical techniques and pediatric neurointensive care have improved the morbidity and mortality associated with encephalocele. The presence of hydrocephalus, the amount of brain tissue within the sac, and the presence of concurrent anomalies are factors that influence outcomes of children with encephalocele(14)(15)(16)(17)(10)(18)(19).

1.2. Statement of the problem

Encephalocele excision and repair is a commonly performed neurosurgical procedure. However, managing it presents challenges for both neurosurgeons and the families involved. Encephalocele can have significant neurological consequences for children and can place an additional burden on their families. Although there have been global studies on the outcomes of surgically treated children with encephalocele, survival rate at specific time and factors associated with it were not studied widely across the globe, particularly in our region. In resource-limited regions such as sub-Saharan Africa, including Ethiopia, there is a lack of studies and limited knowledge about the overall and operative survival rates of children born with encephalocele. Consequently, it is crucial to address the following questions regarding children born with encephalocele in Ethiopia:

- What is the survival rates of children operated for congenital encephalocele?
- What are the factors that affects survival outcomes
- What are the patterns of encephalocele occurrences

1.3 Justification of the Study

This study will help to narrow the knowledge gap regarding the presentation, complications, and operative outcomes of surgically treated children with congenital encephalocele and identify factors that are affecting neurodevelopmental outcomes. Ultimately, this research intends to improve the standard of care and quality of life of children with encephalocele, establish a foundation for future studies, and the results may also influence policymakers for implementation.

2. Literature review

Encephalocele is a complex congenital malformation of central nervous system characterized by protrusion of cranial contents through skull defect. The presentation is diverse due to variable anatomic location and composite of the sac, though swelling over head is most common presentation. Even though there is variation, worldwide estimated prevalence is 0.8–4 per 10,000 live births (20). Encephalocele may be associated with multiple problems including difficult airway, associated congenital malformations, seizure, hydrocephalus, and hemodynamic disturbance affecting perioperative management (21). Despite advancements in surgical techniques and critical care, optimal management of encephalocele continues to be debated. Surgical repair of encephaloceles is complex and usually requires a multidisciplinary approach. The goals of surgery aims to repair sac and prevent further herniation, reconstruct cranial defect, and minimize neurological deficits, the timing and approach for intervention significantly influence patient outcomes(11)(3)(5).

2.1 Patterns of Encephalocele Occurrence

Encephaloceles exhibit variation in occurrence and patterns among different populations. Numerous studies have reported varying incidence rates and distribution of encephaloceles across different regions worldwide. In countries like the USA, Europe, and Australia, occipital encephaloceles are the most common, whereas frontoethmoidal encephaloceles are frequently observed in Southeast Asia(5)(12). Additionally, a study conducted in Pakistan found occipital encephaloceles to be the most common type, while a similar report from Thailand indicated predominance of occipitoparietal encephaloceles(7)(20). In eastern and western Africa, occipital encephaloceles were reported as the most frequent location, with some ethnic variations observed as demonstrated by a study conducted in Uganda by Benjamin C et al(21). On the other hand Loubna Rifi et al in morocco reported frontoethmoidal encephalocele as the most frequent encephalocele encountered(22). These variations suggest that the occurrence and distribution of encephaloceles might be influenced by both genetic and environmental factors(6)(3).

2.2 Survival outcome of children with Encephalocele

The outcome of encephalocele surgery varies depending on the content and location of the sac, as well as the presence of hydrocephalus and associated malformations(23). In comparison to MMC, encephalocele has a more favorable outcome. The surgical procedure involves the removal and repair of the sac, ensuring a watertight closure of the Dura with or without reconstruction of any bony defects using either autologous bone or allografts such as titanium mesh(17)(14). Recently, there has been a report of a minimally invasive surgical approach using endoscopy for the repair of frontoethmoidal and basal encephaloceles(24).

In a study conducted in North America, a series of encephalocele cases reported good surgical outcomes, with a survival rate of 95% and 48% of patients achieving normal neurological development however, in Spain mortality rate of 36% was reported by Juan F et al (25) . Gyang Markus Bot et al' series of 14 cases of giant encephaloceles operated on in Nigeria demonstrated no in-hospital mortality, and the size of the sac was not found to be associated with survival (26). The largest study in Eastern and Central Africa, conducted in Uganda by Benjamin et al, included 110 children who underwent surgical excision of encephalocele at a pediatric neurosurgical center. The study showed a survival rate of 87% at 1 year, which subsequently decreased to 61% at 5 years(21). The possible causes of death were related to infections and associated anomalies, which were found to be 8 times higher than what was reported in North America and Canada. A report by N. munyi et al in Kenya similarly showed a survival rate of 89%, and half of the patients had comparable physical and mental development to those without encephalocele(27). This difference in terms of survival could possibly be attributed to the quality of service delivery and the setup of the medical facilities, as well as the presence of associated anomalies and the extent of brain tissue within the sac(28).

A prospective cohort study in our pediatric neurosurgery center by Abenezer et al on surgical treatment of 228 children with neural tube defect revealed no mortality at admission ,overall better perioperative outcome comparing to previous retrospective series conducted by Samuel et al at a the same center (29)(30).

Recent advancements in diagnostic and interventional technologies have facilitated the utilization of minimally invasive surgeries employing endoscopes for the treatment of anterior encephalocele and fetal anomalies during the prenatal period. The existing literature indicates that these procedures yield outcomes that are generally comparable to those achieved through open trans cranial surgery, while also offering the advantages of reduced hospitalization duration, lower treatment costs, and improved cosmetic results. Nonetheless, the establishment of long-term follow-up protocols and the implementation of systematic studies are necessary prerequisites for further generalization of these findings. Antonio et al. documented a case of fetal surgery performed to address an isolated occipital encephalocele, which demonstrated no significant postoperative complications and posed no risks to the mother. The authors emphasized the adaptation of the procedural techniques from the MOMS trial, thereby laying a solid foundation for future clinical applications(31)(24).

2.3 Perioperative complication in children with Encephalocele

Various complications have been reported in the literature regarding the repair of encephalocele during the perioperative period. In a study conducted by Ahmed et al, 30 patients with congenital hydrocephalus (HCP) were examined, and seizure disorder was found to be a common perioperative complication. Additionally, the occurrence of seizure was identified as a negative prognostic factor for outcomes, followed by HCP (23). Another series of cases involving patients with occipital encephalocele at a tertiary hospital revealed that HCP was the most frequent

complication (60%) following encephalocele repair, along with wound infection and shunt-related complications(32).

In the published literature from Southeast Asia, a study on 52 children with frontoethmoidal encephalocele who underwent an extra-cranial approach to repair the encephalocele demonstrated a high rate of postoperative cerebrospinal fluid (CSF) leak (rhinorrhea) as compared to the conventional trans cranial approach. However, the incidence of meningitis was rare (5)(33).Complications, such as hydrocephalus, wound-related issues, CSF leak, infection, and seizures, have been reported in different studies. It is crucial to carefully plan and manage the perioperative period in order to minimize these complications and improve overall outcomes(28)(3)(34).

2.4 Hydrocephalus in Children with Encephalocele

Hydrocephalus is a frequent complication in children with encephalocele, affecting up to 70% of cases. This risk is higher with posterior encephalocele compared to anterior. While it can appear soon after birth, hydrocephalus often develops after surgical closure of the encephalocele, significantly impacting both short-term and long-term neurological outcomes. Studies support this association. Stephanie L. et al. found that 24% of children with encephalocele develop hydrocephalus, with about half eventually requiring shunts(4). Similarly, Samuel Berchi et al. reported a 30% overall shunt dependence, with posterior encephalocele showing a higher risk than anterior(10).

Other research explores specific types of encephalocele. A Brazilian study revealed symptomatic hydrocephalus in 50% of children with occipital encephalocele, especially when combined with anomalies like Dandy-Walker malformation or pre-existing ventriculomegaly(35). Notably, the sac size did not influence post-operative hydrocephalus risk. Ehab Ahmed et al. confirmed this, finding no increased risk of hydrocephalus after repairing a large occipital encephalocele, highlighting the importance of encephalocele location(36). Ventriculoperitoneal shunting remains the preferred treatment for hydrocephalus in these cases, although Endoscopic Third Ventriculostomy (ETV) has also been reported in some instances(21)(10)(4)(37).

2.5 Neurodevelopmental outcome of children with Encephalocele

Various degrees of cognitive deficits and physical disabilities have been observed in children with encephalocele. The severity of these impairments depends on factors such as the amount of brain tissue within the sac and the presence of hydrocephalus, among other factors(38). There is a limited amount of worldwide data available on the assessment of cognitive, motor, and developmental milestones in children with encephalocele, and the reports vary. A report from the United States by Benjamin et al. indicated that 48% of the cases assessed according to the diagnostic and statistical manual for developmental disability showed normal development, while 25% had severe developmental delays(25). In a study conducted in India, the neurologic

and cognitive development of children who had undergone surgery for encephalocele was assessed. The study found that 27.8% of the children experienced motor delay, 41.1% had verbal impairments, and 58.8% had cognitive deficits. Another study, which evaluated the developmental progress of 94 children with encephalocele based on CDC developmental milestone guidelines, revealed that 67% had normal development, while 14.9% experienced severe neurodevelopmental delay(10)(39). The presence of hydrocephalus and amount of neural tissue within the sac was identified as an important prognostic factor for neurodevelopmental delay. In a series of patients with encephalocele in Egypt, a relatively higher rate of developmental delay (46%) was reported(40).

3. Objective of the study

3.1. General Objective

- The purpose of this study was to assess the operative survival outcomes of children born with congenital encephalocele at ZMH, a pediatric neurosurgery affiliated center.

3.2 Specific Objective

- To assess the operative survival rates of children with congenital Encephalocele at specific times.
- To determine factors those are affecting operatives survivals.
- To determine the patterns of Encephalocele occurrences
- To determine rate of CSF diversion in children operated for congenital EC.
- To Asses Seizure occurrence in children operated for congenital EC

4. Research methodology

4.1 Study Area and Period

The study was conducted at Zewditu memorial hospital (ZMH) in Addis Ababa Ethiopia, which is AAU Neurosurgery affiliated hospital in the period of September 2023 to July 2024 GC. ZMH is government owned hospital in Addis Ababa, Ethiopia. It has around 180 beds out of which neurosurgical unit has 30 beds and it is primarily a pediatric neurosurgery affiliated center. Neurosurgical Services such as neuro-oncology, pediatric neurosurgery, spine surgery, and emergency operations are done on a daily basis. Totally, 14 Neurosurgeons and 40 neurosurgery residents are working in both hospitals.

4.2. Study design

- This is an institutional based retrospective cohort study (chart review vs cross-sectional) study design of congenital encephalocele who were operated at Zewditu memorial hospital (ZMH) between January, 2018 and June, 2024

4.3 Source and Study of population

4.3.1 Source of Populations

- All pediatric patients who were admitted to ZMH, pediatric neurosurgery center between January 2018 and June 2024.

4.3.2 Study populations

- All pediatric patients with congenital EC who were admitted to and operated at ZMH, pediatric neurosurgery center between January 2018 and June 2024.

4.4 Eligibility Criteria

4.4.1 Inclusion Criteria

- All patients with the diagnosis of congenital encephalocele who were admitted and operated within the specified period

4.4.2 Exclusion Criteria

- Patients with incomplete clinical data were excluded from the study.

4.5 Sampling Technique and Sample size determination

4.5.1 Sampling Technique

- A Simple convenient sampling technique was used among patients that were operated at ZMH from January 2018 to June 2024 GC.

4.5.2 Sample size determination

- Our study population was below the estimated sample size; hence we had included all patients into the study.

4.6 Variables

4.6.1 Independent Variable

- Socio-demographic data ; age, Sex , Birth status
- Clinical data ; presentation ,HC ,Location of sac, Size and Contents of the sac ,Hydrocephalus, type of and urgency of surgical Interventions ,duration of hospital stay
- Other malformations including Intracranial and systemic anomalies**.**

4.6.2 Dependent variables; Operative survival outcomes of Encephalocele

- Operative survival of f Encephalocele occurrence, Final status of the patient

4.6.3 Operational definitions

- Congenital anomalies; Malformation that occur before birth.
- Operative outcome; conditions of patients after surgery as written on chart (alive and died).
- Shunt dependence; operated on patients that developed hydrocephalus and required shunting for survival.

- Giant Encephalocele; an Encephalocele having a size greater or equals to the size of head measurement

4.7 Data collection procedures

4.7.1 Data collection tools

A pretested, structured questionnaire in English was prepared by the principal investigator. All the necessary data was collected by trained data collectors and supervised by the principal investigator. All relevant data were retrospectively obtained from the recorded medical files. Information regarding age, sex, gestational age(birth status), location of EC sac, size of EC sac, content of sac imaging data, presence of preoperative ventriculomegaly, symptomatic hydrocephalus, perioperative findings, associated CNS and systemic extra cranial malformations, shunt dependency, seizure, and final status as alive or dead were recorded.

The status of the EC was described as Intact if there was no skin decoration, ruptured if CSF was leak from the sac, and ulcerated if there was skin sloughed but no CSF leak from the sac. Contents of the sac of the EC were confirmed by detailed imaging report. The EC sac was categorized into two groups: posterior and anterior. The posterior lesions included, occipitocervical, occipital ECs, parietal and, whereas the anterior group included sincipital (nasofrontal, frontoethimoidal) and basal,).objective neurologic findings were obtained in all patients and motor status noted as deficit if strength of extremities was recorded below 5 and intact if it was 5. Perioperative complication, duration of hospital stay, shunt dependency for HCP, duration of follow up, seizure occurrence, and survival time or final status (alive or died) in the documented in patients' medical files was collected.

Brain imaging including brain CT scan, brain MRI or Transfonatanel-Ultrasound (TF-US) was performed preoperatively to characterize the sac, rule out associated CNS anomalies and ventriculomegally. If Ventriculomegaly was diagnosed according to the Reports of patients' imaging results extracted from their files the patients. If hydrocephalus was symptomatic CSF diversion/VPS was considered or had been routinely followed up in the referral clinic for asymptomatic ones. Information related to death was obtained from hospital`s medical record files. For cases of encephalocele with no death record, survival was censored at the end of the follow-up period (June 30, 2024).

4.7.2 Data handling and quality control

The collected data were entered, cleaned, stored, and checked for its completeness and internal consistency. Every day, filled-out questionnaires were checked for completeness by both the data collector and supervisors. Further rechecking was done before data entry**.**

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4.7.3 Data processing and analysis

Data coding, cleaning, verification, and editing was done before entry into SPSS. Collected data entered, and statistical analyses were performed using Statistical Package for Social Science version 25 (IBM Corporation). The participant's dependent and non-dependent variables were presented using the relevant descriptive statistics. Continuous variables were presented as the median (IQR) or Mean-SDV Categorical variables were described as frequency and constituent ratio).The survival probability was estimated for all cases of encephalocele, from time of surgery to 1 month, 12 month, and 24 month, 4 years of age and during the overall study period using the Kaplan–Meier method. We also estimated Kaplan–Meier survival curves for possible prognostic factors, including birth status, gender, location and content of sac, neurologic deficit, and presence of associated congenital anomalies, post op infection, hydrocephalus and duration of hospital stay. Survival Analysis with life table performed to determine the probability of survivals of children with congenital Encephalocele at specific time in month. Kamlan-Mieir curve analysis was done to estimate graphic survival function and compare difference in survival between two variables. The log-rank test to examine the variation in survival by time of surgery and possible prognostic factors. We estimated hazard ratios and 95% CIs in relation to possible prognostic factors taking into account other covariates by means of the Cox proportional hazards model (Cox regression model).

5. Ethical Clearance

Ethical clearance for the study was obtained from Addis Ababa University's College of Health Sciences Ethical Committee, and the study was conducted after getting an official support letter from the department of surgery and neurosurgery unit at each hospital. The participants' names were not being mentioned, and all relevant information was kept confidential. The investigator was responsible for ensuring this by keeping all documents discreet.

6. Results

Between the period of January 2018 and July 2024,142 cases were identified from Zewditu memorial hospital pediatric neurosurgery center patient`s medical file data base. Sixteen among the 142 patients were excluded due to incomplete clinical data. The characteristics of the encephalocele were ascertained from Imaging and clinical findings.

6.1 Demographic characteristics

From a total of 126 cases, 51 (40.5%) were males and 75 (59.5%) were females. At presentation, the mean age in days was 130 +- 59. Based on the region of residence, Most of were from Addis Ababa city (38.9%) and Oromia region (38.1%).

Table 1 Demographic characteristic findings in patient with EC, ZMH

Variables	Frequency	Percentage
Patient's age group		
<1 month	82	65.1%
1-12 months	35	27.8%
>1 year	9	7.1%
Gender		
Male	51	40.5%
Female	75	59.5%
Residency region		
Addis Ababa	49	38.9%
Oromia	48	38.1%
Amhara	20	15.9%
SNNPR	6	4.8%
Afar	2	1.6%
Benshangul Gumuz	1	0.8%

6.2 Perinatal and Maternal findings

The median age of the mothers was 27, ranging from 19 to 44 years. Ninety-two (73%) mothers had ANC followup, while 34 (27%) had no clinic visit during pregnancy. Ninety (71.4%) pregnant mothers were supplemented with folic acid, whereas 36 (28.6%) were not supplemented with folic acid. Prenatal diagnoses of congenital encephalocele were not mad in 105(83.3%) mothers. Majority of the patients (90.5%) were term at delivery.

Table 2 Perinatal findings of patients with Congenital EC, ZMH

Variables	Frequency	Percentage
Mother's age group		
18-25	46	36.5%
26-30	44	34.9%
31-35	23	18.3%
36-40	10	7.9%
41-45	3	2.4%
18-25	46	36.5%
ANC follow up		
Yes	92	73%
No	34	27%
Folic Acid Supplementation		
Yes	90	71.4%
No	36	28.6%
Was The diagnosis made Prenatal		
Yes	21	16.7%
No	105	83.3%
Mode of delivery		
SVD	120	95.2%
CS	6	4.8%
Birth Status		
Preterm	114	90.5%
Term	12	9.5%
Place of delivery		
Health Care Facility	119	94.4%
Home	7	5.6%
Maternal Comorbidity		
No Comorbidity	123	97.6%
HTN	1	0.8%
DM	1	0.8%
HTN and DM	1	0.8%

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6.3 Preoperative clinical characteristics

All patients presented with craniofacialswelling. Posterior encephalocele was the most common type accounting (87.3%). Among these, 67.5% were occipital encephalocele.
At presentation, the sac of encephalocele was intact in 93 patients (73.8%). The sac contained CSF only in 84 patients (66.7%) and dysplastic/neuronal tissue in 42 patients (33.3%).The median size of the sac was 7 cm, ranging from 2 to 20 cm.At admission, 111 patients (88.1%) had a Glasgow Coma S cale (GCS) score of 15, and 15 patients (11.9%) had a GCS score of 13-14.
Motor strength of the extremities was intact in majority of the patient's patients 98 (78.8%). Diagnosis of associated congenital anomaly was mad in 42(33.3%). Corpus callosum agenesis 12(33.3%) was most frequent malformation followed by Chiari Type-II Malformation10 (27.8%).

Table 3 Clinical characteristic findings in patient with congenital EC, ZMH

Variables	Frequency	Percentage
Type of Encephalocele		
Posterior	110	87.3%
Anterior	16	12.7%
Size of Encephalocele in centimeter		
1-5	41	32.5%
6-10	54	42.9%
11-15	23	18.3%
16-20	8	6.3%
Status of Encephalocele sac		
Intact	93	73.8%
Ruptured	9	7.1%
Ulcerated	24	19.0%
Content of Encephalocele		
Neural tissue Absent	84	66.7%
Neural tissue present	42	33.3%
Preoperative Ventriculomegaly		
Yes	39	31.0%
No	87	69.0%
Preoperative motor Strength		
Intact (Grade 5)	98	77.8%
Incomplete(1-4)	28	22.2%
Congenital Anomaly		
Yes	40	31.7%
No	86	68.3%

Table 4 Congenital anomalies associated with EC, ZMH

Variables	Frequency	Percentage
CNS Congenital Anomalies (n-36)		
Chiari II malformation	10	27.8%
Chiari III malformation	1	2.7%
Corpus callosum agenesis	12	33.3%
DWM	5	13.9%
MMC	2	5.6%
Arachnoid Cyst	2	5.6%
*Others	4	11.1%
Systemic Congenital Anomalies (n-8)		
ASD	3	37.5%
VSD	3	37.5%
TOF	1	12.5%
Cleft palate	1	12.5%

***Others; dermoid cyst, lipoma, Holoprosencephaly, porencephalic cyst**

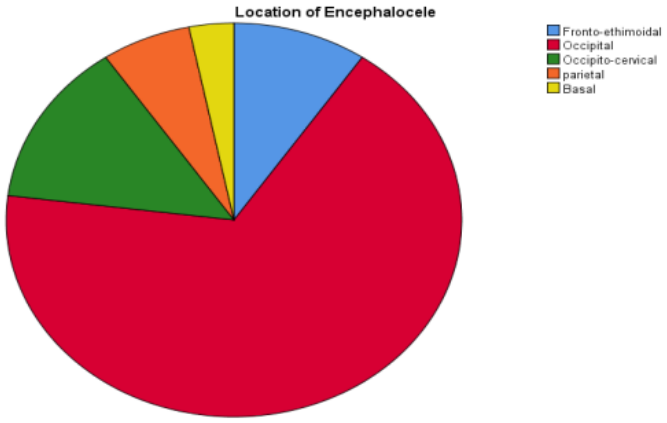

Figure 1 Encephalocele distribution by location, ZMH

6.4 Operative and post-Operative findings

The surgical approach was open trans-cranial in 124 cases (98.4%) and a combined trans-cranial with endoscopic approach in 2 cases (1.6%). This involved excision of the sac with or without resection of dysplastic neuronal tissue, followed by watertight dural closure and multilayer closing of the scalp. Most patients were operated on an elective basis were 121 (96%). Twenty-three patients (18.3%) developed post-operative infections. Among these, CNS focus was most common12 (9.5%).There was reoperation during the same admission in 15 cases (11.9%). The median duration of hospital stay was 4 days, ranging from 3 to 58 days.

Table 5 Operative and post-operative findings in patient with EC, ZMH

Variables	Frequency	Percentage
Type of Surgery		
Excision and repair	124	98.4%
Repair and bony defect reconstruction	2	1.6%
Type of Surgical Approach		
Open trans-cranial	125	99.2%
Combined Trans cranial and Endoscopic	1	0.8%
Occurrence of Post op infection		
Yes	23	18.3%
No	103	81.7%
Focus of post op infection(n-)		
CNS	13	52%
Wound	6	24%
Chest	6	24%
Post op CSF leak		
Yes	10	7.9%
No	116	92.1%
Reoperation on the same admission		
Yes	15	11.9%
No	111	88.1%
Reason for reoperation (n-15)		
Hydrocephalus	6	40%
CNS infection/Ventriculitis	4	26.7%
CSF leak	3	20%
Wound failure	2	13.3%

6.5 Follow up and operative survival outcomes

The median follow-up time in months was 27, ranging from 1 to 80 months. Overall, 15(11.8%) patients died and 111(88.1%) patients survived. Of those who died, 2 were passed away before discharge due to sepsis and surgery-related complication, and the rest (13 patients) died later due CNS or chest focus infection, and cardiorespiratory arrest related to congenital malformations. Seizures developed in 23 (18.3%). Patients operated for Encephalocele out of these, 2 patients had seizure preoperatively. Overall, 32 (25.4%) patient's required CSF diversion for symptomatic hydrocephalus. Ventriculoperitoneal shunt (VPS) was inserted for 28(87.5%) patients and ETV for 4 patients (12.5%). There were VPS revisions for malfunction or infection in 4 (3.2%) patients.

Table 6 Overall outcomes of patients with EC, ZMH

Variables	Frequency	Percentage
Final status of the patient		
Alive	111	88.1%
Died	15	11.9%
Follow up CSF diversion		
Yes	32	25.4%
No	94	74.6%
Type of CSF diversion(n-32)		
VPS	28	87.5%
ETV	4	12.5%
Seizure occurrence		
Yes	23	18.3%
No	103	81.7%

The survival probability of children operated for congenital Encephalocele was 75% at 6.5 years. Independent categorical variables were analyzed with Kaplan Meier log-rank test to compare differences in survival and possibly predict their association with outcomes. There were no significant differences in survival between males and females. Preterm deliveries were possibly associated with higher relative risk for survival. Neural tissue absent within the sac had better survival compared to a sac contained neuronal tissue within. Presence of congenital anomalies along with encephalocele showed decreased survival compared to these that had no anomalies. There was difference in survival among locations of encephalocele. Patients who had post-operative infection had higher hazard, however it was not statically significant. Preoperative motor strength deficit showed lower survival rate compared to no deficit. Cox proportional hazard model analysis (cox regression model) revealed significantly lower survival for children with congenital encephalocele who had their sac contained neuronal tissue, had associated congenital anomalies and occipito-cervical location of the sac.

Table 7 Cumulative survival of patients with EC at specific time in month, ZMH

Time in months	Survival probability (%)
At surgery	100
1 month	98
6 month	97
1 year	95
2 year	92
4 year	85
6.5 year	75

.

Table 8 Survival of children with EC by selected demographic and clinical characteristics, ZMH Sept 2024-Oct 2024

Characteristics	TotalNumbers	No.death (Mortality)	Overall survival (%) [95% CI]	Log-rank test of overall survival ; P-value
Gender				
Male	51	6	88.2 (60.573, 77.435)	0.828
Female	75	9	88 (51.882, 61.632)	
Birth status				
Term	114	12	89.5(64.425, 75.272)	0.062
Preterm	12	3	75(26.390, 43.295)	
Content of EC				
Abscent	84	1	98.8(77.132, 80.922)	0.001
Present	42	14	66.7(37.354, 59.349)	
Type of EC				
Posterior	109	13	88.1(62.264, 74.253)	0.902
Anterior	17	2	88.2(47.293, 66.535)	
Preoperative ventriculomegally				
Yes	39	8	79.5(52.587,72.475)	o.148
No	87	7	92.0(55.750,63.420)	
Presence of anomalies				
Yes	40	14	66(37.264,59.339)	0.001
No	86	1	98.8(77.169,80.910)	
Preoperative motor strength				
Intact(grad-5)	98	7	92.9(69.349,78.216)	0.011
Incpmpelete(grade 1-4)	28	8	71.4(43.117,66.664)	

Table 9 cox proportional hazard analysis of selected Bivariate, ZMH

Variables	p-value	Exp(HR)	95%CI Lower, upper
Age of patient in days	.474	.999	(0.995, 1.002)
Location of Encephalocele	.851	1.044	(0.665, 1.639)
Size of Encephalocele in cm	.069	1.091	(0.993, 1.199)
Occurrences of post op Infection	.130	0.436	(0.149, 1.278)
Reoperation at same admission	.077	0.354	(0.112, 1.117)
Duaration of hospital stay in days	.058	1.088	(1.049, 1.128)

Table 10 Cox proportional hazard analysis in patients with EC with selected Covariates, ZMH

Independent Variables	P-Value (sign)	Exp(B)	95% CI for Exp(B) Lower, upper bond
Birth status	.308	2.189	(.485, 9.883)
Location of Encephalocele	.890	1.070	(.412, 2.774)
Size of Encephalocele in CM	.477	.958	(.850, 1.079)
Content of Encephalocele	**.010**	0.061	(0.007, 0.514)
Presence of preoperative Ventriculomegally	.340	2.370	(.403, 13.921)
Preoperative motor Strength C	.795	1.256	(.226, 6.995)
Presence of congenital anomalies	**.006**	.049	(.006, .430)

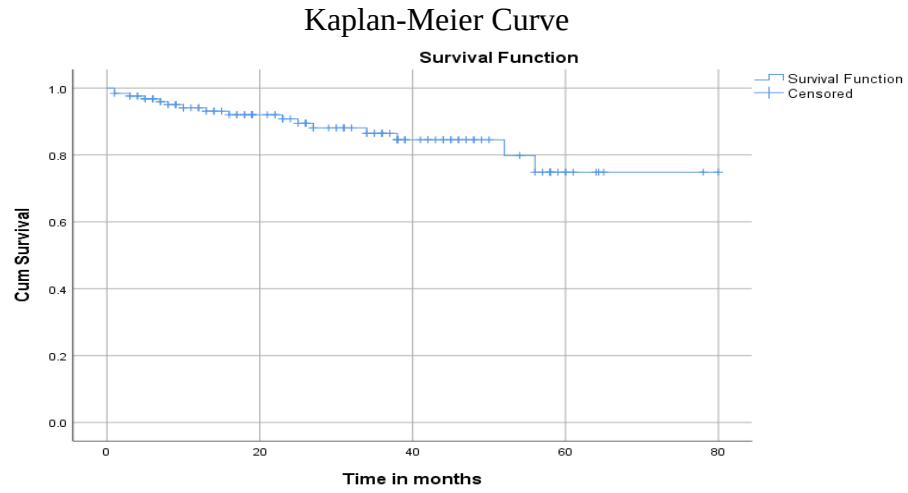

Figure 2 Survival curve of patients with Encephalocele, ZMH

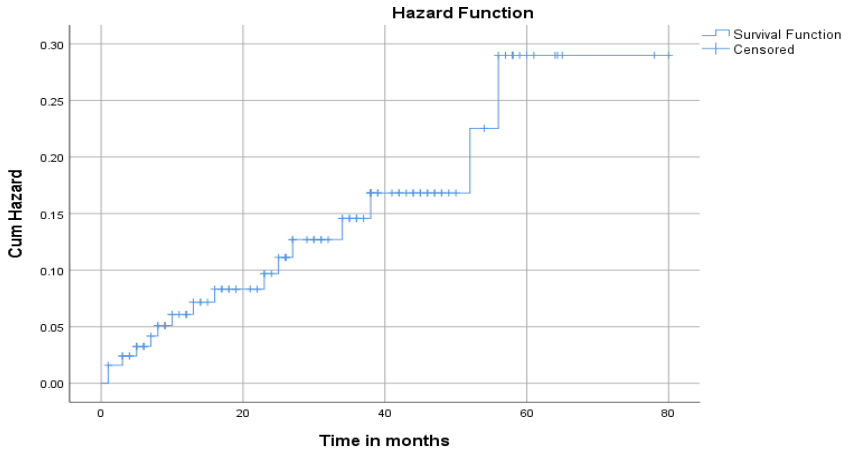

Figure 3 Hazard curve of children with Encephalocele, ZMH

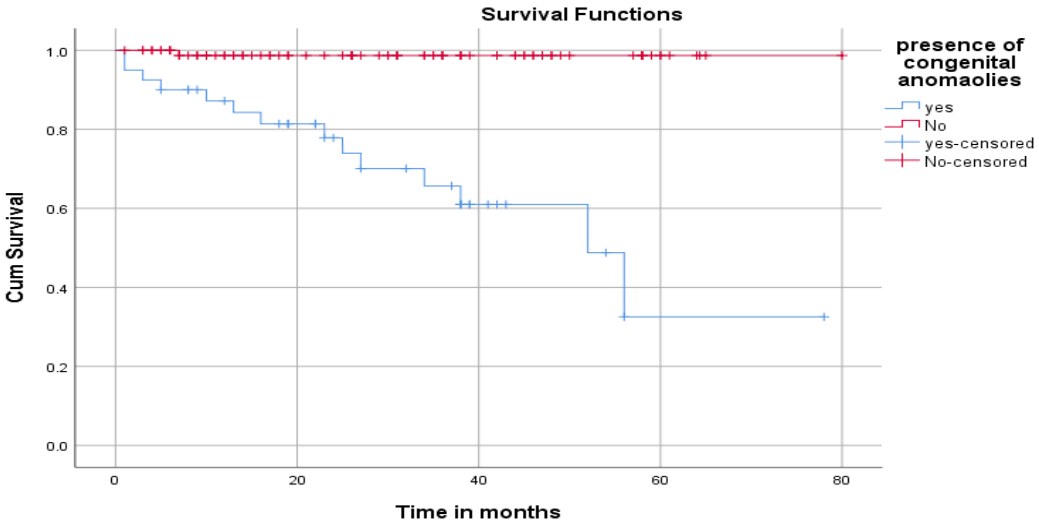

Figure 4 Survival of children with EC by presence of congenital Anomalies, ZMH

7. Discussion

Encephalocele is a complex congenital malformation of central nervous system characterized by protrusion of cranial contents through skull defect. The precise pathophysiologic mechanism of encephalocele is not fully understood, however it is hypothesized to be the result of a defect in the separation of the ectoderm and neuroectoderm during the embryonic stage of brain development, particularly between the 5th and 9th weeks of gestation(8)(4)(28). Encephalocele excision and repair is a commonly performed neurosurgical procedure. The surgery is aimed to minimize further herniation of neural tissue, risk of meningitis and neurologic deficit .The outcome of encephalocele depends on certain factors like content and location of the sac, as well as the presence of hydrocephalus and associated anomalies(41) (5)(42). However, deference still exists in survival rate among children operated for congenital Encephalocele across a regions(23).

7.1 Survival outcomes of children with Encephalocele

Previous study showed different survival rate for children with congenital encephalocele. A cohort in Australia showed that, the survival probability of children with Encephalocele at 1-month was 65% and at 6 years-year was 54.5%(43) .Other study in USA, reported that the survival probability was ranging from 77.1% at age of 1 day to 67% at 20 years(44). Other series of patients' in eastern Africa showed survival probabilities of 87% at 1 year, and 61% at 5 years(21). In our study, The survival probability of children operated for congenital Encephalocele was 98% at 1 month, 92% at 2 years, 85% at 4 year and 75% at 6.5 years with mean survival time of 65.5 months (95% CI: 63.102, 73.996).The relatively higher survival probability in our cohort might be related to that fact that most cases had short follow up time compared to others.

The overall outcome of children with encephalocele was better than these who had spinal dysraphism as it was reported in literature. A study conducted in north America had reported survival rate of 95% for children with encephalocele however, another series of patients with encephalocele in Spain showed mortality rate of 36% compared to 84% to 50% in Australia (25)(16). Previous report in east Africa revealed that there was 40% mortality rate at 5 year follow up(21). In contrast to these results, a cohort in India reported no mortality in 27 patients operated for congenital Encephalocele at mean follow up of 7.2 year (Anil). In our series the overall survival was 88.1% at 6.5 follow up (mortality of 11.9%). out of these 2 patient died at

the same admission due to surgical elated complication and sepsis, and 13 of them they died later during of follow period. The cause of death was related to congenital malformations, sepsis, and cardiorespiratory arrest. Our results are compatible to other reports in literature. The difference in outcomes might be explained by degree of brain dysplasia and presence of associated anomalies. A prospective cohort study in our pediatric neurosurgery center on surgical treatment of 228 children with neural tube defect reported overall better outcome comparing to previous retrospective series conducted at a the same center (29)(30).

7.2 Survival outcomes and demographic characteristics

Most literatures had reported female predominance in Encephalocele (21)(17)(20). In our study, 75 (59.5%) were females and 51 (40.5%) males. We observed that the mean age at presentation was 130 days, ranging from 2 days to 15 years. A previous study has shown age at surgery ranging from 2 month to 14 years, compared to another study which reported age range from 3 days to 36 months(3)(17). Studies have reported female gender to be associated with lower survival rate comparing males(10)(21). In our series, females had lower probability of survival compared to males, but it was not statistically significant.

The optimal age for surgery in children with encephalocele was not yet determined. Most literatures advocate operative repair of encephalocele at the time of diagnosis made so as to minimize CNS infection, while other author author still recommended delaying surgical intervention beyond the neonate period to allow gaining weight to tolerate surgery and anesthetic related complications (3)(41).In our study, 68.3% patients presented within the age of 1 month, 27 (21.4%) within 1-12 months and 13 (10.3%) patients were above the age of 1 year. A study in Kenya reported age at presentation ranged from one day to 15 years(27).In previous cohort majority of patients were operated at their neonatal period which was compatible with our results(35).We observed that, age at surgery did not affect operative survival of patients with congenital encephalocele.

Literatures report most patients with congenital Encephalocele were term at delivery. A cohort of 102 patient with encephalocele showed 95(94.7%) of patients were term at delivery compared to our study which revealed 114 (90.5%) term, and 12 (9.5%) preterm. Other study reported 84.7% of patients with encephalocele were term at delivery. Operative outcomes of preterm infants with Encephalocele have shown increased hazard(44).In our study, birth status (gestational age at birth) was associated with possible worse operative survival; however it was not statistically significant on cox proportional hazard model of analysis. Life table of children operated for congenital Encephalocele at specific time in months

7.3 Survival outcome and Encephalocele characteristics

The occurrence of encephaloceles varies geographically, with occipital encephaloceles (66%–95%) being more common in the USA, Europe, and the Middle East, while sincipital encephaloceles are more prevalent in Southeast Asia (6)(7). In eastern and western Africa, occipital encephaloceles were reported as the most frequent location. In other study, occipital region were predominant (66.7%) , occipitocervical (20%), parietal(6.7%) , frontoethmoidal(3.33%) and (3.33%) Basal (21).Other study in Egypt showed 90% of encephalocele were in the occipital region, (3.33%) parietal, and (13.33%) were in the frontal region(23). According to our study, occipital encephalocele was the most common accounted for 69.8%, followed by occipito-cervical in 11.1% , fronto-ethmoidal in 12(9.5%), parietal 8.7% and basal in 3.2%. Another study have shown occipital (48%) most common site followed by sincipital (30%), and parietal le (22%)(21). On the other hand a cohort in morocco reported frontoethmoidal encephalocele as the most frequent encephalocele encountered(22). These variations suggest that the occurrence and distribution of encephaloceles might be influenced by ethnic, genetic and environmental factors(6)(3). Site or location of the Encephalocele has been associated with outcomes(10). In our study occipital-cervical location was associated with increased hazard, however on cox regression model it was statistically significant (P=0.89).
At presentation, the sac of encephalocele was intact in 93 patients (73.8%) ulcerated in 24 (19%), and ruptured in 9 (7.1%). A cohort in Pakistan reported 18% of patients were admitted of sac rupture with cerebrospinal fluid (CSF) leakage(20). Other study showed 17.6% of children had CSF leakage at admission(17). In our study, sac rupture at presentation did not show effect on survival.

Suwanwela -Suwanwela reported size of the enceohalocele from 2cm to a size greater than a child`s head circumference(9). A study in USA showed that the size of the encephalocele ranged from 0.5 to 23 cm in maximal diameter, with a median size of 3 cm(4). Another report from India showed size of Encephalocele sac as large as 58 cm(32). In our study
the mean size of the Encephalocele was 7.95 cm, ranged from 2 to 20 cm. The mean head circumference was 36.1 cm (range 31 - 42) in reports. Our finding is almost similar to others study. Size of the sac was reported to be associated with worse outcomes in some and no association in others(32). However, size of encephalocele did not show significant effect on operative as reported previous cohort(36). In our study, the size of the sac had no effect on operative survivals.

The sac contained neuronal tissue in 67%, while in 23 (33%), neural tissue was not present(4).Other study revealed that, 42.2% of the Encephalocele had contained neural tissue(10). In another study in Egypt, neural tissue was presented in 63.33% of patients(23). In our study, we observed that the sac contained neuronal tissue in 42 patients (33.3%), whereas neuronal tissue was abscent in 66.7% of patients. A study in Iran showed that 42.2% of patient with encephalocele had neuronal tissue within the sac. Comparing to others, our cohort showed less proportion of sac with neuronal neuronal tissue which could be related to the fact that the types of imaging modalities we had at the time of preoperative evaluation. In many literatures, content of the sac was correlated with operative outcomes(23)(10)(43)(44).In our study , cox

proportional hazard model had demonstrated statistically significant association between presences of neuronal tissue within the sac of encephalocele and operative survival.

7.4 Survival outcomes and presence of associated congenital malformations

Previous study showed that congenital anomalies can accompany encephalocele in 60% of patients(42). Another study reported, 43 patients (61%) had associated cranial anomalies (22 with only cranial anomalies and 21 with both cranial and extra cranial anomalies), 7 patients (10%) had only extra cranial anomalies. Other study in Egypt revealed that among cranial anomalies, 6 (20.0%)Of patients had corpus callosum agenesis, 2 (6.67%) of patients had Dandy–Walker malformation and 2 (6.67%) of patients had preoperative hydrocephalous, and among extra cranial anomalies 2 (6.67%) of patients had cleft lip, 2 (6.67%) of patients had Klippel Feil syndrome, 1 (3.33%) had microtia, 1 (3.33%) of patients had polycystic kidney and 2 (6.67) of patients had cardiac anomalies(23). In our study, diagnosis of congenital anomaly was mad in 40(31.7%).Thirty-six patients had associated CNC anomalies including; 12(9.5%) corpus callosum agenesis, 10(7.9%) Chiari Type-II Malformation, 5(4%) Dandy walker malformation, 2(1. Chiari-Type 3 malformation, 2 arachnoid cyst,2 Menigiomyelecele(MMC) and 4(3.2% others (dermoid cyst,lipoma,Holoprosencephaly,porencephalic cyst), and 8(6.3%) patients had Systemic anomalies including 7(5.6%) cardiac anomalies, and 1 cleft palate. Previous study has shown poor survival in patients with associating congenital anomalies(45)(28)(46)(44).In our, cox proportional hazard model showed statistically significant correlation between presence of congenital anomalies and survival rate($p<0.05$).

7.5 Survival outcome and Preoperative motor deficit and ventriculomegaly

The severity of neurological abnormality depends on the degree of cerebral or cerebellar dysplasi a associated with the encephalocele or other intracranial congenital anomalies. Approximately tw o-thirds of the patients with encephalocele showed little or no disability (1).
In our study, 88.6% ofhad intact motor strength, whereas 21.4% of the patients had some degree of incomplete motor deficit. According to our study, preoperative motor deficit was associated with worse operative survival; however, it was not statistically significant ($p = 0.795$).
Previous literature reviewed hydrocephalus as a frequent complication in children with encephal ocele, with varied incidences ranging from 21% to 70% of cases(40)(3). Another series of patien ts reported that 35 patients (32%) required treatment for hydrocephalus, out of which 2 patients were treated before encephalocele repair(21). In our study, 39 (31%) patients had preoperative ve ntriculomegaly, out of which only 1 was symptomatic and required VPS placement before encep halocele repair. Overall, 31 patients (24.6%) required CSF diversion for symptomatic hydrocep halus.The presence of hydrocephalus was described as significantly related to overall morbidities and mortalities. In our study, the presence of preoperative ventriculomegaly was not statistically significant in operative survival.

7.6 Survival outcome and Perioperative complications

Various complications have been reported in the literatures including hydrocephalus, wound-related issues, CSF leak, infection, and seizures, have been reported in different studies (28)(3)(34). A series of patients in India reported post-operative complications in 20% of patients operated on for encephalocele, with CSF leak being the most frequent postoperative complication, recorded in 9.3% of patients(3). In a series of 59 patients in Brazil, CSF leak and surgical wound infection were reported in 11.8%(35). Another study showed 2 (6.67%) patients had wound infections, 3 (10.0%) patients had CSF leak from the wound, 2 (6.67%) patients had sac rupture with CSF leak, 5 (16.67%) patients had postoperative hydrocephalus, and 6 (20%) had seizures ,India. In our cohort, twenty-three patients (18.3%) developed post-operative infections, including 12 (9.5%) with a CNS focus, 6 (4.8%) with a chest focus, and 5 (4%) with a wound site focus. We observed almost comparable complication with prior results. In our study, the occurrence of post-operative infections was associated with worse operative survival. However, on the Cox proportional hazard model, post-operative infections were not statistically significant in the operative survival outcomes. Duration of hospital stay varies in different literatures. Prior study demonstrated the average duration of hospital stay in range of 1 to 35 days, with median of 7 days(28). The median duration of hospital stay was 4 days, ranging from 3 to 58 days. In our study, seizures developed in 18.3% patients operated for Encephalocele out of these, 2 patients had seizure preoperatively at presentation. In another cohort, seizure was noted in 20% of patients operated for encephalocele(23).The occurrence of seizure in our study was almost comparable to reports in literature.

8. Conclusion

Our study demonstrated that surgery for congenital encephalocele is a common practice in our institution. The presence of neuronal tissue within the sac associated congenital malformations, were factors statistically correlated with worse operative survival outcomes. Despite the challenges, our study's overview of the survival outcomes of children operated on for congenital encephalocele was almost comparable to other results. We believe that our finding can serve as a foundation for future prospective and long-term studies. This may also help decision-makers and caregivers to plan for subsequent follow-up and long-term care in patients with congenital Encephalocele.

9. Limitations of the study

Our study was an institution-based retrospective chart review, which can be prone to bias. It was also conducted in a single center. There were also inconveniences with chart-keeping and a lack of complete clinical evaluation and recording of relevant information. As a result, we were not able to assess functional outcomes in all patients. Some of our patients also had a short follow-up period, which could affect our analysis and results.

10. Recommendations

For the future it is important to improve patient's medical files keeping and relevant clinical data should be documented properly. Families should also be encouraged to have regular and longer follow-up so as to pick problems. For the future, we also recommend conducting a multi-center prospective and long-term study on encephalocele with particularly emphasis on functional outcomes.

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Annex I. Questionnaire

1. Socio demographic Data

1.1 Card no_____ 1.2 sex _____1.3 age (in days)_____

1.4 Phone Address_____ 1.5 Region of Residence; _____

1.6 Hospital = ZMH

2. Maternal and Perinatal History

2.1. What is the age of mother?;____ 2.2. did she has ANC follow up? a. yes b. no

2.3 If yes for the above question, was she supplemented with folic acid? a. yes, b. no

2.4. Was the diagnosis of encephalocele mad prenatally? a. yes b. no

2.5 What was the mode of delivery? a . SVD b. CS

2.6. What was the Birth status? a . term b preterm

2.7. Where was the Place of delivery? a.health care facilty b. home delivery

2.8. Did the mother have known comorbidity? A. DM b. HTN c. Epilepsy, d. Others specify;__

2.9. had the mother been taking any drugs/medication? A . yes b. no

2.10. if yes ; specify it;_____

3. Preoperative Clinical Data

3.1 Patient's symptoms at presentation?

a. craniofacial swelling , b. Nasal cavity swelling, c. discharge from the swelling

3.2 What associated symptoms the patient had?

a. vomiting b. respiratory distress c. fever d. failure/ decreased breast feeding e. seizure

3.3 was there derangement in vital signs ? a. yes , b. no 3.4. if yes for the above , mention it;____

3.5. Head circumference (in cm)?;__ 3.6. Type of Encephalocele? A. anterior, b. Posterior

3.7. Lactation of encephalocele; ____ 3.8. Size of EC sac in cm;____

3.9. Content of EC sac; a. CSF only b. CSF with neural tissue

3.10. Status of EC sac; a. intact b. ulcerated c. ruptured

3.11. Associated ventriculomegally? A. yes b. no

3.12. If yes for the above, did the patient require CSF diversion before surgery? A. yes , b. no

3.13. what type of head Imaging the patient Had? A. CT , b. MRI C.TF-US d. no image

3.14. GCS of the patient? ; ____

3.15. Pupillary response to the light? A. symmetrically reactive, b. asymmetrically reactive

3.16. dis the patient follow light? a. yes, b. no

3.17. Strength of extremity?;_____/5

3.18. Tone of extremity? A. norm tonic, b. hypertonic, c. hypertonic

3.19. Presence of Associated Congenital anomalies

3.20. Associated CNS malformations, mention it; _____

3.21. Associated systemic congenital malformations, mention it; _____

3.22. Derangement in baseline investigations; a. Yes, b. No

3.24. If yes for the above question specify it. _____

4. Operative Data

4. 1. Duration of surgery (in minutes); ____

4.2. Surgery performed.

a. Sac excision and repair

b. Sac excision and repair with bone defect reconstruction

4.3. Type of surgical approach?

a. Open Trans cranial b. Endoscopic c. Combined

4.4. Was VPS performed at same surgery? A. Yes , b. No

4.5. Was there any Intra-op Incident? A. Yes, b. No

4.6. If yes for above question specify it; _____

4.7 Urgency of the surgery? A. Elective b. Emergency.

4.8. Post op patient disposal? a. NICU, b. Ward

5. Post-Operative Data before Discharge

5.1. Did the patient develop post op infection? A. Yes, b.No

5.2. If yes, what is the focus of infection? A. CNS, chest, c. GUT, d. wound e. others epecify;___

5.3. Did the patient develop post op CSF leak? A. Ye, b. No

5.4. Did the patient develop Post repair HCP requiring CSF diversion? A. Yes, b. No

5.5. If yes for the above, what type of CSF diversion was done?

a. VPS b. ETV c. EVD

5.6. If EVD was inserted, what was the indication?; ____

5.7. Was there reoperation before discharge? A. Yes, b. No

5.8. If the response is yes, what was the cause of reoperation? Mention;___

5.9. Condition of the patient at discharge?

a. improved, b. same, c. worsened , d. died

5.10. If died, what was the cause of death? Mention; _____

5.11. Duration of hospital stay?(in days); _____

6. Follow up Data

6.1. Is the patient Alive? A .Yes, b. No

6.2 if not, what is the cause of death? Mention; ___

6.3 If alive, what is condition of the patient?

a. Improved b. same c. Worsened

6.4. Duration of patient's t follow up (in months) ;____

6.5. Did the patient required CSF diversion on follow up? a. Yes, b. No

6.6. If the response is yes, what type of CSF diversion was done?

a. VPS b. ETV

6.7 Did the patient develop seizure? A. yes, b. no