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UNDERSTANDING THE DEMOGRAPHIC AND CLINICOPATHOLOGIC CHARACTERISTICS OF ENCEPHALOCELE AT A TERTIARY INSTITUTION
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M. Magoha, S. Apondi and S. Alinoor

ABSTRACT

***Background*: Encephaloceles are neural tube anomalies marked by either an incomplete formation of skull bone or a failure in the fusion of the skull, leading to the protrusion of intracranial structures. These anomalies are classified according to their specific locations, which may include occipital, parietal, sincipital, or basal regions.**

Risk factors include genetic predisposition, ethnicity, infections, folate deficiency, and exposure to environmental contaminants. Detecting this abnormality can be done through medical imaging or a physical examination.

***Objective*: To document the clinical pattern, management, and demographic characteristics of patients with encephaloceles in Kenya between the years 2015 to 2021.**

***Methods*: In this retrospective cross-sectional study, we analysed 27 patient files with confirmed diagnoses of encephaloceles. We obtained the files from the records department at Kenyatta National Hospital.**

***Results*: The mean age of participants was 2.8 years with the range being from 0-6 years. 44% of these patients were female while 56% were male. 70% of the patients had occipital encephaloceles, 26% had frontal encephaloceles, 2% had sincipital encephaloceles while 2% were uncategorized.**

***Conclusion*: Encephalocele affects children in Kenya. With Males being affected 1.3 times more than females. Occipital encephaloceles had the highest proportion at 70% which is in line with international studies. All the patients were diagnosed at birth as opposed to prenatal diagnosis which is the standard, which would greatly enhance patient outcomes and quality of life.**

INTRODUCTION

Encephaloceles are rare neural tube defects (NTDs) characterized by partial skull bone absence or fusion failure, allowing brain matter, meninges, and cerebrospinal fluid to protrude[1]. They are categorized as frontal or occipital encephaloceles[2], with the majority (75%) located in the occipital region. These can be further classified as meningoceles, meningoencephaloceles, or meningo-encephalo-cytoceles based on their contents.

The global prevalence of encephaloceles is between 0.8 to 3.0 out of 10,000 live births, placing encephaloceles as a rare disease. In Africa, as of 2020, the prevalence of encephaloceles has been reported as 2.0 out of 10,000 live births following a systematic review and meta-analysis. In Kenya, few studies have been conducted to determine the prevalence of NTDs and one study by Githuku et al found that between 2005 and 2010 only 7% of surgical records at KNH were encephaloceles[3].

Encephaloceles can occur as isolated conditions or coexist with serious disorders, such as Meckel Gruber syndrome, which features multiple abnormalities. These defects may be associated with genetics, ethnicity, infections (e.g., TORCH), environmental factors (e.g., organic mercury), and maternal dietary deficiencies (folate and B12 vitamins)[4–6].

Folic acid supplementation has effectively reduced NTDs in developed countries. In contrast, Middle-Income countries like Kenya face a higher encephalocele prevalence due to dietary deficiencies, leading to the introduction of folic acid supplementation in 2012 in accordance with WHO recommendations[7].

Diagnosis involves prenatal ultrasound and postnatal clinical examination, with MRI for additional assessment. Treatment for encephalocele involves a multidisciplinary team. Surgery achieves dural closure, sac resection, skull defect repair and reconstruction often using autologous bone grafts or artificial mesh. Postoperative complications include hydrocephalus and infections such as meningitis[8].

Prognosis varies; some patients develop normally, while others may experience mental and physical impairments[9]. Patients with frontal encephaloceles without added complications tend to have a better outlook. Conversely, those with posterior encephaloceles face a 55% survival rate, with outcomes worsening when complications are present. Approximately 75% of surviving infants may exhibit some degree of mental deficiency[10].

This study aims to investigate the clinical patterns and management of encephaloceles in Kenya, bridging the gap in understanding local prevalence, characteristics, and treatment outcomes. By doing so, we aim to contribute valuable insights to the field of encephalocele management, ultimately improving the care and outcomes of affected individuals in Kenya.

METHODS

This study investigates encephaloceles among children in Kenyatta National Teaching and Referral Hospital (KNH), located in Nairobi County, Kenya. This hospital is the largest referral facility in East and Central Africa, catering to patients from across the nation. The data for this study was extracted from records spanning from 2015 to 2021.

We selected participants from the hospital's newborn unit, pediatric wards, pediatric intensive care unit, and the child health clinic. The inclusion criteria included a confirmed diagnosis of encephalocele, determined

through clinical examinations performed by pediatricians or neurosurgical specialists, and complemented by CT scan or MRI imaging, either at birth or during follow-up. We included individuals who met these criteria within the chosen time frame and were not terminated pregnancies or adults over 18 years of age. Patients with prenatal diagnoses alone were excluded from the analysis.

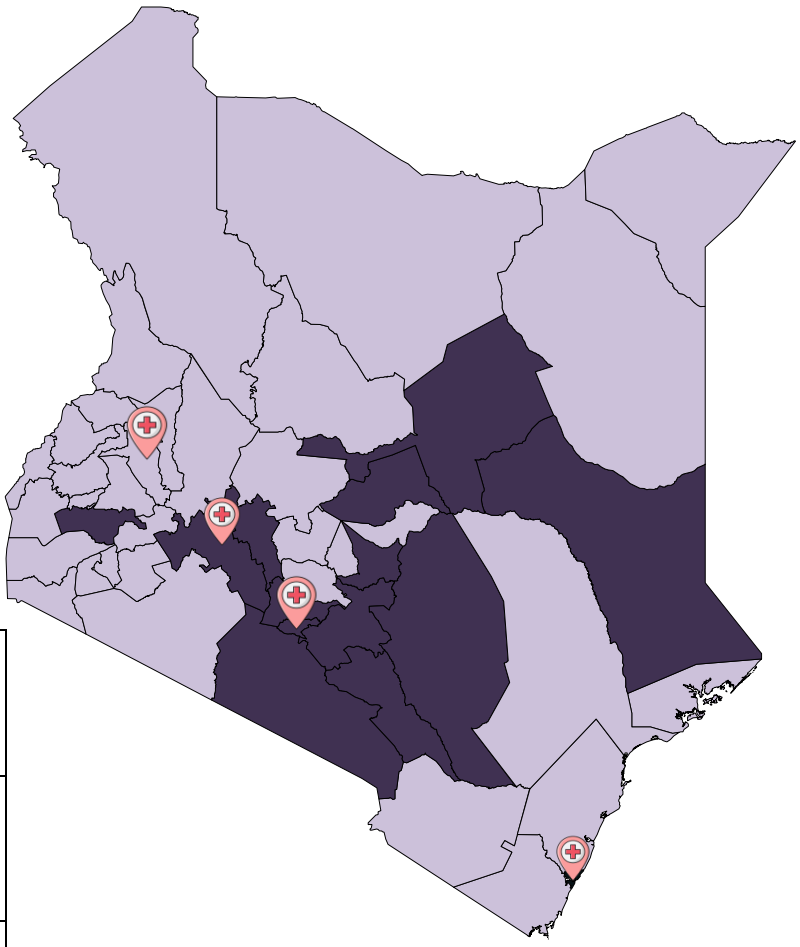
Data collection was carried out at the records department using a structured questionnaire and the findings analyzed using the current version of Excel and SPSS (27).

Ethical Consideration

Ethical approval was provided by the KNH-UON ethics and research committee. Data was collected from the patient files with permission from the involved departments and the patient's consent was not required.

RESULTS

We obtained forty-two patient files and 27 met the inclusion criteria. Of these 55.6% (15/27) were male and 44.4% (12/27) were female. The 27 files were analysed, and the data collected was filled in closed-question questionnaires. The age range of the patients was from 0-6 years with the mean age being 2.8 years. Regarding demographic characteristics, 30% (8/27) of the patients were from Nairobi, 19% (5/27) were from Kiambu, 7% (2/27) were from Garissa, 7% (2/27) from Makueni, 7% (2/27) from Machakos, Meru, Nyandarua, Embu, Kitui, Kisumu, Isiolo, Kajiado and Nakuru each had 1 patient (4%). 70% of the patients were Bantus, 19% were Nilotes and 11.5% were Cushites.



	Public facilities with neurosurgical care.
	Counties that reported encephalocele cases
	Counties that did not report encephalocele cases.

Figure 1. Map showing counties that reported Encephalocele cases and Encephalocele managing centres in Kenya

Encephalocele types

Using the Suwanella and Suwanella classification, 19 (70%) of the patients had occipital encephalocele, 7 (26%) had frontal and 1 (4%) were uncategorized[11].

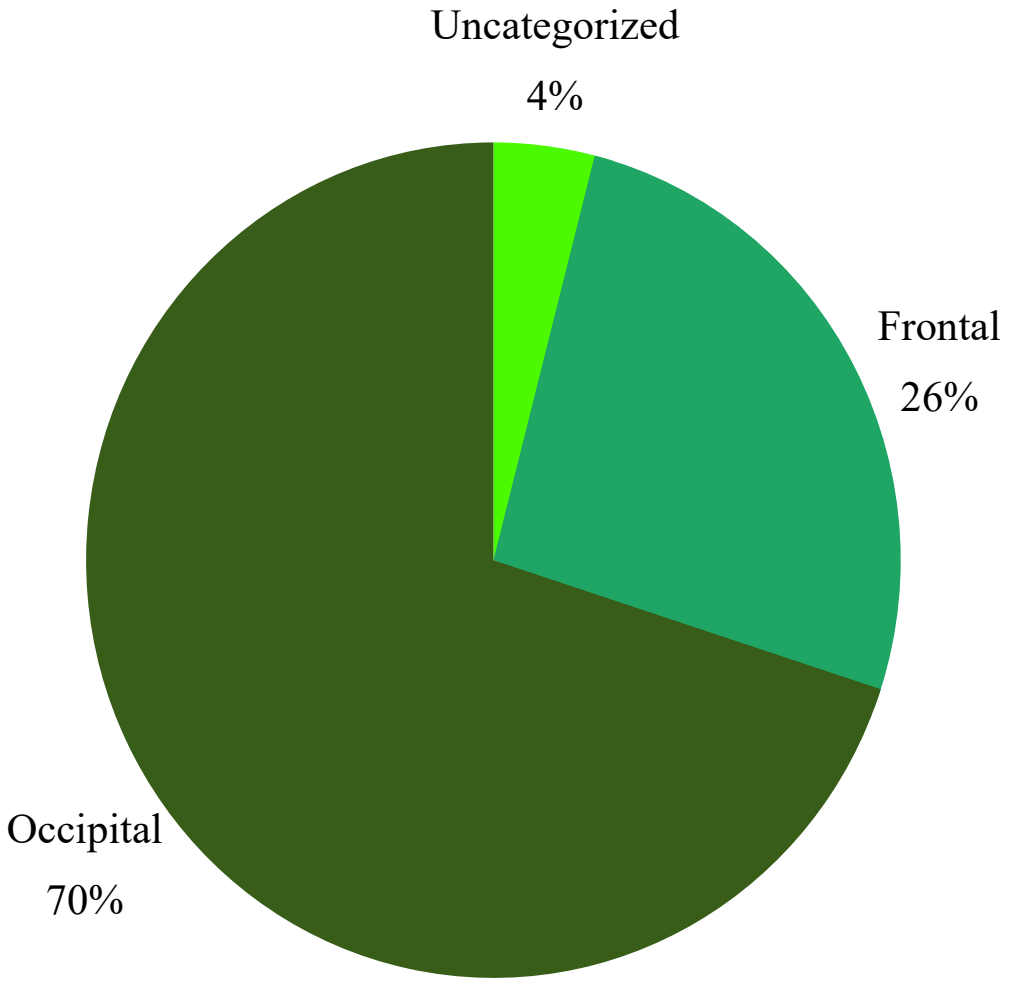


Figure 2. Pie chart showing the Encephalocele types

Management

The patients that underwent surgery were 55% while 44% of the patients were managed through non-surgical symptomatic management. 22% of the entire cohort could not afford surgery due to financial constraints. 1% of the patients died while awaiting surgery. The surgeries done were both single-stage and two-stage encephalocele repair. With the single stage being the most prevalent due to occipital encephaloceles. 15% of patients developed hydrocephalus and underwent ventriculoperitoneal shunting in the standard fashion.

Association Test

We examined the association between encephalocele prevalence and ethnicity in Kenya. The chi-square tests revealed that there was no association between the different types of encephaloceles and the various ethnic groups in Kenya.

Table 1

Showing the Association Cross tabulation

		VAR00002			
		bantu	cushites	nilotes	Total
Frontal	Count	4	1	2	7
	%	21.1%	33.3%	40.0%	25.9%
NA	Count	1	0	0	1
	%	5.3%	0.0%	0.0%	3.7%
Occipital	Count	14	2	3	19
	%	73.7%	66.7%	60.0%	70.4%
Total	Count	19	3	5	27
	%	100.0%	100.0%	100.0%	100.0%

Table 2

Showing the Chi-Square Tests

	Value	**df**	**Asymptotic Significance (2-sided)**	**Exact Sig. (2-sided)**
Pearson Chi-Square	1.153[a]	4	.886	.863
Likelihood Ratio	1.390	4	.846	.863
Fisher's Exact Test	2.571			.763
N of Valid Cases	27			

DISCUSSION

The patient population in this investigation showed a discernible gender distribution, with males experiencing a higher prevalence (56%) than females (44%). This finding differs from previous studies, which have generally shown that females are 4.5 times more likely to develop an encephalocele and 1.9 times more likely to develop an occipital encephalocele[1]. However, the incidence of anterior encephalocele is similar in males and females. NTDs, such as craniorachischisis, thoracic spina bifida, anencephaly, and encephaloceles, while males tend to have a higher incidence of spina bifida affecting the lower spine are more common among female patients[12].

Patients visiting the hospital originated from 13 counties with the largest percentage of (29.63%) coming from Nairobi County. Notably, patients travelled from distant regions, including Garissa, to seek care at KNH. This underscores the role of KNH as one of the primary public facilities in Kenya for managing encephaloceles. The majority (70%) of encephaloceles diagnosed at KNH were of the occipital variety, aligning with existing research that suggests approximately 70% to 90% of encephaloceles involve the occipital region[10].

There was no significant association between encephalocele types and ethnic groups in Kenya in this study, however studies in other countries have found that the prevalence of encephalocele varies according to ethnic background and geographical origin. This might have been due to the sample size. In The United States, encephaloceles are more prevalent in black community, while in Australia, they are more common among Aboriginal communities. Similarly, in Uganda, infants of Bantu origin exhibit a higher proportion of occipital encephaloceles than those of Nilotic origin[13–15].

55% of patients received surgical treatment at the hospital, which aligns with the established practice of surgical management as the mainstay for encephaloceles, the procedure involves repair of bone defect, dural closure and removal of non-functional brain tissue and excess skin[1]. Additionally, endoscopic endonasal repair has emerged as a safe and effective option, particularly for infants under one year of age, and is increasingly favoured for therapeutic intervention in most cases[16].

A significant portion of our patient cohort did not receive surgical treatment due to financial constraints. According to the *Kenya Household and Health Expenditure and Utilization Survey - KHHEUS* (2013), 17% of Kenyans reported having some type of insurance, but only 3% of those in the lowest wealth quintile had coverage. Only 29% of persons in urban areas and 11% in rural areas have any type of insurance, with the National Hospital Insurance Fund (NHIF), a government-owned organization, providing most of the health coverage. This highlights a critical issue regarding healthcare access and the economic burden associated with congenital abnormalities. Many patients are forced to pay out of pocket which is not sustainable due to the expensive nature of specialized surgeries. Birth defects can entail substantial medical and nonmedical costs. Further investigation is warranted to understand the economic impact on society and determine the cost profiles across affected individuals' lifespans[17].

One particularly concerning aspect of our findings is the fact that all these patients were diagnosed at birth, whereas encephaloceles are typically detectable in utero by the 13th week of pregnancy[18]. 74% of these patients were referral cases while 26% were diagnosed at birth in KNH. This underscores the need for improved folic acid supplementation, prenatal screening, and early detection all over the country to ensure timely intervention and optimal outcomes for affected individuals.

Study Limitations

Due to this study being retrospective with a paper-based medical filing system, there was limited data on prenatal care and supplementation, postoperative complications, number of clinical visits post-surgery, and further imaging. This inhibited our ability to meet our last specific objective which was to analyse postoperative complications, number of clinics, post-operative visits and further imaging, a prospective study with electronic paper records will go a long way to enhance the knowledge about encephalocele prevalence.

CONCLUSION

Encephalocele is a rare condition commonly found in regions marked by prevalent dietary deficiencies, particularly in low- and middle-income countries like Kenya. They are primarily managed through surgical intervention, with 55% of cases being treated surgically. However, financial constraints pose a significant challenge to surgical treatment accessibility. These findings underscore the need for additional research, neurosurgical expansion, and initiatives to enhance prenatal

diagnosis while addressing economic barriers associated with birth abnormalities. It is important to note that three other public facilities in the country offer neurosurgical care, apart from private institutions. This might have reduced the number of patients presenting to KNH. There is currently no ongoing study regarding encephaloceles that has been reported at the ethics committee in our institution. A multi-centre study might give a clearer picture of encephaloceles in Kenya. These efforts aim to ultimately improve care and outcomes for encephalocele patients in Kenya.

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