Original Article

The Pattern of Distribution of Encephalocele in University of Port Harcourt Teaching Hospital - A Three Year Experience

DR AMADI CE, MBBCH, PROF. EGHWRUDJAKPOR PO, MBBS, DMS, FICS
Neurosurgical Unit, Department of Surgery University of Port Harcourt Teaching Hospital, Port Harcourt, Nigeria

ABSTRACT
BACKGROUND: Encephalocele is a congenital anomaly that results from failure of complete neural tube closure during foetal development. It is a known cause of mortality and morbidity in infants. This study was carried out to highlight its distribution pattern in University of Port Harcourt Teaching Hospital over a three-year-period.
METHODOLOGY: This is a retrospective study of children with encephalocele admitted from January 2007 to December 2009. The following information were obtained from their medical records: sex, age at diagnosis, distribution pattern, place of origin, detailed antenatal history, maternal occupation/level of education, family history, associated anomalies and outcome of surgery.
RESULTS: 17 cases (10 females and 7 males) were seen over this period. 12 presented as frontal encephalocele while 5 were occipital. Their ages at diagnosis were: prenatal (determined by abdominal ultrasound) 5, 0-6 months 11, and 7-12 months 1. 9 of 17 mothers were unbooked. Pregnancy was uneventful in all cases. None had family history of encephalocele. 5 had multiple anomalies while 12 had only encephalocele. 10 patients had surgery, of which 9 were successful. 1 died in the immediate postoperative period. 7 patients did not have surgery. Among these, 3 died before surgery while the parents of 4 children refused operation. 10 mothers had primary education, 5 secondary, while 2 had attained tertiary education.
CONCLUSION: Encephalocele may be frontal or occipital. The distribution pattern of our cases was in favour of frontal location, with slight female preponderance.
KEYWORDS: Distribution Pattern; Encephalocele; Neural Tube Defects; Port Harcourt; Experience.

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INTRODUCTION

Encephalocele is one of the neural tube defects (NTDs) - a group of congenital disorders that occur as a result of failure of the surface ectoderm to separate from the neuroectoderm with consequent failure of complete fusion of the neural tube. This results in a bony defect in the skull, that allows herniation of the meninges or/and brain tissue through it. The occipital region is the most common site of this type of neural tube defect (75%) in the United States and Western Europe.[1,2] With antenatal ultrasound scanning, it may be seen as herniation of a spherical, fluid-filled structure through a calvarial defect beyond the calvarial confines that may be more correctly diagnosed as a meningocele or encephalocele, with the earliest reported ultrasonographic diagnosis being made at the 13th week of gestation.[1]

Several other anomalies have been reported in association with encephalocele, including spina bifida, congenital heart disease and congenital *tallipes equino varus*, amongst others. In one study of patients with encephalocele, other malformations and/or chromosomal anomalies were observed in at least 60% of the subjects.[3-5] Currently, most cases are diagnosed prenatally.[6-9] Maternal serum alpha-fetoprotein levels are elevated in only 3% of patients, because most encephaloceles are covered with skin.[10] Thus, ultrasonographic diagnosis seems more reliable. Postnatally, infants may have associated cerebrospinal fluid (CSF) rhinorrhea and recurrent meningitis.[10] In another report, a female preponderance was found among all infants with neural tube defect (including encephalocele), but a male preponderance in foetuses delivered spontaneously before week 20, indicating selective male late foetal deaths.[11]

In this article, we report the outcome of a study on the distribution pattern of encephalocele in University of Port Harcourt Teaching Hospital with reference to sex distribution, location (whether it is occipital or frontal), maternal occupation and level of education, and the outcome of surgical intervention.

METHODOLOGY

This is a retrospective study done in the University of Port Harcourt Teaching Hospital, a tertiary Medical Centre located in the oil rich Delta of the South-South region of Nigeria noted for high incidence of neural tube and other congenital defects which many think may be related to the high level of environmental pollution and degradation resulting from intense oil exploration and exploitation activities.[2]

The case files of children with encephalocele presenting to the hospital through the Special Care Baby Unit (SCBU) and neurosurgical out patients' clinic respectively from January 2007 to December 2009 were retrieved from the medical records department of the hospital; and the following pieces of information

Correspondence: Professor Patrick O. Eghwrudjakpor, Department of Surgery, University of Port Harcourt Teaching Hospital
P. M. B. 6173, Port Harcourt, Rivers State, Nigeria, Telephone: +234-806-930-1611, +234-807-236-8681, E-mail: patejakpor@yahoo.com

obtained: sex, age at diagnosis, distribution pattern (frontal or occipital), place of origin (state, local government area), antenatal history (whether the mother was booked or unbooked, drug history, and significant events during pregnancy, if any), maternal occupation and level of education, family history of encephalocele or other neural tube defect, other associated anomalies and outcome after surgical intervention. The data collated were analyzed using SPSS statistics 17 software.

RESULTS

A total of 17 cases of encephalocele were seen from January 2007 to December 2009. 9 of them were from Bayelsa State, 5 from Rivers State, 2 from Akwa Ibom and 1 from Delta State. Among these, 10 were females while 7 were males, giving a female:male ratio of 1:0.7. All the cases were less than one year old at time of diagnosis. 5 were diagnosed in the prenatal period by ultrasonography and 12 after delivery. Among the latter, only 2 had skull x-ray. None of the patients had a CT scan because they could not afford it. 11 of the 17 cases were located at the frontal part of the head while 6 were in the occipital region (Table 1, Figs. 1, 2 and 3).

5 of the 17 infants (29.4%) also had other congenital anomalies including hydrocephalus, microcephaly, congenital heart disease, omphalocele and *tallipes equino varus*. 12 of the 17 cases did not have associated anomaly.

Figure 1. Occipital Encephalocele with Microcephaly

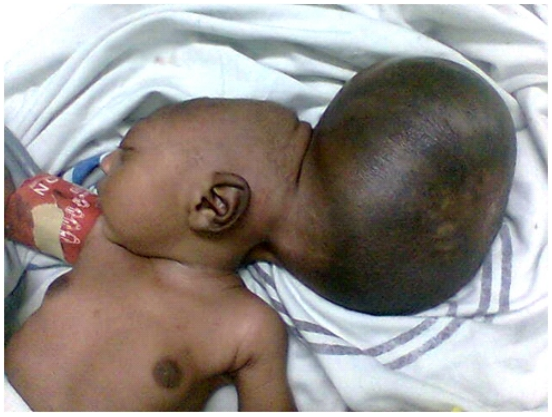
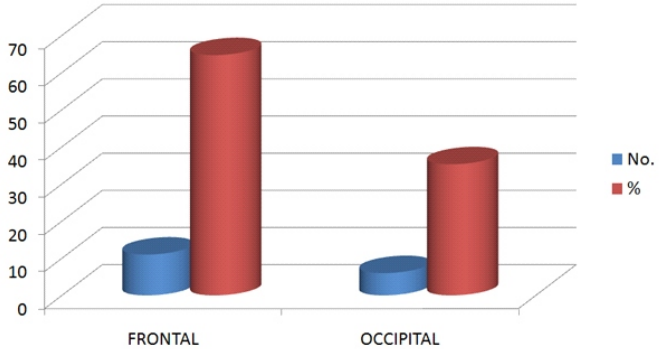


Figure 3. Position of Encephalocele



There was no history of encephalocele or other neural tube defects in the families of any of the affected babies.

8 of the mothers were booked for antenatal care. 3 of them however defaulted in their antenatal visits. The remaining 9 mothers were not booked. In all the cases, the antenatal period was uneventful. However, it was noted that none of the mothers took folic acid before conception.

10 of the mothers had attained primary level of education, 5 had secondary while 2 had tertiary education (Fig. 4). 11 of the mothers were petty traders, 2 were civil servants, 1 was a complete housewife and 3 were students.

Surgery was performed in 10 of the 17 children. Operation was performed by standard methods and involved excision of herniated brain tissue where present, repair and closure of the defect and removal of the redundant sac. Surgery was successful in 9 of the 10 cases. 1 patient died soon after surgery. The main postoperative complication noted was cerebrospinal fluid leakage. This was observed in 3 cases. 7 of the 17 patients did not have surgery. 3 died before operation could be performed and the parents of the remaining 4 children rejected surgery.

There was marginal yearly increase in the number of cases seen during the period of the study, with 4 cases being seen in 2007, 5 in 2008 while 8 cases were seen in 2009.

Figure 2. Fronto-Nasal Encephalocele

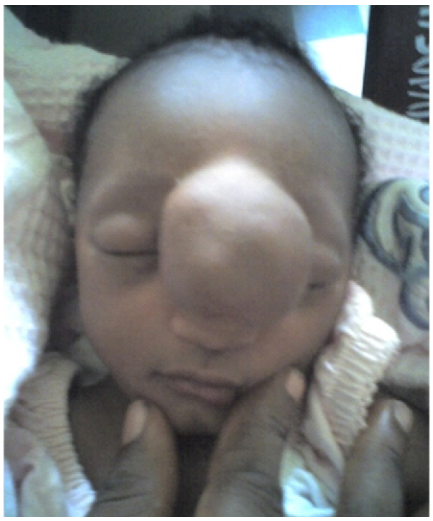


Figure 4 Mothers' Level of Education

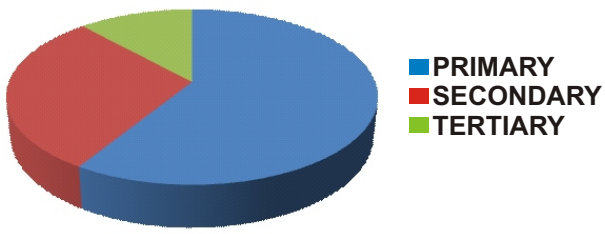


Table 1: Position of Encephalocele

POSITION	NO. OF PATIENTS	PERCENTAGE
FRONTAL	11	64.7
OCCIPITAL	6	35.3
TOTAL	17	100.0

DISCUSSION

Encephalocele (or cephalocele) is one of the neural tube defects, a group of disorders that affects the developing nervous system. They have varying incidences depending on prevailing conditions.[2] Anecephaly and spina bifida are the most common with encephalocele being seen less frequently.[2] There are 1-4 cases of encephalocele per 10,000 live births, and in foetuses that have been spontaneously aborted before 20 weeks' gestational age, it is believed to be the predominant neural axis anomaly.[12-15]. Our finding of 17 cases in 3 years in the UPTH which serves Rivers State and three neighboring States with a total population of over 15 million is in keeping with its rare occurrence.

Encephaloceles (or cephaloceles) are commonly classified according to their 1. Contents, and 2. Location.

1. Depending on the contents, a cephalocele may be:

a. Meningocele - where it contains only meninges and cerebrospinal fluid
b. Meningoencephalocele which contains meninges, cerebrospinal fluid and herniated brain
c. Encephalocele - containing only herniated brain, and
d. Meningoencephalocystocele - where the herniated part of the brain contains a cavity which communicates with the ventricle.

2. Classification according to the location of the cephalocele is based on the relation of cranial defect to the coronal suture (whether it is anterior or posterior to it).

a. In Frontal (anterior) encephalocele, the cranial defect is located anterior to the coronal suture. They are generally divided into those that are visible (sincipital encephalocele), and those that are not visible (basal encephalocele). Sincipital encephaloceles are further subdivided into four groups depending on the affected bones. These include: 1. Frontal, 2. Frontonasal, 3. Frontoethmoidal and 4. Nasopharyngeal encephaloceles. On the other hand, basal encephaloceles may be subdivided into five groups: 1. Transethmoidal (intranasal), 2. Sphenoethmoidal (posterior intranasal), 3. Transsphenoidal (sphenopharyngeal), 4. Spheno-orbital and 5. Sphenomaxillary encephaloceles.
b. In Occipital (posterior) encephalocele, the defect is posterior to the coronal suture. These are further grouped into the *Cephalocele occipitalis superioris* (where the defect lies above the external occipital protuberance) and the *Cephalocele occipitalis inferioris* where the defect lies below the external occipital protuberance. Whereas it is the occipital lobe that may be involved in the former, it is the cerebellum that is usually involved in *Cephalocele occipitalis inferioris.* Where the bony defect extends down to the foramen magnum, the condition is known as *Cephalocele occipitalis magna.*

In Western populations, 85% of these lesions occur in the occipital region, whereas in Southeast Asian and Africa, frontal lesions are relatively more common.[13,15,17]. Our finding of 11 cases of frontal and 6 cases of occipital encephaloceles is in keeping with other reports from Africa[18]

In this study, we found a female preponderance with a ratio of 1:0.7 (F:M). This is in keeping with another study from Nigeria[19] and a study from Lund, Sweden, where a female excess was also found among all infants with neural tube defects.[11]

None of the mothers of the affected babies in this study had taken periconceptional folic acid. Several studies have shown that neural tube defects can, to a large extent, be prevented by periconceptional use of this micronutrient whereas failure to use the substance before conception predisposes to it.[20-26] It is possible, therefore, that some of our cases could have been prevented if the mothers had been aware of, and had taking folic acid before they became pregnant.

It is worth noting that majority of the mothers in this study had only achieved a primary level of education. Obviously, this group of persons is less likely to be informed about the benefits of folic acid in prevention of NTDs. It is also worth noting that in an earlier study reported by us, we found that even among many well educated women in our society, the level of awareness of this information is

comparatively poor[2].

Regarding the socioeconomic status of the study population, we observed that majority of children were from a relatively low socioeconomic background as 11 of 17 mothers were petty traders, 3 were students and 1 was an unemployed housewife. Only 2 of them were comparatively well placed civil servants.

The surgical outcome for those operated upon was generally satisfactory. Though, they have not been followed up for long enough, the main complication noted was cerebrospinal fluid leakage in 3 cases. Only 1 of the 10 patients operated upon died in the immediate post operative period. This patient also had associated severe microcephaly preoperatively. Of the 7 patients that did not have surgery, consent for surgery was not obtained for 4 due to their families' cultural beliefs concerning head operations, while 3 died because of associated severe congenital anomalies. 5 of the 17 patients had associated congenital anomalies, which is about 30%. This is less than what was reported by other authors in a study where malformations and/or chromosomal anomalies were observed in at least 60% of patients with encephalocele.[3-5]

Some of the challenges encountered in the course of this study include difficulty having access to case files of the patients, and a significant number (five) of those operated upon being lost to follow-up; thus rendering thorough assessment of their post operative status over long periods difficult.

CONCLUSION

The distribution pattern of encephalocele in University of Port Harcourt Teaching Hospital over the three years period of January 2007 to December 2009 is in favor of frontal encephalocele with a slight female preponderance.

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