ENCEPHALOCELES: THE KIJABE EXPERIENCE

Authors:
Munyi N\textsuperscript{1,2}, Poenaru D\textsuperscript{1}, Bransford R\textsuperscript{1}, Albright L\textsuperscript{1}

Affiliation:
1. Bethany Kids at Kijabe Hospital, Kijabe, Kenya.
2. School of Medicine, University of Nairobi
INTRODUCTION

- Encephaloceles are congenital lesions consisting of a herniation of intracranial contents\(^1\).
- The herniation may consist of:
  - Meninges only (meningocele),
  - Brain matter (encephalocele)
  - Meninges + brain matter (meningoencephalocele)
  - Meninges + brain matter + ventricle (hydroencephalomeningocele)\(^1\)

---

Are they NTDs?
- There has been controversy as to whether they are all truly NTDs.
- Studies have reported that only a minority are associated NTDs ²
- Rowland et al 2002 found that with folic acid fortification, prevalence decreased for spina bifida but not significantly for encephaloceles or anencephaly ³.

Epidemiology

- Are rare and the incidence varies with geographical location and race. ⁴
- Overall incidence is 0.8 to 3.0 per 10,000 live births. ⁵, ⁶
- Occipital type (85% of encephaloceles) is most common in N. America and W. Europe with an incidence varying between 1 in 3000 to 1 in 10,000 LB ⁷, ⁸
- 70% of occipital encephaloceles are in females ⁷
- 15-20% are associated with NTD
- Anterior encephaloceles are mostly found in SE Asia, Russia and Central Africa. The incidence ranges from 1 in 3500 to 1 in 5000 ⁴, ⁷

---

CLASSIFICATION

Are classified according to the anatomical location of the skull defect. These include:

Sincipital
- Frontoethmoidal
  - nasofrontal
  - nasoethmoidal
  - naso-orbital
- Interfrontal
- Craniofacial cleft

Basal
- Intranasal
- Spheno-orbital
- Sphenomaxillary
- Spenopharyngeal

Convexity
- Occipital
- Parietal
- Saggital
- Occipitalcervical

Atretic
RATIONALE OF STUDY

- Though encephaloceles are rare congenital malformations, they are associated with severe morbidity and mortality if untreated.
- Most reported series of encephaloceles originate in the West, where resources for their treatment are radically different than in Africa.
- This study seeks to find out the presentation, management and complications of encephaloceles in an African setting as well as answer the question: Can encephaloceles be successfully managed in a resource-poor setting?
Design and Methodology

- A retrospective study of patients seen and managed at Kijabe hospital between January 1998 and August 2006.
- **Inclusion criteria**: any patient with an encephalocele.
- **Exclusion criteria**: any patient who’s records were not available.
- Data collected: Biodata, Type of encephalocele, Associated anomalies, US and CT features, Surgical approach used, Intra-OP and Post-OP complications and Follow up outcomes.
- Data Analysis: Calculation of Means, Drawing of frequency charts and distribution graphs done.
RESULTS

- Total number of patients: 53
  - 23 male; 30 female.
  - Charts of seven of the patients were unavailable and hence excluded from the analysis.

- Age Distribution: 1 day - 15 yrs
  Mean: 24 months
Types of encephaloceles

Distribution:
- Basal: 0%
- Complex: 2%
- Missing: 4%
- Sincipital: 33%
- Convexity: 61%

Detailed distribution:
- Occipital: 53%
- Parietal: 4%
- Sagittal: 0%
- Occipitocervical: 4%
- Frontoethmoidal: 24%
- Interfrontal: 9%
- Basal: 2%
- Complex: 6%

Convexity encephaloceles:
- Occipital: 88%
• Of those with Hydrocephalus (HC) :
  • 9 had occipital encephaloceles
  • 3 had sincipital encephaloceles
  • 1 was not classified
26 px had additional Sx.

17/26 (65%) had VPS insertions.
Complications (Intra-op, post-op & late)

- **6/49** had intra-operative complications
- **20/49** had post operative complications.
  - 2 deaths, CSF leak was most common complication  
    **6/20 (30%)**, Others: seizures, wound infection, shunt malformations, HC
- **10/47** had late post-operative complications
  - 2 deaths, 3 recurrences, 1CP, 2 HC, 1 infection, 1 frontal swelling
Follow up and outcomes

Follow up outcomes

- Other: 4
- CP and blind: 2
- Death: 5
- MR: 2
- Blind: 1
- Developmental delay: 5
- Doing well: 30

Series 1
Discussion

- Our sample size one of the largest case series in literature (over 8 yr period)
- Missing population based data therefore no conclusion on incidence can be made
- Female preponderance: our study 61% vs other studies: 70%
- Contrary to other studies: occipital type is most common in our set up.
- Most of our patients had mainly CSF with little brain tissue: good prognosis
Discussion II

- Age at presentation: earlier presentation than in Thailand study (70% present under 6 months in our study, in Thailand most present between 6-12 months)
- Imaging: inaccessible due to low socioeconomic status
- Various surgical approaches have different indications.
- Complications: most common CSF leak - similar to other studies.
Conclusions

- Diagnosis can be done on clinical basis.
- Encephalocele repair is generally a clean procedure hence no need for routine pre-operative medication.
- External approach is used for occipital but intracranial gives better outcome for frontal
- Most common associated anomaly is HC
- Encephaloceles generally have good outcome even in an African setting.
ERROR: stackunderflow
OFFENDING COMMAND: ~
ERROR: stackunderflow

STACK:

~