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CASE REPORT

# Encephalocele: A Case Report at Yaoundé Central Hospital, Cameroon

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## ABSTRACT

An encephalocele is characterized by congenital herniation of the brain tissue and/or meninges through a skull defect. The underlying cause is complex and not fully understood. The exact worldwide frequency is not known. Microcephaly with the presence of neural elements in the encephalocele is associated with poor prognosis. We report a case of a second twin premature baby born at 31 weeks gestation by cesarean section with an occipital encephalocele and died after 48 hours.


**Keywords:** Encephalocele, Neural tube defects, Twin pregnancy.

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A 21-year-old woman second gravida was admitted in our labor room for uterine contractions at 31 weeks. For her first pregnancy, she gave birth to a boy weighing 3200 gm who is actually in good health. The current pregnancy was not well followed up. She did three antenatal consultations by a nurse. She did not perform any ultrasound during the pregnancy. She has been taking iron and folic acid since 20 weeks gestation. She did not report any other medications taken during this pregnancy. Her HIV test was negative at 30 weeks of pregnancy. Her husband was a 54-year-old trader.

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On physical examination, we discovered a twin pregnancy with the first twin in transverse presentation. We did an emergency cesarean section. The first twin was a boy weighing 1550 gm and the second one was a girl weighing 2240 gm with an occipital encephalocele associated with polyhydramnios. The second twin was transferred to the Neonatology Unit where she died 48 hours later.

## DISCUSSION

Encephaloceles are congenital malformations characterized by a sac-like protrusion of the brain and/or its covering membranes through an opening in the skull. A substantial proportion of children, especially those born with a large encephaloceles, are physically and intellectually disabled. The etiology of encephalocele is considered to be complex, and in most cases the causes of this condition remain elusive. Trisomy 13 is a cause for a few encephaloceles (1–5%).[1] Our case was a black woman aged 21 years. Wen et al[2] found that encephaloceles were significantly more common among the offspring of Hispanic women than offspring of White women [adjusted prevalence ratio: 1.91, 95% confidence interval (CI) 1.34–2.70]. In addition, compared to the offspring of women 25 to 29 years of age, encephaloceles were more common among the offspring of women 20 to 24 years of age (adjusted prevalence ratio: 1.52, 95% CI 1.01–2.27). Dadmehr et al[3] did not find any significant risk factor in the case group as compared with controls, although the families of patients with encephaloceles had a better economic status ($p = 0.03$) and the fathers had a higher mean age. In our case, the father was 54 years old. The fetus was a female sex. Encephalocele is more common in female fetus than male.[3-7] Meanwhile, Raja et al[8] found 76% male and 24% female prevalence.

The encephalocele is detectable at 12 to 13 weeks on ultrasound using the vaginal probe. Ultrasound shows on a fetus of paracranial. In 75% the mass is occipital, in 12% the mass is frontal, and in 13% it is parietal. The mass may be fluid filled, entirely brain filled, or filled with both fluid and brain. Polyhydramnios may be present. Placenta is normal.[1] In our case, the diagnosis was made at delivery, and the women did not perform any ultrasound during the follow-up of her pregnancy. Even the twin pregnancy was discovered in labor room.

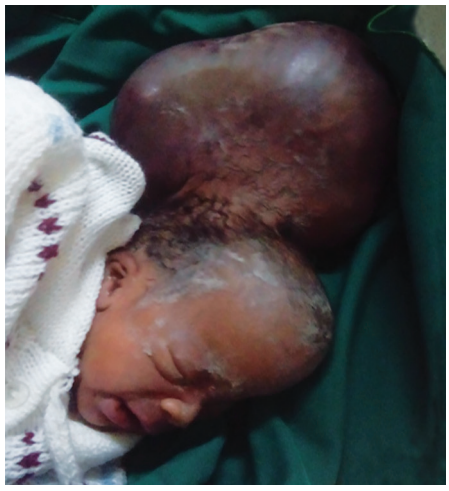

**Fig. 1:** Fetus at 31 weeks gestation with occipital encephalocele

In our case, it was an occipital encephalocele (Fig. 1). The delivery management depends on the size of the defect, the amount of herniated brain, and associated abnormalities. Nonaggressive management is recommended when severe microcephaly is present. In cases with normal head size, cesarean section might improve prognosis by avoiding trauma to the herniated brain tissue.[1] Our fetus was delivered by cesarean section but the indication was twin pregnancy with the first fetus in transverse presentation. The prognosis depends on the associated brain malformation. The size is not important prognostically because a large size may not contain neural elements. Microcephaly with the presence of neural elements in the encephalocele is associated with poor prognosis. After delivery of a child with encephalocele, cranial scan or magnetic resonance imaging should be performed to define the defect and to delineate any associated central nervous system abnormalities.[1] An occipital encephalocele is a congenital neurologic condition with an extremely high morbidity and mortality in spite of the treatments rendered pre- and postoperatively with a mortality rate of 29% after surgery.[4] Kotil et al[6] found 33.3% of mortality rate. Seizures and hydrocephalus are the more common neurologic complications requiring intervention.[1] In our case, the baby died 48 hours in neonatal without any further investigation or treatment because the family was unable to pay for the care. The intake of folic acid in periconceptional period can decrease the neural tube defect's risk up to 0.6 per 1000 births.[9]

## CONCLUSION

Encephaloceles remain rare in our practice. If the fetus has an occipital encephalocele associated with microcephaly, the pregnancy should be terminated because of poor prognosis. The most suitable solution in our low-setting resource is a policy of prevention with folic acid treatment before and during pregnancy.

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