

Texas Children's Hospital, Department of Surgery and Division of Neurosurgery, Baylor College of Medicine

BACKGROUND

In utero repair of fetal posterior cephaloceles (meningocele and encephalocele) is being performed based on the premise that fetal surgery prevents progressive herniation of neural tissue and brain damage during pregnancy.¹ However, the extent to which progressive herniation occurs during pregnancy, specifically from prenatal diagnosis to after delivery, is not well known.

PURPOSE

The objective of this study was to describe the natural history of patients with fetal cephaloceles focusing on the incidence of progressive herniation.

METHODS

We conducted a retrospective cohort study of all patients referred to our center for posterior fetal cephalocele between 2006 and 2021. All patients had prenatal *and* postnatal MRI. Progressive herniation (primary outcome) was defined as an increase in the absolute volume of neural tissue within the cephalocele of >5% *or* new herniation of a critical structure into the cephalocele. Total brain and cephalocele volumes were calculated to determine herniation progression from prenatal to postnatal MRI. The presence of hydrocephalus, epilepsy, and developmental delay (secondary outcomes) were collected at 1 year of age.



Fig 1A: Axial T2-weighted MRI of a fetus at 22 weeks gestation demonstrating a low occipital cephalocele containing part of the enlarged dorsally herniating right occipital horn and right occipital lobe



Fig 1B: Axial T2-weighted MRI of the same patient at 1 day of life demonstrates progressive herniation/deformity involving the occipital lobes, cerebellum, and brainstem with distortion and likely herniation of the midbrain and diencephalon.

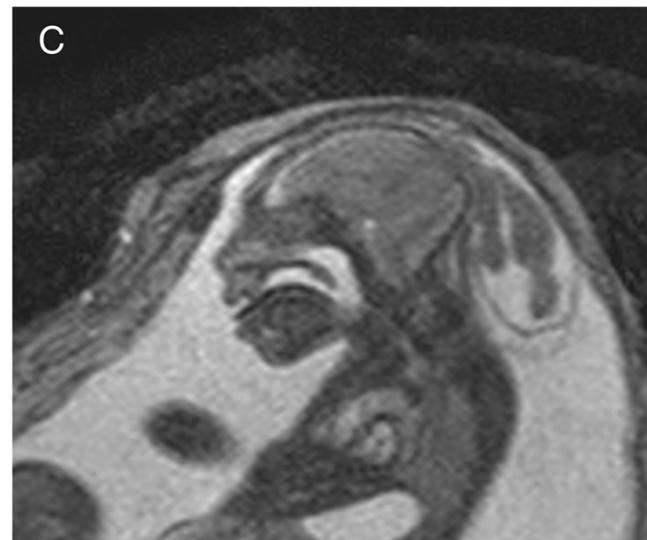


Fig 2A: Sagittal T2-weighted MRI of a fetus at 22 weeks gestation demonstrates a large parieto-occipital cephalocele with herniation of significant parts of the bilateral parieto-occipital lobes and global microcephaly.



Fig 2B: Axial T2-weighted MRI of the same patient at 1 day of life demonstrates no commensurate change in bilateral cerebral herniation compared to prenatal imaging.

RESULTS

- Twenty patients met all study criteria
- Ten patients demonstrated progressive herniation from prenatal to postnatal MRI (50%; 95%CI 0.27, 0.73)
- Three patients with progressive herniation were diagnosed with a meningocele prenatally and had an encephalocele postnatally
- Two patients without progression had meningocele identified prenatally that regressed and became atretic by birth
- Both prenatal hindbrain herniation ($p=.03$) and prenatal microcephaly ($p=.05$) were predictive of progressive herniation
- The rates of hydrocephalus (44%), epilepsy (44%), and developmental delay (66%) were not associated with the occurrence of progressive herniation in this study.

CONCLUSION

As defined in this study, progressive herniation was not a rare event (50%). Fetal hindbrain herniation and fetal microcephaly were associated with the occurrence of progressive herniation. These results support further investigations into why progressive herniation occurs in utero, and if progressive cerebral herniation in utero plays a significant role in determining clinical outcome.

REFERENCES

1. Cavalheiro S, Silva da Costa MD, Nicácio JM, et al. Fetal surgery for occipital encephalocele. *J Neurosurg Pediatr.* Sep 11 2020:1-8.