THE INCIDENCE OF PROGRESSIVE HERNIATION OF FETAL CEPHALOCELES

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Background: In utero repair of fetal encephaloceles is being performed based on the premise that fetal surgery prevents progressive herniation of neural tissue during pregnancy. However, the extent to which progressive herniation occurs during pregnancy, specifically from prenatal diagnosis to after delivery, is not well known. The objective of this study was to describe the incidence of progressive herniation in patients with fetal cephaloceles (meningocele and encephalocele).

Materials/Methods: We conducted a retrospective cohort study of all patients referred for fetal occipital cephalocele between 2006 and 2021. All patients had an occipital lesion with a prenatal and postnatal MRI. Brain and cephalocele volumes were calculated to determine progression. Before analyzing the data, progressive herniation was defined as an increase in the absolute brain volume within the encephalocele of >5% or new herniation of a critical structure into the cephalocele (e.g., brainstem, cerebellum, cerebrum, dural sinus, or spinal cord). The presence of hydrocephalus, epilepsy, and developmental delay was collected at 1 year of age.

Results: Twenty patients met the study criteria. Ten demonstrated progressive herniation from prenatal to postnatal MRI (50%; 95%CI 27%, 73%). Of these, three patients had meningocele only on their prenatal MRI. Two meningoceles became atretic at birth. Both prenatal hindbrain herniation (p=.03) and prenatal microcephaly (p=.05) were predictive of progressive herniation. The rates of hydrocephalus, epilepsy, and developmental delay were not associated with the occurrence of progressive herniation in this study.

Conclusions: As defined in this study, progressive herniation was not a rare event. Potential prenatal risk factors for progressive herniation included hindbrain herniation and microcephaly. These results support further investigations to determine if progressive herniation is clinically significant.

Images / Graph / Table

Figure 1.
a) Axial T2-weighted MRI of a female fetus at 22 weeks gestation demonstrating a low occipital encephalocele containing the herniated right occipital horn and a small portion of the right occipital lobe (patient 12). b) Axial T2-weighted MRI of the same patient at 1 day of life demonstrates progressive herniation involving the occipital lobes, cerebellum, and brainstem with distortion and likely herniation of the midbrain and diencephalon.