EDITORIAL

Fetal repair of encephaloceles

Nalin Gupta, MD, PhD

Department of Neurological Surgery, University of California, San Francisco, California

DURING the past 20 years, the role of fetal intervention for myelomeningocele has moved from an experimental intervention to an accepted and proven option for the condition.¹ The technical challenges that needed to be solved included successful access and closure of the gravid uterus, intraoperative fetal monitoring, and strict control of early labor. Fetal surgery for a variety of conditions is now performed at many centers across the world, and other conditions have been evaluated for potential intervention.

In this report, Cavalheiro et al. describe their experience with a small group (n = 9) of fetuses with occipital encephaloceles who underwent early surgical repair during mid-gestation.² Encephaloceles that had brain tissue greater than 20% of the volume of the cyst were excluded from fetal repair. The surgical repair as described appears to be similar to what would be done in the postnatal setting: early identification of the meningeal layers, preservation of the neural tissue if possible, and then closure of the dura mater and skin. In addition, an absorbable plate was used to repair the bony defect. The authors, who have extensive experience with fetal neurosurgical procedures, are to be congratulated for developing and extending the techniques to allow repair of fetal encephaloceles. Although they state, “there is no great difficulty in repairing the OE,” the fragile nature of fetal tissues does require attentiveness to detail.

Although the fetuses who had surgical repair were evaluated in terms of the need for shunting, reversal of microcephaly, and an assessment of developmental outcome, this small cohort should be viewed primarily as a demonstration of feasibility of the technical procedure. Of the 9 children, 2 had developmental delay. This is encouraging, but the caveat is that fetuses with greater than 20% of the cyst volume occupied by neural tissue were excluded. Fetuses with a larger amount of neural tissue

within the encephalocele would be expected to have a poorer outcome. In their review of a large cohort of children with encephaloceles in the postnatal period, Lo et al. noted that nearly half had normal development.³ The critical question is whether long-term functional outcomes in this restricted group are superior to those of children who undergo repair after term delivery. This is particularly important since the maternal and fetal risks, which are well defined for open fetal interventions, must be balanced by clear benefits to the child. The role of fetal intervention of hydrocephalus is a cautionary example, as small groups of these patients with fetal hydrocephalus treated by shunt placement have had generally poor outcomes.⁴

<https://thejns.org/doi/abs/10.3171/2020.5.PEDS20234>

References

1. Adzick NS, Thom EA, Spong CY, et al. A randomized trial of prenatal versus postnatal repair of myelomeningocele. *N Engl J Med*. 2011;364(11):993–1004.
2. Cavalheiro S, da Costa MDS, Nicácio JM, et al. Fetal surgery for occipital encephalocele. *J Neurosurg Pediatr*. Published online September 11, 2020. doi:10.3171/2020.3.PEDS19613
3. Lo BW, Kulkarni AV, Rutka JT, et al. Clinical predictors of developmental outcome in patients with cephaloceles. *J Neurosurg Pediatr*. 2008;2(4):254–257.
4. Pisapia JM, Sinha S, Zarnow DM, et al. Fetal ventriculomegaly: diagnosis, treatment, and future directions. *Childs Nerv Syst*. 2017;33(7):1113–1123.

Disclosures

The author reports no conflict of interest.

Correspondence

Nalin Gupta: nalin.gupta@ucsf.edu.

INCLUDE WHEN CITING

Published online September 11, 2020; DOI: 10.3171/2020.5.PEDS20234.

Response

Sergio Cavalheiro, MD, PhD,¹ Marcos Devanir Silva da Costa, MD, PhD,¹ Jardel Mendonça Nicácio, MD,¹ Patricia Alessandra Dastoli, MD, PhD,¹ Italo Capraro Suriano, MD, PhD,¹ Mauricio Mendes Barbosa, MD, PhD,² Hérbene Jose Milani, MD,² Stéphanno Gomes Pereira Sarmiento, MD,² Tereza Cristina Carbonari de Faria, PT, PhD,¹ and Antonio Fernandes Moron, MD, PhD²

¹Department of Neurosurgery, Universidade Federal de São Paulo; and

²Department of Fetal Medicine, Hospital e Maternidade Santa Joana, São Paulo, Brazil

We would like to thank Dr. Gupta for his inspiring and detailed editorial. As he pointed out, in fetal translational neurosurgery, it can take a long time to transform an idea into a successful surgical procedure that is capable of improving the quality of life of patients and is accepted and reproducible worldwide. Such progress has occurred in the treatment of fetal myelomeningocele, as demonstrated by the Management of Myelomeningocele Study, a major milestone in the development of fetal neurosurgery.

We agree with Dr. Gupta that these advances in fetal neurosurgery should be made with caution and supported by detailed fetal evaluation with the consideration that other malformations could benefit from intrauterine treatment, such as hydrocephalus and perhaps fetal tumors. The improvement of less-invasive techniques for fetal surgeries has been associated with lower maternal risks and vaginal deliveries; these improvements are very welcome and should be continued.

Unlike myelomeningocele, encephalocele might be associated with genetic diseases that must be ruled out

before the fetal surgical procedure. The presence of herniated tissue greater than 20% of the volume of the cyst is not a contraindication to the procedure and was only considered to standardize our sample. As the pregnancy progressed, the herniated content gradually increased;¹ performing the fetal surgical procedure should be considered at gestational ages below those proposed for the cases of myelomeningocele.

Despite some limitations and criticisms, the treatment of fetal occipital encephalocele followed the same principles as fetal surgery for the treatment of Chiari malformation type II.² The previous experience of our team with this kind of surgery was essential for performing surgery to treat occipital encephalocele. We should emphasize that the surgeries were performed after detailed parental counseling and their request to respect their cultural beliefs.

This initial experience presents the possibility of the intrauterine treatment of a major fetal malformation with an impact on the child's life. We hope that other fetal neurosurgery teams can be motivated in this direction so that a multicenter experience can be developed and improve our understanding of fetal encephalocele treatment.

References

1. Smith AB, Gupta N, Otto C, Glenn OA. Diagnosis of Chiari III malformation by second trimester fetal MRI with postnatal MRI and CT correlation. *Pediatr Radiol*. 2007;37(10):1035–1038.
2. Adzick NS, Thom EA, Spong CY, et al. A randomized trial of prenatal versus postnatal repair of myelomeningocele. *N Engl J Med*. 2011;364(11):993–1004.

INCLUDE WHEN CITING

Published online September 11, 2020; DOI: 10.3171/2020.6.PEDS20415.