EDITORIAL

Separation of craniopagus twins

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THE authors have detailed a very rare case of a set of craniopagus twins who were born at 28 weeks' gestational age and shortly after birth required an emergency surgical separation due to the rapid and unexpected death of one twin.¹ The authors provide a comprehensive literature review and point out that this appears to be a case with the youngest known age for separation where one twin survived the surgery and at 12 months' follow-up appears to be doing well. The medical details and surgical planning are well detailed along with the complexity of this type of frontal angular craniopagus type. The surgical planning was clearly quite thoughtful and focused on the details of an emergency separation. Based on the weight (1250 g) and age of the surviving child and surgical blood loss this was clearly a tour-de-force and the successful outcome excellent. The birth of craniopagus twins is a very rare event, with only 2 or 3 sets being born around the world per year who are candidates for possible separation surgery. The real difficulties in this case were the small size of the survivor and the rapid death of the other twin, which precluded doing any extensive neuroradiological workup that would have assisted in the surgical planning. These circumstances and the fact that the medical team was able to save the one child make this paper a useful addition to the literature. I am personally aware of 4 cases of craniopagus twins in which surgical separation was attempted during the past 13 years and in which both twins died despite surgical intervention, so the outcome in this case clearly exceeds the standards in this rare disorder.

In reviewing this paper it is important to appreciate that a staged separation would not have been possible in this case due to the death of one child and the rapid deterioration of the surviving twin. The authors detail the issues with the vascular anatomy as it pertains to the anatomical conjoining. In most cases the vascular sharing only involves the venous portion of the brain anatomy. In this case, however, there was also crossover of the arterial circulation, and this is important to appreciate as it will affect the surgical separation planning.

Also common in these types of conjoined twins is the presence of an anomalous venous vascular lake called a circumferential venous plexus. This structure can only rarely, if ever, be separated, owing to its "lake-like" configuration. The entire structure is typically given in its entirety to one twin during a staged separation.

In separating craniopagus twins the cerebral anatomy can be quite complex—in particular, when sorting out the brain interfaces. It has been our experience that there is often a good deal of fused brain, which might not be so apparent on the MR images. In the nonfused brain there is clearly seen a dural membrane between the 2 brains. Having said this, the imaging studies can often be misleading as there appears to be CSF in the spaces between the 2 brains, giving the impression that the brains have an open interface; in our experience this is rarely the case. During surgery the surgical team will find opposing and often fused brain cortex that will need to be separated.

In our review of the literature and now with 7 sets of craniopagus separations done with staged procedures, the risk of hydrocephalus has been markedly reduced. It would appear, however, that a single-stage approach to separation leads to a significant increase in the risk of CSF shunting in one if not both children. Because hydrocephalus can lead to CSF leaks and an increased risk of meningitis and seizures, a focus on reducing hydrocephalus in single-stage emergency craniopagus separations can clearly benefit these children.

This case report clearly details the multiple risks involved in the care of these types of craniopagus twins. The surgical treatment in this case where one twin died early is well outlined, and such cases clearly constitute neurosurgical emergencies. With a capable multidisciplinary team and rapid early intervention there is a possibility of significantly reducing the surgical morbidity in the survivor or survivors.

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References

1. Nejat F, Habibi Z, Goudarzi M, Azad MS, Moradi E, Heidari V, et al: Emergency separation of craniopagus twins: case report. **J Neurosurg Pediatr** [epub ahead of print July 14, 2017. DOI: 10.3171/2017.1.PEDS16306]

Disclosures

The authors report no conflict of interest.

Response

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We read with great interest and respect the editorial written by Professor Goodrich. We appreciate his comments, which originate from his considerable experience with craniopagus surgery and which have added so much to the value of this case report. According to our literature review, this case is unique for the very young age and low weight of the twins. For our success in this complex surgery, without the extensive preoperative investigation that is mandatory for this kind of operation, we are indebted to the experience of the chief surgeon (F.N.) for observing the unsuccessful separation of the famous Iranian twins Laleh and Ladan, to the very high speed of surgery at all stages of separation or closure, and to the precise support of the very careful anesthetic and neonatology teams.

The death of one twin allowed us to use dura and skin from that twin to close the anatomical layers in the living twin, which increased the speed of surgery dramatically. Separation and closure of dura and skin are difficult stages of craniopagus surgery that require time before and during surgery for planning the incisions and providing the necessary flaps for perfect closure.

Due to the very thin skin in this premature neonate, we

could not use bone from the dead twin to close the cranial defect at the site of splitting. Cranioplasty at the time of separation may prevent skull deformity subsequent to rapid head growth in the presence of a large skull defect, which might be particularly likely to result in deformity if the separation is performed when the twins are extremely young, as in our case.

The skull deformity that we confronted just several days after surgery occurred very early compared with the successful separations reported so far in the literature, as the patients in those cases were mostly older than ours and did not have brain and head growth as fast as our patient's. Due to the fast growth of our premature neonate's body and head associated with progressive enlargement of the intracranial cyst and large skull defect, gradual brain herniation occurred through the defect, leading to progressive torticollis and cranial deformity. We used some external pressure with compressive dressing, but as the skin was very thin, we could not apply much pressure. Because the defect was in the vertex, however, we wanted to apply pressure on the bulging mass from above. We used a soft handmade hat secured with tape under the patient's chin, but this system interfered with feeding and caused some skin erosion on the neonate's chin. As this approach was uncomfortable for him, it was withdrawn.

The progressive torticollis and skull deformity continued despite CSF shunting from the intracranial cyst to decrease the brain herniation. Recently, when the patient was 18 months old, he underwent cranial remodeling surgery, which required an extensive craniotomy. A helmet was prescribed for the patient to wear beginning 2 weeks after surgery to attain a better cranial shape.

Long-term follow-up after separation of conjoined twins is necessary for observation and follow-up of all problems related to cranial deformity, hydrocephalus, probable mental or motor delay, and associated extracranial anomalies. Timely management of these complications can affect the prognosis and long-term outcome.