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CASE REPORT

Pediatric/Craniofacial

Use of Multidirectional Cranial Distraction Osteogenesis for Cranial Expansion in Syndromic Craniosynostosis

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Summary: Patients with syndromic craniosynostosis often require a large amount of cranial expansion to avoid intracranial hypertension, but the surgical procedure remains controversial. A patient of severe syndromic craniosynostosis with multiple bony defects and anomalous venous drainage at the occipital region was treated by multidirectional cranial distraction osteogenesis (MCDO) at the age of 8 months. Distraction started 5 days after surgery and ceased on postoperative day 16. The distraction devices were removed 27 days after completing distraction. After device removal, the increase of intracranial volume was 155 ml and the cephalic index was improved from 115.5 to 100.5. The resultant cranial shape was well maintained with minimal relapse at postoperative 9 months. In cases of syndromic craniosynostosis with multiple bony defects and/or anomalous venous drainage at the occipital region, expansion of the anterior cranium by MCDO is a viable alternative to conventional methods. *(Plast Reconstr Surg Glob Open 2017;5:e1617; doi: 10.1097/GOX.0000000000001617; Published online 22 December 2017.)*

Patients with syndromic craniosynostosis display considerable impairment of skull growth and often require cranial expansion to prevent the development of intracranial hypertension within the first year of life. However, the best approach for cranial expansion in this setting remains controversial. Although fronto-orbital advancement (FOA) was a typical option,[1] this procedure is plagued by high rates of relapse.[2]

Distraction osteogenesis was introduced in 1998 as a means of expanding the cranium in patients with craniosynostosis,[3] and this method has continued to gain in popularity. Distraction osteogenesis has some theoretical advantages over 1-stage surgical cranial remodeling.[3,4] Gradual expansion of the soft-tissue envelope allows greater gains in intracranial volume and minimizes relapse, and the bone flaps remain attached to dura mater, which reduces the risk of extradural abscess and supports postoperative bone repair and growth. FOA with distraction osteogenesis in particular has been widely used to achieve sufficient expansion of anterior cranium in syndromic craniosynostosis.[4] Recently, posterior cranial vault distraction osteogenesis (PCVDO) has been advocated for syndromic craniosynostosis to greatly improve intracranial volume.[5–7] Unfortunately, the posterior cranium in such patients is often thin, riddled with multiple bony defects (copper beaten skull), and subject to anomalous venous drainage, all of this jeopardizing the safety and efficiency of PCVDO and heightening postoperative complication rates.[8]

Multidirectional cranial distraction osteogenesis (MCDO) is a viable alternative to conventional distraction procedures.[9] Herein, we report a case of syndromic craniosynostosis with anomalous venous drainage treated by MCDO.

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CASE REPORT

Preoperative Findings

An 8-month-old boy with Pfeiffer syndrome underwent ventriculoperitoneal shunting at the age of 6 months (**see Figure, Supplemental Digital Content 1**, which displays preoperative frontal view of the patient, ***http://links.lww.com/PRSGO/A633***). Three-dimensional computed

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tomographic (CT) images showed a cloverleaf skull with multiple bony defects and a Mercedes-Benz pattern of craniosynostosis affecting occipital region. A CT venogram also disclosed an enlarged occipital emissary vein and a prominent suboccipital venous network (Fig. 1). (**See Figure, Supplemental Digital Content 2**, which displays pre- and postoperative 3-dimensional computed tomographic imaging, confirming sufficient expansion and rapid bone formation/fusion without relapse, ***http://links.lww.com/PRSGO/A634***.) (**See Figure, Supplemental Digital Content 3**, which displays preoperative 3-dimensional computed tomographic venogram, ***http://links.lww.com/PRSGO/A635***.)

These findings made us reluctant to perform PCVDO, so MCDO was undertaken as an initial cranial surgery to expand the anterior cranium.

Surgical Procedure

The MCDO surgical procedure was performed as described previously.[9] Briefly, osteotomies of anterior cranium were performed, creating rectangular bone flaps and a supraorbital bar (**See Figure, Supplemental Digital Content 4**, which displays schemas and intraoperative views of MCDO, ***http://links.lww.com/PRSGO/A636***). An ultrasonic bone scalpel (Sonopet; Stryker Corp, Kalamazoo, Mich.) was used for safe osteotomy, without dural tears. Except for supraorbital bar, all dural attachments to bone flaps remained individually intact, preserving their vascular supply. Traction pins were fixed in each bone flap and in the supraorbital bar. Upon closure of wounds, a helmet-type frame was fixed by anchoring pins on the temporal bones. Wires secured in traction pinholes were then passed through holes in the frame, ultimately fixing the wires to frame-mounted distractors. Total procedural duration, including time expended for setting the device after wound closure, was 194 minutes. The amount of blood transfused was 140 ml (18.67 ml/kg).

Quantitative Assessment

The patient underwent 3-dimensional CT scans preoperatively, just after device removal, and 9 months postoperatively. Pre- and postoperative cephalic index and intracranial volume were calculated by DICOM image viewer (OsiriX; Pixmeo, Bernex, Switzerland).

Postoperative Course

Distraction was initiated on day 5, when the bone pieces were shifted outward, and the wires lost appropriate tension. Distraction was continued at a rate of 1.5 mm/d and terminated on postoperative day 16. During the activation period, the traction rate was adjusted, depending on cranial shape and tension of the wires. Although a 6-week consolidation was scheduled, the anchor pins loosened, forcing device removal 27 days after completing distraction. Still, sufficient anterior cranial expansion was achieved, and the resultant cranial shape was sustained at 9 months (Fig. 2). (**See Figure, Supplemental Digital Content 5**, which displays postoperative photograph of sufficiently expanded anterior cranium, ***http://links.lww.com/PRSGO/A637***.)

The cephalic index was improved from 115.5 to 100.5 and 100.0 immediately after distraction and at postoperative month 9, respectively, with corresponding intracranial volume expansion from 682 ml to 837 ml and 881 ml.

DISCUSSION

In treating syndromic craniosynostosis, the best method and optimal timing of cranial expansion are open to controversy. Specifically, it is debatable whether the first cranial expansion should involve anterior or posterior skull. Although no direct comparisons between FOA with distraction osteogenesis and PCVDO have been published, it is reasonable to presume the superiority of PCVDO on grounds that posterior cranium generally exceeds anterior cranium in cross-sectional area.[6,7]

Because anomalous venous drainage at the occipital region is a frequent feature of syndromic craniosynostosis,[10] preoperative CT venogram is necessary for PCVDO. In our previous report, preoperative CT venogram was not performed for MCDO routinely because posterior cranium was not an object for operation at that time.[3] In this

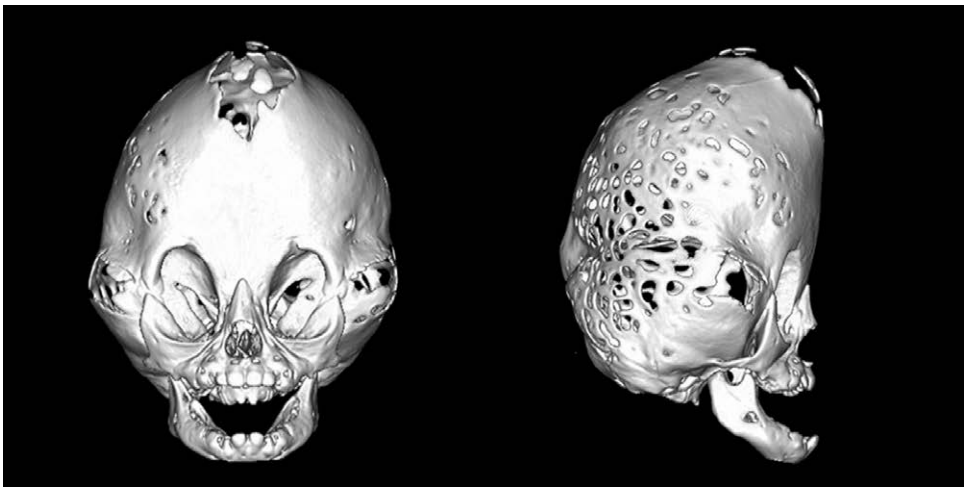

Fig. 1. Preoperative 3-dimensional CT imaging.

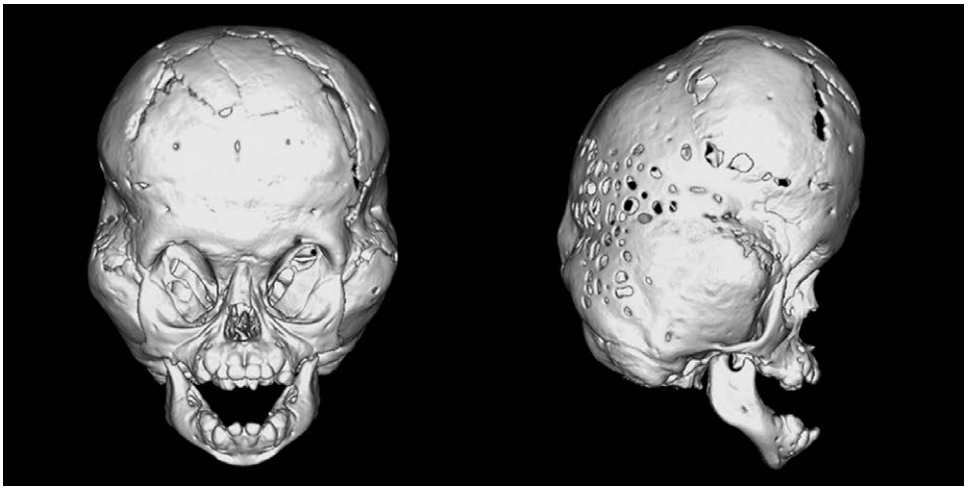

Fig. 2. Postoperative 3-dimensional CT imaging.

patient, however, anomalous venous drainage prevented safe and efficient PCVDO, and thus, we selected MCDO as a first cranial surgery for anterior cranial expansion.

MCDO has distinct advantages over unidirectional cranial distraction osteogenesis (UCDO). Because the surface area of a quarter sphere is twice its cross-sectional area, the volumetric change per unit distance of distraction in MCDO is theoretically twice that of UCDO. (**See Figure, Supplemental Digital Content 6**, which displays approximation graphic comparing UCDO and MCDO, ***http://links.lww.com/PRSGO/A638***.) Another benefit is that distances between bone flaps after distraction are narrower in MCDO than in UCDO, so earlier bone fusion and thereby a shorter consolidation period is expected. In addition, both direction and distance of bone-flap mobilization in MCDO can be more flexibly controlled, thus facilitating to achieve desired cranial shaping.

One disadvantage of MCDO (vs UCDO) is the extent of surgical invasiveness, because a minimum of 5 bone flaps are created in MCDO. The added osteotomies result in longer operative time and greater need of transfusion.

Loosening of the anchor pins and traction pins is a characteristic complication in MCDO.[9] To solve this problem, we have used poly-L-lactic/polyglycolic acid (PLLA-PGA) plates and succeeded to acquire much more stability of anchor pins and traction pins recently.

Our case suggested that MCDO is a viable alternative to the conventional methods for the treatment of syndromic craniosynostosis particularly in patients with multiple bony defects and anomalous venous drainage at the occipital region. Decompression of the brain by expansion of the anterior cranium will result in improvement of the multiple bony defects in the posterior cranium. Then, we can expand the posterior cranium at the second surgery.

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