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CASE REPORT
Craniofacial

Craniocerebral Disproportion Corrected With Biparietal Distraction Osteogenesis

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Summary: Ventriculoperitoneal shunt therapy represents a lifesaving neurosurgical intervention in pediatric patients with hydrocephalus. Although necessary, research indicates that shunting may be associated with a spectrum of craniocerebral disproportion from frank craniosynostosis to later presenting neurofunctional symptoms and headaches. The surgical management of craniosynostosis is generally treated according to provider and institutional preference; however, shunt therapy presents difficulties for typical vault remodeling procedures. In this report, we describe the utility of a biparietal distraction osteogenesis configuration for the treatment of craniocerebral disproportion in 2 patients. A parallel arrangement of an anterior and a posterior set of 2 distractors allows for a doubling of the typical expansion of the parietal diameter. This method effectively alleviates the scaphocephaly without disruption of the shunt site. We propose our repair for the indication of sagittal craniosynostosis in the shunted patient. *(Plast Reconstr Surg Glob Open 2025;13:e6747; doi: 10.1097/GOX.0000000000006747; Published online 1 May 2025.)*

Craniocerebral disproportion (CD), in the form of shunt-related craniosynostosis, is a known, though poorly understood, sequelae of ventriculoperitoneal (VP) shunt therapy. Diagnosis is a result of visible cranial deformity, commonly in the sagittal suture,[1] or by recognition of symptoms, such as headaches and neurofunctional deficits. Treatment includes some form of cranial vault remodeling (CVR) or distraction osteogenesis (DO) to accommodate continuing neurological expansion as well as alleviate a resultant dyscosmetic cranium. The ideal cranial reconstruction approach is debated, especially considering a concurrent VP shunt.[2–7] The authors aim to report the successful treatment of 2 CD cases with biparietal distraction osteogenesis (BDO) therapy, with and without a shunt.

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CASE PRESENTATION NO. 1

A 12-month-old infant with a VP shunt secondary to intraventricular hemorrhage, shunt revision for infection, G-tube dependency, and intermittent sixth cranial nerve palsy presented to the plastic surgery clinic with scaphocephaly. Physical examination demonstrated palpable sagittal sutural fusion, confirmed on head computed tomography (Fig. 1). The patient's parents consented to cranial DO. Virtual surgical planning (VSP) was performed, and 3-dimensional cutting guides were developed for the surgery. The patient was admitted to the hospital for surgical treatment followed by distraction.

CASE PRESENTATION NO. 2

A 12-year-old boy with a medical history of idiopathic intracranial hypertension refractory to acetazolamide, chronic headache, slit ventricle syndrome, and papilledema presented to the clinic. Head imaging revealed distention of the optic nerve sheaths bilaterally and craniosynostosis of the sagittal and bilateral lambdoidal sutures. Two lumbar punctures also revealed increased opening pressures. Following VSP, the patient underwent BDO placement with inpatient distraction to follow. (**See figure, Supplemental Digital Content 1**, which displays the

Disclosure statements are at the end of this article, following the correspondence information.

Related Digital Media are available in the full-text version of the article on www.PRSGlobalOpen.com.

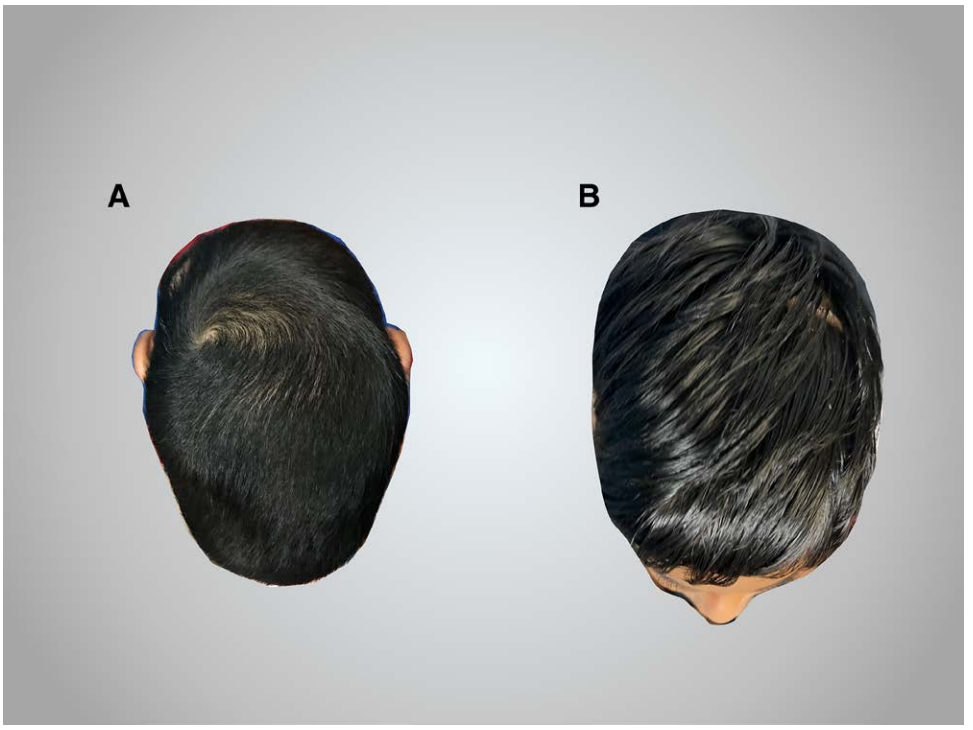

Fig. 1. A photograph of preoperative (A) and postoperative (B) cranial profile in case 1.

VSP models of case 1 and case 2, **http://links.lww.com/PRSGO/E3**.)

SURGICAL MANAGEMENT

After a bicoronal incision and avascular plane dissection, the scalp and pericranial flaps were reflected. The computer-generated osteotomy/distraction device endplate guides were placed and marked. Assisted by the osteotomy guides, the co-neurosurgeon mobilized bilateral parietal bone flaps. In the first case, an osteotomy also disconnected the occipital bone from the fused sagittal suture, leaving a peninsular sagittal bone section; barrel stave osteotomies were also made in the occipital region and bent outward using a Tessier bone bender to accommodate further posterior expansion (Fig. 2). Four 30-mm craniomaxillofacial distractors (DePuy Synthes, Monument, CO) were then anchored securely with titanium pins and absorbable pegs from this peninsular sagittal bone to the bilateral, osteotomized biparietal bone flaps (Fig. 3). The pericranial and scalp flaps were reflected over these devices, puncture incisions were made, and the distractor arms were externalized through the incisions. Before flap and scalp closure, the surgical site was copiously irrigated with saline, triple antibiotic solution, and Betadine, and a subgaleal Jackson-Pratt drain was placed for wound drainage. Finally, both flaps were closed in a staged manner.

POSTOPERATIVE COURSES

Both patients tolerated the procedure well, and the devices were advanced approximately 1.05 mm in the mornings for 14 days postsurgery. This resulted in roughly 30–32 mm of total distraction distance. Daily dressing care included a saline-diluted hydrogen peroxide rinse, antibiotic ointment, and a petroleum-impregnated bacteriostatic dressing. The case 1 patient also received a 10-day postdischarge course of prophylactic cephalexin. Neither patient experienced postoperative complications. After 3 months of osseous consolidation, surgical device removal was performed. The case 1 patient showed improvement in cranial shape, resolution of intermittent sixth cranial nerve palsy, progress in language acquisition, improved oral intake, and weight gain over 14 months of follow-up. The case 2 patient reported durable resolution of headaches and improved concentration, mood, and general affect over 9 months of follow-up.

DISCUSSION

Sandler et al[8] argued convincingly for the acceptance of the umbrella term "CD" for the various denominations of the same conceptual entity, such as slit ventricle syndrome, idiopathic CD, or postshunt craniosynostosis. In contrast to syndromic craniosynostoses, patients with CD demonstrate no identifiable genetic basis for their disease.[9] Therefore, it is most likely caused by a disruption in what Bryant et al[9] termed "the mechanotransduction pathways," thought to influence normal sutural physiology. In the shunted cranium, the decrease in fluid volume may affect a loss of tension in the collagenous dural and periosteal layers, signaling the loss of growth potential in the developing cranium, as a proxy of normal sutural closure. Regardless of discussions of etiology, in our experience,

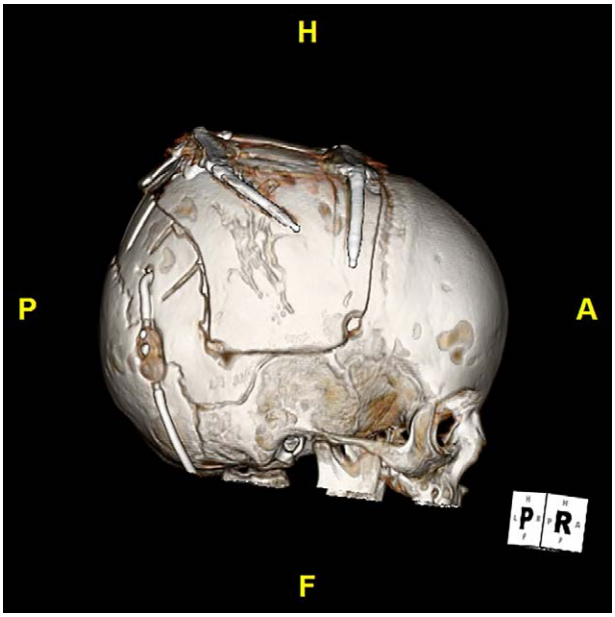

Fig. 2. A photograph of the sagittal view from a 3-dimensional reconstructed computed tomography showing the mobilized parietal bone, occipital barrel stave osteotomies, and an undisturbed VP shunt in case 1. H, A, F, and P represent head, anterior, feet, and posterior, respectively, as orientation directions in the original medical imaging software.

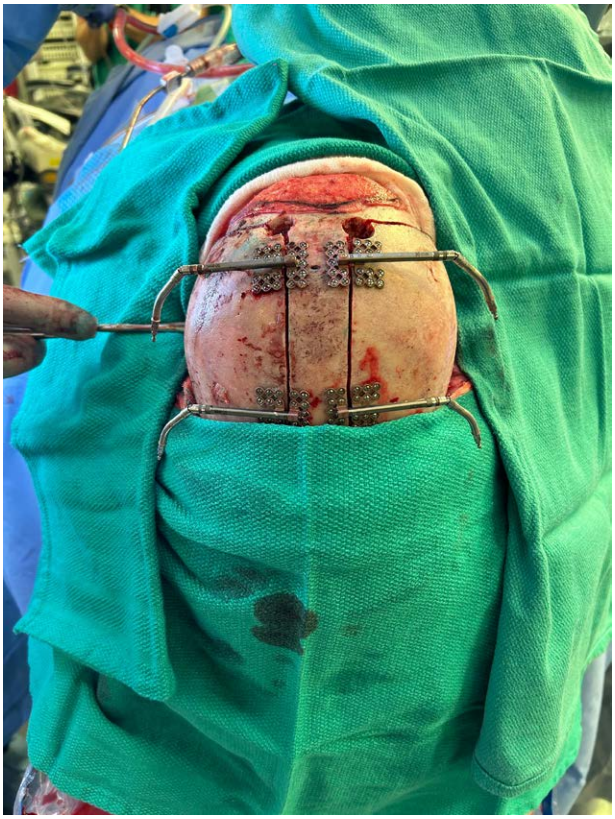

Fig. 3. A photograph of intraoperative cranial distractor layout in case 2.

BDO treatment may be indicated for the whole spectrum of CD, shunted or otherwise.

Traditional CVR is commonly recommended for the indication of CD.[4,5,8] Comparing posterior vault DO to CVR, transcutaneous distractors raise concern for potentially increased risk of infection and shunt infection/revision. However, externalization and manipulation of the shunt during distractor device placement present a potential confounder, especially in comparison to our BDO approach, which avoided exposure during surgery and the implication of the posterior shunt site in the distraction field.[4] Fastidious distractor hygiene may also reduce the risk of infection. Besides potential risks, posterior vault DO has potentially better volume expansion compared with traditional CVR, as well as decreased blood loss and operative time.[6] Finally, the necessity of cranial vault stability due to concurrent shunt therapy may be compromised by CVR.[5]

Spring-assisted DO has also been used in this setting with a potentially reduced risk of infection due to total subcutaneous implantation.[2,3] However, infection risks of externalized DO may be addressed, yet springs lack the control and anchoring offered by incremental linear distractors, presenting their own unique complication profile. DO repair along a biparietal vector may then represent the ideal repair by providing stability in mechanical connections while increasing cranial vault volume. Descriptions of DO along this vector in the settings of an anteriorly based shunt[7] and nonsyndromic plagiocephaly[10] have been written; however, our approach performed well in the setting of CD, especially with a posteriorly based shunt.

CONCLUSIONS

Two cases of CD were treated uniquely with BDO. Cranial restriction and recurrent headaches were resolved postoperatively in respective cases. By arranging 2 sets of expanders in series, our technique doubles the potential biparietal expansion. With the stability afforded by the sagittal bone peninsula, a BDO arrangement may afford greater control over cranial expansion while protecting a posterior shunt site, a potentially ideal repair for the indication of CD.

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DISCLOSURE

The authors have no financial interest to declare in relation to the content of this article.

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