Posterior Distraction Osteogenesis in Syndromic Craniosynostosis: A Case Series

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ABSTRACT

Posterior distraction osteogenesis is now being considered as an alternative to conventional cranial vault remodeling (CVR) techniques in the management of craniosynostosis. Gradual displacement of the posterior skull in distraction osteogenesis offers greater volume expansion, decreased soft tissue tension, less blood loss, and reduced risk of relapse. We present two patients who underwent successful posterior distraction osteogenesis as part of the correction for their syndromic craniosynostosis.

Both patients were diagnosed with syndromic bilateral coronal synostosis at birth. Patient 1 exhibited a tall head with a small anterior fontanelle and a flattened forehead and occiput. Patient 2 exhibited a tall head, large open anterior fontanelle, asymmetric flatness of the occiput, and midface hypoplasia. Both underwent a two-stage correction consisting of posterior distraction followed by fronto-orbital advancement. Distraction was successful in both patients, resulting in significant expansion of skull volume and normalization of head shape with no increase in intracranial pressure. Thus, posterior cranial vault distraction osteogenesis may be a better alternative to conventional CVR in the management of certain types of craniosynostosis.

INTRODUCTION

Craniosynostosis is defined as the premature fusion of one or more cranial sutures. This condition, especially when more than one suture is involved, may potentially lead to increased intracranial pressure (ICP) and disruption of normal brain development, ultimately requiring surgical correction.¹,² Bilateral coronal suture fusion is often seen in cases of syndromic craniosynostosis. This leads to a skull shape that is described as turribrachycephaly, or one that is tall in the vertical dimension and short in the antero-posterior (AP) plane.³ The brain is unable to expand and grow in the AP plane, and instead is forced to grow upward, often through an enlarged open anterior fontanelle. Conventional cranial vault remodeling (CVR) techniques in the treatment of this type of skull shape involves...
direct horizontal displacement of the posterior vault (Figure 1). This method requires exposure and craniotomy of the occiput, followed by reshaping and repositioning of the osteotomized bone fragments in the desired configuration. Results with CVR are often suboptimal for a variety of reasons: scalp tension prevents adequate AP expansion of the skull, open craniotomy carries risk for significant blood loss, and there is potential for relapse with supine positioning of the patient postoperatively.

![Figure 1. Comparison of conventional posterior vault reconstruction and posterior distraction osteogenesis. A. Posterior cranial vault reconstruction (CVR) involving craniotomy and direct advancement of osteotomized segment; B. Posterior distraction osteogenesis involving placement of distraction devices and gradual advancement of occipital bone.](image)

Physicians are now considering cranial vault distraction osteogenesis as an alternative to the open CVR approach. Posterior cranial vault distraction is especially appealing and involves the gradual repositioning of the entire occipital bone. The concept of distraction osteogenesis was first developed by Gavril Ilizarov in Russia for the treatment of long bone fractures with extensive bony gaps that needed to be bridged. The utilization of distraction in the craniomaxillofacial skeleton is universally attributed to Joe McCarthy, who was the first to apply the technique to mandibular lengthening in 1992. The five phases of the technique consist of osteotomy, latency, distraction, consolidation, and remodeling. The gradual retro-movement of the osteotomized fragments offers greater control in the expansion process, and the consolidation period helps prevent the fragments from shifting back to their original positions as new bone is deposited into the osteotomy gap. The theoretical advantages with this technique are: greater volume for expansion in the posterior cranial vault, which would allow the brain to reposition more inferiorly without the need for any application of external pressure; dura remains attached to the endocranial surface of the vault bone, which reduces blood loss; and the soft tissues and scalp are gradually stretched, thereby reducing the tendency for relapse. Once the posterior vault is expanded, and the height of the skull reduced, any remaining concomitant anterior deformities can be addressed through a conventional fronto-orbital advancement. In this case report, we present two patients with syndromic bilateral coronal craniosynostosis and turribrachycephaly who underwent successful posterior distraction osteogenesis as part of the correction of their skull deformities.

**CASE PRESENTATION**

**Patient 1**

The patient was first seen as a 4-month-old male, born at 39 weeks gestation, and was the product of an uncomplicated pregnancy and vaginal delivery. At birth, he was noted to have an abnormal head shape, with no signs of increased ICP or other medical conditions. His family history was significant for craniosynostosis in both his mother and paternal uncle, leading to the diagnosis of a syndromic craniosynostosis. A 3-di-
imensional head computed tomography (3D head CT) revealed bilateral coronal synostosis (Figure 2A).

![Figure 2](image)

**Figure 2.** Status post-placement of bilateral distraction devices for bilateral coronal synostosis in patient 1. A. Head CT without contrast; B. Skull is tall with a small anterior fontanelle and a flattened forehead and occiput.

Physical examination showed a tall head with a small anterior fontanelle and a flattened forehead and occiput (Figure 2B). Due to the height of the vertex and the flattening of the posterior cranium, a two-stage correction was undertaken. Stage 1 consisted of a posterior cranial vault distraction to decrease the height of the skull as well as to increase the AP dimension (Figure 2A). Distraction began at a rate of 1 mm/day on both sides and was completed at 36 mm bilaterally, at which time the consolidation period began (Figure 3A).

![Figure 3](image)

**Figure 3.** Patient 2 posterior distraction progress. A. Distraction completed at 36 mm bilaterally; B. Status post-distraction device removal and fronto-orbital advancement. Vertex is lowered and posterior cranial volume expanded; C. Anterior view of improved head shape following recovery.

Two months after surgery, the patient presented to the emergency department with evidence of infection at his left distractor site. A head CT showed a 1 cm abscess on the left scalp superior to the ear without intracranial involvement. His abscess was drained through the left distractor site, and his distraction devices were removed. A bacterial culture grew 1 Proteus, pan susceptible. He was given intravenous vancomycin and ceftriaxone for 3 days after surgery, followed by 3 weeks of oral cephalixin.

Four months after the initial distraction, the patient’s bicoronal incisions were healed but widened above the left ear at the former site of the infected distractor. There was no evidence of continued infection. Most notably, his vertex was significantly lowered compared to the portions of the skull anterior to the distraction devices (Figure 3B). At this time, stage 2 was performed which consisted of a conventional fronto-orbital advancement to correct the flattened forehead. The surgery was uncomplicated, and the patient was discharged on post-operative day 5. He has done well since then with no evidence of any sequelae from the infection (Figure 3C).

**Patient 2**

The patient was an 8-month-old male diagnosed with Antley-Bixler syndrome, a rare craniosynostosis syndrome characterized by turribrachycephaly, midface hypoplasia, humeroradial synostosis, and other abnormalities. Due to hydrocephalus, he had a right frontal ventriculoperitoneal shunt placed prior to his transfer to our institution.

Physical examination revealed a severe cranial deformity, involving a tall head, large open anterior fontanelle, flatness of the occipital region bilaterally with the right side more pronounced than the left, retruded orbital rims and a shallow midface (Figure 4A, 4B).
A 3D CT scan of the head showed bilateral coronal synostosis and severe posterior positional plagiocephaly, along with a very small foramen magnum (Figure 5A). A CT venography (CTV) showed a normal transverse sinus and a large vein circling the foramen magnum.

The patient underwent a two-stage correction of his skull deformity. First, posterior cranial vault distraction was undertaken through a biparieto-occipital craniotomy and insertion of 3 posterior cranial vault distraction devices. Two distraction pins were placed on the left side and 1 was placed on the right side (Figure 5A-C). The post-operative healing was uncomplicated. There was intermittent bleeding at his pin site, for which iodine dressing was used over the pin insertion site. By post-operative day 4, he was deemed suitable for discharge.

Following discharge, left-sided distraction proceeded at a rate of 1 mm/day. Due to the asymmetry of the right side, right-sided distraction began 10 days later at a rate of 2 mm/day (1 mm twice daily). One month following surgery, the distraction process was complete with the left side fully distracted at 23 mm and the right side at 40 mm.

The distraction significantly expanded the patient’s skull volume, allowing his previously compressed ventricles to normalize in size. He had no signs of increased ICP and demonstrated a sunken anterior fontanelle. The flattened occiput appeared much less noticeable and the forehead began to spontaneously assume a more rounded overall shape (Figure 6). Removal of the distractors and fronto-orbital advancement will be performed following the consolidation period.
DISCUSSION

Posterior distraction is a novel and promising method of treating craniosynostosis as it allows bone to expand over distances that are difficult to accomplish with conventional procedures.\textsuperscript{5,9} Unlike open CVR, this technique provides tension-free scalp closure, and the soft tissue has sufficient time to adapt while the bone is gradually advanced posteriorly.\textsuperscript{5} The overall volume of the posterior cranium is greater than any other section of the skull and expansion in this region creates enough room for the brain to spontaneously reposition in a posterior inferior direction, thus ameliorating the overly tall vertex. The distractors also prevent relapse secondary to supine positioning, which is especially common in the early postoperative phase of CVR when the brain has not yet expanded to fill the surgically-created gap.\textsuperscript{5} In a study conducted by White et al.,\textsuperscript{5} five syndromic patients underwent posterior distraction, with a mean advancement measuring 24 mm. Similarly, Steinbacher et al.\textsuperscript{7} reported a mean advancement of 23 mm (range 19-32 mm) for 8 patients. Due to the limitations of scalp closure in CVR, it is unlikely that this 2 to 3 cm magnitude of expansion is achievable using conventional means.\textsuperscript{7} In both patients presented in this study, their cranial volumes were substantially increased (maximum distance of 36 mm in patient 1 and 40 mm in patient 2), surpassing the average reported by White et al. Although further follow-up is required, neither patient is currently showing any signs of relapse.

Another advantage of posterior distraction osteogenesis is the ability to leave the dura attached to the osteotomized bone, allowing the bone to retain its vascular supply, thereby maintaining bone viability and decreasing blood loss.\textsuperscript{5,10} Steinbacher et al.\textsuperscript{7} compared the results of posterior vault distraction to CVR and found that distraction produced less blood loss (reduction of 8-10%) and shorter length of admission (Table 1).

In a retrospective evaluation of the safety of posterior CVR, Chen et al.\textsuperscript{11} reported a 28% complication rate in a 50-person cohort. The intraoperative complications were often life-threatening, consisting of dural tears and bleeding, sagittal sinus injury, or transverse sinus injury.\textsuperscript{11} In this case study, both patients had successful surgeries with no intraoperative complications, signs of increased ICP, or signs of neurodevelopmental impairment.

Posterior distraction osteogenesis also has a significant indirect effect on anterior fossa expansion, probably due to the calvarial plasticity in children younger than 2 years old and the horizontal axis of the distraction.\textsuperscript{5,12} The posterior procedure leaves the anterior fossa intact thus preserving the anterior calvarium for future procedures, such as a fronto-orbital advancement. This is especially important in syndromic patients who carry a high risk of recurrent ICP elevation along with often severe midface deformi-

Table 1. Comparison of Distraction Posterior Vault Remodeling to Conventional Posterior Vault Remodeling in Syndromic Patients.

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<tr>
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<th>Distraction PVR</th>
<th>Conventional PVR</th>
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<tbody>
<tr>
<td>EBL, % of TBV</td>
<td>52</td>
<td>60.6</td>
<td>0.026</td>
</tr>
<tr>
<td>Operative time, minutes</td>
<td>229.5</td>
<td>245</td>
<td>NS</td>
</tr>
<tr>
<td>Length of admission, days</td>
<td>3.25</td>
<td>5.6</td>
<td>0.0006</td>
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\textit{PVR: posterior vault remodeling; EBL: estimated blood loss; TBV: total blood volume; NS: not significant.}
ty. In the case of patient 1, fronto-orbital advancement was performed after completion of posterior distraction, with a better overall head shape and more stable results when compared to conventional CVR. Patient 2 has similar plans for anterior cranium and orbit reconstruction in the near future.

The current standard protocol involves a distraction rate of 1 mm/day and a consolidation period of several months. However, a recent study by Nowinski et al. adopted a modified time scheme defined by a shortened latency (48 hours), increased distraction rate (2 mm/day), and reduced consolidation time (4 weeks). By decreasing the length of time that the distractors were in place, Nowinski and colleagues managed to reduce the risk of complications such as infection, trauma to external parts, loosening, and skin deterioration. While the use of distractors may carry the risk of infection, as in patient 1 of our report, early removal of the devices may reduce infection potential. Patient 2 underwent right-sided distraction at the rate of 2 mm/day, and the accelerated rate neither compromised outcome nor led to infection or other complications. Furthermore, reducing the number of distractors to a minimum may also prevent device-related complications.

Compared to conventional CVR, distraction osteogenesis carries the disadvantage of limited ability to reshape contour abnormalities, such as flattened or bulging zones. While the craniotomized segment is advanced, the contour of the distracted bone remains largely unaltered. Consequently, there is heavier reliance on secondary molding from the brain. However, over time, gross head shape can clearly be modified using the distraction technique, as illustrated by the final skull shapes of the patients in this study. Treatment time is prolonged, and removal of the distraction devices requires a second procedure. However, compared to the significant decreases in operative time, blood loss, and overall morbidity, these limitations do not outweigh the long-term benefits of posterior vault distraction.

For patients with significant skull deformities carrying a high risk of needing multiple cranial vault expansions for increased ICP, such as those with syndromic craniosynostosis, posterior vault expansion confers powerful advantages. In conclusion, posterior cranial vault distraction osteogenesis may be a better alternative to conventional cranial vault reconstruction in the management of certain types of craniosynostosis and subsequent skull deformities.

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LEARNING POINTS

- Posterior distraction osteogenesis offers greater expansion of the posterior skull compared to conventional cranial vault reconstruction techniques in the correction of syndromic craniosynostosis.

- Through gradual repositioning of the posterior cranial bone, posterior distraction osteogenesis provides sufficient time for soft tissue adaptation, thereby providing tension-free scalp closure and decreased risk of relapse.

- The two-stage approach consisting of posterior distraction followed by fronto-orbital advancement may provide better aesthetic results than conventional cranial vault reconstruction.

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REFERENCES


