A combined frontoorbital and occipital advancement technique for use in total calvarial reconstruction

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The management of infants with bilateral coronal synostosis and resultant brachyturricephaly poses a significant therapeutic challenge. The application of total calvarial reconstruction to the treatment of this problem has represented a major recent innovation that has substantially improved the cosmetic results in this patient population. However, rigid fixation of the reconstructed calvaria is often required to maintain the correction achieved and to provide protection for the underlying brain. The requirement for extensive fixation constitutes a significant disadvantage for the use of this procedure in infants and young children. In this report, the authors describe an approach to the treatment of this problem that incorporates a series of tongue-in-groove osteotomies to provide increased stability to advancements of both the frontal and occipital regions in conjunction with cranial height reduction, while minimizing the need for metallic fixation. With this approach, the reconstructed skull is sturdy enough to resist the compressive force applied by the weight of the child's head immediately after surgery, but retains the ability to expand progressively. The authors have found the cosmetic results to be extremely gratifying. In this article they present their experience with this technique in seven children.

**Key Words** • calvarial reconstruction • coronal synostosis • craniofacial abnormality • lambdoid • craniosynostosis

The management of children with bilateral coronal synostosis constitutes a challenging problem because in many instances the cosmetic deformities result not only from the effect of frontoorbital retrusion, but also from a severe associated growth restriction of the posterior calvaria and compensatory changes in the surrounding cranial vault. The combination of anterior and posterior calvarial growth restriction produces a markedly foreshortened skull; because compensatory growth occurs laterally and upwardly, these patients characteristically exhibit a brachyturricephalic cranial shape.

Although such patients have long been treated with frontoorbital advancement techniques, the cosmetic results are sometimes disappointing because these procedures address only the forehead anomalies without treating the abnormal calvarial height or the severe occipital flattening. Not uncommonly, multiple operations are required to achieve an acceptable cranial appearance. Accordingly, the development by Persing, et al., of a technique for achieving total calvarial reconstruction represented a major advance in the management of this problem. However, when this procedure is undertaken in infants or young children, the surgeon is confronted with opposing problems. On the one hand, the construct requires some degree of stability to prevent loss of correction and injury to the underlying brain. On the other hand, the construct must have sufficient potential mobility to allow future calvarial expansion.

In an effort to address these seemingly divergent goals, we have developed a modified calvarial expansion procedure that provides some element of rigidity to the reconstructed forehead and occiput, while at the same time preserving the potential for cranial expansion. We have applied this technique to seven children who were treated at our institution during the last 2 years and have witnessed gratifying cosmetic results.

**Clinical Material and Methods**

*Preoperative Evaluation*

All patients with complex craniosynostosis admitted to our institution are examined by a multidisciplinary craniofacial team. A general clinical examination is performed to identify stigmata of syndromic synostosis as well as any underlying medical problems. An ophthalmological evaluation is also performed to rule out papilledema and impairments of ocular motility. In patients with
evidence of syndromic bilateral coronal synostosis, a complete skeletal survey is obtained to define any associated bone anomalies. Computerized tomography (CT) scans of the brain, calvarial vault, and cranial base, supplemented by three-dimensional reconstructions, are then performed.

Patient Selection

The seven patients who were selected as candidates for total calvarial reconstruction instead of conventional forehead advancement shared several common features. First, each child had severe brachycephaly with a horizontal cephalic index \( \leq 72 \) to 88 (illustrated in Fig. 1 left), based on normative data derived from a racially mixed population. Second, these patients were all severely turricephalic with a vertical cephalic index approximating 100, well above the normal range (Fig. 1 right). Third, on three-dimensional CT reconstructions, these patients not only had obvious coronal synostosis with a profoundly foreshortened anterior cranial fossa, but also evidence of synostosis at the lambdoid sutures with severe occipital flattening. The clinical features of the seven patients are summarized in Table 1.

In the five patients who had not undergone prior craniofacial surgery, total calvarial reconstruction was performed between 6 and 12 months of age. This management plan represented a compromise between the desire to perform the operation early to incorporate the maximum period of brain growth and relieve any potential restriction to adequate brain development, and the realization that many children with complex craniofacial disorders who undergo operation during the first weeks of life develop recurrent synostosis within 12 months of surgery and may therefore require a second operation in order to achieve an acceptable outcome. Moreover, because the blood volume of a neonate is substantially smaller than that of a 6-month-old infant, the operation is probably better tolerated in the slightly older age group.

Two other children had severe, progressive brachy turri cephaly after initial anterior (Case 6) or staged anterior and posterior (Case 3) reconstructive procedures. Both of these children also had evidence of increased intracranial pressure (ICP). The patient in Case 3 had papilledema and evidence of a beaten copper appearance on her skull x-ray films. The patient in Case 6 experienced periods of vomiting and irritability without obvious papilledema.

Prior to operation, all seven children underwent lateral flexion and extension views of the cervical spine to rule out vertebral anomalies or cervical hypermobility, which would preclude the use of the approach outlined below. No such abnormalities were detected in these children.

Operative Technique

Each child was placed in a modified prone position as outlined by Persing and colleagues. A bicoronal incision was made and the skin flaps were dissected forward to expose the orbital region and backward to expose the foramen magnum region. Bilateral frontal and occipital craniotomies were performed using a high-speed drill (Midax Rex, Fort Worth, TX), leaving a midline strut of bone to protect the sagittal sinus and torcular as well as biparietal struts, which were used in subsequent height adjustments. Bilateral osteotomies of the orbital roof, beginning 5 to 10 mm posterior to the orbital margin, were extended medially just in front of the crista galli, above the frontonasal suture, and laterally just above the frontozygomatic suture moving inferiorly and through the sphenoid ridge

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**TABLE 1**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (mos) at Operation, Sex</th>
<th>Syndrome</th>
<th>Prior Operations</th>
<th>Operating Time (min)</th>
<th>Blood Loss (ml)</th>
<th>Length of Hospitalization (days)</th>
<th>Length of Follow Up (mos)</th>
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<tr>
<td>1</td>
<td>9, F</td>
<td>Saethre–Chotzen</td>
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<td>580</td>
<td>350</td>
<td>7</td>
<td>23</td>
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<tr>
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<td>12, M</td>
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<td>565</td>
<td>500</td>
<td>12</td>
<td>20</td>
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<td>3</td>
<td>25, F</td>
<td>Crouzon’s</td>
<td>anterior &amp; posterior calvarial reconstructions at 3 &amp; 9 mos</td>
<td>555</td>
<td>1250</td>
<td>25</td>
<td>19</td>
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<tr>
<td>4</td>
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<td>475</td>
<td>200</td>
<td>6</td>
<td>14</td>
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<td>360</td>
<td>150</td>
<td>7</td>
<td>11</td>
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<tr>
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<td>21, F</td>
<td>Jackson–Weiss</td>
<td>anterior calvarial reconstruction at 6 mos</td>
<td>420</td>
<td>450</td>
<td>5</td>
<td>10</td>
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<tr>
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<td>7, F</td>
<td>Crouzon’s</td>
<td>none</td>
<td>440</td>
<td>500</td>
<td>6</td>
<td>8</td>
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</tbody>
</table>

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**FIG. 1.** Graphs depicting preoperative and postoperative horizontal (left) and vertical (right) cephalic indices in each of our seven patients, specified by case number. Normal ranges for each index (median ± two standard deviations), based on measurements in normal subjects compiled by Haas, are indicated by the space between the broken lines.
superiorly using the high-speed drill; they were compe-
ited using a straight osteotome. The orbital bar osteotomies
were continued posteriorly to incorporate a “tongue” of
the squamosal portion of the temporal bone, which would
play a role in the bone advancement (see below). If possi-
ble, the periosteum was left attached to the orbital bar
to facilitate vascularization and thus minimize the risk of
bone resorption. Because the lateral portion of the sphe-
noid bone was characteristically thickened and displaced
anteriortly and superiorly, this abnormal bone was partial-
ly resected.

The orbital bar was recontoured using wedge-shaped
partial thickness cuts through the posterior surface of the
bone to facilitate remodeling (Fig. 3 upper right, inset).
The bar was then advanced forward and inclined down-
ward slightly to correct not only the flattening but also the
abnormal angulation of the lateral orbital margin. Because
the entire anterior cranial base was small in these patients,
the bones on both sides medially and laterally were ad-
vanced at least 15 mm and, in some cases, as much as 25
mm (Fig. 3 upper right). In these severely affected pa-
tients, an effort was made to achieve as much of an ad-
vancement as the bone geometry and soft tissues would
allow. Bone grafts were secured to the posterior surface of
the orbital bar in the midline and over each orbit to main-
tain the bar in its advanced position. The advanced orbital
bar was also held forward using a tongue-in-groove tech-
nique (Fig. 3 lower right). The tongue of the orbital bar
was positioned in a groove that was fashioned within the
temporal bone inferior to the synostotic coronal suture to
discourage posterior displacement of the bar while al-
lowing further spontaneous advancement consistent with
brain growth. Twenty-eight-gauge wire was used to keep
the midline bone graft opposed to the posterior surface of
the orbital bar and to maintain the contour of the lateral
orbital angle. Elsewhere, absorbable (2-0 Vicryl) sutures
were used to secure the orbital bar in place.

Barrel stave cuts were made in the remaining occipital
bone, and the bones were greenstick fractured outward to
expand the skull posteriorly. The expansion was stabilized
by bridging each of the barrel staves to a bandeau of
occipital bone that had been excised, recontoured, and
replaced in a tongue-in-groove fashion into the remaining
occipital bone, posterior to the site of the lambdoid suture
(Fig. 3 lower right). By appropriately positioning the ban-
deau, the occipital bone could be advanced by at least 15
mm. The bandeau was held in place using absorbable
sutures except in the midline, where one or two of the bar-
rel staves were held in opposition to the bone strut using
28-gauge wire. In the two older patients, wire was also
used to secure the tongue-in-groove construct.

A fiberoptic ICP monitor (Camino Laboratories, San
Diego, CA) was placed through the right parietal bone
into the brain parenchyma and the cranial height was re-
duced by resecting a rectangular wedge from the biparietal
strips and gently tightening the bone edges together using
28-gauge wire (Fig. 4). If indicated, the vertex of the skull
was also shifted posteriorly by sliding the struts backward
before repositioning. Because the combined frontal and
occipital advancements substantially increased the anter-
posterior dimension of the calvaria before the height
reduction was performed, ICP often remained in the nor-
mal range during the height reduction. However, in those
patients in whom ICP did increase significantly during
this maneuver, great care was taken to avoid sustained ele-
vations above 20 mm Hg by tightening the wires down
gradually over a period of up to 45 minutes. In all cases,
ICP after completion of the height reduction was less than
15 mm Hg.

The frontal and occipital bones were then recontoured
and often reoriented to correct the brachycephalic shape of
occipital advancement technique

the calvarial vault by narrowing the abnormally wide appearance of the forehead and occiput (Fig. 4). The recontoured frontal bone segments were connected to the orbital bar using a combination of 28-gauge wire and sutures. A solid connection in conjunction with continuous bone resurfacing was employed to avoid cosmetically unacceptable gaps or irregularities. However, the bones were left free-floating toward the vertex so as not to restrict subsequent anterior skull growth. In the occipital region, the recontoured bone plates were sutured to the bandeau using 2-0 absorbable sutures and left free-floating toward the vertex. Thus, no metallic fixation was used in the vertex region to allow subsequent expansion of the cranial vault.

Postoperatively, the patients were maintained in a soft, padded helmet for approximately 1 month to encourage skull remodeling in an anteroposterior direction and to protect the construct in the event of a fall.1,11

Discussion

The techniques described in the current report were developed as a result of our frustration with the suboptimal cosmetic results achieved with anterior cranial vault remodeling for syndromic bilateral coronal synostosis associated with severe brachycephaly, and the recognition that, with such a limited approach, a significant percentage of children (such as those in our Cases 3 and 6) manifest cranial volume restriction that results in increased ICP during early childhood. The approach that we have adopted represents a modification of previous techniques used to correct brachycephaly.1,9,10 The notable alterations are the way in which the frontal and occipital bones are advanced and held in position to maintain the cosmetic correction while still allowing for progressive calvarial expansion. In particular, the advanced frontal and occipital bars are stabilized using a combination of a tongue-in-groove osteotomies and both absorbable and nonabsorbable sutures. This avoids the “loss of correction” that can occur after free-floating or weakly buttressed forehead and occipital advancements. The fixation this incision, healing was slow and she was maintained on intravenous antibiotics and local wound care as an inpatient. The incision eventually healed without the need for further intervention. The patient in Case 2 had a slightly increased length of hospitalization as a result of a flare-up of asthma. There has been no other morbidity and no mortality in these children.

On average, the frontal and occipital bones were each advanced by at least 1.5 to 2 cm and the height was reduced by 1.5 cm. Children had an expansion of 3.5 to 5 cm in their occipitofrontal circumference. Thus, the patients had a dramatic and immediate correction of their cosmetic deformity. In all cases, the horizontal and vertical cephalic indices were corrected to within normal range. However, it should be emphasized that we did not use these measures intraoperatively to guide the reconstruction, but instead tailored the procedure based on aesthetic grounds to provide the optimal cosmetic correction. Comparisons between preoperative and postoperative cranial indices are depicted in Fig. 1 left and right.

In view of the substantial objective changes in the patients’ calvarial anatomy, the families of these children rated the cosmetic result as good to excellent in each case. Two children have mild residual occipital flattening and two have hypertelorism and midfacial hypoplasia, as a result of their underlying syndrome, which will be corrected, if needed, when the children reach 4 to 5 years of age. Of critical importance is the fact that all seven children have been followed for more than 6 months after surgery and have demonstrated progressive cranial growth on serial measurements, which has followed normal percentile curves in each case, without recurrence of the turriccephalic head appearance. Representative preoperative and 12-month postoperative lateral CT reconstructions of one child are shown in Fig. 5 upper left and right illustrating the degree to which the turriccephaly and anteroposterior foreshortening can be corrected. Axial CT images obtained before and after reconstruction (Fig. 5 lower left and right) demonstrate the normalization of the horizontal cephalic index.

The results in our seven patients are summarized in Table 1 and Fig. 1. The median blood loss for the operation was 450 ml, approximately two to three times the typical blood loss in an anterior calvarial procedure. Only one patient (Case 3), who had undergone two prior reconstructive procedures, had a blood loss that exceeded 500 ml. Operating time including patient positioning and skin preparation ranged from 360 to 580 minutes (median 475 minutes), approximately twice the length of an average anterior calvarial reconstruction. The median hospital stay was 7 days, which is comparable to the length of stay for patients who undergo strictly frontoorbital advancement. The patient in Case 3, who had a prolonged hospitalization, had a superficial infection of her incision line. Because she had undergone two previous operations through this incision, healing was slow and she was maintained on intravenous antibiotics and local wound care as an inpatient. The incision eventually healed without the need for further intervention. The patient in Case 2 had a slightly increased length of hospitalization as a result of a flare-up of asthma. There has been no other morbidity and no mortality in these children.
achieved is sturdy enough to resist the compressive forces applied by the weight of the child’s head, but not so rigid that it restricts further calvarial growth. Our observation in all seven children, each of whom was followed for more than 6 months, that postoperative head growth remained within normal percentile curves indicates that the degree of fixation achieved does not severely limit further calvarial growth, at least in the short term.

Our rationale for performing the operation in previously untreated patients at 6 to 12 months of age rather than during the neonatal period was based both on the supposition that the lengthy procedure was likely to be safer in an older infant with larger blood volume and the observation that patients sometimes develop recurrent synostosis within 12 months after a calvarial reconstruction. This recurrence is less likely to be of cosmetic or functional significance if the operation is performed later in infancy. The rationale for not deferring the operation further was based on our concern that the calvarial growth restriction in these children might ultimately hamper their brain growth and that with the development of progressively worsening brachycephaly, the chances of achieving a favorable cosmetic result would be diminished.

An additional advantage of the technique outlined here is that significant height reduction can be achieved without excessive increases in ICP. Because the anterior and posterior bars are each advanced by at least 15 mm, it has often been possible to immediately reduce cranial height as much as approximately 15 mm without increasing the ICP above 20 mm Hg. However, we agree with Persing, et al.,10 that ICP should generally be measured during the course of the height reduction because some patients will experience transient elevations in pressure and, in such cases, the height reduction must proceed slowly and carefully to avoid sustained increases in ICP above 20 mm Hg. Several years before adoption of our current techniques, a patient with Crouzon’s disease, who had severe residual deformity after several frontoorbital advancements, underwent height reduction without ICP monitoring at our institution and suffered a partial sagittal sinus thrombosis and a subsequent venous infarction, resulting in profound mental deterioration.

Several other technical issues are of importance in ensuring patient safety during these extensive procedures. First, because blood loss often approaches one blood volume, intraoperative monitoring of hemodynamic status with a central venous line and an arterial line is essential. Hematocrit, platelet, and clotting parameters are also measured frequently during the operation to guide blood replacement. Two units of cross-matched blood (often obtained from family members who wish to provide “directed” donation) is immediately available in a refrigerator outside the operating room. Second, because of the possibility of air embolism, a precardial Doppler ultrasound probe is placed and end-tidal CO2 is measured during surgery. Fortunately, we have not encountered this problem in any of our cases. Third, because of the high potential for hypothermia in small children, which can lead to hemodynamic compromise, a circumferential heating blanket is maintained around the trunk and lower extremities and rectal temperature is measured throughout the operation. Finally, all patients are monitored after surgery in an intensive care unit for at least 24 hours.

As a final caveat, it should be emphasized that management of patients with bilateral coronal synostosis must be individualized. Children who exhibit little or no brachycephaly may be adequately treated with frontoorbital advancement alone. Conversely, children with synostosis of multiple sutures, such as those with kleeblattschädel, may require calvarial expansion during the first few days of life to relieve their severely increased ICP.11 Because the calvarial bone in such neonates is often diffusely abnormal, conventional advancement techniques are often infeseable and alternate approaches must be used.12 In addition, patients who exhibit radiographic evidence of cervical hypermobility are not suitable candidates for the modified prone position because of the potential for injury to the cervical spinal cord or stretching of the vertebral arteries. Such patients are better managed in a staged fashion using alternative positioning techniques.5

It is also important when considering the performance of total calvarial reconstruction to be aware of the disadvantages of this approach in comparison to more limited procedures. First, blood loss and operating time are doubled. Second, the potential for serious morbidity and life-threatening bleeding are increased. Third, the consequences of a severe wound infection are more serious because this could potentially require removal of the devascularized bone grafts over both the anterior and posterior calvaria. Finally, the frontoorbital advancement component of the procedure is technically more demanding when performed with the patient in the prone rather than
Occipital advancement technique

the supine position because the brain tends to fall forward and obscures the exposure of the frontal floor during the orbital osteotomies. In view of the advantages and disadvantages of the various techniques available for calvarial reconstruction, decisions regarding the management of patients with complex craniosynostosis problems are best made by a multidisciplinary craniofacial team coordinated by both a plastic surgeon and a neurosurgeon.

Summary

In summary, our technique of total calvarial reconstruction has provided gratifying cosmetic results with an acceptable increase in the overall complexity of the operation in comparison to conventional frontoorbital advancement procedures. These technical modifications have enhanced the stability of the reconstruction without restricting future calvarial growth and, potentially, may improve the quality of the cosmetic result, although long-term follow-up study will be required to assess the latter point more conclusively. We feel that the issue of timing of the surgery is critical to achieving an acceptable cosmetic and functional outcome with low morbidity. By delaying the operation until 6 to 12 months of age in children without evidence of increased ICP, we have been able to reconstruct the skull to nearly adult dimensions, allowing the potential for continued calvarial growth. At this age, with the aforementioned caveats, we have found the operation to be relatively safe and well tolerated.

References


Manuscript received June 14, 1995. Accepted in final form September 13, 1995. Address reprint requests to: Ian F. Pollack, M.D., Department of Neurosurgery, Children’s Hospital of Pittsburgh, 3705 Fifth Avenue, Pittsburgh, Pennsylvania 15213.