The “clamshell” craniotomy technique in treating sagittal craniosynostosis in older children

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Object. Although most patients with sagittal craniosynostosis are recognized and treated in infancy, some children are not referred to craniofacial centers until later in childhood. In this paper the authors describe a novel operative technique for calvarial reconstruction in older children with previously untreated sagittal craniosynostosis.

Methods. The authors report a clinical series of eight patients who were treated using novel single-stage calvarial reconstruction, and they assess the complications and outcomes. The patient is placed supine for the procedure, which consists of a coronal incision, bifrontal craniotomy without orbital osteotomy, and multiple interlocking midline parietooccipital osteotomies and recontouring. Fixation is achieved using a bioabsorbable plate system. Cranial indices were calculated from measurements obtained before and after the reconstructive procedures. Preoperative, intraoperative, and postoperative photographs and three-dimensional computed tomography scans are presented for review.

Between November 2003 and April 2005, the authors treated seven boys (age range ~1–10 years, mean age 4.2 years) with uncorrected sagittal craniosynostosis and one with bicoronal and sagittal synostosis. The mean operating time was 5.13 hours (range 4.3–8 hours), with a mean blood loss of 425 ml (range 200–800 ml). As a percentage of the estimated circulating blood volume, the mean operative blood loss was 33.5% (range 17–57%). The mean hospital stay was 4.9 days. The cranial index significantly improved from a mean of 65.6 to 71.3% (p = 0.001). No acute or delayed complications have been noted. Follow-up examinations performed at an average of 12 months (range 1–17 months) have confirmed early patient and family satisfaction.

Conclusions. An approach of aggressive calvarial reconstruction with multiple interleaving osteotomies crossing the midline achieves improvements in biparietal narrowing. Combined with a bifrontal reconstruction, early outcomes are excellent, with an acceptable amount of intraoperative blood loss and no significant complications.

KEY WORDS • craniosynostosis • sagittal synostosis • delayed repair • calvarial reconstruction • pediatric neurosurgery

Craniosynostosis is a common malady in pediatric craniofacial surgery and affects approximately one in 2100 children.7 Whereas most instances represent sporadic cases involving a single cranial suture, the condition may develop secondarily due to systemic disorders or as part of a syndrome, such as Crouzon, Apert, or Pfeiffer, which represent the most common forms of its syndromic manifestation.6 The sagittal suture is most frequently involved, representing approximately 56 to 58% of cases of single-suture nonsyndromic craniosynostosis.2,11 The hallmarks of sagittal craniosynostosis include frontal bossing and biparietal narrowing producing a prominent sagittal ridge, giving the toddler a so-called scaphocephalic head shape, named so after its resemblance to a boat keel (from Greek “skaphos” meaning a boat or bowl).

Although most cases of sagittal craniosynostosis are recognized and treated in infancy, some children are not referred to craniofacial centers until later in childhood. By this time, the child’s cranial vault has developed abnormally resulting in severe skull base deformity.9 Subsequently the child’s skull will be significantly thicker and less pliable to reshaping. After 1 year of age, cranial defects due to aggressive surgical remodeling procedures may not reossify. Finally, there is a sharp decline in the driving mechanism that leads to closure of the sutures.3,14 These additional difficulties present a particular challenge to the surgeon when treating this subset of patients with sagittal synostosis.

Researchers at many centers have reported their experience and various techniques in the treatment of infants with sagittal craniosynostosis,1,5,11,15,17 yet relatively few series have been published in which the authors address the surgical management of this problem in previously surgically
untreated older children. It should be noted that there is currently no consensus about the indications for treatment of these patients, nor for the surgical techniques used. We describe our operative technique for calvarial reconstruction in eight older children who had previously untreated sagittal craniosynostosis.

### Clinical Material and Methods

#### Preoperative Evaluation

A total of eight patients underwent this procedure between November 2003 and April 2005. Patient ages ranged from approximately 1 to 10 years (mean 4.2 years), and all were boys. The patients were evaluated by the plastic surgery and neurosurgery teams; medical history recording and physical examinations were performed at the initial visit. Cranial indices were calculated using caliper measurements taken before and after the reconstruction procedures, and preoperative digital photographs were taken for later comparison. In one patient with failure to thrive and bicoronal and sagittal suture involvement, an additional referral was made for genetic counseling and testing.

Each patient underwent a preoperative neuroimaging evaluation in which a bone-window algorithm and 3D reconstructed CT scans of the head were obtained to delineate the extent of the sutural fusion and to evaluate the patency of the remaining sutures. Furthermore, the reconstructed CT scans were used to aid in the operative planning for the reconstructive procedures. Laboratory testing including complete blood counts and clotting studies was also performed. Two to four units of packed red blood cells were cross-matched prior to the procedure. All patients' parents signed informed consent for the procedures as well as for photographic documentation for clinical management and research publication.

#### Intraoperative Positioning and Monitoring

Intraoperative monitoring consisted of placement of a central venous catheter or two large-bore peripheral venous catheters, an arterial catheter, and an end-tidal CO₂ monitor to assess the patient’s volume status as well as for possible air emboli. An intravenous antibiotic agent (cefazolin) was administered to all patients prior to making the skin incision. The patients were placed supine, with a molded vacuum bean bag placed under the occiput, neck, and shoulders. Horizontal gel rolls were positioned under the legs. Additional foam padding was placed at all pressure points, and the head was maintained in flexion taking care not to kink the reinforced endotracheal tube. Care was taken to provide enough flexion to access the occipital “bullet” (that is, the osseous bulge at the base of the skull) but not to impede the endotracheal tube or restrict cranial venous outflow (Fig. 1). During the procedure, frequent communication with the blood bank was maintained to ensure that an adequate amount (at least one circulating blood volume) of blood products was available.

### Surgical Technique

An ear-to-ear coronal incision is marked in a zig-zag fashion posterior to the coronal suture. A small band of hair is shaved in the region of the planned incision (Fig. 1C), and a sterile preparation and draping technique is performed in which the entire cranial vault from the supraorbital ridge to below the inion is exposed. The scalp incision is made using a Colorado microdissection needle cautery (Stryker-Leibinger, Portage, MI), and the anterior and frontal scalp flaps are dissected, preserving the pericranium to prevent excessive oozing. Once the cranial vault is exposed, the planned osteotomies and frontal craniotomy incisions are drawn on the surface (Fig. 2A). Bur holes are placed bridging the sagittal sinus near the coronal and lambdoid sutures (lamba and bregma), as well as the frontotemporal region just above the lesser wing of the sphenoid bone bilaterally (Fig. 2B). First, a bifrontal craniotomy is performed, just anterior to the coronal sutures and 1 cm above the orbital ridge bilaterally. The frontal bone is removed and taken to the back table of the operating room for recontouring (Fig. 2C) while the biparietal and occipital osteotomies are performed.

Although the pericranium had been left intact on the frontal bone, at this point separate lateral pericranial flaps are brought down from the midline, extending anteriorly to the coronal suture and posteriorly to the lambdoid suture, exposing the squamosal sutures inferolaterally at their base. To widen the biparietal constriction, multiple interleaving barrel-stave osteotomies are marked 15 to 18 mm apart, alternating as they cross the midline. These are returned to the region of the lambdoid suture, where their orientation is...
turned to provide some flattening of the occipital bullet (Fig. 3). As each osteotomy is performed, sequential dissection of the dural superior sagittal sinus is performed and bleeding points can be secured using bipolar cautery or GelFoam (Pfizer Inc., New York, NY).

All barrel-stave sections are fractured and recontoured as necessary, and bioabsorbable plating material is used to secure them into their new position. This allows for 10 to 15 mm of lateral expansion, leaving a checkerboard pattern of small cranial defects near the midline (Fig. 3). Generally the frontal bone is bisected in the midline, rotated 180˚, and fixed in place by using bioabsorbable plating material to correct the frontal bossing (Fig. 4). The previously repositioned lateral pericranial flaps are tacked back into place, and the galea and scalp are closed over two bulb-suction drains. A sterile head-wrap dressing is placed providing gentle compression.

One patient (Case 8, Fig. 5) underwent noninterlocking parietal barrel-stave osteotomies that did not cross the midline: split-thickness parasagittal autografts were used for the cranial expansion. Although this procedure minimized the residual cranial defects, the reconstruction was more involved and time consuming than the simpler plating and reconstruction method for interleaving osteotomies crossing the midline.

Postoperative Care and Follow-up Course

Intravenous cefazolin is continued after the procedure until discharge from the hospital. For the first 24 hours, serial hematocrit levels are checked every 6 hours, and drainage fluid is closely monitored while the child is in the intensive care unit. The head wrap is loosened on postoperative Day 1, when most patients are transferred out of the intensive care unit to the surgical ward. Operative drains are typically removed after 36 hours. Acetaminophen, ibuprofen, and narcotic agents are given routinely for pain control. A 3D CT reconstruction of the head is obtained prior to discharge from the hospital. Patients are routinely followed up.

Fig. 2. Intraoperative photographs. A: After the cranial vault is exposed, planned osteotomies are marked on the surface. B: Bur holes are placed bridging the sagittal sinus near the coronal and lambdoid sutures, as well as the frontotemporal region just above the lesser wing of the sphenoid bone bilaterally. C: After the bifrontal craniotomy is performed 1 cm above the orbital rim, the frontal bone is removed and taken for recontouring.

Fig. 3. Intraoperative photographs (A and C) and 3D reconstructed CT scans (B and D) showing the midline and occipital reconstruction. A: Occipital osteotomies. B: Image obtained immediately postoperatively, showing the occipital reconstruction. C: Midline osteotomies. D: Image obtained immediately postoperatively showing the midline reconstruction.
Fig. 4. Frontal reconstruction. A and B: Intraoperative photographs showing frontal bone inversion to correct frontal bossing. C: In some cases, patients had a split frontal craniotomy to correct bossing as can be seen in this postoperative 3D CT reconstruction.

Fig. 5. Preoperative, immediately postoperative, and 1-year follow-up 3D reconstructed CT scans obtained in all eight patients. Two patients have not yet undergone 1-year follow-up CT scanning.
Delayed sagittal synostosis repair

Table 1

Characteristics and operative details in eight patients who underwent calvarial reconstruction for sagittal synostosis*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs)</th>
<th>Weight (kg)</th>
<th>ECBV (ml)</th>
<th>Diagnosis</th>
<th>EBL (ml)</th>
<th>% ECBV</th>
<th>Duration of Stay (days)</th>
<th>OR Time (hrs)</th>
<th>Preop Cranial Index (%)</th>
<th>Postop Cranial Index (%)</th>
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<tr>
<td>1</td>
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<td>8.4</td>
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<td>29.8</td>
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<td>4.5</td>
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<td>78</td>
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<tr>
<td>2</td>
<td>1.2</td>
<td>5.2</td>
<td>442</td>
<td>pansynostosis</td>
<td>250</td>
<td>56.6</td>
<td>5</td>
<td>4.3</td>
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<tr>
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<td>sagittal CS</td>
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<td>76</td>
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<td>33.5</td>
<td>4.9</td>
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* CS = craniosynostosis; EBL = estimated blood loss; ECBV = estimated circulating blood volume; OR = operating room.

2 to 3 weeks postoperatively and then undergo additional examinations at 3 and 6 months, then annually.

Results

Patient characteristics and operative details are provided in Table 1. All patients had sagittal craniosynostosis with typical clinical features (Figs. 5 and 6); one (Case 2) also had multiple suture synostosis causing cranial constraint and concern for elevated intracranial pressure. The mean operating time was 5.13 hours (range 4.3–8 hours), and the mean blood loss was 425 ml (range 200–800 ml). As a percentage of the estimated circulating blood volume, the mean operative blood loss was 33.5% (range 17–57%). The average hospital stay was 4.9 days (range 4–6 days). The cranial index improved from a mean of 65.6 to 71.3% (p = 0.001, paired Student t-test). No acute or delayed complications have been noted in any patient, and the follow-up examinations at an average of 12 months (range 1–17 months) have confirmed early patient and family satisfaction.

Discussion

Experience has shown that the optimal time for surgical intervention for the treatment of craniosynostosis is early in the natural history of the disease (when the child is < 9 months old).1,8,17 The challenge then often becomes early recognition and diagnosis of the disorder, and referral to the appropriate specialists. However, this initial screening task is performed by general family practitioners or pediatricians who may have limited experience in recognizing the early signs of craniofacial abnormalities, particularly in cases in which there are more subtle defects but no other associated anomalies.8 These nonsyndromic single-suture synostoses are usually the most amenable to surgical intervention when identified early. Delays in diagnosis, as well as initial refusal by some parents to proceed with surgical correction in infancy, result in a small subset of patients with uncorrected craniosynostosis necessitating the development of calvarial reconstruction techniques.

Due to calvarial maturity, more advanced cases of sagittal synostosis are not amenable to treatment using less extensive techniques, such as endoscopic strip craniectomy and placement of a molded orthosis,2 or a simple barrelosteotomy.12 Because of the natural history of craniosynostosis and the abnormally growing calvaria causing elongation of the skull base, techniques incorporating a total calvarial reconstruction are required to attain a reasonable cosmetic appearance in these patients. The complexity of the surgical intervention needed for these patients requires cooperation among a dedicated craniofacial team, including neurosurgeons, plastic surgeons, and anesthesiologists who are familiar with the intraoperative complexities and pitfalls associated with these procedures.

Several authors have described surgical techniques for the correction of delayed sagittal synostosis.3,4,10,14,16 The authors of previous reports on total calvarial vault reconstruction have indicated that there is a potential for morbidity due to operative blood loss (that is, 0.5–5 times the amount of the normal circulating blood volume).3,4,10,14,16 We have herein described a technique of aggressive total calvarial reconstruction that we recommend for the correction of delayed sagittal synostosis. Patient and family satisfaction has been uniformly high, and this technique has resulted in significantly less blood loss (17–57% of the circulating blood volume) than that reported in previous series. Blood loss is minimized by limiting the subperiosteal dissection and by meticulous adherence to hemostasis. No acute or delayed complications have been noted, although the potential for cranial defects to occur at the expansion sites remains; long-term follow-up success of the procedure has not yet been confirmed. Despite the high level of satisfaction, we routinely inform families that they should not expect this procedure to completely normalize their child’s head shape. In our experience with this technique, improving the frontal contour (reduction of bossing) and moderate widening of the biparietal diameter have been the greatest accomplishment. We caution families not to expect significant narrowing in the anteroposterior dimension.

Conclusions

An approach of aggressive calvarial reconstruction with multiple interleaving osteotomies crossing the midline leads to improvements in biparietal narrowing. Combined with a bifrontal reconstruction, early outcomes associated with this technique are good; the amount of intraoperative blood loss is acceptable and there are no significant complications. Although these early results could be expected to correlate
Fig. 6. Preoperative and postoperative frontal, lateral, and vertex photographs obtained in all eight patients. The age at which each operation was performed is shown in the left-hand column.
with long-term outcome due to the calvarial maturity in this population of older children, continued follow-up examinations of these patients will be required to fully validate the usefulness of this technique.

References


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