Nonendoscopic, minimally invasive calvarial vault remodeling without postoperative helmeting for sagittal synostosis

Clinical article

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Object. Multiple surgical procedures have been described for the management of isolated nonsyndromic sagittal synostosis. Minimally invasive techniques have been recently emphasized, but these techniques necessitate the use of an endoscope and postoperative helmeting. The authors assert that a safe and effective, more “minimalistic” approach is possible, avoiding the use of endoscopic visualization and routine postoperative application of a cranial orthosis.

Methods. A single-institution cohort analysis was performed on 18 cases involving infants treated for isolated nonsyndromic sagittal synostosis between 2008 and 2010 using a nonendoscopic, minimally invasive calvarial vault remodeling (CVR) procedure without postoperative helmeting. The surgical technique is described. Variables analyzed were: age at time of surgery, sex, estimated blood loss (EBL), operative time, intraoperative complications, postoperative complications, length of stay, pre- and postoperative cephalic index (CI), clinical impressions, and results of a 5-question nonstandardized questionnaire administered to patient caregivers regarding outcome.

Results. Eleven male and 7 female infants (mean age 2.3 months) were included in the study. The mean duration of follow-up was 16.4 months (range 6–38 months). The mean procedural time was 111 minutes (range 44–161 minutes). The mean length of stay was 2.3 days (range 2–3 days). The mean EBL in all 18 patients was 101.4 ml (range 30–475 ml). One patient had significant bone bleeding resulting in an EBL of 475 ml. Excluding this patient, the mean EBL was 79.4 ml (range 30–150 ml). There were no deaths or intraoperative complications; one patient had a superficial wound infection. The mean CI was 69 preoperatively versus 79 postoperatively, a statistically significant difference (p < 0.0001). Two patients were offered helmeting for suboptimal surgical outcome; one family declined and the single helmeted patient showed improvement at 2 months. No patient has undergone further surgery for correction of primary deformity, secondary deformities, or bony irregularities. Complete questionnaire data were available for 14 (78%) of the 18 patients; 86% of the respondents were pleased with the cosmetic outcome, 92% were happy to have avoided helmeting, 72% were doubtful that helmeting would have provided more significant correction, and 86% were doubtful that further surgery would be necessary. Small, palpable, aesthetically insignificant skull irregularities were reported by family members in 6 cases (43%).

Conclusions. The authors present a nonendoscopic, minimally invasive CVR procedure without postoperative helmeting. Their small series demonstrates this to be a safe and efficacious procedure for isolated nonsyndromic sagittal synostosis, with improvements in CI at a mean follow-up of 16.1 months, commensurate with other techniques, and with overall high family satisfaction. Use of a CVR cranial orthosis in a delayed fashion can be effective for the infrequent patient in whom this approach results in suboptimal correction.

key Words • pediatric neurosurgery • craniosynostosis • sagittal suture • calvarial vault remodeling

THE optimal surgical management of isolated nonsyndromic sagittal synostosis remains unclear. While the gold standard has been open CVR with large scalp incisions and extensive bone work resulting in significant blood loss, both historic and current literature provide evidence that limited procedures can yield acceptable results with less morbidity.5,8,12,13,17 Despite the long history of research into the many methods of treating this condition, there is still considerable disagreement regarding how limited these techniques can become while still yielding satisfactory outcomes. Two significant questions arise when currently addressing the limitations of minimally invasive surgery for the treatment of isolated nonsyndromic
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sagittal synostosis. First, is endoscopy required for the safe removal and release of bone? Second, must a postoperative molding helmet be used? Since late 2008, the senior author (T.A.M.) has used a minimally invasive technique for sagittal synostectomy and calvarial release that requires no endoscopy and no postoperative helmiting. In this paper we publish results for our initial group of patients with 6 or more months of follow-up.

Methods

Patient Variables

A search of institutional databases for patients who underwent minimally invasive procedures for the treatment of isolated nonsyndromic sagittal synostosis between 2008 and 2010 yielded 18 patients, all treated by the senior surgeon (T.A.M.). Approval was obtained from the institutional review board at Cincinnati Children’s Hospital Medical Center to perform this analysis.

A retrospective chart review of these 18 cases was conducted. Data extracted included patient age at time of surgery, sex, weight, and preoperative CI. Intraoperative data included length of surgery, EBL, and complications. Postoperative data included hospital length of stay and complications. Outpatient parameters included length of follow-up, CI at last follow-up, description of clinical findings (cosmesis, contour, and bony defects), incidence of helmiting, and incidence of revision surgery. Preoperative imaging was generally performed by the referring physician and few postoperative imaging studies were acquired to evaluate new or persistent calvarial irregularities and deformities.

In addition, all families were administered a 5-item nonstandardized survey in the conduct of patient care. No members of the surgical team participated in this process. During a routine clinic or telephone follow-up session, a parent or care provider was asked the following 5 questions: 1) “Are you pleased with the cosmetic outcome?” 2) “Are you happy to have avoided a molding helmet?” 3) “Have you wondered if a helmet would have made the head shape better?” 4) “Are you worried that repeat surgery will be needed?” and 5) An open-ended question to elicit comments and concerns.

The CI was determined by measurements of plain radiographs, CT scans, standardized photographs, or by direct measurement of the head during patient examination. The CI was determined in the standard manner, dividing the euryon-to-euryon cranial width by the glabella-to-opisthocranion cranial length.

Surgical Technique

Following the induction of general anesthesia, nasotracheal intubation was performed. Supine or modified sphinx positioning technique was used. For the latter, the body was cradled in a well-padded, vacuum-solidified beanbag with the head and neck minimally extended and stabilized laterally. This optimized exposure to the entire vertex and occipital region for surgery. All patients received appropriate intravenous and arterial catheters as well as Foley bladder catheters. Nine (50%) of our patients received intraoperative tranexamic acid infusions to limit bleeding, dosed at 100 mg/kg for an initial bolus and then 10 mg/kg as a continuous drip until skin closure was completed.

After the scalp was prepared with antiseptic solution and small aliquots of 0.4% lidocaine with epinephrine at 1:200,000 were instilled, 3 knife incisions were made, each 3 cm in length: 2 transversely at the anterior and posterior fontanels and 1 longitudinal over the midline, halfway between the anterior and posterior incisions (Fig. 1A). Skin hemostasis was achieved using bipolar cautery, and incisions were then deepened with needle monopolar cautery through the galea, leaving the periosteum intact.

Using Metzenbaum scissors, the plane between the galea and periosteum was well developed for the width of 1.5–2 cm to both sides of the midline between the anterior and posterior wounds. The epidural space was entered at the fontanels, using curettage and punch osteotomes to create sufficient working space. Using a blunt instrument such as a brain ribbon, the epidural space on both sides of the fused sagittal suture was developed. Under direct visualization, straight Tessier cartilage scissors were introduced into the epidural and subgaleal planes, making bilateral parietal bone cuts (Fig. 1B) to create a strip containing the fused sagittal suture measuring 2–3 cm in width (Fig. 1C). The cartilage scissors were then used to release the coronal sutures (through to the perion) and lambdoid sutures (to the asterion). The middle incision was used as an access point for the creation of 3–4 barrel-stave osteotomies oriented radially toward the sphenoparietal and squamosal sutures (Fig. 1D). These cuts were typically made with partial visualization superiorly and palpation of the scalp inferiorly, with the blunt scissors blades gently dissecting within the subgaleal and

Fig. 1. The nonendoscopic, minimally invasive CVR procedure. A: Three 2–3 cm scalp incisions are planned: at the anterior fontanel, the posterior fontanel, and midway between. B: The Tessier cartilage scissors are introduced into the subgaleal and epidural spaces. C: A 3-cm wide strip craniectomy is made with the scissors. D: Bilateral barrel-stave parietal bone osteotomies are made with the scissors. Additional osteotomies are made along the lambdoidal suture and occipital bone, if necessary.

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epidural planes. The rostral edges of the barrel staves were trimmed as necessary to prevent overlap of the bone “fingers.” To allow reduction of the occipital bulb when necessary, bilateral cuts are made 2 cm off the midline from the lambda to the inferior aspect of the bulb and the segment is greenstick fractured.

Prior to 2009, the strip was not routinely replaced. Based on the presence of small bony defects more than 6 months postoperatively, the bone was replaced in most subsequent cases, suturing it to the dura in a fashion identical to open technique previously described by the senior author. Hemostasis was obtained using bone wax and/or Floseal Hemostatic Matrix (Baxter). The scalp incisions were then closed in 2 layers using interrupted 4-0 Vicryl (Ethicon) suture for the galea and a running 5-0 Monocryl (Ethicon) for the skin; they were then covered with a sterile dressing. For no patient was an endoscope used. No patient was prescribed a postoperative molding helmet.

Results

Patient Population

Eighteen patients (11 male and 7 female infants) were treated between 2008 and 2010. The mean duration of follow-up was 16.4 months (range 6–38 months). All patients had both clinical appearance and preoperative imaging (plain skull radiographs or CT) consistent with this diagnosis. The patients’ mean age at time of surgery was 2.3 ± 0.69 months and their mean weight was 5.6 ± 0.63 kg (Table 1).

Perioperative Care

The mean surgical time (from incision to the placement of a dressing) was 111 minutes (range 44–161 minutes; generally less than 75 minutes in recent cases). The mean length of stay was 2.3 days (range 2–3 days). The mean EBL was 101.4 ml (range 30–475 ml). Calculated as a fraction of estimated blood volume (kg × 80) the mean was 23% (range 7%–108%). One patient had significant bone bleeding without laboratory evidence of coagulopathy, resulting in an EBL of 475 ml. The mean EBL and fractional blood loss values for the other 17 patients were 79.4 ml (range 30–150 ml) and 18% (range 7%–40%), respectively. Sixteen patients (89%) required an intraoperative transfusion of packed red blood cells, with a mean weight-adjusted volume of 25 ml/kg (range 0–87.3 ml/kg). All transfusions were initiated by the anesthesia service and were not based on any specific criteria; our anesthesia service maintains a low threshold for transfusions in order to avoid complications of anemia. There were no deaths or serious complications. There were no dural lacerations or violations of the sagittal sinus. There were no other intraoperative or postoperative complications.

Clinical, Morphometric, and Cosmetic Outcomes

The mean CI for the 17 patients with complete mea-

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<th>Table 1: Demographic and clinical characteristics of 18 patients who underwent nonendoscopic, minimally invasive CVR*</th>
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* EBV = estimated blood volume; FU = follow-up; LOS = length of hospital stay; NA = data not available.
† Weight-adjusted volume.
‡ Denotes use of tranexamic acid.
§ This patient was prescribed a molding helmet for acquired posterior plagiocephaly.
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surement data sets was $69 \pm 7.7$ (SD) preoperatively versus $79 \pm 4.4$ postoperatively; this is a highly statistically significant difference ($p < 0.0001$, paired Student t-test). All CIs used in the postoperative analysis were obtained at the patient’s last visit (a mean of 16.1 months after surgery).

After 2008, no significant bony defects were appreciated beyond the 1st postoperative year. One patient had a superficial wound infection requiring oral antibiotics but not surgical revision. Minor palpable bony irregularities were noted by the examining health care provider at last follow-up in 6 patients. These were felt to be cosmetically insignificant. Figure 2 presents photographs of a male patient treated at 3.7 months of age with follow-up at 38 months after the procedure. Improvement in the CI, frontal bossing, temporal and parietal pinching, and mild turriccephaly are demonstrated.

Based on the senior neurosurgeon’s impression of incomplete correction, 2 patients were offered helmeting. The first patient had a preoperative CI of 68.0 and a postoperative CI of 74.3 and at 6 months postoperatively demonstrated persistent scaphocephaly, based on subjective assessment, as well as posterior brachycephaly, which persisted to the 9-month follow-up visit. The patient’s family declined further intervention. The second patient had a preoperative CI of 79.7 and a postoperative CI of 81.3 but had a significant posterior brachycephaly both pre- and postoperatively. This patient’s family elected to trial a remodeling helmet and the patient has demonstrated significant improvement in his posterior brachycephaly. No patient to date has undergone further surgery for correction of primary deformity, secondary deformities, or bony irregularities.

Clinical Questionnaire

We were able to further assess outcome in 14 (78%) of 18 cases through a series of questions presented to family members by a non–neurosurgeon care team member. In the majority of cases the family members were: pleased with the cosmetic outcome (12 [86%] of 14 cases), happy to have avoided helmeting (13 [93%]), doubtful that helmeting would have provided more significant correction (10 [71%]), and doubtful that further surgery would be necessary (12 [86%]). Minor, palpable, aesthetically insignificant skull irregularities were reported in 6 cases (43%).

Discussion

Based on an incomplete understanding of the pathophysiology of craniosynostosis and the considerable clinical variability of deformities that result, the surgical management of infants with these cranial anomalies remains as much an art as a science. During the past 50 years, many procedures have been proposed and presented, with many originators and champions presenting their case series in attempts to demonstrate superiority. Early authors believed that it was important to release fused sutures to prevent high intracranial pressures, allowing more morbidity to be tolerated. Since it became clear that most patients with single-suture synostosis did not suffer from elevated intracranial pressure, the indication for operative correction of isolated nonsyndromic sagittal synostosis became primarily an aesthetic issue, placing responsibility on surgeons to provide effective deformity correction with minimal morbidity.

After Lannelongue reported the first surgical attempt at curing sagittal synostosis, strip craniectomies became widely used to treat this disease. The popularity of the strip craniectomy waned in the 1980s and 1990s as multiple studies showed improved results using several

Fig. 2. Preoperative (A–C) and postoperative (D–E) photographs of a male patient who underwent a nonendoscopic, minimally invasive CVR procedure at the age of 3.7 months. He was 42 months old at the most recent follow-up visit and has shown improvement in CI, temporal and parietal pinching, and frontal bossing.
of our approach. However, in 1998, Jimenez and Barone began a reversal of this trend when they reported on their experience with endoscopic strip craniectomies followed by molding helmets. Over the past decade, this method has enjoyed further empirical support for its safety and aesthetic outcomes. 

Despite conclusive evidence of patient benefit with a minimally invasive approach to INSS, a paucity of compelling data exists to justify the de facto use of an endoscope for safety and efficacy. Similarly, robust evidence to support the need for routine postoperative helmeting to assure acceptable cosmetic outcomes is lacking. Although one group has reported on the use of the microscope rather than endoscope use, patients in this series received helmeting which makes it difficult to differentiate the effect of surgical technique versus orthosis on outcome. A single study presents the outcomes of patients treated with helmets versus those not treated with helmets following minimally invasive cranial remodeling techniques. Although differences were reported in favor of the helmet-treated group, the number of non–helmet-treated patients was extremely small (6), the surgical technique unclear, and the clinical and cosmetic relevance of these differences in CI uncertain. Furthermore, treating infants with helmets following open craniosynostosis and craniofacial procedures remains uncommon, supporting a hypothesis that cranial orthosis application is not essential to produce good cosmetic outcomes. Finally, the time, effort, and financial, physical, and emotional impacts of helmet application on the patient and family are substantial.

The senior author has had extensive experience with open, biparietal-widening, occipital-reduction CVR without postoperative helmeting in 3- to 6-month-old infants affected by isolated nonsyndromic sagittal synostosis. Recently we transitioned into a minimally invasive approach, performed at a slightly younger age range with avoidance of large bicoronal incisions and complete scalp dissections. Because the extent of bony remodeling was nearly identical to that of the open approach, we believed that helmeting was not required in the majority of patients and should remain an option rather than a required part of treatment. This review of our first 18 cases in which patients were so treated offers an opportunity to perform a critical appraisal of our approach.

Our general surgical technique is based on the approach of Jimenez with limited scalp incisions and dissection that differentiate it from an open CVR approach. We have incorporated a third incision at the vertex to allow for the performance of accurately directed, radially oriented osteotomies in the parietal bone. This maneuver is essential to afford proper lateral expansion of the parietal regions over time and produces no significant increased negative effects on the patient. We combine direct visualization of the calvaria using loupe magnification with nonvisualized but palpated direction of Tessier cartilage scissors into the subgaleal and epidural spaces. Given the consistent anatomical definition of these planes and the use of a blunt-tipped instrument, we have not placed patients at increased risk; this is borne out by our complete lack of inadvertent durotomies or scalp injuries.

Proper education of caregivers and careful postoperative follow-up are requisite for good cosmetic outcomes. Head positioning maneuvers are reviewed and positioning devices such as Z-Flo fluidized positioners (Sundance Enterprises Inc.) are provided to the caregivers during the initial hospital stay. Strategies for managing calvarial asymmetries are reviewed at each clinical follow-up visit.

Our surgical and postoperative outcomes were consistent with others who have presented their series of endoscopically assisted sagittal synostosis procedures. We have encountered no procedure-related complications or immediate postoperative complications. Scalp swelling and postoperative pain are minimal due to limited dissection; this allows prompt discharge from hospital on non-narcotic analgesics.

Morphometric and cosmetic outcomes are also consistent with results reported in published endoscopic series. All of our patients were followed up for at least 6 months (mean of 16.1 months). Therefore, we believe our correction is robust and will be sustained, as asserted in the neurosurgical literature, wherein a sustainable correction is generally achieved around 6 months after surgery. We achieved our primary objective by significantly improving the CI into the normocephalic range (∼74–83) for the group as a whole (mean value) and for the majority (83%) of patients individually. Three patients persisted with mildly scaphocephalic CIs; however, only one was perceived by health care providers and parents as remaining cosmetically undercorrected. Only one of our patients had a brachycephalic CI, and this may have been related to positional plagiocephaly; he is now benefiting from helmet correction.

Although cognitive and social scientists correctly assert that parental impressions following surgery are biased toward expressing positive outcomes, we believe that our families presented valid opinions about cosmetic outcomes. Based on the results of a nonstandardized questionnaire, most were very pleased with the appearance of their child’s head shape. The overwhelming majority was pleased to have avoided helmeting. Of the few parents who remained uncertain about the need for possible future surgery, their concerns were based on minor bony deficiencies and protuberances, not the overall head shape.

As acknowledged by family members and the senior neurosurgeon, 2 patients had suboptimal outcomes: one with persistent scaphocephalic deformity and another with moderate posterior brachycephaly. Even when helmet application was recommended, one family refused this treatment. The other patient was treated with a helmet and has demonstrated an excellent and rapid correction. While our data are too limited to make definitive conclusions about the role of helmeting as a “salvage” intervention, we believe our strategy is sound, based on the overall experience with postoperative helmeting in this patient population.

Finally, we assert that the simplicity of this technique cannot be overemphasized. This procedure is especially germane for surgeons unskilled in the use of endoscopic equipment or when endoscopic equipment is unavailable or infeasible. The instrument requirements are minimal and inexpensive (basic surgical instruments and Tessier cartilage scissors). Indeed, surgeons practicing in under-

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developed countries can reasonably conduct “minimally invasive” craniosynostosis procedures with truly minimal resources, benefiting patient populations that might otherwise have been denied the opportunity for deformity correction.

Conclusions

We present a nonendoscopic, minimally invasive CVR procedure without postoperative helmeting for the correction of isolated nonsyndromic sagittal synostosis. Our analysis of this small series demonstrates the safety and efficacy of this technique. We assert that a “minimallistic” surgical and postoperative approach to craniosynostosis is valid. This can potentially further decrease the surgical, physical, psychological, and financial impacts on patients and families. Further investigations of this technique applied to larger groups of patients, and for other types of craniosynostosis, are warranted.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Maugans. Acquisition of data: both authors. Analysis and interpretation of data: both authors. Drafting the article: both authors. Critically revising the article: both authors. Reviewed submitted version of manuscript: both authors. Approved the final version of the manuscript on behalf of all authors: Maugans. Statistical analysis: both authors. Administrative/technical/material support: Maugans. Study supervision: Maugans.

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