Cranial vault remodeling for sagittal craniosynostosis in older children

S. Alex Rottgers, M.D.,1,2 Peter D. Kim, M.D., Ph.D.,1,3 Anand Raj Kumar, M.D.,1,2 James J. Cray, Ph.D.,1,2 Joseph E. Losee, M.D.,1,2 and Ian F. Pollack, M.D.1,3

1Children’s Hospital of Pittsburgh of UPMC; 2Division of Pediatric Plastic Surgery, University of Pittsburgh Division of Plastic and Reconstructive Surgery; and 3Division of Pediatric Neurosurgery, University of Pittsburgh Department of Neurosurgery, Pittsburgh, Pennsylvania

Objective. Sagittal craniosynostosis is the most common form of craniosynostosis and is commonly treated within the first year of life. Optimal treatment of patients older than 1 year of age is not well characterized. The authors reviewed cases of sagittal craniosynostosis involving patients who were treated surgically at their institution when they were older than 1 year in order to determine the rate of intracranial hypertension (ICH), potential to develop nonhealing cranial defects, and the need for various surgical procedures to treat the more mature phenotype.

Methods. A retrospective chart review was conducted of all cases in the Children’s Hospital of Pittsburgh Neurosurgery Database involving patients who underwent cranial vault remodeling for scaphocephaly after 1 year of age between October 2000 and December 2010.

Results. Ten patients were identified who met the inclusion criteria. Five patients underwent anterior two-thirds cranial vault remodeling procedures, 3 patients underwent posterior vault remodeling, and 2 patients underwent 2-staged total vault remodeling. All patients had improved head shapes, and mean cephalic indices improved from 65.4 to 69.1 (p = 0.05). Six patients exhibited signs of ICH. No patients with more than 3 months of follow-up exhibited palpable calvarial defects.

Conclusions. Patients with sagittal synostosis treated after 1 year of age demonstrate increased rates of ICH, warranting diligent evaluations and surveillance to detect it; rarely develop clinically significant cranial defects if appropriate bone grafting is performed at the time of surgery; and achieve acceptable improvements in head shape.

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Key Words • craniosynostosis • sagittal craniosynostosis • scaphocephaly • dolicocephaly • cranioplasty • craniofacial deformity

Craniosynostosis is the early fusion of one or more of the cranial sutures, which results in restricted skull growth perpendicular to the involved suture and compensatory growth parallel to it. The ultimate result of this is a pathological head shape and potentially elevated intracranial pressure, which occurs in 14% of cases of single-suture fusion.14 Sagittal craniosynostosis is the most common form craniosynostosis. It occurs in 1 in 5000 children and affects boys more frequently than girls.3 Sagittal suture fusion results in an elongated, or scaphocephalic, head shape.

Both minimally invasive strip craniectomy and more extensive cranial remodeling procedures for sagittal synostosis within the 1st year of life have been described. Debate exists regarding the relative benefits of each.4–8,11,13,15,18 Regardless of the technique employed, most surgeons advocate for operative intervention within the 1st year of life. As scaphocephaly is usually apparent at birth, this is often accomplished. Less commonly, children will present after 1 year of age or require delayed interventions due to associated comorbidities. These older patients with scaphocephaly are a poorly characterized population with unique considerations regarding their risk of intracranial hypertension (ICH), potential to develop nonhealing cranial bone defects, and need for more extensive operations to achieve an aesthetic head shape. We report our experience with scaphocephalic children who underwent cranial vault remodeling for scaphocephaly after 1 year of life, highlighting these issues.
Methods

The neurosurgery patient database of the Children’s Hospital of Pittsburgh of UPMC was queried for all patients with the diagnosis of nonsyndromic sagittal craniosynostosis who underwent cranial vault remodeling procedures at an age greater than 1 year between October 2000 and December 2010. Patients with multisuture synostosis were excluded if the remodeling procedure performed was intended to address abnormal head shape resulting from the other fused sutures and did not address the finding of scaphocephaly. A retrospective review was performed of the medical records of all patients. Factors studied included: 1) patient demographic characteristics, 2) intraoperative blood loss and transfusion rates, 3) ICU and hospital length of stay, 4) type of calvarial remodeling performed, 5) signs and symptoms of ICH, 6) postoperative complications, 7) the incidence of residual cranial bone defects, and 8) changes in head shape as measured by the cephalic index (CI).

The CI was calculated from anthropomorphic measurements obtained in the clinic and measurements taken from available CT scans. This value is defined as the widest dimension of the cranium (eurion-eurion [eu-eu]) divided by the maximal length of the cranium (glabella-opisthocranion [g-op]) multiplied by 100. The CT measurements were made using 3D reconstructions when available. When they were not, 2D CT scans were analyzed. In these cases, lateral scout views or sagittal reconstructions of 2D CT scans were used to measure the g-op dimension, while axial cuts were used to measure the eu-eu dimension. Preoperative and postoperative CI values were compared using a paired t-test. The assumption of normality was confirmed using the Shapiro-Wilk test. Comparisons were only made between pre- and postoperative measurements derived from the same imaging modality to maintain internal consistency for each individual. Single measurements without a corresponding pre- or postoperative value attained with the same modality were not analyzed. In instances in which multiple measurement modalities were available, anthropomorphic measurements (actual measurements of the patient’s head) were prioritized. Any 3D CT reconstructions were prioritized next as they limited possible distortions caused by rendering a 3D structure in 2 dimensions. Finally, 2D CT scans were analyzed if 3D reconstructions were not available.

Surgical Approach

A defect-targeted strategy was employed in designing each patient’s reconstruction. Patients were assessed to determine their degree of scaphocephaly, the severity of their occipital constriction and bulleting, frontal bossing, temporal narrowing, and hypotelorism. A procedure was designed to adequately address all significant pathology. All procedures were carried out through a zig-zag-shaped bicoronal incision.

For patients with pathology that was predominantly anterior with frontal bossing and temporal narrowing, an anterior two-thirds cranial remodeling procedure was performed, including bifrontal and biparietal craniotomies with reshaping and interpositional bone grafting to widen the frontal bone, trimming of the parietal bones to decrease the anterior-posterior (AP) diameter, and barrel staving with out-fracture of the temporal bones to widen the skull base (Fig. 1). If hypotelorism and/or superior orbital rim retraction was present, a supraorbital expansion and/or advancement was completed by removing the supraorbital bandeau, cutting it in the midline, and placing an interpositional bone graft at the radix. Those patients with more pronounced occipital restriction and bulleting were treated with posterior cranial remodeling with occipital craniotomies, flattening of the occipital bone through barrel staving, parietal craniotomies and trimming to decrease the AP diameter, and barrel staving with out-fracture of the parietal and temporal bones to release constriction. Patients with severely altered anterior and posterior head shapes were treated with a staged procedure addressing each region sequentially (Fig. 2).

Our bone-grafting technique of choice was previously described and was employed in the last 8 patients in this series to fill residual bone defects after remodeling. Briefly, demineralized bone matrix, particulate bone material, and blood were mixed and protected in a sandwich between 2 layers of resorbable mesh to achieve protected bone regeneration.

Results

Thirteen patients were initially identified from the database who had sagittal craniosynostosis and had un-
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dergone a cranial vault remodeling procedure at an age greater than 1 year. Two patients had inadequate records available and were excluded. One additional patient who underwent cranial vault expansion at 4.86 years of age for multisuture craniosynostosis was also excluded.

The remaining 10 patients presented at an average age of 1.75 years (range 0.93–3.14 years). Nine patients were boys. Eight patients underwent single-stage cranial vault procedures (5 anterior and 3 posterior). Three of the patients treated with anterior approaches underwent a frontal orbital expansion as part of this procedure. Two of the patients undergoing posterior approaches had Chiari malformation Type 1 and underwent concurrent decompression procedures. Two patients were treated with a 2-staged anterior and posterior approach (Fig. 2). The patients’ average age at the time of cranial vault remodeling was 2.64 years (range 1.01–5.26 years). The second procedure was delayed 10.8 and 6.3 months for the patients undergoing staged reconstructions (Table 1). The average follow-up period was 2.24 years (range 0.2–9.3 years). All neurosurgical procedures were performed by a single surgeon (I.F.P.). Eight of the 10 patients were operated on by a single craniofacial surgeon (J.E.L.).

The average hospital length of stay for 12 operations completed in 10 patients was 4.1 days, with an average ICU stay of 1.3 days. Procedures were completed with an average surgical time of 5.4 hours. The average estimated blood loss volume was 544 ml. Intraoperative cell salvage was used for 7 patients in 9 cases, and an average volume of 187 ml of autologous blood was returned to the patients. Seven patients received packed red blood cell transfusions intraoperatively during 8 procedures. The average transfusion volume was 423 ml. Two of these patients and 1 additional patient received postoperative transfusions.

Intracranial Hypertension

Six patients exhibited clinical evidence of ICH during their preoperative evaluation (Table 1). In 4 patients, dilated fundoscopic examinations revealed papilledema. Four patients complained of headaches that were attributed to ICH. Of the 6 patients with clinical evidence of ICH, 2 had preoperative lumbar punctures performed under general anesthesia to confirm the presence of ICH; in both cases the pressure was greater than 30 mm Hg.

Despite the high incidence of ICH in our cohort, it was not commonly apparent at initial presentation. Often, diligent screening or longitudinal follow-up was needed to make the diagnosis. Three patients presented with a

Fig. 2. Staged total cranial remodeling. Anterior and posterior approaches to cranial remodeling may be employed to target the regions of the head with the greatest pathology. In individuals with significant anterior and posterior pathology, we favor a staged approach for total cranial remodeling. A: Preoperative images showing significant anterior and posterior pathology with frontal bossing, bitemporal narrowing, and occipital constriction with bulleting. B: Seven-month postoperative images following anterior cranial remodeling showing improvement in frontal bossing, bitemporal narrowing and scaphocephaly, but persistent occipital bulleting. C: Thirteen-month postoperative images following a second-stage posterior cranial remodeling showing improvement of the occipital bulleting with further improvement of scaphocephaly.
TABLE 1: Summary of clinical characteristics in 10 patients who underwent cranial vault remodeling after the age of 1*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Op</th>
<th>Age at Op (yrs)</th>
<th>Preop CI</th>
<th>Postop CI</th>
<th>Evidence of ICH</th>
<th>Significant Cranial Defects</th>
</tr>
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<tbody>
<tr>
<td>1</td>
<td>ant approach w/ FOE</td>
<td>1.2</td>
<td>none</td>
<td>72.1</td>
<td>papilledema, headaches, confirmed w/ LP</td>
<td>no</td>
</tr>
<tr>
<td>2</td>
<td>ant approach</td>
<td>3.0</td>
<td>73.7</td>
<td>68.3</td>
<td>copper beaten skull, headaches, confirmed w/ LP</td>
<td>no</td>
</tr>
<tr>
<td>3</td>
<td>ant approach w/ FOE</td>
<td>4.9</td>
<td>63.3</td>
<td>68.3</td>
<td>papilledema, headaches</td>
<td>no</td>
</tr>
<tr>
<td>4</td>
<td>staged ant/pst approach</td>
<td>3.4; 4.3</td>
<td>63.9</td>
<td>71.0</td>
<td>none</td>
<td>no</td>
</tr>
<tr>
<td>5</td>
<td>staged ant/pst approach</td>
<td>1.2; 1.8</td>
<td>none</td>
<td></td>
<td>no</td>
<td>no</td>
</tr>
<tr>
<td>6</td>
<td>pst approach w/ CM decompression</td>
<td>5.3</td>
<td>66.2</td>
<td>68.1</td>
<td>papilledema, headaches</td>
<td>no</td>
</tr>
<tr>
<td>7</td>
<td>pst approach</td>
<td>2.7</td>
<td>65.3</td>
<td>69.2</td>
<td>papilledema</td>
<td>no</td>
</tr>
<tr>
<td>8</td>
<td>ant approach w/ FOE</td>
<td>1.0</td>
<td>60.3</td>
<td>68.0</td>
<td>papilledema</td>
<td>no</td>
</tr>
<tr>
<td>9</td>
<td>ant approach</td>
<td>1.6</td>
<td>63.0</td>
<td>68.0</td>
<td>none</td>
<td>present at 3-mo FU</td>
</tr>
<tr>
<td>10</td>
<td>pst approach w/ CM decompression</td>
<td>2.2</td>
<td>67.4</td>
<td>68.1</td>
<td>headaches</td>
<td>present at 3-mo FU</td>
</tr>
</tbody>
</table>

* ant = anterior; CM = Chiari malformation; FOE = frontal orbital expansion; FU = follow-up; LP = lumbar puncture; pst = posterior.

Chief complaint of scaphocephalic head shape and were subsequently found to have either papilledema or a Chiari malformation associated with occipital headaches, which motivated the decision to operate. The families of 2 patients initially declined surgery to correct the patient's head shape, but the patients later required surgery due to changing clinical conditions and development of ICH symptoms. One of these families was motivated toward operative management by development of posterior headaches associated with a known Chiari malformation. The other family declined surgery for sagittal synostosis when the patient was 9 months old, but the patient returned at 4 years of age with subsequent fusion of the coronal sutures and development of “copper beaten skull” radiographic findings, headaches, listless behavior, and intracranial pressure greater than 30 mm Hg on lumbar puncture. Finally, one patient underwent an extended strip craniectomy at 2.1/2 months of age and during follow-up presented with severe headaches at 3 years old. He was noted to have papilledema on ophthalmological examination; ICH was confirmed by lumbar puncture.

Intracranial hypertension was successfully treated by the described procedures in 5 of 6 patients. The patient previously treated with an extended strip craniectomy underwent an anterior two-thirds vault remodeling procedure at 3 years of age with temporary symptom alleviation and improvement of his papilledema. Unfortunately, his headaches and papilledema recurred and he underwent shunt insertion.

Three Type 1 Chiari malformations were identified in these patients. One patient had papilledema and occipital headaches that motivated surgical intervention and another had occipital headaches alone. Both patients were treated with concurrent posterior cranial vault remodeling and Chiari decompression. The third patient had papilledema but a more significant anterior cranial vault deformity. This patient underwent anterior two-thirds remodeling without Chiari decompression and papilledema resolved postoperatively.

Head Shape

Preoperative and postoperative CI measurements were obtained from the medical records in 8 cases. In 1 case, pre- and postoperative anthropomorphic measurements were available; in 3 cases, pre- and postoperative 3D CT scans were available for measurements; and in 4 cases only 2D axial CT scans were available. Surgical intervention significantly improved the CI from a preoperative mean of 65.4 to 69.1 postoperatively (p = 0.013, t = 3.284, the assumption of normality was met) (CI ranges: mesocranic 76–80.9, dolichocranic 70–74.9, hyperdolichocranic < 70). The CI measurements increased by 5.7%. Seven of 8 patients showed CI improvement (Table 1). The individual without improvement had developed ICH following extended strip craniectomy in infancy. His head was not significantly scaphocephalic preoperatively (CI 73.7), and the surgical priority was vault expansion for ICH treatment.

Nine of 10 showed complete correction of their most significant cranial pathology (Fig. 2). One patient who underwent an anterior correction maintained significant occipital constriction and bulleting. This patient’s most recent clinic visit was only 3 months after surgery. The possibility of staged posterior remodeling was discussed with the family, but the patient did not have ICH as of this writing and the family was not interested in additional interventions.

Cranial Defects

Two of 10 patients had palpable cranial bone defects on physical examination at their last follow-up appointment (Table 1). In both cases, the follow-up appointment was only 3 months after surgery and long-term follow-up findings were not yet available. In 4 additional patients, skull defects were visualized on CT scans, but the defects were small. Since these defects are not palpable, their clinical significance and likelihood of requiring correction are small. No revision bone-grafting procedures have been performed.
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Complications

No death, postoperative bleeding, CSF leak, or sagittal sinus injury were documented. In 3 cases, small dural tears were discovered and treated intraoperatively with primary closure. One patient received a narcotic overdose while under observation in the ICU but suffered no long-term sequelae. Two patients were readmitted to the hospital because of fever, but in both cases viral illness was diagnosed when imaging and culture findings were negative. Two minor wound infections were treated successfully with oral antibiotics without operative intervention. Finally, 1 patient developed a small partial-thickness scalp injury that healed with hypertrophic scarring of this region.

Discussion

Surgical corrections performed for sagittal synostosis management are highly variable, and the optimal approach is controversial. There are proponents of early interventions with open or endoscopically assisted strip craniectomies. These procedures are performed in isolation or with molding of the skull with postoperative helmeting or placement of springs to actively distract the suture site. Others have advocated open remodeling of the cranial vault either through an anterior or posterior approach, while still others have advocated total calvarial remodeling either in a single or multiple staged procedures. Although many case series have been reported in the literature, optimal treatment has yet to be fully defined.

Strip craniectomy techniques are performed in the early postnatal period to take advantage of the higher growth velocity to correct residual skull deformities, whereas advocates of more extensive remodeling procedures wait longer to improve the safety profile relative to blood loss and to decrease the potential for deformity relapse due to abnormal growth. Regardless, all of these procedures are typically performed before patients reach 1 year of age for presumed improved aesthetic and functional outcomes. First, although the controversy regarding mental outcomes related to craniosynostosis surgery is far from settled, the knowledge that patients presenting after 1 year of age have more ICH and inferior developmental outcomes has motivated surgeons to operate before this age. Second, large calvarial defects can heal spontaneously in infants, but this ability begins to diminish late in the 1st year of life, raising the risk of persistent skull defects.

Because sagittal synostosis can be detected at birth, it is commonly diagnosed prior to 1 year of age. Surgical treatment of this condition at older ages can be successful, although patients who present later may be at higher risk of ICH, are less likely to experience healing of ungrafted cranial defects, and often require more extensive cranial remodeling procedures.

With regard to the risk of ICH, our findings support those previously seen by Arnaud and associates. Patients who present later with scaphocephaly are at a higher risk for ICH (60% in our small series). This may be due to a selection bias inherent in the population prone to present and undergo surgery at an older age, or it may represent the ultimate outcome of restricted suture growth. It does highlight the need for a greater degree of suspicion in approaching these patients, especially since 50% of our patients with ICH did not present with this complaint initially, and only diligent evaluation uncovered it. It also underscores the need for longitudinal care of all patients with craniosynostosis until they have completed their cranial growth, regardless if operative management is undertaken. One of our patients presented at 9 months of age and the family declined operative intervention until ICH developed later in childhood and fusion of the coronal sutures was noted on repeat imaging. Another patient underwent an extended strip craniectomy at 2 1/2 months of age and subsequently developed ICH that did not respond to cranial remodeling and required shunt placement. Development of ICH after strip craniectomy is rare, occurring in 1.5% of cases in one series, but it underscores the importance of ongoing follow-up in patient management. Families must be educated regarding the symptoms of ICH and patients must be seen annually for a dilated fundoscopic examination to rule out papilledema. When present, papilledema and other symptoms must be followed regularly postoperatively to document their resolution.

When operating on older patients with sagittal craniosynostosis, the potential for development of nonhealing calvarial bone defects is higher than when operating on those less than a year old. In our series, no patients with significant follow-up were found to have clinically significant or palpable defects in their cranium. We achieved this result through the aggressive use of bone-grafting techniques. All bone chips and bone dust resulting from the remodeling procedures should be kept and used in the grafting procedure. These were mixed with allogeneic demineralized bone matrix when defect size demanded. Additional technical improvement can be achieved in large calvarial defects by protecting the healing bone graft from the dural pulsations that can fracture and destroy immature regenerating bone. This is achieved by creating a “sandwich” of absorbable mesh above and below the bone-grafting material; the mesh will ultimately be resorbed after calvarial healing is complete.

Our results indicate that a tailored operative approach adequately and reliably addressed each patient’s pathology and improved both their CI and cranial shape. Our results demonstrated a significant improvement in CI with operative intervention. While the 5.7% increase was less impressive than that reported in other series, this may be partially explained by the high incidence of ICH in our cohort. While the goal of calvarial remodeling should always be to normalize head shape, one must be cognizant of the need to expand the cranial volume. With a 60% incidence of ICH in our cohort, we may have tempered how vigorously we decreased the cranial AP diameter in these cases. Surgeons should aggressively decrease the AP diameter to improve CI when remodeling the scaphocephalic cranium, but must do so without compromising the goal of resolving ICH. Regardless of this, all but one of our patients showed marked improvement in CI, and all families were uniformly pleased with the outcomes. Additionally, “normal CI” is somewhat of a misnomer. The CI is widely variable between ethnic groups, and with an ethnically heterogeneous population, particularly one as small as ours, the idea of normalizing the CI is erroneous. The surgeon’s goal should...
be to decrease the AP dimension and widen the head to make the patient’s skull more closely approximate a normal appearance in light of his or her ethnicity and other features.

While the most easily standardized, CI should only be one outcome measurement of scaphocephaly correction. Other abnormalities such as temporal narrowing, frontal bossing, and occipital constriction can be more noticeable and problematic than an abnormal CI. Addressing these deformities can be the most important component of management. Our strategy followed an algorithm previously described to target interventions to the area of greatest pathology. The pathological findings in children with craniosynostosis may be widely variable, and not all patients require the same operation. Significant frontal bossing, temporal narrowing, and hypotelorism may be addressed through an anterior approach, while occipital findings of constriction and bulleting may be addressed through a posterior approach. Both interventions afford the opportunity to narrow the AP dimension and out-fracture the skull to improve the CI. If a patient presents with significant anterior and posterior pathology, a total calvarial remodeling may be indicated. We elected to perform this procedure in a staged manner in the cases described here, after discussion with the families about the pros and cons of single versus 2-staged interventions. When taking this approach, one must involve the family in the consent process to identify their understanding of the deformity and to discern which components of it are most troubling to them. As one of our cases demonstrates, for a deformity that may warrant consideration of both anterior and posterior remodeling, the family may opt for and be satisfied with a less-extensive procedure, provided the most significant areas of pathology are addressed.

Conclusions

Surgical treatment of patients with sagittal craniosynostosis after the age of 1 is uncommon and carries with it special considerations. Whether due to selection bias or the evolution of progressive growth restriction, these patients are at increased risk for ICH. Following patients longitudinally until cranial growth is complete to document absence of ICH is essential. Individuals older than 1 year of age are at increased risk for developing nonhealing cranial defects, but the use of protected bone regeneration techniques can prevent this. Significant improvement in head shape can be achieved in this population, and these patients may benefit from having less postoperative growth to complete, which may lead to less recurrence of the phenotype. When using a defect-targeted approach to the anterior, posterior, or total calvaria, involvement of the patient’s family in planning is essential to assess their willingness to consider a multi-staged treatment plan and to identify the pathological findings most concerning to them.

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: Pollack, Kim, Kumar. Acquisition of data: Rottgers. Analysis and interpretation of data: Rottgers, Cray. Drafting the article: Rottgers. Critical revision of the article: Pollack, Kim, Kumar, Cray, Losee. Statistical analysis: Cray. Administrative/technical/material support: Losee. Study supervision: Pollack, Losee.

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Manuscript submitted April 17, 2011. Accepted May 25, 2011. Address correspondence to: Ian F. Pollack, M.D., Children’s Hospital of Pittsburgh, 4401 Penn Avenue, Faculty Pavilion Room 4129, Pittsburgh, Pennsylvania 15224. email: POLLACI@upmc.edu.