Surgical Correction of Severe Scaphocephalic Deformities

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Sagittal synostosis may result in severe skull deformities. Characteristic components of the deformity include extreme elongation, frontal and occipital bossing, temporal pinching, and angulatory apical skull deformation. Conventional strip craniectomy often fails to correct these complex problems completely in severe early or late cases of sagittal synostosis. Techniques for total calvarial vault reconstruction have previously been reported, but a single large series has not been presented. Eighteen consecutive patients ranging in age from 3 months to 5 years (mean = 12 months) with severe early and late scaphocephalic skull deformities underwent total calvarial vault reshaping. All children required transfusions ranging from 250 to 1,100 mL. Operative times averaged 6 hours, and hospital stay ranged from 4 to 7 days. There was no perioperative mortality. Two patients experienced transient syndrome of inappropriate secretion of antidiuretic hormone, which responded to fluid restriction. One patient was noted to have a 2-cm parietal craniectomy defect 9 months after operation. Microscrews, which were used in all 18 patients, had to be removed in 2 patients when they became palpable. Excellent aesthetic results were noted in all 18 patients up to 36 months of follow-up.

Key Words: Sagittal synostosis, scaphocephalic deformities

Sagittal synostosis, with its characteristic oblong calvarial shape (scaphocephaly), is the most common type of craniosynostosis [1-3]. The severity of the calvarial deformity varies from slight cranial elongation with sagittal ridging to extreme elongation, with a large occipital shelf and pronounced frontal bone bossing. Persing, Jane, and Edgerton [4] suggested that the deformity is not limited to the sagittal suture but also involves the basis of the skull, which in part contributes to the degree of the cranial dysmorphology. Many neurosurgeons and craniofacial surgeons consider strip craniectomy of the sagittal suture to be the standard of surgical treatment for early sagittal synostosis [5]. Some investigators reported excellent results when the initial deformity is mild and surgery is performed within the first few months of life [5-6]. Others, however, reported unsatisfactory results in children of any age when the scaphocephalic deformity is severe, particularly when associated with large occipital shelves and marked frontal bone bossing [7-9].

At Scottish Rite Children’s Medical Center, we have found strip craniectomy to provide unsatisfactory results, even when performed within the first few months of life in infants with marked scaphocephalic deformities (Fig 1). Because of its high failure rate, strip craniectomy in children with scaphocephaly who are diagnosed after 18 months of age is generally not indicated. Children presenting with late scaphocephalic deformities often have marked frontal bossing, large occipital shelves, temporal pinching, and severe angulatory deformities at the apex of the skull (Fig 2). Because the majority of the brain growth has already taken place and the deformity is fixed, strip craniectomy has little or no effect on outcome [10]. To improve surgical results, Marchac performed near-total calvarial vault reconstruction using a transposition technique in older patients with scaphocephaly [11]. Persing, Jane, and Edgerton described a technique for complete one-stage correction of severe scaphocephalic deformities in 1989 but did not report a large series or an analysis of their results [4]. Marsh and Vannier [12] discussed a group of patients with scaphocephaly and quantitated their radiographic results but did not focus on perioperative problems or technical nuances of surgical management. We recently reported our initial results using a modification of their technique in patients older than 18 months [13]. Herein, we report the results of total calvarial vault reconstruction in 18 consecutive patients over a 3-year period.
A modified surgical approach is recommended to both maximize protection to the frontal branch of the facial nerve and minimize dissection of the fat pad (Fig 4). As described previously, elevation of the coronal flap in the temporal region proceeds in the subaponeurotic plane to a level approximately 2 cm above the zygomatic arch. The middle temporal artery and vein can be seen in some patients, coursing within the fat pad deep to the superficial layer of the temporal fascia. The superficial layer of the fascia is incised below this level, and dissection proceeds inferiorly immediately on the undersurface of the fascia to the periosteum of the zygomatic arch. The superficial temporal fat pad is maintained in its normal location, and dissection within the fat is minimized. At the conclusion of the reconstructive procedure, the transected superficial layer of the deep temporal fascia is repaired.

**CONCLUSION**

We presented 6 patients with obvious temporal contour deformities after bicoronal flap elevation and exposure of the lateral craniofacial skeleton. The deformity results from atrophy or prolapse of the superficial temporal fat pad. In the future, the surgical approach to the lateral craniofacial skeleton will have to be done not only with the frontal branch of the facial nerve in mind but also the superficial temporal fat pad. Meticulous dissection of the fascial layers, preservation of the fat pad in its normal anatomical location, and avoidance of the middle temporal artery are critical.

**REFERENCES**

Surgery for Scaphocephalic Deformities

Materials and Methods

Eighteen patients with scaphocephaly (10 males, 8 females) ranging in age from 3 months to 5 years (mean = 12 months) were surgically treated over a 36-month interval. Nine were younger than 12 months (group 1), and 8 were older than 12 months (group 2). Those patients in group 1 with early sagittal synostosis had severe scaphocephaly, which was not expected to improve with strip craniectomy alone. Patients in group 2 were older and had moderate to severe scaphocephaly, requiring definitive calvarial remodeling. Follow-up ranged from 12 to 36 months. All patients in both study groups had moderate to severe scaphocephaly with marked frontal bossing, large occipital shelves, bitemporal narrowing, and angulatory apical skull deformities. There were no syndromal cases. Two of the patients had calvaria-shaped abnormalities primarily related to sagittal synostosis but were also found to have other areas of craniosynostosis: lambdoid (n = 1) and metopic (n = 1) synostosis. The cause of the synostosis was unknown in all but 1 patient, who had suffered a series of two skull fractures crossing the sagittal suture at 10 and 18 months of age.

Preoperative Evaluation

All patients underwent a complete physical examination including a detailed neurological examination before operation. All patients had anteroposterior (AP) and lateral skull x-ray films and computed tomographic scans of the skull and brain with three-dimensional reconstruction of the axial images. In addition, flexion/extension views of the cervical spine were obtained before surgery to rule out occult spine disorders.

Surgical Techniques

Severe Early Sagittal Synostosis: Group 1

After general anesthesia is induced, the endotracheal tube is secured to the mandible with a circummandibular wire. Two large-bore intravenous catheters are placed. A Foley catheter is inserted to monitor urine output. The patient is then placed in the prone position, with the head position maintained using Olympic Vac positioning system (Sizell, Olympic Medical, Seattle, WA) chin...
support. The neurosurgical and craniofacial portions of the procedure are carried out in sequential fashion.

First, a bifrontal craniotomy is performed with a supraorbital osteotomy below the frontal boss. The posterior osteotomy is placed in front of the coronal suture, and the frontal bone is carefully removed. The two lateral temporoparietal bone segments are removed, staying just lateral to the sagittal suture. The occiput is removed, keeping the posterior osteotomy below the occipital shelf. A burr hole is placed adjacent to the midline of the occiput to facilitate stripping of the dura mater. Barrel-stave osteotomies are performed in vertical fashion along the remaining temporal bone and occipital bone and infraverted to permit lateral expansion of the cranium (Figs 3 and 4). Plication sutures are placed diagonally in two rows in the bulging frontal dura mater. The frontal bone is split into halves and expanded in the transverse plane with radial osteotomies and interpositional bone grafts. A wedge of bone is removed from the frontal bone at its junction with the supraorbital osteotomy to allow posterior rotation of the frontal bone. Microplates (Howmedica Corp., Newark, NJ) are used to link the two halves of the frontal bone and the supraorbital osteotomy site. The occiput is split in half, allowing for lateral expansion, in the same fashion as the frontal bone.

While the craniofacial surgeon reconstructs the frontal and occipital bones on a side table, the neurosurgeon inserts an intracranial pressure monitor through the right side of the large sagittal bone strip, between the osteotomized frontoparietal and occipital bone segments. This central strip serves as the anchor for the AP correction.

Two small drill holes are placed in the central portion of the occipital and frontal bone plates. The central bone segment covering the sagittal sinus is then dissected free of the dura and sagittal sinus. A section of the central strip is removed according to the amount of AP correction deemed necessary. This averages 2.5 cm. A corresponding strip of the lateral temporoparietal bone segments is also removed. Small drill holes are placed through the central sagittal bone strip. The frontal bone segment is replaced using four-hole microplates along the supraorbital rim.

Posteriorly, the occipital bone is replaced in a similar manner. A 28-gauge wire is then passed through the osteotomized occipital and frontal bone segments and attached to the central sagittal bone strip. While carefully monitoring the intracranial pressure, the right and left wires are slowly twisted on the frontal segment until bone contact occurs between the frontal bone and the central sagittal strip. As AP correction proceeds, bulging of the dura mater laterally is noted. The occipital correction is carried out by slowly twisting the wires, advancing the occipital bone segment toward the central sagittal strip until bone contact is achieved. Intracranial pressure is maintained at 16 mm Hg with normocarbia. The AP correction is carried out slowly, often over 45 to 60 minutes.
Two small holes are drilled into the lateral temporoparietal bone segments, which are sutured to the dura mater with a horizontal mattress-type suture.

After all of the osteotomized segments have been replaced, attention is turned toward the anterior temporal fossa area. Preoperatively, this area is often pinched in appearance. To correct these depressions, the excess bone that has been harvested from the calvarium is cut into two rectangular strips, which are bent with bone-contouring forceps and fit into the temporal fossa to prevent a hollowing deformity. They are then attached with microscrews to the lateral orbital rims. To fill out the temporal fossa further, bilateral advancements of the temporals muscle are carried out. The large pericranial flap, which is elevated at the time of the craniotomy, is replaced, covering the frontal bone and lateral orbital regions. Next, the entire complex is copiously irrigated with an antibiotic solution. A small closed-system drain is then placed through a separate stab incision in the posterior occipital flap. The galea is closed using absorbable sutures, and the skin is closed with staples. A standard neurosurgical head dressing is applied.

Postoperatively, the patient is extubated in the recovery room and sequential neurological examinations are performed. The patients are placed in the intensive care unit for approximately 48 hours. Careful monitoring of serum sodium is carried out over the first 72 hours. Transfusions of donor-directed blood, usually from the parents, is begun intraoperatively and continues postoperatively for the first 24 hours until the hematocrit stabilizes. The neurosurgical head dressing is kept in place for approximately 4 days, at which time the drain output has usually been reduced to less than 10 mL per day. The head dressing and the drain are removed at the same time. The patient is discharged home in the care of the parents with no special care instructions. The skin staples are removed 1 week postoperatively.

Correction of Late Sagittal Synostosis: Group 2

Preoperative evaluation, surgical preparation, and postoperative care are similar to those in group 1 patients. Only the points that differ in surgical technique are discussed.

The main difference in technique for these patients is that the portion of the calvarium between the frontal and occipital bones is removed in three transverse bands of approximately equal widths (Figs 5 and 6). The lateral osteotomies for these bands are made just above the squamosal suture. The osteotomy across the midline is accomplished via three slightly off-center central drill holes to protect the sagittal sinus. In contrast to the early cases, in which the bone over the majority of the
sagittal sinus is left intact, in the late cases, the bone is removed in its entirety. Each one of the three central bony segments is taken to a side table, where multiple partial-thickness osteotomies are made with a cutting burr on the inner surface of the bone. Next, gentle green-stick fractures are made along each one of the partial-thickness osteotomies with the bone-contouring forceps. An average of 1.5 cm of bone is removed from the anterior and posterior skeletal bands to allow AP correction. Microplates are then placed on the lateral borders of each one of the bands. Small drill holes are placed at the anterior and posterior bands to allow for wiring to the frontal and occipital bone plates. The central band is secured with microplates to the lateral temporal bone to allow it to act as a central post. Next, the anterior and posterior bands are replaced. The frontal bone is connected to the anterior strut with a 28-gauge wire, as is the occipital bone to the posterior strut. With careful attention to the intracranial pressure, the frontal bone is then slowly brought posteriorly to meet the anterior band. Lateral dural bulging into the barrel-stave osteotomies will be noted. The occiput is repositioned in the same fashion.

RESULTS

Eighteen children underwent total calvarial reconstruction for scaphocephaly over the last 3 years. All children required blood transfusions ranging from 250 to 1,100 mL (mean = 400 mL). The child with 1,100-mL blood loss had hypocalcemic rickets and extremely vascular bone. There was one sagittal sinus disruption, which accounted for a 1,000-mL blood loss in a second child. There was no perioperative mortality or long-term morbidity. The 2 children with the greatest blood loss and fluid shifts experienced hypovolemia postoperatively secondary to syndrome of inappropriate secretion of antidiuretic hormone. Both responded to fluid restriction. The operative times ranged from 300 to 390 minutes (mean = 6 hours). The hospital stay ranged from 4 to 7 days (mean = 5 days). Nine months after the operation, 1 child was noted to have a 2-cm bony parietal defect, which was thought to represent bone graft resorption. This defect slowly decreased in size, and no reoperation is contemplated. Two children had microscrews removed when they became palpable in the scalp 1 year after surgery.

The treatment goal of correction of the severe scaphocephalic deformity was accomplished in each case. Occipital and frontal deformities were significantly improved in all patients (Figs 7-9). Computed tomograms were not routinely performed postoperatively; however, they were obtained on representative cases from groups 1 and 2 to verify the calvarial changes. These radiographic studies confirmed our intraoperative impression of a decreased AP dimension with a concomitant increase in bitemporal width (Fig 10).

DISCUSSION

True to Virchow's rule, synostosis of the sagittal suture by whatever mechanism results in compensatory skull deformation [14-16]. In addition to the ovoid elongation of the head, a central vault deformity, significant frontal bossing, and an occipital shelf are almost always encountered. This is particularly true in those patients not treated in early infancy. The variety of surgical techniques that have been proposed confirm the fact that previous attempts have been largely unsuccessful in correcting severe and late scaphocephalic deformities [8,9,17]. These techniques rely on passive temporal expansion of the cranial vault by the growing brain to increase the bitemporal width while decreasing AP length. We believe that these so-called passive techniques result in release of the synostosis but often fail to correct the occipital or frontal bone deformities totally. The strip craniectomy types of procedures are best reserved for cases of sagittal synostosis treated in the first 3 to 7 months of life, which are of mild to moderate severity. It is now widely accepted that even single-suture synostosis has a corollary deformity at the base of the skull [15,16]. Whether the initiating event is a problem with one of the synchondroses at the base of the skull or whether the base of skull deformity is secondary to synostosis remains controversial [12]. McCarthy and colleagues advocated that the base of skull be addressed, as much as possible, in the surgical treatment of craniosynostosis [18,19]. We believe that strip craniectomy, which relies on passive mechanisms without definitively addressing the complete deformity, is likely to lead to an incomplete correction in the more severe early, as well as the late, cases of sagittal synostosis. Marsh and co-workers quantitatively evaluated two techniques of surgical management of sagittal synostosis in 22 infants [20]. The first 11 infants underwent extended craniectomy, whereas the next 11 infants were treated with subtotal calvarectomy with remodeling. Both operations were performed preferentially at 3 months of age. Both the perioperative and postoperative cephalic indexes were significantly different from those preoperatively for each group. Although the infants who underwent extended strip craniectomy moved into the normal range perioperatively, their cephalic indexes remained below the normal range. Cephalic indexes of both groups appeared to drift toward values indicative of scaphocephaly over the first postoperative year. Nonetheless, most values for children who had undergone total calvarial vault reconstruction remained within the normal range, whereas those for infants who had undergone extended craniectomy remained below the normal range.
Marchac and Renier reported a transposition technique to correct severe scaphocephalic deformities [11]. Although their technique addresses the total deformity, it relies on the presence of appropriately shaped skull segments for transposition, which may not always be available. We prefer to reshape the affected skull segments, returning them to their original location [12]. Although the base of the skull cannot be approached through current surgical maneuvers, we advocate as complete a correction of the vault deformity as possible, addressing the frontal bossing, occipital shelving, central steeple deformity, and temporal pinching.

Moss and others proposed that the dural abnormality in craniosynostosis is an active rather than passive participant in accentuating the skull deformity [21,22]. This may be a particularly important mechanism in the young patient in whom bone is being actively deposited on the endosteal or dural surface. To address this theoretical concern, we plicate the dura mater anteriorly, which results in visible bulging of the temporal areas. Plication of the dural sack permits its passive compliance with the newly contoured frontal, occipital, temporal, and parietal bones.

The role of rigid fixation in craniofacial surgery remains controversial [23,24]. We favor the use of rigid fixation and have found that microplate and screw fixation offers three-dimensional freedom in designing the osteotomies and allowing a semirigid construct, which can be molded by relatively minor pressures, such as those generated by tightening the anterior frontal and posterior occipital wires. We believe that it is important not to plate across natural growth lines such as the coronal sutures, lambdoid sutures, or even nasofrontal sutures when they are not involved in the deformity [24]. In children older than 5 years, the majority of calvarial growth
has already been achieved. Further growth and remodeling of the skull occur by periosteal resorption and endosteal deposition; therefore, the entire calvarium can be reconstructed with less concern for plate and screw placement [10]. In infants, abundant room between the lateral bone plates and the occiput, as well as the frontal bone, is purposely left to allow for growth (see Figs 3 and 4). This is an important technical consideration in the growing child that may prevent an iatrogenic synostosis. No evidence of head growth retardation, as measured by head circumference, has occurred in any of our 16 patients. In addition to the technical advantages of rigid microfixation, it has been shown experimentally that rigid fixation enhances bone revascularization and decreases bone resorption, an obvious advantage when using extensive bone grafts [25,26]. In our series, microplates or screws had to be removed in 2 patients because they became visible or palpable. To date, no infections associated with these metallic devices have been noted. We have not had any cases of inward migration of plates through the skull with intradural penetration.

The technical feasibility of total calvarial reshaping for severe scaphocephaly has been firmly established, but questions remain regarding the indications for these more radical measures. The argument can be made that surgery for sagittal synostosis is primarily an aesthetic

Fig 9 Same patient as in Figure 8, seen 16 months after correction. (A) Note complete correction of frontal and occipital deformities. (B) Note improved head shape with correction of elongation and transverse narrowing.

Fig 10 Computed tomographic scans of patient with late sagittal synostosis. (A) Preoperative scan with anteroposterior length-width ratio of 1.48 : 1. Note the elongated head with narrow transverse dimension. (B) Postoperative scan with anteroposterior length-width ratio of 1.26 : 1. Note that this is consistent with clinical improvement in head shape.
procedure. Renier and colleagues demonstrated that up to 13% of patients with single-suture synostosis have significant increases in intracranial pressure [27]. In our series, only 1 of the 16 patients had a copper-beaten appearance of the inner table of the skull, suggesting the presence of chronically elevated intracranial pressures. Many of the older patients related painful psychological experiences at the hands of their peers. Cruel name-calling such as “banana head,” “boat head,” and so on was often mentioned by both the patients and their families as a cause of psychological trauma. Adding to their self-consciousness and further stigmatizing them was the inability to wear standard headgear for a variety of athletic activities. In a society such as ours, which places high value on physical appearance, the aesthetic abnormalities associated with sagittal synostosis cannot be overemphasized [8]. Although we do not undertake detailed psychological testing in the older child, the majority of our patients expressed a great deal of satisfaction in their improved appearance. The potential advantages from a psychological as well as a functional point of view, however, must be weighed against the potential morbidity and mortality of these extensive procedures. Total calvarial reconstruction may result in significant blood loss and risk of damage to the sagittal sinus, which occurred in 1 of our patients. Infections complications, which have not arisen in our experience, could be devastating with widespread calvarial loss.

Total calvarial vault reshaping can be safely performed and is the procedure of choice in older patients with scaphocephaly. Total calvarial vault reconstruction should also be considered in place of strip craniectomy in infants with severe scaphocephaly.

REFERENCES

Commentary on Surgery for Scaphocephalic Deformities

In this report, Burstein and colleagues outline their experience in the treatment of severe scaphocephalic deformities. The techniques that they use have as basic tenants total cranial vault reshaping, active reduction of abnormally elongated skull shape, dural plication in regions of abnormal dura and bone prominence, and microplate fixation. Although each of these techniques has been described previously by others [1–3], the description of total cranial vault remodeling by another craniofacial team, noting their success and endorsement of the techniques is worthwhile.

The authors have divided their patient population into two groups: early and late. Patients in the early group are younger than 12 months, and those in the late group are older than 18 months. Similarly, previous authors [2,3] believed it was important to distinguish between different age groups regarding bone remodeling techniques to achieve the best possible reconstruction. This distinction is necessary because of variations in bone thickness and malleability with age. In the very young child, the malleable bone allows more ready and subtle variation in reshaping compared with the child older than 1 year, whose bone is more brittle and resistant to reshaping. These authors and others [2,3] have found that, to reshape a mature bone more fully, selective weakening in the endocranial surface of the cranium, by the creation of endocranial channels, or kerfs, is worthwhile. Additionally, if the periosteum is left on the external table of the skull while remodeling, despite microfracture of the skull bone, orientation of the microfractured fragments is more often maintained because of the tethering effect of the periosteal attachments.

The authors report active reduction of skull length using intracranial pressure recording (maintaining normocarbia) so as to approximate, intraoperatively, the intracranial pressure levels that are likely to be seen postoperatively. I believe the authors rightly emphasize that skull molding and active reshaping are beneficial, but this requires a slow remodeling process to avoid brain injury [2].

The authors report that blood loss in their patients ranged from 250 to 1,100 mL, and 1 patient sustained sagittal sinus disruption intraoperatively. This is a relatively rare event, but the risk is increased if the sagittal suture is partially patent, as has been reported to occur previously [4]. Caution should be exercised when stripping the parasagittal bone from the underlying dura in these circumstances [1].

The authors reinforce the merits of plate fixation in aligning bone fragments, particularly anteriorly at the orbital rim. Although better fixation is achieved with this approach, it also must be cautioned that, in the very young child, plate and screw apparatus will “migrate” through the skull bone, as successive layers of bone are added to the external surface of the skull, while resorption continuously occurs on the endocranial surface of the skull. Selective use of microfixation in these regions is likely to yield the best results induced by the plates.

Although the authors report increased intracranial pressure occurring in 13% of patients with single-suture synostosis, Renier indicated a slightly lower rate (8% of the time in patients with scaphocephaly), and in these patients intellectual performance was not judged to be subnormal [5]. It is, therefore, inconclusive what the overall signification of increased intracranial pressure is in patients with single-suture stenosis. This question requires further study.

The authors are to be commended for their advocacy of the total cranial vault reshaping approach, particularly in the older child with more severe deformity. Linear craniectomy in patients with sagittal synostosis may yield significantly fewer improvements in cranial shape, even in infancy. As quantitative studies now demonstrate even in young children, return to normal skull shape is seen more frequently in patients who undergo a more active skull remodeling process compared with more limited, passive cranial reshaping techniques [6].

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