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 base 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.

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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.

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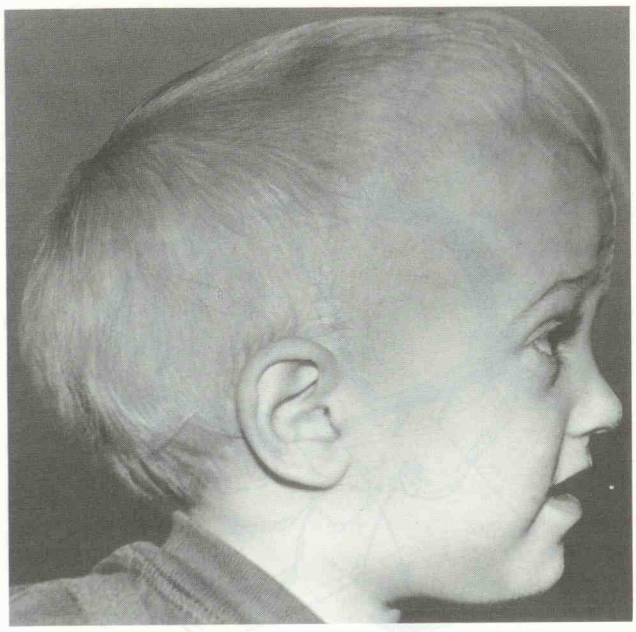


Fig 1 Eighteen-month-old patient who underwent sagittal strip craniectomy at 5 months of age. Note the persistent occipital and frontal bossing with scaphocephalic head shape.

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 18 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.

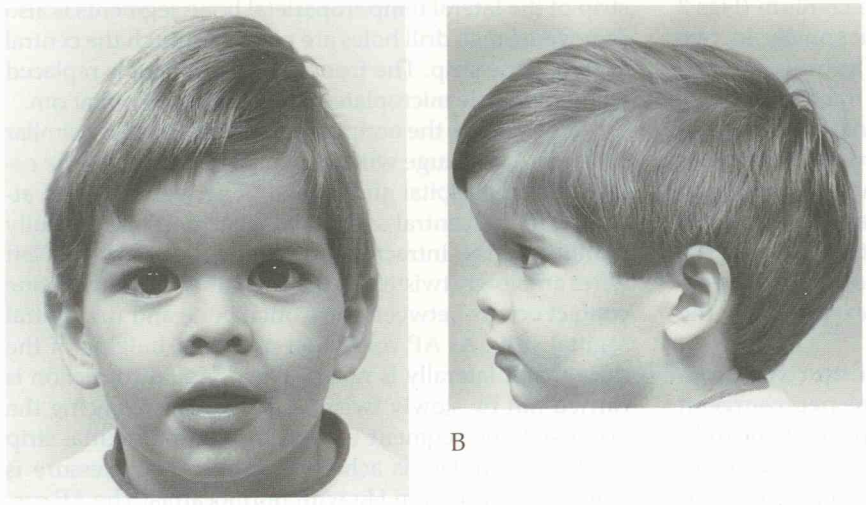
Preoperative Evaluation

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 circumandibular 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



A B

Fig 2 Four-year-old with late deformity. (A) Note frontal temporal pinching with a very acute angle at skull apex. (B) Note frontal bossing.

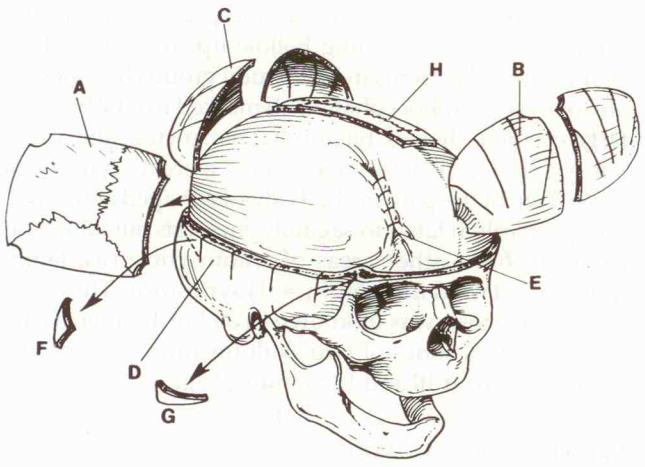


Fig 3 Diagram of surgical correction for severe early sagittal synostosis. (A) Temporoparietal bone segments. (B) Frontal bone split. (C) Occipital bone split. (D) Barrel-stave osteotomies for lateral expansion. (E) Dural plication. (F, G) Wedges of bone removed to allow tilting of frontal bone and occiput inward. (H) Central strip, which will be shortened at stippled line.

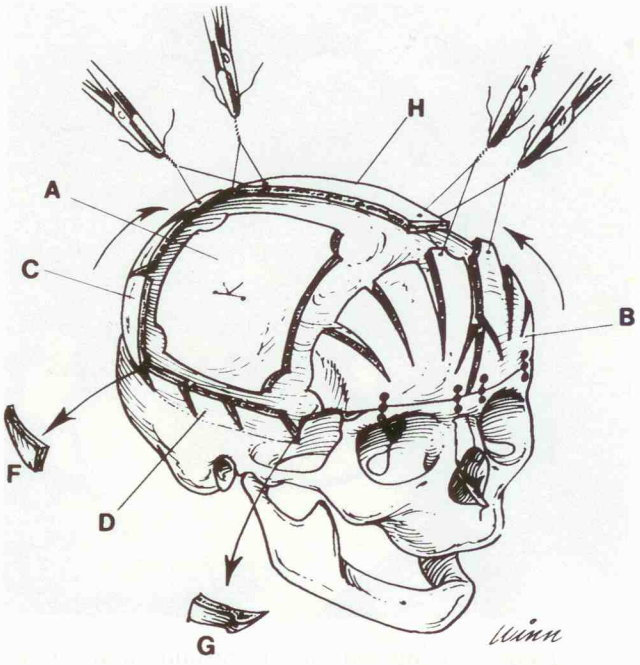


Fig 4 Diagram of surgical correction for severe early sagittal synostosis. (A) Temporoparietal bone segments. With bony segments replaced (see Fig 3), occipital (C) and frontal (B) bones have been split and expanded. Slow twisting brings occipital bone split (C) and barrel-stave osteotomies (D) toward central strip (H). (F, G) Wedges of bone removed to allow tilting of frontal bone and occiput inward.

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 infractured 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.

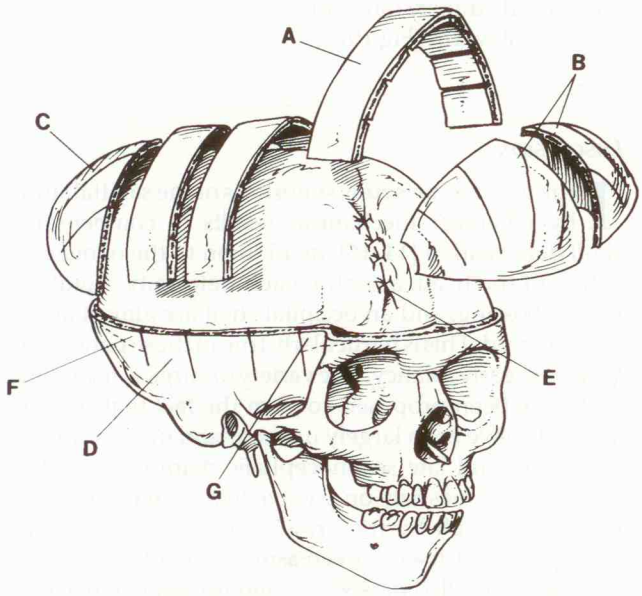


Fig 5 Diagram of surgical correction for late sagittal synostosis. (A) Central bone strips with partial-thickness osteotomy to allow expansion. (B) Frontal bone. (C) Occipital bone. (D) Lateral osteotomies. (E) Dural plication. (F, G) Areas of bone to be removed to allow inward pivot of frontal and occipital bones.

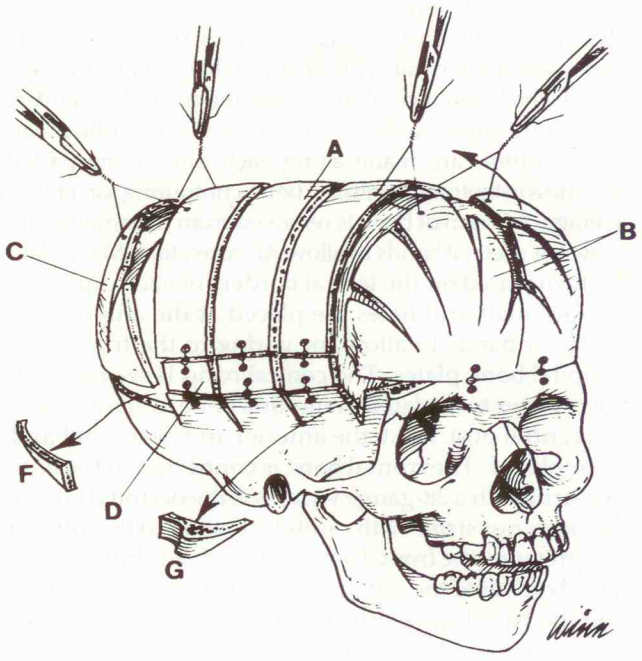


Fig 6 Diagram of surgical correction for late sagittal synostosis. Central bone strips (A), expanded with bone grafts (D) to increase transverse skull dimension. Frontal bone (B) and occipital bone (C) are brought forward to stabilize central bone strips (A), decreasing the anteroposterior dimension. (F, G) Areas of bone to be removed to allow inward pivot of frontal and occipital bones.

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 temporalis 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