

Total calvarial reconstruction for sagittal synostosis in older infants and children

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✓ Premature closure of the sagittal suture is the most common form of craniosynostosis, but this condition occasionally goes unrecognized until the child is too old to undergo procedures that depend upon continued calvarial growth for success. As the entire calvaria is affected and thus misshapen by sagittal synostosis, late correction involves total calvarial reconstruction. The extensive nature of this undertaking has precluded its utilization despite the presence of significant deformities. Adapting the techniques and experience gained from craniofacial surgery, the authors performed total calvarial reconstruction on nine children with sagittal synostosis and subsequent scaphocephaly diagnosed after the age of 1 year. In each case the goals of shortening the anteroposterior length, widening the biparietal diameter, and reducing frontal and occipital deformities were met. Morbidity consisted of acute blood loss, postoperative hyponatremia, and in one case a residual skull defect. The rationale for this procedure and the techniques utilized are discussed.

KEY WORDS • sagittal synostosis • craniosynostosis • scaphocephaly • operative techniques

SAGITTAL synostosis is the most common form of craniosynostosis and is usually easily recognized, even in early infancy. Unfortunately, either because of a failure to diagnose the condition early or because calvarial growth precipitates worsening of an initially mild deformity, some children with sagittal synostosis are not considered for surgical correction until after the age of 1 year. At this age, the calvaria has reached 85% of its adult size and procedures that depend upon continued calvarial growth for success, such as strip craniectomy, are of little value. Persing, *et al.*,¹⁶ have shown that single suture synostosis causes deformity of the entire calvaria. Thus, correction of "late" sagittal synostosis would involve a total reshaping of the cranial vault. The extensive nature of this intervention and concerns with associated morbidity and even mortality have prevented corrective surgery in the older child with sagittal synostosis.

Since the development of craniofacial surgery in the 1960's by Tessier, significant advances have lowered morbidity and mortality rates and allowed the application of these techniques to an increasing variety of conditions. Persing, *et al.*,¹⁶ previously described an innovative procedure for the correction of late sagittal synostosis derived from their extensive experience in craniofacial surgery. We describe our operative approach using a modification of their techniques and

our results in the treatment of children with late sagittal synostosis.

Clinical Material and Methods

Patient Population

Since October, 1990, we have performed total calvarial reconstruction on nine children (six boys and three girls) ranging in age from 16 months to 5 years (Table 1). The characteristic stigmata of sagittal synostosis were present in each case: an elongated cranium, biparietal narrowing, and a protuberant forehead and occiput. Diagnosis was made by clinical criteria and confirmed by plain film radiography and computerized tomography (CT). Ten procedures were performed, as one child with a markedly elongated head required a two-stage reconstruction.

Operative Procedure

With the patient under general endotracheal anesthesia, either two large-bore peripheral intravenous lines or a central venous catheter is placed, as well as an intra-arterial line and a Foley catheter. The hair is shaved and the child is turned to the prone position with the neck hyperextended. Preoperatively, neutral and hyperextension lateral cervical spine radiographs are obtained to rule out anomalies that would preclude

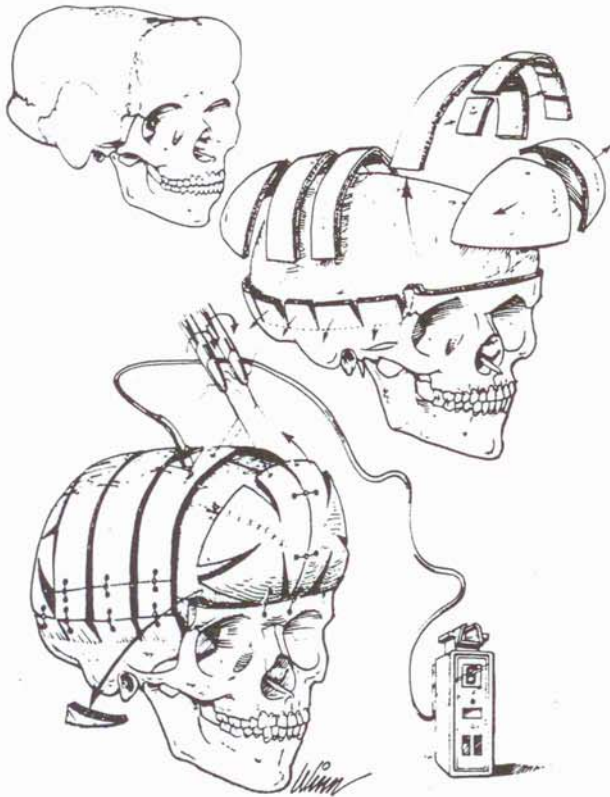


FIG. 1. Drawing illustrating the technique of total calvarial reconstruction for treatment of sagittal synostosis in the older infant or child.

prolonged use of cervical hyperextension. This position is maintained with the use of a Vac Pac positioning system.* All pressure points, including the chin and cheeks, are well padded. After sterile preparation of the head, an incision is made across the vertex, and the subgaleal plane is dissected to allow elevation of the skin and galea below the occipital shelf posteriorly and at the level of the supraorbital ridge anteriorly. The pericranium is incised just above and following the contour of the temporalis muscle bilaterally and then across the vertex. This allows elevation of the pericranium in two flaps, one posteriorly and the other anteriorly. The temporalis muscle is elevated to complete the exposure of the cranial vault.

A bifrontal craniotomy is then performed with the supraorbital osteotomy positioned below the frontal boss (Fig. 1). The posterior osteotomy is placed in front of the coronal suture. The posterior frontal and parietal bones are removed in three bands of approximately equal width. The lateral osteotomies for these bands are performed just above the squamosal suture, and the osteotomy across the midline is accomplished via a midline burr hole. The occiput is then removed, with the posterior osteotomy positioned below the occipital

* Vac Pac positioning system, size 11, manufactured by Olympic Medical, Seattle, Washington.

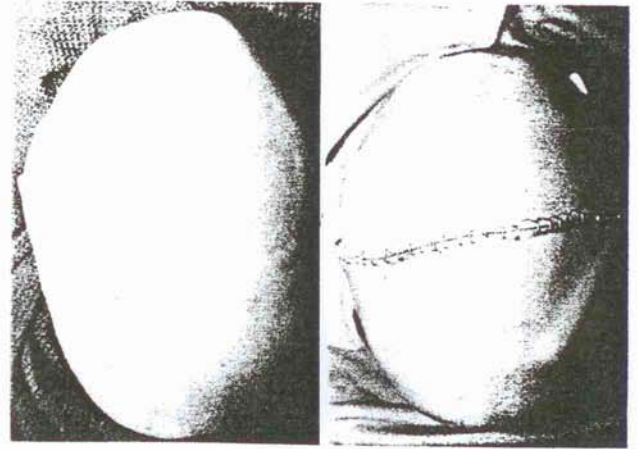


FIG. 2. *Left:* Preoperative photograph in Case 8, superior view with the nose to the bottom of the picture, demonstrating a markedly elongated cranium. Posterior asymmetry is caused by associated lambdoid synostosis. *Right:* Photograph taken immediately after surgery showing that the cranium is shortened in anteroposterior length and is wider biparietally and that the posterior plagiocephaly has been corrected.

shelf. A burr hole is placed in the midline of the occiput to facilitate stripping the dura from the bone.

The dura is dissected away from the temporal bone, and vertical cuts are made to allow out-fracturing of the temporal bone (barrel-stave osteotomies). Plication sutures are placed diagonally in the bulging frontal dura which reduces the anteroposterior length and the lateral frontal bulge of the dura. As the frontal sutures are placed, the parietal dura begins to bulge into the space created by the barrel-stave osteotomies.

As each segment of bone is removed, it is handed to the craniofacial plastic surgeon for reshaping. The frontal bone is split vertically in the midline, radial osteotomies are cut, and the bone is bent outward to correct the frontal boss. Microplates and screws are utilized to rejoin the two halves of the frontal bone. A wedge of bone is removed from the inferolateral frontal bone to allow it to tilt backward when repositioned on the supraorbital ridge, thus reducing the anteroposterior length. A similar technique is utilized to correct the occipital shelf. The parietal bands are grooved along their inner surface ("kerfing") and bent laterally with Tessier bone benders. Approximately 2 to 3 cm of bone is cut from the bands and used as graft material. These 2 to 3 cm constitute the anteroposterior reduction of the calvaria.

The biparietal bands are then placed back over the vertex. Bone is grafted laterally to close the gap created by the more convex bands and the out-fractured temporal bone. This procedure constitutes the biparietal widening. The bands and their grafts are fixed rigidly with microplates and screws; the parietal bone serves as a central pillar for fixation of the occiput and frontal bones.

The deformity of sagittal synostosis is frequently worse either frontally or in the occiput. If the forehead

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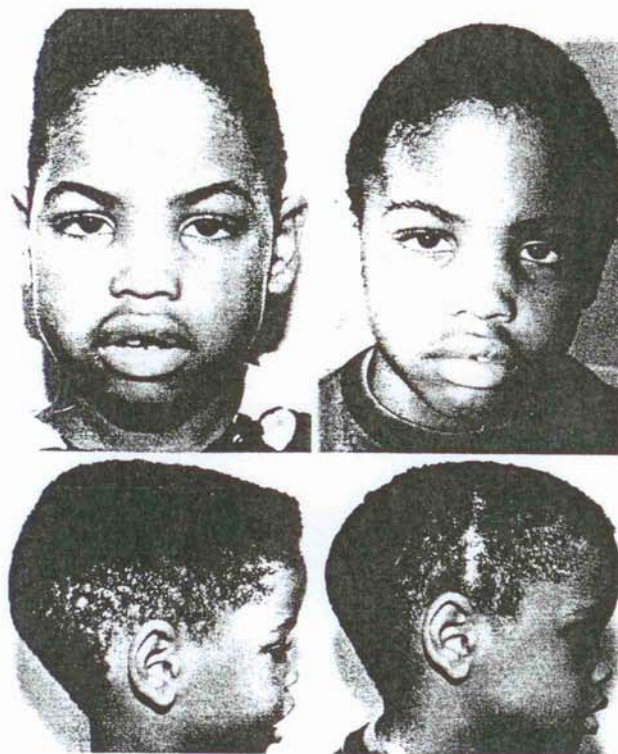


FIG. 3. *Upper and Lower Left:* Preoperative photographs in Case 7 demonstrating the long narrow cranium indicative of sagittal synostosis. *Upper and Lower Right:* Photographs taken 3 months after total cranial reconstruction showing that the cranium is both shorter and wider.

is considered to be the major cosmetic problem, the occiput is first reattached to the central pillar, and *vice versa* if the occiput is worse. This attachment is also rigidly fixed with microplates and screws. Finally, the area of greatest deformity is replaced, being attached inferiorly at the supraorbital ridge or the occiput with microplates. An intracranial pressure (ICP) monitor† is placed through a twist-drill hole in the central pillar as far forward as possible and the ICP is monitored as wires are slowly twisted between the last segment and the central pillar. This pulls either the frontal or the occipital bone toward the central segment and serves to reduce the anteroposterior length. Normocarbica is maintained during this phase of the procedure and twisting is stopped if the ICP rises above 15 mm Hg. Further tightening of the wires is resumed when the pressure decreases. We have found that the dural plication sutures afford much of the correction, and an acceptable skull shape is usually possible without undue pressure from the last segment of bone attached.

Any remaining skull defects are filled with the previously obtained graft material; in older children, it is possible to split the bone if further graft material is needed. The pericranium is loosely repositioned over the vertex, and the temporalis muscle is attached either

† Monitor manufactured by Camino Laboratories, San Diego, California.

TABLE 1

Clinical characteristics and surgical data of patients with late sagittal synostosis

Case No.	Age (yrs.) Sex	Hospital Stay (days)	Duration of Surgery (mins)	Blood Loss (cc)	Morbidity*	Associated Abnormalities
1	2.5, M	6	390	1000	SSS tear, SIADH, skull defect	—
2	2, M	5	330	1100	SIADH, seizure	hypophosphatemic rickets
3	4, M	4	380	250	—	—
4	1.5, F	7	270	350	—	—
		5	125	150	—	—
5	2, F	4	300	300	—	developmental delay
6	3, M	5	300	500	—	—
7	5, M	5	315	200	—	—
8	3, M	5	300	400	—	lambdoid synostosis
9	1.5, M	5	275	250	—	—

* Abbreviations: SSS = superior sagittal sinus; SIADH = syndrome of inappropriate antidiuretic hormone secretion.

to the pericranium or to holes placed in the bone. The galea and skin are closed in two layers. Although a compressive head dressing has been used previously, subgaleal swelling has been significant; we now use a closed-system drain in the subgaleal space.

Results

Ten procedures were performed on nine patients. One child had such an elongated head that her occiput touched her upper back when she was placed in the prone position with the neck hyperextended. An initial anterior reconstruction with the patient in this position was followed 1 month later by a posterior reconstruction with the patient in the prone head-down position.

Complications

The duration of surgery ranged from 275 to 390 minutes (average 324 minutes) in those children whose deformity was repaired in one stage (Table 1). Blood loss was quite significant in most cases, ranging from 200 to 1100 cc (average 500 cc). One child with a 1100-cc blood loss had hypophosphatemic rickets and extremely vascular bone, and one child with a 1000-cc blood loss had disruption of the superior sagittal sinus. Total blood transfusion ranged from 200 to 1000 cc (average 485 cc) in single-stage repairs. Morbidity was restricted to the two children with the highest amount of blood loss. Both had hyponatremia postoperatively secondary to the syndrome of inappropriate antidiuretic hormone secretion. One child became obtunded and the other suffered a generalized seizure. Both responded to correction of the hyponatremia and neither had permanent sequelae. The hospital stay ranged from 4 to 7 days (average 5 days).

Deformity Resolution

In each case the preoperative goal of shortening and widening the calvaria was accomplished (Figs. 2 to 4).

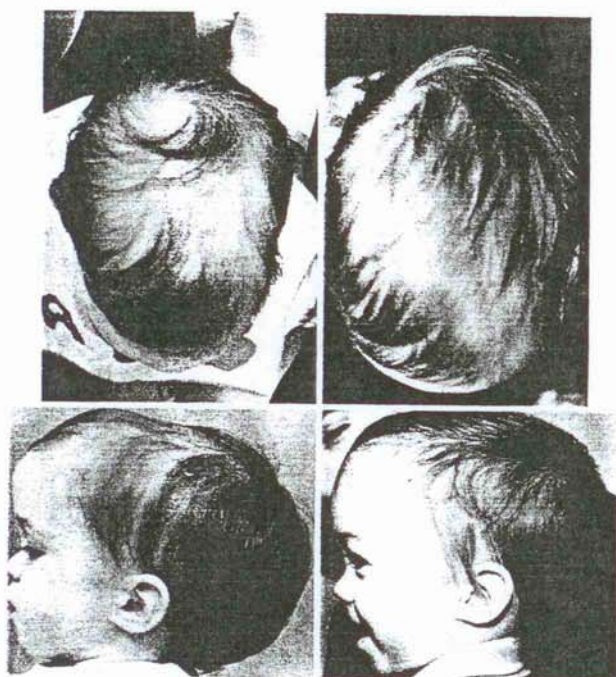


FIG. 4. *Upper and Lower Left:* Preoperative photographs in Case 2. The cranium is markedly elongated and narrow biparietally, and the forehead is protuberant. *Upper and Lower Right:* Photographs obtained 4 months after surgery. The cranium is shorter, wider, and as viewed from above, much rounder.

Additionally, deformities of the forehead and occiput were significantly ameliorated. Palpable irregularities in the recontoured bone, present in the first postoperative months, have smoothed considerably over time. One child developed a 2-cm skull defect in the parietal area 9 months postoperatively. This is being followed with the expectation that new bone growth will bridge the gap. There were no deaths and, with the possible exception of the previously described skull defect, no permanent morbidity.

Change in Head Shape

Confirmation of the change in head shape was obtained by CT in one case (Fig. 5). A ratio of the calvarial anteroposterior length:width was acquired from both pre- and postoperative scans. The preoperative ratio was 1.48:1 (15.1:10.2 cm), indicating an elongated head consistent with sagittal synostosis. The postoperative length:width ratio was 1.26:1 (14.2:11.3 cm), representing an improvement. Postoperative CT was not routinely performed, as cosmetic improvement was clinically apparent in all patients.

Discussion

Rationale for Surgery

It is certainly appropriate to question the rationale of performing such an extensive procedure on children who are usually normal except for head shape. Few would question surgical correction in infancy by either

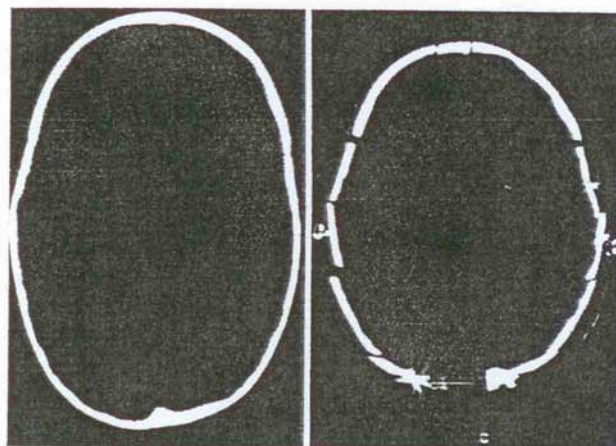


FIG. 5. Computerized tomography (CT) scans in Case 9. *Left:* Preoperative CT scan revealing marked elongation of the calvaria, with a narrow biparietal diameter. *Right:* Postoperative CT scan at the same level showing improvement in the contour, with an improved length:width ratio.

strip craniectomy or one of the many other procedures that have been utilized.^{1-3,5-12,14,18-20,22} What changes for the older children is the risk to benefit ratio: is the anticipated change in head shape worth the risk of undergoing a procedure involving total reshaping of the calvaria? It is difficult to understate the potential negative effects on peer acceptance and self-image of a child with an abnormally shaped head. Shillito and Matson¹⁸ described the case of a 7½-year-old boy who developed a duodenal ulcer because his second grade classmates made fun of his elongated head. Surgery was not performed because of his age. Mullan¹³ reported a 4¾-year-old boy with severe sagittal synostosis. After playmates began making fun of his cranial deformity, he became withdrawn and would hide from visitors. Following a three-stage reconstructive procedure, he had "regained his former self-confidence." Parents of the two oldest children in our series requested surgical intervention because their children were being called "banana head" and "boat head" by other children.

A report from Australia by Barritt and colleagues⁴ studied the families of children with sagittal synostosis and described a high incidence of anger and anxiety among those parents who elected no surgical correction for their child. Two marriages have "foundered" because of disputes regarding the child's deformity and lack of treatment. Nine of the 34 untreated children have been teased about their deformity and, in four cases, the child could no longer cope with the school environment. Our society places great significance on physical appearance and the importance of correction of the scaphocephalic deformity to psychosocial development should not be ignored.

Intracranial Pressure

Renier, *et al.*,¹⁷ measured ICP preoperatively in a series of children with craniosynostosis and reported abnormally increased pressures (> 15 mm Hg) in three

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(13%) of 23 children with scaphocephaly. While we did not monitor ICP preoperatively, one 15-month-old child showed evidence of abnormally raised pressure. In this child, closure of the lambdoid as well as the sagittal suture was demonstrated on CT and a significantly beaten-copper skull was visualized on plain films. His development and examination were normal and papilledema was not present. Thus, although not common, increased ICP can occur with sagittal synostosis, especially if other sutures are involved, and its presence would dictate surgical intervention.

Growth Patterns

Virchow's rule of craniosynostosis²¹ states that growth of the calvaria is restricted perpendicular to the closed suture and a compensatory increase occurs parallel to it. Persing, *et al.*,¹⁶ have expanded upon these observations to describe how single-suture synostosis can produce deformity of the entire calvaria. In the case of sagittal synostosis, biparietal narrowing takes place as growth is restricted across the sagittal suture, and the frontal boss and occipital shelf are created by increased bone deposition along the coronal and lambdoid sutures, respectively. This abnormal growth pattern creates the scaphocephalic deformity which becomes more pronounced as calvarial growth continues. Thus, the distortion continues, albeit slowly, during early childhood until the skull has reached adult dimensions.

Because the entire calvaria is involved in late sagittal synostosis and simple opening of the closed suture and continued cranial growth will not correct the deformity, a procedure designed to totally reshape the calvaria is necessary. To allow access to the entire cranial vault, we have adapted the prone position with cervical hyperextension as described by Park, *et al.*¹⁵ The Vac Pac positioning system, which molds to the child, is used to maintain this position.

Surgical Goals

The goals of the procedure are to shorten the anteroposterior (sagittal) dimension of the calvaria while widening the biparietal diameter. Additionally, correction of the frontal boss and occipital protuberance are important. We have accomplished these goals utilizing the techniques described by Persing, *et al.*¹⁶ The importance of frontal dural plication and the use of ICP monitoring should be emphasized. We have found that a significant shortening of anteroposterior length, correction of the frontally widened dura (equivalent to the frontal boss), and widening of the brain and dura into the space created by the temporal bone out-fractures occur following the bilateral frontal dural plication. As a result, less compression is needed when the bone is replaced. The anteroposterior skull length reduction is performed by removing 2 to 3 cm from one of the biparietal struts (usually the last one replaced). The bone gap thus created is closed by gradually twisting wires to bring either the frontal or the occipital bone (depending upon which is thought to constitute the worst preoperative deformity) to the strut. The ICP is watched closely during this anteroposterior reduction

by placement of an ICP monitor in the right frontal lobe. If the ICP rises above 15 mm Hg, tension is released from the wire and the tightening is begun again only after the pressure decreases. It is important that this process is performed under conditions of normocarbia to reflect adequately the postoperative ICP. By slow reduction with frequent stops for equilibration, we have been able to accomplish adequate anteroposterior shortening in every case.

As older children do not have the bone regenerative capability of infants, all bone defects and gaps must be filled. This is accomplished by utilizing the bone removed from the biparietal strut as graft material. If the bone is thick enough, splitting it through the diploë will increase the amount of graft material available. We have not had to harvest graft material from the ribs, iliac crest, or other sources.

Complications

Intraoperative blood loss has been significant, averaging 500 cc. The magnitude of the bone work and the prolonged nature of the procedure are responsible for most of the loss. In one case the superior sagittal sinus was opened with a craniotome during removal of a biparietal strut, resulting in a blood loss of several hundred cubic centimeters until hemostasis was achieved. Prior to this complication, we had tried to minimize burr hole placement to reduce the amount of skull defects that would require grafting. We now place a burr hole in the posterior midline of each biparietal strut to facilitate dissection of the dura from the overlying bone. Enough graft material has been available to fill the holes thus created. Significant blood loss (1100 cc) also occurred in a child with hypophosphatemic rickets due to the very vascular nature of his bone. Both of these children developed a syndrome of inappropriate antidiuretic hormone secretion and were treated with concentrated saline solution administration and fluid restriction; there were no permanent sequelae in either case. This syndrome was probably precipitated by the large fluid shifts during the surgical procedure and occurred 14 to 48 hours postoperatively. In an effort to prevent this complication, we administer isotonic saline solutions postoperatively and enforce mild fluid restriction ($\frac{2}{3}$ to $\frac{3}{4}$ maintenance). Sodium levels are checked frequently and symptomatic hyponatremia is aggressively treated with infusion of hypertonic saline solutions.

A potential risk of this procedure is bone resorption and development of skull defects. This occurred in only one case in our series, with the skull defect remaining relatively small. In an effort to prevent this complication, pericranium is pulled over the skull construct as far as possible and, along with the dura, should insure adequate blood supply to the devascularized bone. It is hoped that rigid fixation with microplates and screws will reduce the bone resorption that can occur secondary to movement of the bone plates.

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