Variants of sagittal synostosis: strategies for surgical correction

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Sagittal synostosis is discussed with respect to the variations seen with the deformity. The morphological spectrum ranging from marked frontal bossing to prominent occipital bulging is described. Surgical techniques have been specifically designed for these variants. These techniques provide an immediate correction of scaphocephaly, and achieve a reduction of the specific deformity with morbidity comparable to that associated with conventional operations. The lack of large areas of craniectomy and the avoidance of synthetic materials are cited as additional advantages of these techniques. The importance of altering the surgical approach to the specific clinical problem is underscored. Two illustrative cases of sagittal synostosis variants are described, and recent experience with the modified operative techniques in treating these and similar cases is discussed.

KEY WORDS • craniosynostosis • craniectomy • cranioplasty • sagittal suture • scaphocephaly • skull

Sagittal synostosis is a deformity that probably poses little threat to normal brain development. Surgical treatment must therefore be justified and evaluated on cosmetic grounds. The goal of operative management is the restoration of an esthetically pleasing skull contour, not simply synostectomy. Optimal results require that consideration be given to several factors, including the timing of surgery, the specific deformity affecting the patient, and the advantages and disadvantages of the various operative techniques.

The importance of early surgery in the correction of craniosynostosis has been repeatedly emphasized in the literature. The consensus has been that operation after the first 6 months of life is met with less favorable results than surgery performed earlier. When surgery is performed after 1 year of age, little reliance can be placed on further brain growth to achieve a normal skull shape. It is therefore particularly important in planning surgical correction for the older infant that an interventional change in skull shape be achieved.

As important as the timing of surgery is an assessment of the cosmetic problem on an individual basis. Much heterogeneity is seen in any large group of patients with sagittal synostosis, and this must be considered in operative planning. A partial or localized area of synostosis may occur along the sagittal suture, which produces only a localized area of depression in the calvaria without appreciable scaphocephaly (Fig. 1). Clearly, the surgical management of this variant is quite different from that required in a patient with general-

Fig. 1. Left: Schematic lateral projection of skull demonstrating an area of localized sagittal synostosis posterior to the coronal suture. Note the relative depression at the site of synostosis as well as the compensatory frontal bossing. Right: Oblique projection to demonstrate normal skull configuration (broken line) in comparison to localized sagittal synostosis.
ized fusion of the suture with a scaphocephalic deformity. In patients with complete synostosis of the suture, the resultant skull deformity may vary considerably owing to differences in the form in which compensatory growth occurs. This compensatory growth, which is perpendicular to the fused sagittal suture, may occur such that there is a preponderance of frontal bossing, a marked occipital bulge, or a mixture of both. We have presumed that this is a result of disproportionate growth at the coronal or lambdoidal sutures and may reflect the site of initial synostosis, whether anterior or posterior. Still other patients may exhibit an exceptionally narrow biparietal or intertemporal diameter as an additional aspect of a deformity resulting from sagittal synostosis. In these latter two subtypes, the operation performed should be modified to allow for expansion of the dura at these sites. This can most often be accomplished by slightly larger areas of craniectomy in the appropriate regions.

The decision as to which technique to use depends upon the age of the patient and the particular deformity. For instance, in the first 6 weeks of life simple synostectomy may be adequate for the three major morphological variants seen with synostosis; namely, anterior compensation with frontal bossing, posterior compensation with occipital bulging, and localized synostosis. Exceptionally narrow bitemporal or biparietal diameters are considered subtypes of these three main categories. With increasing age, synostectomy becomes increasingly ineffective in the correction of these deformities, with the degree of ineffectiveness being related to the overall severity of the condition. For example, mild scaphocephaly probably is effectively treated with synostectomy performed up to 6 to 8 months of age. Severe frontal bossing, on the other hand, may need special treatment by 3 months of age, in spite of the tendency for frontal prominence to decrease with age both in normal children and in those with scaphocephaly.

It is, therefore, our belief that although no one surgical technique will be adequate for all, one surgical attitude will be. This attitude involves an emphasis on correction of the specific deformities harbored by the individual patient.

A previous report from this institution has described a surgical technique for the immediate correction of sagittal synostosis which we have termed the "pi procedure" as this describes the shape of the initial bone removal. This operation has the advantages of immediate alteration in skull contour, avoidance of early postoperative suture reclosure, and the elimination of large craniectomy defects. There is also the possibility that this operation will yield superior cosmetic results in older infants given the dynamic skull shortening involved.

The purpose of the present report is to describe two modifications of the pi procedure which retain all of the above advantages while yielding superior cosmetic results. These techniques were used in patients with two variants of sagittal synostosis; namely, sagittal synostosis with exceptional frontal bossing, and sagittal synostosis with marked occipital bulge.

**Summary of Cases**

**Clinical Material**

Our series was derived from the last 23 cases of isolated synostosis of the sagittal suture treated in our institution. Twelve patients were treated with the standard pi procedure while eight were thought to have sufficient occipital deformity to warrant the reverse pi procedure.

**Fig. 2. Standard pi procedure.** A: Initial bone flaps as removed with a craniotome. Note the sites of original burr-hole placement as well as the extension of flaps into the temporal region. B: Full extent of bone removal. An additional craniectomy is made across the coronal suture and into the temporal region bilaterally. The craniectomy also extends across the lambdoid suture posteriorly (arrow). C: Undermining the dura along the sagittal strip. Holes have been made in the frontal bone as well as in the sagittal strip for placement of wires. Bone flaps are replaced by suturing them to dura. D: Immediate reduction of the anteroposterior diameter with wires. An allowance is made for a compensatory bulge in the temporal region, further restoring a normal skull contour.
Variants of sagittal synostosis modification to be described. One child presented with marked frontal bossing with localized sagittal synostosis and was treated with the pi procedure with the frontal modification also to be described. Two additional children who presented at 3 and 4 years of age, respectively, were treated with acrylic overlay cranioplasty and will therefore not be considered further. The follow-up period ranged from 6 months to over 5 years.

Operative Procedures

All patients being prepared for craniosynostosis surgery have placement of an intravenous line suitable for the administration of blood products. Following the induction of anesthesia, the patient's head is positioned on a well padded headrest with emphasis on preventing excessive cutaneous or, in the case of the prone position, ocular pressure. We are presently using a horseshoe headrest generously padded with orthopedic soft roll. The head is further supported by a sling of tape brought transversely across the headrest. Also, for the prone position, the patient's face is protected with adhesive foam padding applied prior to turning prone. By exercising these precautions we have avoided any pressure injury in our series of patients.

Sagittal Synostosis With Occipital Bulge

For correction of sagittal synostosis with occipital bulge, the patient is positioned prone. A bicoronal incision is used which extends slightly posteriorly at the vertex. The scalp flaps are reflected to expose the calvaria from the coronal suture to the lambda. The pericranium is left intact to avoid bone bleeding. Burr holes are then placed bilaterally approximately 1.5 cm lateral to the midline and 1.5 cm anterior to the lambdoid suture (Fig. 3A). Two L-shaped bone flaps are created with the craniotome, extending parasagittally to a point anterior to the coronal suture with the lateral limbs extending along the lambdoid suture. The dura is then carefully dissected from beneath the lambdoidal and coronal sutures and the craniectomy is performed across these sutures. Bone removal is further carried across the midline posteriorly to complete the craniectomy which is in the shape of the Greek letter pi (π) (Fig. 3B). The dura is then separated from beneath the sagittal suture anteriorly. This can be accomplished easily and without excessive bleeding in infants in whom the suture has prematurely fused. Two holes are then made in the sagittal strip as well as in the occipital bone using towel clips or alternatively the high-speed drill (Fig. 3C). Wires are threaded into the holes and shortened. The anteroposterior dimension may be safely shortened by as much as 10 mm. The dura will then bulge into the areas of parietal craniectomy, thus creating a more normal contour with greater biparietal width. The bone removed may then be reshaped if necessary and replaced by suturing it to the dura (Fig. 3D). Conventional scalp closure is then carried out. Drains are not used. Postoperatively, the infant is closely observed for the first 24 hours, with serial determinations of hematocrit and transfusion of blood as necessary.

Sagittal Synostosis With Marked Frontal Bossing

For correction of localized or complete sagittal synostosis with marked frontal bossing, the patient is positioned supine as in the standard pi procedure. A standard bicoronal incision is made, and the scalp flaps are reflected. Parasagittal burr-holes are placed and bone is removed with the craniotome and rongeurs, incorporating the sagittal suture and extending across the lambdoid and coronal sutures (Fig. 4A). Bone is also removed in a bicoronal fashion (Fig. 4B). In this case, however, the areas of frontal bossing are removed with the craniotome, leaving a midline strip of bone, and the bone that has been removed is reshaped. The frontal dura is plicated and the bone is replaced by suture to the dura (Fig. 4C). Wires are then inserted laterally in the frontal bone (Fig. 4D) to decrease frontal bossing and achieve some degree of shortening.

Results

There has been no operative mortality with the pi or reverse pi procedures, or with the pi procedure with

Fig. 3. Correction of sagittal synostosis with occipital bulge. A: Areas of bone removed with the craniotome. Note the placement of burr holes. B: Full extent of bone removal. The craniectomy extends across and along the lambdoidal suture. Note the anterior extent of the craniectomy crossing the coronal suture. C: Placement of wires in the sagittal strip and occipital bone. Prior to wire placement the dura was dissected from beneath the sagittal bone strip to facilitate shortening and prevent kinking of the sagittal sinus. D: Reduction of the anteroposterior diameter with wires. Removed bone is replaced by suturing to dura.
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Frontal modification. No reoperations were performed or even considered desirable in this series, either for correction of persistent cosmetic deformity or for premature suture reclosure. We have recently seen a patient who presented with a primarily occipital deformity that we corrected with a reverse pi procedure. The child now has a mild degree of frontal bossing, but the parents do not believe that it warrants correction.

One patient required a minor operative procedure for the removal of an extruded wire. This patient exhibited slight, but transient, asymmetry of skull growth following unilateral wire removal, although no skull asymmetry was noted at a follow-up visit at 18 months of age. This patient represents the only case of significant, although reversible, morbidity in this series. Only two patients were treated entirely without transfusion either intraoperatively or in the immediate postoperative period. These patients were Jehovah’s Witnesses and their pre- and postoperative hematocrits were 33.7% and 28% in the first case, and 33% and 29% in the second case.

Case Reports

Two illustrative cases of the modified procedures are presented.

Case 1

This 1-month-old baby girl presented with scaphocephaly, first noted at birth. The child weighed 6 lb 5 oz at birth, the product of a full-term pregnancy with no complications. The patient had been in good health since birth but was noted to have an abnormal-shaped head.

Examination. On examination, the child was noted to be normal except for an abnormal-shaped head, which was scaphocephalic with a prominent posterior sagittal ridge. The head circumference was 39 cm. The anterior fontanel was patent and soft. A sagittal ridge was palpable along the posterior sagittal suture, and there was marked bulging in the occipital region. The biparietal diameter of the skull also appeared slightly narrow (Fig. 5). The patient was neurologically normal.

Operation. Surgery was performed as previously described for posterior sagittal deformity. Skull shortening of approximately 6 mm was obtained. The patient was discharged on the 6th postoperative day in satisfactory condition. The skull contour was considered to be normal.

Case 2

This 6-month-old baby boy was in good health until 2 months of age when his mother noted that his head

FIG. 5. Case 1. Left: Lateral preoperative photograph demonstrating sagittal synostosis with prominent occipital bulge. Note the normal configuration of the forehead. Right: Postoperative photograph demonstrating a normal occipital contour. Arrow indicates the location of the incision.
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![Image](image_url)

Fig. 6. Case 2. Upper Left: Lateral preoperative photograph demonstrating localized sagittal synostosis with an area of depression in the coronal region and marked frontal bossing. Compare with Fig. 1. Upper Right: Postoperative photograph obtained 1 week after surgery demonstrating marked diminution of frontal bossing. Lower Left: Preoperative skull radiograph demonstrating marked frontal bossing with a localized area of sagittal depression posterior to the coronal suture. The scaphocephaly in this patient was more apparent on the skull radiograph than on physical examination. Lower Right: Postoperative lateral skull radiograph obtained 10 months postoperatively demonstrating reduction of frontal bossing as well as a significant decrease in anteroposterior skull diameter. Note the correction in the localized area of sagittal depression as well as the growth of new bone into the areas of previous craniectomy.

was elongated. Physical examination and skull films performed by his family physician suggested the diagnosis of sagittal synostosis.

Examination. Physical examination revealed the child to be well developed and well nourished. The head was elongated, with a palpable sagittal ridge and a marked localized depression. There was profound frontal bossing (Fig. 6). The remainder of the examination was normal except that the patient was noted to be diffusely hyperreflexic.

Operation. The child underwent surgery on the 2nd hospital day, consisting of the pi operation modified for frontal bossing. He was discharged on the 7th postoperative day following an unremarkable postoperative course. The cosmetic result was considered to be excellent.

Discussion

Several operative approaches for the correction of sagittal synostosis have been described. Earlier techniques generally consisted of strip craniectomy, either incorporating or running parallel to the fused suture. Good results can be obtained with these methods, although recurrent synostosis is not uncommon. Subsequent attempts to prevent this complication have included painting the dura with caustic agents, the application of polyethylene films to the bone edges, or the interposition of Silastic material into areas of craniectomy. The first technique may cause injury to subjacent brain as suggested by Pawl and Sugar and Marlin, et al. The latter two techniques are cumbersome and carry with them a theoretically greater risk of infection. Recently, more extensive craniectomy has been advocated as a means of preventing premature re-fusion. Greater bone removal, however, has the disadvantage of generating significant parental anxiety as well as potentially increasing susceptibility to minor head trauma.

The pi technique, first described in 1978, accomplished the cosmetic goals of the operation without premature suture re-fusion, the use of synthetic materials, or residual large craniectomy defects. We have hypothesized that the separation of dura from the inner table, as is done in this operation, along with the active shortening performed, changes the relationship between the vault, clivus, and skull base, and thus "cures" the disease, preventing premature re-fusion. We assume that the disease consists of an abnormal base transmitting anomalous growth information to the vault through the dura as originally suggested by Moss.

Despite satisfactory results using the pi operation, as originally described, in over 22 children, it became apparent that sagittal synostosis was a heterogeneous entity. Particularly common variants are sagittal synostosis with a marked occipital bulge and sagittal synostosis with excessive frontal bossing. We have, therefore, modified the original pi operation to make it more applicable to sagittal synostosis with occipital bulge and sagittal synostosis with excessive frontal bossing. Over the past 5 years we have used the pi procedure and the modifications described here for the treatment of sagittal synostosis in patients presenting under 1 year of age. We have observed immediate postoperative improvement in skull contour in all patients without the presence of large bone defects and without utilization of polyethylene or Silastic material. No incidence of premature postoperative re-fusion of the sagittal or other sutures has been noted. These operations can routinely be accomplished in under 1½ hours with a rate of morbidity that is consistent with the cosmetic nature of the procedure.

We recommend that each case of sagittal synostosis be assessed on an individual basis with special regard to morphological variants within the disease entity. Specific aspects of each deformity should be noted, especially the degree of frontal or occipital bossing or areas of localized depression due to local synostosis, and these aspects should then be considered in selection of the operative approach. Our experience suggests that
either the pi procedure or its modifications described here produce consistently excellent cosmetic results with immediate correction of the deformity.

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