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# Total Cranial Vault Repair for Sagittal Craniosynostosis

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Sagittal craniosynostosis is the most commonly encountered form of craniosynostosis, accounting for an estimated 50 to 60% of isolated nonsyndromic craniosynostoses. It results from the premature fusion and subsequent growth arrest at the sagittal cranial suture. The general appearance of the cranium is one that is disproportionately long and narrow. This condition is termed scaphocepahly or dolichocephaly. Depending on the timing, location, and extent of the premature sutural closure, infants can present with a variety of skull shapes and forms.

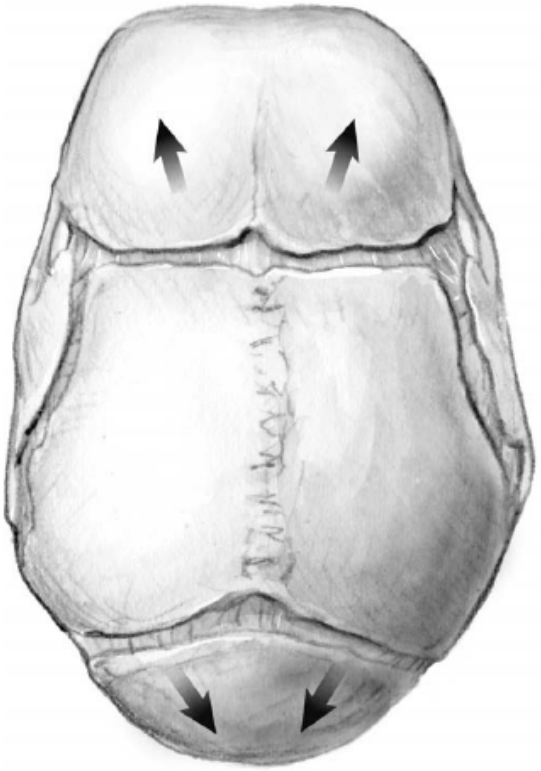

**Figure 44–1** Sagittal craniosynostosis. The deformity includes a disproportionately long and narrow skull. Compensatory growth can occur at the coronal and metopic sutures, causing frontal bossing or at the lambdoid sutures, resulting in an occipital knob.

Normal skull growth during the first year of life is marked by a tendency toward roundness, where the skull width grows at a greater rate than the skull length. By contrast, in sagittal synostosis, the ratio of skull width to skull length remains constant. This is an important consideration when recommending surgery to patients. It can be stated that, in fact, untreated sagittal synostosis appears to get "worse" with age. This occurs first because the natural tendency to roundness is prevented and second because in absolute terms the head is longer than normal. In addition to the elongated and narrow skull shape, there is often an accompanied occipital knob if posterior sagittal fusion predominates or significant frontal bossing is seen when anterior closure is most prominent. Complete early closure of the sagittal suture leads to both occipital and frontal compensations (**Fig. 44–1**). In most cases, the orbit and midface are unaffected in this process.

Most patients with sagittal craniosynostosis do not have generalized elevated intracranial pressure (ICP). The reported incidence of intracranial hypertension (ICH) ranges from 13.8 to 25% based on preoperative ICP monitoring. Preoperative single photon emission computed tomography studies have shown abnormal cerebral perfusion underlying the fused suture in over two-thirds of patients and positron emission tomography scans demonstrate diminished cerebral glucose metabolism. Children with sagittal synostosis also have an increased incidence of language and learning deficits. Although there is no definitive proof that surgical correction improves cognitive outcome, children in whom surgical intervention was performed >1 year of age experienced an increased incidence of cognitive delay compared with those undergoing surgery at <1 year.

## Patient Selection

Diagnosis is based on the characteristic physical appearance of an infant with a disproportionately long and narrow skull. Premature fusion leads to a palpable ridging along the length of the fused portion of the suture, and this finding can also be helpful. Plain skull films provide useful information regarding the suture patency, but confirmation is most readily obtained using three-dimensional computed tomography of the skull.

### Indications and Timing

The surgical goals are to improve the overall appearance of the skull and to provide adequate skull volume to relieve possible elevated ICP. Simply put, because sagittal synostosis is characterized by a long and narrow skull, the skull must be made shorter and wider. In addition, compensatory changes in the frontal and/or occipital regions, when present, should also be addressed.

Although subject to debate as to the exact timing, age at operation depends on the surgical technique. Craniofacial procedures such as the strip craniectomy or the endoscopic cranial release procedures that use a postoperative molding helmet to effect the change in skull shape are more often performed prior to 6 months. Our opinion is that the total cranial vault technique provides superior surgical control over the cosmetic result. We have found that at 6 months, infants are able to tolerate the volume loss of this more invasive technique well and the bone remains supple enough to manipulate during the reconstruction. Because the volume of the brain almost triples in the first year of life, subsequent growth of the brain can be utilized as an additional factor to help maintain the improved skull shape following surgical release of the fused suture. Early correction also spares the child emotional or psychological trauma over his/her appearance, before the child reaches the age of self-awareness at 5 years old (or younger).

An exception to our standard practice of performing surgery at 6 months is in the setting of ICH. In infants who appear to have elevated ICP, surgical correction is performed at the earliest possible date. The diagnosis of ICH based on clinical and radiographic examination can be difficult. The presence of papilledema on funduscopic examination is an excellent clinical marker for ICH. However, its absence does not exclude elevated ICP. Suture diastasis and sellar erosion as evident on plain skull radiographs are more sensitive indicators of raised ICP than the findings of cerebral digital markings seen on the inner table of the calvaria (beaten-copper markings). There are no definitive CT findings that correlate with ICP, but obliteration of the basal cisterns are a concerning finding. On occasion, an intracranial monitor is needed to accurately record pressures; however, this is generally reserved for those instances where the child has symptoms suggestive of ICH or where early correction has not been done and a decision to operate would be influenced by the presence of increased ICP.

## Preoperative Preparation

Once the decision has been reached to proceed with surgery, a preoperative workup consisting of routine blood tests including a complete blood cell count, electrolyte panel, and a prothrombin time and a partial thromboplastin time are performed. Due to the potential for significant blood loss, a type and screen are obtained, and compatible donors among relatives are encouraged to donate for donor-directed intra- and perioperative transfusions. There is some evidence that pre- and postoperative treatment with recombinant erythropoietin may help reduce the need for postoperative blood transfusion, but this is not part of our routine practice.

At least two large-bore (≥20 gauge) intravenous lines are required for access due to the potential for significant blood loss or fluid shifts during surgery. An arterial line is placed as well as a central line to monitor the total body intravascular volume for both operative and postoperative fluid management. The central line also obviates peripheral venous access issues postoperatively on the ward and significantly impacts postoperative comfort and the ease of care. A Foley catheter is useful to record urinary output, and a thermistor is used to record core body temperatures. A Doppler monitor is placed over the heart to monitor blood flow and is used to detect the possibility of unanticipated intraoperative air embolism. Steroids and anticonvulsants are not routinely used. Prophylactic antibiotics are given just prior to the incision but are not continued postoperatively.

In young children, the hair is clipped to allow the surgeon full visualization of the degree of the skull deformity, in order that the surgical correction can be tailored accordingly. This also helps facilitate the scalp closure and postoperative wound care by the nursing staff and the parents.

## Operative Procedure

### Positioning

Patient positioning depends on the specific deformity encountered; whether there are both frontal and occipital abnormalities, primarily frontal, or primarily occipital. The vast majority of patients are placed either in a modified prone ("sphinx") or supine position. The sphinx position is used for patients who have significant occipital as well as frontal compensatory changes in whom we plan to remove both an occipital and frontal flap. This position can give access from the supraorbital rims (although an orbital rim advancement would be difficult) to the foramen magnum. It also allows the surgeon excellent visualization of the complete cosmetic correction, a view not afforded if the surgery is performed in staged supine and prone positions. The supine position is used for scaphocephalic patients who primarily have frontal bossing without significant occipital deformity. This provides access from the supraorbital rims to just posterior to the lambdoid sutures. Access to the occipital bone is limited and requires lifting the infant's head, a maneuver that must be performed with care and close interaction with the anesthesiologist. On rare occasions in which the forehead appearance is normal and the deformity is mainly occipital, we will use a prone positioning. This generally allows access from the foramen magnum to several centimeters anterior to the coronal suture. In the prone and sphinx positions great care must be taken to ensure that the face is well padded to prevent pressure ulceration.

Bilateral tarsorrhaphies are performed, and the scalp and upper face are prepped with Betadine (povidone-iodine) solution and draped. The surgical table is rotated so that the anesthesiologist is positioned along the patient's side at the foot level. The nursing staff and all instrumentation, which have been placed on a single large table, are positioned op-

posite the anesthesiologist, who is also at the foot of the bed. A smaller Mayo stand is positioned over the patient's abdomen, and only those instruments most currently in use are kept for ready access.

### Skin Incision and Flap Elevation

A wavy bicoronal incision is performed extending from just behind one ear across to the opposite side. The scalp flap is elevated anteriorly to expose the supraorbital rims. The full extent of the supraorbital rims with elevation of the periorbita is not necessary. Posteriorly the scalp is elevated past the lambdoid sutures and inferior to the occipital knob, if present. The temporalis muscles are left attached to the scalp flap and are elevated off the underlying bone during the subperiosteal dissection.

### Craniotomy and Cranioplasty

There are several variations of our cranial vault reconstructions for sagittal craniosynostosis. In all, however, the operation begins with removal of a midline biparietal flap that includes the sagittal suture (**Fig. 44–2**). When the anterior fontanelle is still open, this can be used to dissect the dura from the overlying bone along the sagittal, coronal, and metopic sutures. When not present, the bur holes are fashioned on either side of the sagittal suture just posterior to bregma and anterior to lambda. The width of the biparietal flap varies, but is not taken further lateral than the horizontal portion of the parietal bone.

After the midline biparietal craniotomy is performed, a more complete elevation of the dura can be performed at the coronal and lambdoid sutures. Using the craniotome, parallel cuts are made on either side of the coronal and lambdoid sutures, and these sutures are removed at a width of approximately 1 to 2 cm (**Fig. 44–2**). The width of the coronal and lambdoid suturectomies varies depending on how much reduction is felt necessary in the anteroposterior dimension. The two lateral temporoparietal segments of the skull are reshaped through a combination of barrel-stave-like osteotomies and a Tessier bone bender (Lorenz Surgical, Jacksonville, Florida). The bones are still attached inferiorly, but they are outfractured to widen the skull appropriately. The removed midline biparietal flap is tailored and then turned 90 degrees to act as a strut maintaining the newly widened skull (**Fig. 44–3**).

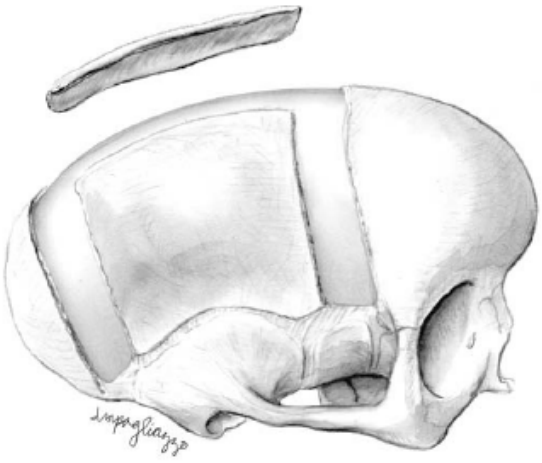

**Figure 44–2** Bone removal. All variations of our cranial vault reconstructions remove a midline biparietal flap and include coronal and lambdoid suturectomies. Depending on the deformity, the frontal and occipital bones are also removed.

The variations of the operation differ according to whether the frontal bone, the occipital bone, both, or neither is removed and reshaped. Patients who have neither frontal bossing nor a significant occipital knob can be managed by simply reapproximating the frontal and occipital bones with the temporoparietal segment that is still attached to the skull base. The amount of shortening is dependent on the width of the lambdoid and coronal suturectomies. The majority of patients, however, require removal of either or both frontal and occipital bones.

Patients who have significant deformities in both the frontal and occipital regions undergo bifrontal and biocipital craniotomies (**Fig. 44–3**). It is should be emphasized that the occipital craniotomy should be taken inferior to the occipital knob into the suboccipital region. If this is not done, a shelf of bone remains that detracts from the apparent shortening of the skull. Tremendous care must be taken to protect the torcula and transverse sinuses during removal of this bone. If a bioccipital flap has been removed, the occipital knob is reshaped by a series of radially oriented bone cuts, allowing the bone to be flattened yet still kept intact. Once reshaped, the bone is gently squeezed toward the temporoparietal segments to simultaneously shorten the skull and cause a bulging of the dura and brain laterally to reinforce the outfractured segments. This squeeze is maintained by rigidly fixing the bones in place using absorbable plates and screws. If forehead correction is also necessary (as it is in the majority of cases) the bifrontal craniotomy is reshaped in a fashion similar to that used for the occipital knob. The skull length is further reduced by a bringing the frontal bone in proximity to the attached temporoparietal segments and fixing the flap using absorbable miniplates. The frontal bone is reattached to the supraorbital rims using either absorbable suture or miniplates. The remaining bone segments from the removed coronal, lambdoid, and sagittal sutures are then loosely attached to adjoining skull segments to fill any gaps with absorbable Vicryl suture. Rigid fixation is used only in areas under stress to maintain the anteroposterior squeeze and the transverse widening.

### Closure Technique

The operative field is copiously irrigated to remove all nonviable debris and bone dust that could act as a nidus for future infection. Because the temporalis muscles were left attached to the scalp flaps, there is no need to reattach them along the lateral temporal ridges. They will naturally return to their orthotopic positions as the scalp flap is closed.

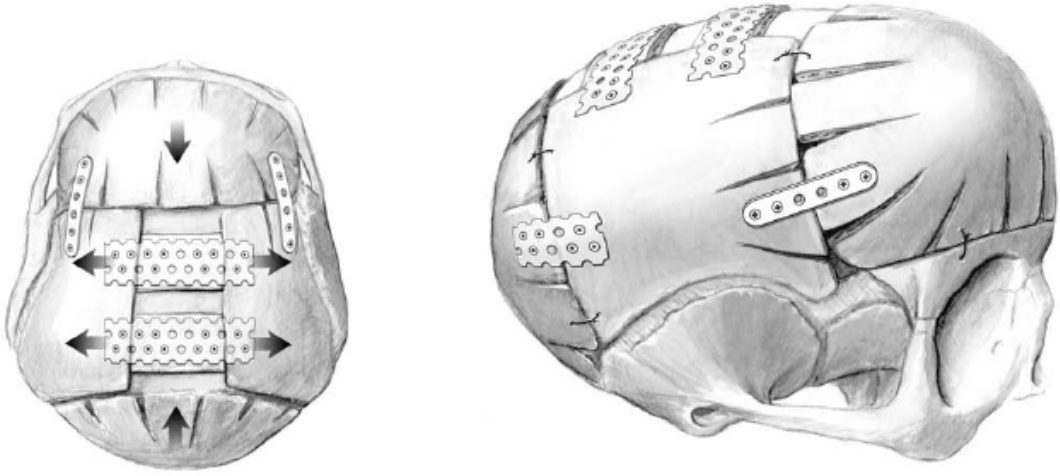

**Figure 44–3** Cranial vault repair. The skull is widened and held in place by bone struts from the removed midline biparietal craniotomy. The occipital and frontal bones are reshaped and brought into proximity to the fixed temporoparietal bone. This shortens the skull and addresses both frontal and occipital compensatory deformities. The amount of shortening is dependent on the width of the coronal and lambdoid suturectomies. Absorbable miniplates and sutures are used to maintain the correction. At major stress points, sturdier miniplates are necessary.

The scalp flap is reapproximated and closed in a two-layer fashion, with buried absorbable sutures in the subgaleal plane, and the skin is closed either using a 4–0 subcuticular absorbable suture or an external running 5–0 fast-absorbing plain gut suture. We have previously placed a subgaleal drain but have abandoned this practice. Although we do believe that it reduces the amount of postoperative swelling, our impression is that it also increases the need for postoperative transfusion. Instead, a snug but nonconstricting dressing is then wrapped in turban fashion over the entire head, and the head is kept elevated at all times to reduce postoperative swelling. Extubation is performed as soon as medically possible.

## Postoperative Management Including Possible Complications

Immediate postoperative care occurs in the pediatric intensive care unit. Vital signs and arterial and central venous pressures are monitored. Laboratory values are obtained for review of evidence of hypovolemia and blood loss. Depending on the degree of blood deficit, blood transfusions are frequently necessary and the concomitant replacement of coagulation factors may be required. Appropriate dosages of intravenous analgesia are given to keep the patient as comfortable as possible while maintaining a neurological examination. On the ward we alternate acetaminophen and ibuprofen in scheduled doses for the first few days. The child is positioned such that the head is elevated at 30 degrees to reduce swelling as much as possible. Commonly, the eyelids will swell shut within 24 hours following surgery. This can cause anxiety for both the parents and the child, and reassurance must be given that the majority of the swelling will resolve in 3 to 4 days.

In most instances, the child is ready for transfer to the regular ward within 24 to 48 hours. A routine CT scan is not performed. Usually, the child will run a slightly elevated temperature between 38°C and 39°C at day 3 or 4 following surgery. A routine fever workup is performed, but only rarely is the fever caused by infection. The child is considered stable for discharge from the hospital once he or she is tolerating a regular diet and eyelid swelling has resolved enough to permit eye opening. A follow-up visit is arranged for 1 week after hospital discharge.

### Complication Avoidance

Close communication with anesthesia cannot be overemphasized. Concerns of excess bleeding should be addressed expeditiously. Care should be taken during the scalp incision and reflection to stop bleeding from the skin edges and the pericranium. During the craniotomy the sources of ongoing blood loss must be recognized and controlled both from bone edges and from the dura. Our tendency is to minimize the amount of bone wax onto bone, as this may hinder osseous growth postoperatively. We have found FloSeal (Baxter Corp., Freemont, California) to be effective in controlling most low-flow bleeding along the bone edges. Particular attention must be paid to emissary veins leading from the sagittal sinus. As the major venous sinuses are exposed, patties should be immediately placed over the sinuses to control bleeding. Bipolar cautery generally controls dural bleeders quite easily. Nevertheless, blood loss occurs during our technique and it has become our practice to transfuse at the onset of the operation, and often we will transfuse a unit of fresh frozen plasma for every unit of packed red

blood cells. Platelet transfusion should also be considered after significant blood loss has occurred but is not necessary in every operation.

The lack of bleeding does not obviate placing patties over exposed sinuses because of the risk of air embolus. Air embolism is a more significant risk in the sphinx position, particularly if the patient is slightly volume depleted. This can be detected by the precordial Doppler ultrasound, as well as by the end-tidal volume gas spectrometer. Treatment includes placing the patient in a Trendelenburg's position and flooding the field with saline to prevent further intake of air into the circulation. A small amount of air is usually tolerated with minimal deleterious effect.

Meticulous attention must be directed in looking for possible tears in the dura mater, which can lead to a persistent cerebrospinal fluid leak, postoperative meningitis, and bone resorption. Dural lacerations can be avoided by careful dissection of the dura from the bone prior to elevation of the flaps. The dura interdigitates with the open coronal, lambdoid, and metopic sutures and should be dissected free prior to attempted elevation of the craniotomies. If recognized, the tear can be easily repaired with a single absorbable suture of 4–0 Nurolon.

The most frequent late complications include infection, resorption of the bone flap or bone grafts, and relapse of the original defect. To prevent infection postoperatively, strict sterile technique must be adhered to and prophylactic antibiotics are used. If osteomyelitis occurs, there is a high risk of loss of the bone grafts. Patients should be followed on an annual basis through adolescence to monitor future growth of the skull and to document any relapse of the original condition, which could require further surgical intervention.

## Conclusion

Cranial vault reconstruction must address the specific deformities encountered in the individual patient. Sagittal craniosynostosis is characterized by a disproportionately long and narrow skull. In addition, compensatory growth can result in frontal bossing and/or an occipital knob. Although all require an operation that shortens and widens the skull, variations are performed to address the patient's specific frontal or occipital deformity.