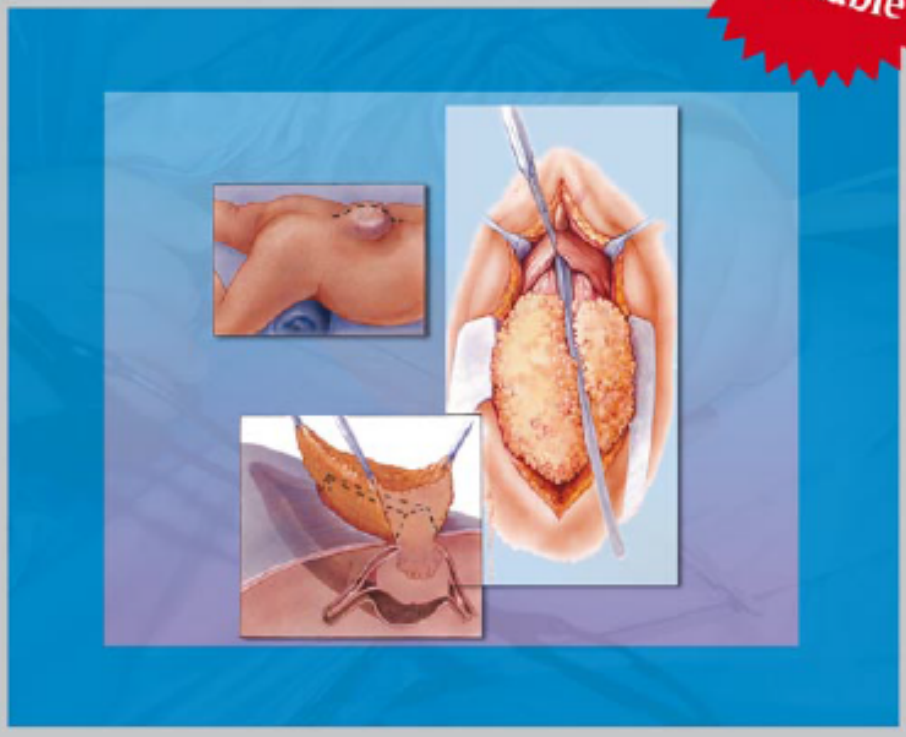
Neurosurgical Operative Atlas

Second Edition

Pediatric Neurosurgery

James Tait Goodrich

CME Credit Available



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Unilateral and Bicoronal Craniosynostosis

Kant Y. K. Lin, John A. Jane Jr., and John A. Jane Sr.

Coronal craniosynostosis is defined as the premature fusion of the coronal suture(s) of the skull; sutural involvement may be either unilateral or bilateral. Because of the position of the coronal suture, the consequences of premature fusion are manifested in the calvaria as well as in the face. As with all forms of craniosynostosis, compensatory growth occurs at the adjacent nonaffected suture sites. This is evident in cases of unilateral involvement at the contralateral coronal suture, where the contralateral side of the metopic and sagittal sutures, and at the ipsilateral squamosal suture growth leading to a unilateral anterior plagiocephaly (**Fig. 3–1**). In bilateral involvement, compensation is noted at both squamosal sutures, as well as at the sagittal suture leading to an overall turribrachycephalic or "tower-shaped" appearance (**Fig. 3–2**). With the resulting skull shapes being so disparate, operative treatments must be geared toward different issues, and the two types will be discussed separately.

Coronal craniosynostosis may be associated with elevated generalized intracranial pressure. The likelihood of this occurrence increases when more than one suture is involved. Bilateral coronal craniosynostosis in often associated with craniosynostosis syndromes, such as Crouzon's or Apert's syndrome. A distinction must be made between the syndromic and nonsyndromic varieties as management and expectations of outcome differ between the two.

Patient Selection

Diagnosis is based on the characteristic medical history and physical examination. Confirmation and more precise delineation of the dysmorphology are obtained from computed tomography of the skull. In particular, three-dimensional reconstruction of the images is useful for presurgical planning. A thorough ophthalmologic examination is indicated both for purposes of detecting intracranial hypertension, as well as to document any orbital axis issues related to the changes in the bony orbit secondary to the stenotic adjacent coronal suture. Often an eyelid ptosis or extraocular muscle imbalance is seen and must be addressed, usually after the bone deformities are corrected. Increasingly sophisticated DNA mapping techniques have resulted in an additional method of diagnosis that is especially useful with inherited forms of coronal craniosynostosis, or when a craniosynostosis syndrome is involved.

Indications and Timing of Surgery

Indications for surgical correction of a unilateral deformity are improvement of overall skull shape with advancement of a recessed forehead and correction of the bossed contralateral forehead, correction of the orbital dysmorphology, which can subsequently allow for correction of the orbital adnexal structures, and possible relief of either generalized or localized intracranial hypertension. Indications for surgical correction of the bilateral deformity are similar but also include the need to correct the overly high or tower-shaped skull and the overall brachycephaly.

Diagnosis of intracranial hypertension can be difficult and is based on "soft" findings such as cerebral markings seen on the inner calvarial table ("copper-beaten" appearance) on plain x-rays, or by late fundoscopic changes seen by slit lamp evaluation. Earlier signs suggestive of increased pressure may be seen with subtle behavioral changes in the child, or with a bulging anterior fontanelle. In the final analysis, a monitoring bolt is needed to accurately record pressures intracranially. Any evidence of elevated pressures is an indication for a more urgent need for surgery.

Although controversial, most surgeons would agree that surgery is best performed before the child has reached the age of 1 year. Our tendency is to perform surgery closer to 6 months of age. Because the volume of the brain almost triples in the first year of life, it would seem prudent to allow the intracranial cavity to accommodate this rapid growth through earlier surgery. The correction should certainly be performed before brain damage has occurred; often, subsequent brain growth can be utilized to help direct future growth and maintain the newly corrected skull and orbit shape following suture release and bony recontouring. Earlier correction also spares the child emotional or psychological trauma over his/her appearance, before the age of self-awareness (5 years or younger).

Preoperative Preparation

Once the decision has been made to proceed with surgery, a preoperative workup consisting of routine blood tests, including a complete blood cell count, electrolyte panel, and a pro-time and prothrombin time, are performed. Because of the potential for significant blood loss, a type and screen

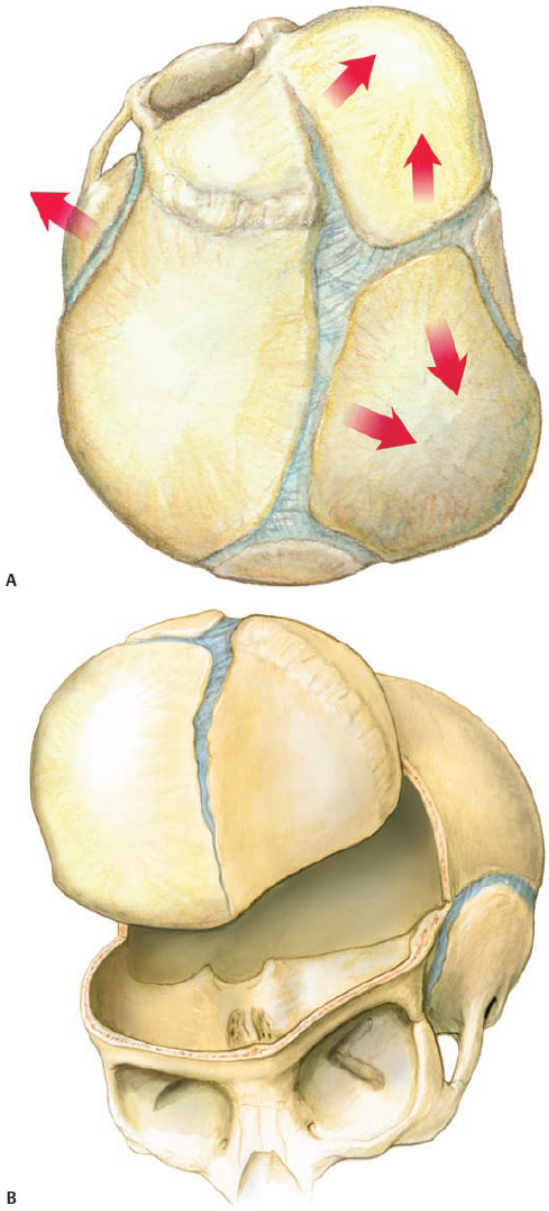

Figure 3–1 Skull deformity in unilateral coronal synostosis. The ipsilateral forehead is flattened and the superior and lateral orbital rims are recessed. **(A)** Compensatory growth (depicted by arrows) occurs at adjacent sutures. Compensatory growth at the metopic and contralateral open coronal sutures causes unilateral frontal bossing. Growth at the sagittal and open coronal sutures leads to a contralateral parietal bulge. **(B)** Skull-base deformity along the anterior cranial fossa also occurs.

are obtained and compatible donors among relatives are encouraged to donate for donor-directed intra- and perioperative transfusions.

The child is brought to the hospital on the day of surgery having been kept NPO for 4 hours prior to the anticipated start time for surgery. 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 and a central line is also helpful to monitor the total body intravascular volume for both operative and postoperative fluid management. A Foley catheter is useful to record urinary output, and a thermistor is used to record core body temperature. 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.

In young children, the hair is clipped to allow the surgeon full visualization of the degree of the skull deformity, so 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.

Once the intraoral endotracheal intubation has been performed, we have found it helpful to secure the tube with either a circummandibular or a circumdental wire, thus obviating the need for taping and allowing full access to the face during surgery. Temporary tarsorrhaphy sutures are also placed for intraoperative corneal protection.

Operative Procedure

Positioning

The patient with unilateral coronal synostosis is placed in a supine position on the operating table with the head resting in slight extension in a Mayfield headrest. The patient with bilateral coronal synostosis is positioned differently, the details of which will be discussed separately from the unilateral deformity. The headring is reinforced with additional soft padding to prevent excessive pressure over bony prominences during the lengthy procedure. The scalp and face are prepped with Betadine (providone-iodine) solution, with emphasis placed on scrubbing the external auditory canals, which tend to colonize with bacteria. The head, face, and neck are then draped to the clavicles, and staples are used to secure the drapes. A 180 degree access to the head and facial region is required, and the surgical table is rotated so that the anesthesiologist is positioned at the patient's side at the foot level. The nursing staff and all instrumentation, which has been placed on a single large table, is positioned opposite 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 standard wavy bicoronal incision is performed extending from just behind one ear across to the opposite side. Care is

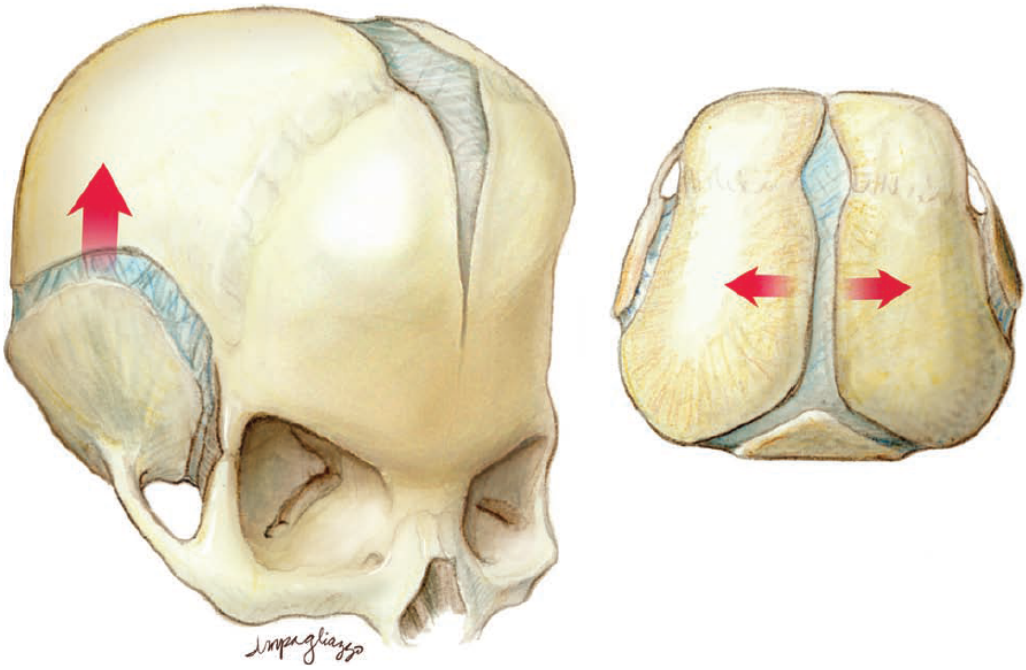

Figure 3–2 Skull deformity in bilateral coronal synostosis. Bilateral coronal synostosis leads to significant bilateral forehead flattening (with a decrease in the overall anteroposterior dimension of the skull) and recession of the orbital rims. Compensatory growth (depicted by arrows) at the squamosal suture causes vertical elongation of the skull. Growth at the sagittal suture causes the skull to widen.

taken to make the incision posterior to the anterior hairline, yet forward enough to allow for access to the orbital region once the scalp flap is dissected. The scalp flap is elevated anteriorly down to the level of the supraorbital rim. The supraorbital neurovascular bundle is preserved and may occasionally need to be freed from its foramen with a thin osteotome. Dissection is then extended laterally down each lateral orbital rim detaching the lateral canthi to the junction with the inferior orbital rim, and medially up to, but not detaching, the insertion of the medial canthal tendons. The nasolacrimal apparati are also carefully preserved. The nasion is exposed during this part of the dissection as well. Inferolaterally, the anterior aspect of the maxilla, the malar eminence, and the anterior aspect of the zygomatic arch are also exposed. The temporalis muscles are elevated off their insertions and left attached to the undersurface of the scalp flap, thus allowing access to the infratemporal hollow. The temporal and sphenoid bones are exposed from the lateral orbital rim close to the junction where the zygomatic arch meets the posterior temporal bone. This area will allow the formation of a tenon extension, once the orbital osteotomies are performed, of the supraorbital bone unit that will be advanced and reshaped to compensate for the temporal narrowing seen in this condition.

Craniotomy and Craniofacial Reconstruction

Unilateral Coronal Craniosynostosis

Emphasis has been placed on the concept that despite unilateral sutural involvement, the deformity is, in almost all cases and occasionally significantly, bilateral. Whereas the ipsilateral side reflects growth restriction, the contralateral side exhibits the effects of compensatory changes driven by the growth of the brain.

A bifrontal craniotomy is performed with the posterior extent of the cuts being posterior to both the fused and nonfused coronal sutures and the anterior cut ~1 cm above the level of the supraorbital rims. Retraction of the frontal and temporal lobes of the brain is then performed, taking care to remain anterior to each olfactory bulb. Three-quarter orbital osteotomies are completed across the orbital roof, superior aspect of the medial orbital wall, lateral orbital wall, and the lateral aspect of the orbital floor into the inferior orbital fissure. Tenon extensions are made extending laterally into the sphenoid and temporal bones. The final remaining cut is made across the nasion just above the nasofrontal suture (**Fig. 3–3**). This forms a single orbital unit that is removed in its entirety to be reshaped. The remaining portion of the

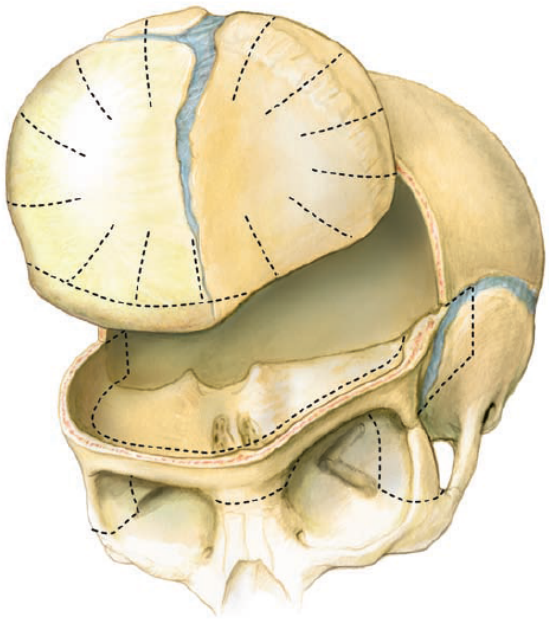

Figure 3–3 Craniotomy and orbital rim osteotomies for unilateral coronal synostosis. The surgery begins with a bifrontal craniotomy that includes both coronal sutures. Radial osteotomies are performed, and the frontal bone is recontoured. Bilateral three-quarter orbital osteotomies are then performed, elevating the visor as a single unit. Dotted lines depict areas of osteotomies.

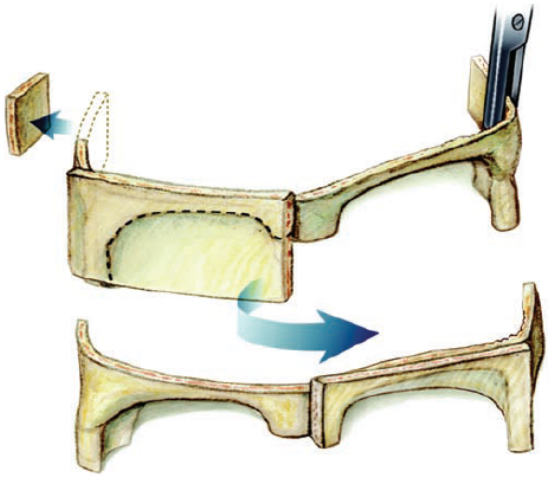

Figure 3–4 Orbital rim reconstruction in unilateral coronal synostosis. During the reconstruction, the ipsilateral superior and lateral orbital rims are advanced and reshaped to match the contralateral side. The contralateral orbital rim often needs to be recessed by removing a portion of the contralateral tenon. A template of bicortical graft is then placed over the contralteral orbital rim and is used as an onlay graft for the ipsilateral orbital rim. Further reshaping of the ipsilateral rim often requires a combination of burring down the inner cortex of the orbital rims, thus softening them enough to use the Tessier bone benders.

abnormally shaped and positioned greater wing of the sphenoid bone is then carefully rongeured medially up to the fused frontosphenoid suture, and into the superior orbital fissure. This will allow for subsequent brain expansion behind the newly configured orbital unit.

The goals for reshaping the orbital unit include: (1) advancement of the ipsilateral lateral orbital rim; (2) advancement of the retruded supraorbital rim in relationship to the inferior orbital rim in the anteroposterior (AP) plane; (3) recreation of the overall shape of the orbit to match the opposite orbit; and (4) recessment of the contralateral lateral orbital rim to take out any compensatory changes. These changes are effected by a combination of burring down the inner cortex of the orbital rims, thus softening them enough to use the Tessier bone benders to reshape the bone in the proper configuration. The recessed portion is given additional projection via advancement of the tenon extension along the lateral temporal bone. A portion of the distal end of the tenon extension on the contralateral side is removed to allow for recessing, again at the temporal bone region (**Fig. 3–4**). Finally, the retruded supraorbital rim and the reshaping of the orbital box are addressed simultaneously by placing an onlay bone graft, harvested from the bifrontal bone piece and fixed with an absorbable lag screw, over the deficient area and burred to the matching configuration of the opposite side (**Fig. 3–5**).

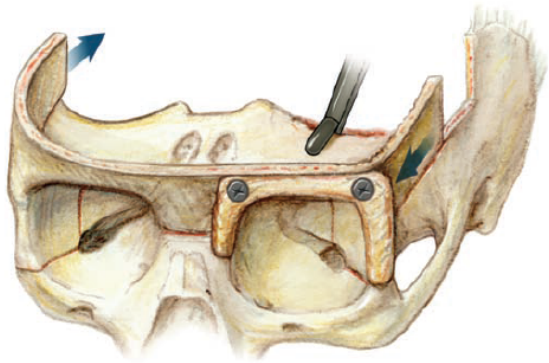

Figure 3–5 Orbital rim reconstruction and advancement in unilateral coronal synostosis. The recessed portion is given additional projection via advancement of the tenon extension along the lateral temporal bone. A portion of the distal end of the tenon extension on the contralateral side is removed to allow for recessing, again at the temporal bone region. The onlay graft is fixed to the ipsilateral orbital rim with lag screws. In addition, a portion of the greater wing of the sphenoid is also removed up to the frontosphenoid suture and into the superior orbital fissure to allow for subsequent brain expansion into the previously constricted space.

The newly configured orbital unit is then returned to its original position, albeit advanced on the affected side and recessed on the opposite side, and secured with 2 mm thick resorbing plates and screws bridging the tenon extensions to the adjacent temporal skull. The segment of frontal bone is also reshaped through a combination of Tessier bone benders, inner and outer cortex burring, and barrel-staven-like osteotomies to match the new curve of the supraorbital unit and to recreate a smooth and symmetric forehead. The segment can be rotated 180 degrees to use the more properly shaped curve of the posterior edge to match the curve of the supraorbital unit if needed. This segment can be secured with either resorbable plates and screws or even absorbing sutures to avoid any possibility of future growth restriction or transcranial migration of any fixation hardware (**Fig. 3–6**).

To prevent early relapse of the deformity, we believe that firmer rigid fixation via plate-and-screw use should be employed but judiciously and only in those areas where significant postoperative pressure can be expected. Prior to closure, lateral canthopexies are performed by attaching the lateral canthi to the orbital rim with permanent sutures anchored through drill holes in the bone.

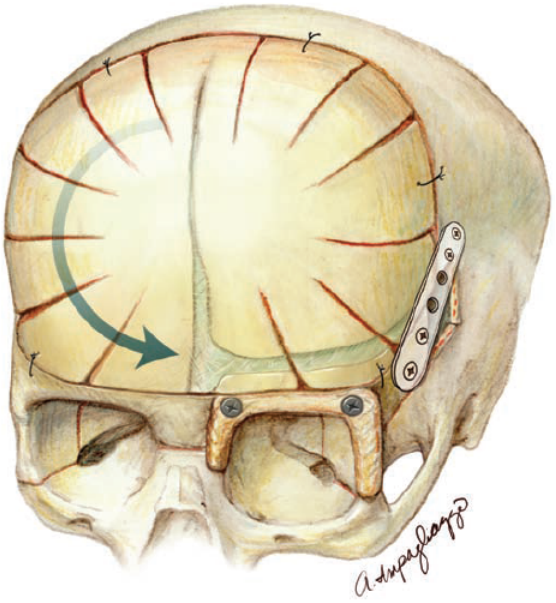

Figure 3–6 Final reconstruction in unilateral coronal synostosis. The orbital rims have been reconstructed using an onlay graft and by advancing the recessed rim and recessing the advanced rim. The orbital rims are attached to the parietal bone using absorbable plates. The segment of frontal bone is also reshaped through a combination of Tessier bone benders, inner and outer cortex burring, and barrel-staven-like osteotomies to match the new curve of the supraorbital unit and to recreate a smooth and symmetric forehead. The segment can be rotated 180 degrees to use the more properly shaped curve of the posterior edge to match the curve of the supraorbital unit if needed. This segment can be secured with either resorbable plates and screws or even absorbing sutures to avoid any possibility of future growth restriction or transcranial migration of any fixation hardware.

Bilateral Coronal Craniosynostosis

The problem with the bilateral deformity is twofold: first, the height of the skull and the recession of both supraorbital rims and lateral orbital rims and second, the brachycephaly that presents a significant problem because it is difficult to correct and failure to correct it will compromise the overall result (**Fig. 3–2**). The issue is whether its correction is warranted in every instance. This can be addressed in two ways: (1) osteotomy and advancement of the single orbital unit consisting of both orbits and the supraorbital bar, as previously described; and (2) expansion of the entire cranial base region that allows for a downward settling of the top portion of the skull along the vertex, thus reducing overall skull height. It is this second goal that necessitates a change in the operative positioning from that used in the unilateral deformity. The patient is placed in a modified prone position, the so-called sphinx position, to correct both the frontal and height abnormalities. Before placing the patient in this position, however, it is important to assess the stability of the cervical spine and the craniovertebral junction by preoperative lateral cervical spine roentgenograms in flexion and extension. Positioning the patient on the operating table is greatly aided by a vacuum-stiffened bean bag to mold the upper body and neck. The face and arms are padded with thick, cushioning foam to prevent pressure sores and compression nerve palsies (**Fig. 3–7**).

Bur holes are placed in the pterion regions bilaterally, and parasagittally in the anterior parietal bone, just posterior to the coronal suture. Similarly, a biparieto-occipital bone graft is outlined with multiple bur holes adjacent to the sagittal and transverse sinuses. Once the bone is elevated both frontally and parieto-occipitally, further dissection epi-

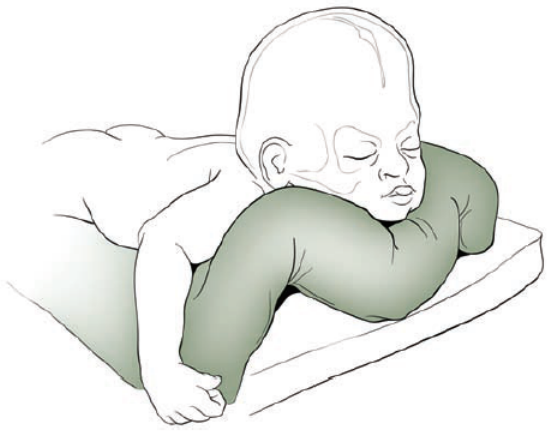

Figure 3–7 Operative positioning for bilateral coronal synostosis. The patient is placed in a modified prone position, the so-called sphinx position, to correct both the frontal and height abnormalities. Before placing the patient in this position, however, it is important to assess the stability of the cervical spine and the craniovertebral junction by preoperative lateral cervical spine roentgenograms in flexion and extension. Positioning the patient on the operating table is greatly aided by a vacuum-stiffened bean bag to mold the upper body and neck.

durally may be performed below the level of the transverse sinus to allow the surgeon to fracture outwardly (in the posterior direction) the occipital bone. The outfractures or barrel-staven-like osteotomies increase the bony capacity by enlarging the perimeter of the skull locally. This allows later brain and dural displacement or settling into this region as the height of the skull becomes reduced secondarily by gravitational forces (**Fig. 3–8**). Barrel-staven-like osteotomies in the occipital bone in the midline and paramedian regions are longer than those placed further laterally, to achieve elongation along the AP axis of the skull, without further widening of the parieto-occiput. The thickened and abnormally elevated superior portion of the greater wing of the sphenoid bone is removed by rongeur in a manner similar to that described for the unilateral deformity. The abnormally convex squamous portion of the temporal bone is left in place but is addressed in a similar manner with barrel-staven-like osteotomies. The bone is reshaped and straightened by radial osteotomies into the center of the convexity and controlled outfracturing of the bone segments with Tessier bone benders. By leaving the temporal bone in place, the overall stability in the lateral portions of the skull is increased. The craniotomized frontal and parieto-occipital bones are reconfigured using similar bone-shaping techniques before being returned to their orthotopic positions. These are then held loosely in their desired positions by fixation with absorbable sutures to the rigidly fixed advanced orbital unit anteriorly, to the radially cut temporal bone laterally, and to the outfractured occipital base bone posteriorly. The vertex of the infant, composed primarily of the fibrous anterior fontanelle, is loosely reattached at the top of the skull with absorbable sutures to the adjoining bones, purposely avoiding any downwardly directly external cinching (**Fig. 3–9**).

We believe that as a consequence of the expansion of the skull-base perimeter, gravitational forces will act to settle the brain and dural into a more inferior overall position, thus having the effect of decreasing the overall height of the skull. This obviates the need for any deliberate downward cinching of the skull vertex as previously described, which did occasionally lead to adverse effects on cerebral circulation.

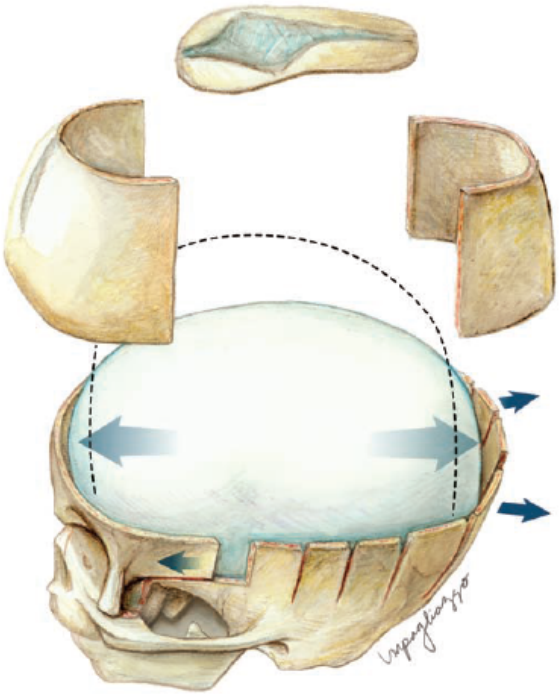

Figure 3–8 Craniotomies and osteotomies for bilateral coronal synostosis. The craniotomized segments include midline biparietal, bifrontal, and bioparieto-occipital flaps. Once the bone is elevated both frontally and parieto-occipitally, further dissection epidurally may be performed below the level of the transverse sinus to allow the surgeon to fracture outwardly (in the posterior direction) the occipital bone. The outfractures or barrel-stavenlike osteotomies increase the bony capacity by enlarging the perimeter of the skull locally. The barrel-staven-like osteotomies in the occipital bone are longer than those placed further laterally, to achieve elongation along the anteroposterior axis of the skull. The abnormally convex squamous portion of the temporal bone is left in place but is addressed in a similar manner with barrel-staven-like osteotomies. The bilateral three-quarter orbital osteotomies are elevated, reshaped, and advanced.

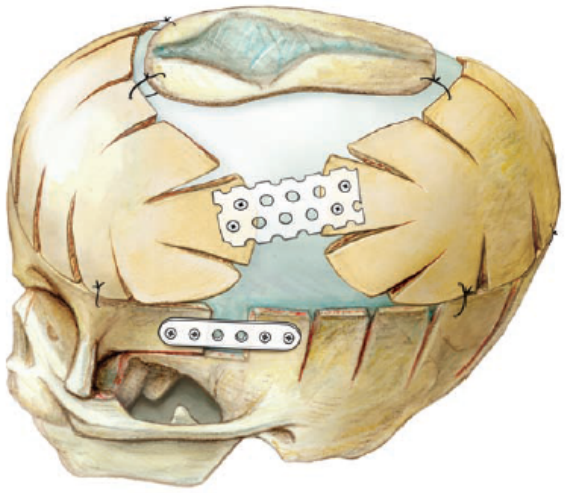

Figure 3–9 Final reconstruction in bilateral coronal synostosis. The craniotomized bifrontal and biparieto-occipital bones are reconfigured using similar bone-shaping techniques before being returned to their orthotopic positions. These are then rigidly fixed to each other and held in their desired positions with absorbable sutures to the rigidly fixed advanced orbital unit anteriorly, to the radially cut temporal bone laterally, and to the outfractured occipital base bone posteriorly. The vertex of the infant, composed primarily of the fibrous anterior fontanelle, is loosely reattached at the top of the skull with absorbable sutures to the adjoining bones, purposely avoiding any downwardly directly external cinching.

Closure

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 never detached from the overlying scalp flap, there is no need to reattach the muscles to their insertions, as they will naturally return to their proper position as the scalp flap is brought posteriorly.

The scalp flap is reapproximated and closed in a two-layer fashion with buried absorbable sutures both in the subgaleal plane and on the skin. A snug but nonconstricting dressing using a sterile surgical towel wrapped as a turban is placed over the entire head, and the head is kept elevated at all times. The tarsorrhaphy sutures and the wire around the endotracheal tube are then removed.

Specialized Instrumentation

A significant advance in instrumentation for pediatric craniofacial surgery occurred with the introduction of resorbable plate-and-screw fixation hardware. These new biomaterials consisting of polymers of polylactic acids are designed to be totally resorbed within 9 to 15 months following implantation. Studies have confirmed that they have tensile strength properties comparable to previously used metallic hardware at the time of their initial use, which allows for the same adaptability when used in three-dimensional calvarial reconstruction. Due to the relatively long retention lifespan of the fixation hardware, the reconfigured and repositioned bone segments can heal with additional support from the plates and screws. Any concern about future translocation of the hardware or restriction of further craniofacial growth is obviated because of predictable resorption of the hardware. High-speed drill and saw systems have made the surgery technically easier.

Postoperative Management Including Possible Complications

Immediate postoperative care is given in the pediatric intensive care unit. Vital signs and central venous pressures are monitored, and 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 analgesia are given intravenously to keep the patient comfortable. The child is positioned on his/her back in bed to keep pressure off the forehead and orbits, and the head is elevated at 30 degrees to prevent excessive postoperative swelling. As a result of the periorbital manipulations, the eyelids commonly 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. 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 the child 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.

Complications

Early postoperative complications arise from intraoperative or perioperative blood loss, which can be compounded by inadequate blood replacement. Tears in the sagittal sinus during the craniotomy can have immediately devastating consequences and must undergo repair quickly. Meticulous attention must be directed when looking for possible tears in the dura mater, which can lead to a persistent cerebrospinal fluid leak. Once recognized, the tear can be easily repaired with a single absorbable suture of 4–0 Nurolon. An air embolism is also a possibility, particularly if the patient is slightly volume-depleted and a sinus is inadvertently entered. 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 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. Injury to the brain itself, as well as the globes, can occur if proper precautionary measures are not taken during the osteotomies, although this risk remains low with an experienced craniofacial surgical team. Pressure sores must be avoided by attention to and protection of the areas where the skull rests during the procedure.

The most frequent late complications include infection and recurrence or relapse of the original defect due to suture restenosis. To prevent infection postoperatively, strict sterile technique must be adhered to and prophylactic antibiotics used up to the time of removal of the drain. If osteomyelitis occurs, there is a high risk of loss of the bone grafts.

Acknowledgment

This chapter is a revision of the chapter, "Treatment of Unilateral or Bilateral Coronal Synostosis" by John A. Persing, M.D., and John A. Jane, Sr., M.D. The chapter appeared in the *Neurosurgical Operative Atlas, Volume # 1,* edited by Setti S. Rengachary and Robert H. Wilkins. The *Neurosurgical Operative Atlas* was published by the American Association of Neurological Surgeons (AANS) from 1991 to 2000.

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