Management of Craniosynostosis

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Learning Objectives: After studying this article, the participant should be able to: 1. Review the etiopathogenesis of craniosynostosis and craniofacial anomalies. 2. Develop a basic understanding of the clinical manifestations and diagnosis of craniofacial anomalies. 3. Describe the surgical principles of managing craniosynostosis and craniofacial anomalies.

Craniosynostosis, or the premature closure of calvarial sutures, results in deformed calvaria at birth. Although the etiology of craniosynostosis is currently unknown, animal experiments and a recent interest in molecular biology point toward interplay between the dura and the underlying brain. This interaction occurs by means of a local alteration in the expression of transforming growth factor, MSX2, fibroblast growth factor receptor, and TWIST. The fused suture restricts growth of the calvaria, thus leading to a characteristic deformation, each associated with a different type of craniosynostosis. Uncorrected craniosynostosis leads to a continuing progression of the deformity, and in some cases, an elevation of intracranial pressure. Clinical examination should include not only an examination of the skull but also a general examination to rule out the craniofacial syndromes that accompany craniosynostosis. Because deformational plagiocephaly, or plagiocephaly without synostosis, occurs secondary to sleep in the supine position during the early perinatal period, the physician should be aware of this abnormality. Treatment for deformational plagiocephaly is conservative when compared with treatment for craniosynostosis, which requires surgery. Appropriate investigations should include genetic screening, radiologic examination with a computerized tomographic scan, and neurodevelopmental analysis. Surgical intervention should be performed during infancy, preferably in the first 6 months of postnatal life, to prevent the further progression of the deformity and possible complications associated with increased intracranial pressure. The principles of surgical intervention are not only to excise the fused suture but also to attempt to normalize the calvarial shape. Long-term follow-up is critical to determine the effect of the surgical outcome. (Plast. Reconstr. Surg. 111: 2032, 2003.)

The calvarial sutures serve two important functions: (1) maintenance of head malleability during passage through the birth canal, and (2) continuance of separation of the calvarial bones during intrauterine and early perinatal life. The sutures serve as growth sites where new bone is deposited in response to the continuing separation of the osteogenic fronts between the opposing bones. Premature closure of any of the calvarial sutures prevents separation of the calvarial bones. Inevitably, this produces a restriction on growth vectors, leading to a morphologic change in calvarial shape. These changes are specific and characteristic for every type of craniosynostosis. However, the sequence of events leading to premature ossification of sutures is unknown. Biomechanical forces and genetically determined local expression of growth factors have been implicated in the etiology of craniosynostosis.

Although Sommering proposed that the calvarial suture was the primary locus of the abnormality, Virchow popularized the concept. Later, Moss conceptualized that the cranial base was the primary locus of the abnormality in children with craniosynostosis and that the altered cranial base transmitted the tensile forces through the dura. This ultimately led to premature closure of the calvarial suture. To study the changes in the cranial base, Babler et al. performed suturectomy in rabbits with and without dural transection. Their findings suggested that suturectomy and transection of the dura did not affect growth more than suturectomy alone. This evidence indicated that the dura played a less important role than believed by Moss. Eaton et al. examined the skulls of North Indian tribes whose members intentionally modified children’s skull shapes by head binding. They concluded

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that a deformity of the cranial vault, either congenital or intentional, alters the structure of the endocranial base and face. This suggested that the endocranial base might not be the primary anomaly in bicoronal and sagittal craniosynostosis.

Over the past decade, the role of the dura in maintaining sutural patency has been extensively studied. Through a series of experiments, Opperman et al. demonstrated that the dura initially plays an inductive role. Later, it assumes a permissive role in maintaining sutural patency through various signaling factors. Levine et al. studied the role of the dura in premature synostosis in rats and determined that the dura generates abnormal signals. These signals could be blocked by a Silastic sheet interposed between the dura and the overlying suture. Most et al. documented an increased level of transforming growth factor-beta and messenger RNA levels locally, indicating an interaction between the dura and overlying sutures at the time of closure. Later, Mooney et al. demonstrated that dura mater transplanted from beneath normally patent sutures to sites of sutural fusion in a naturally occurring craniosynostotic rabbit model successfully kept these sites from re-obliterating. Though the mechanism is not understood, the regional dura mater somehow determines the fate of the overlying suture.

Sutures continue to remain as growth sites where cells undergo proliferation before differentiating into osteoblasts. The calvarial sutures produce new bone at the suture front in response to the expanding neurocranium. As the brain expands, the sutures respond to this stimulus by adding new bone at the suture front. The addition of this new bone helps the suture width remain constant to accommodate the increasing size of the brain. The interaction between the brain and patency of the overlying sutures has been amply proved by clinical evidence showing the premature closure of sutures in the presence of microcephaly, and continuing patency in the presence of hydrocephalus. Secondary craniosynostosis can also develop following placement of a shunt in a child with hydrocephalus (shunt-induced craniosynostosis) because of a drop in intracranial pressure and diminution of neural thrust. The neural thrust and patency of the overlying sutures are closely integrated through the dura. The dura also serves as an intermediary source of signaling, which is mediated by transform-
ittal plagiocephaly with ipsilateral flattening of the occipital region and contralateral bulging. The anterior cranial base deviates toward the fused suture in unicoronal synostosis. Similarly, the posterior cranial base deviates toward the ipsilateral fused lambdoid suture. Because the face grows as a template off the cranial base, uncorrected synostosis leads to asymmetry of the face and occlusal plane.

Closure of multiple sutures can limit the expansion of a neonate’s calvaria and underlying cortex. The continuing expansile forces of the cortex against the fixed volume of the calvaria leads to an increase in intracranial pressure. Blindness and possible mental retardation are two problems associated with intracranial pressure. Diagnosing increased intracranial pressure in children with craniosynostosis has been difficult. Gault et al. discovered the trend by measuring intracranial pressure overnight by using an epidural monitor in children with craniosynostosis. They regarded pressure readings above 15 mmHg as confirmatory for increased intracranial pressure, below 10 mmHg as normal, and between 10 and 15 mmHg as borderline. Using this system, they found that 47 percent of children with multiple-suture craniosynostosis, and 14 percent of those with single-suture craniosynostosis, demonstrated increased intracranial pressure. Thompson et al. using a subdural catheter to measure intracranial pressure, demonstrated that in children with craniosynostosis, 17 percent had increased intracranial pressure, 36 percent had borderline pressure, and 45 percent had a normal reading. Renier also demonstrated that intracranial pressure decreased following surgery. Consequently, there was a direct correlation between the extent of increased intracranial pressure and mental function in a subgroup of 55 children.

Because the precise range of intracranial pressure levels in “normal” children is unknown, the true impact of the studies is still unclear. Regardless, Cohen and Persing offer a good review of techniques and recommendations for measuring intracranial pressure.

**Clinical Examination and Diagnosis**

Although premature fusion of the sutures is a prenatal event, the diagnosis of craniosynostosis is not imminent in the neonatal period. The passage of the head through the birth canal temporarily deforms the head and makes it difficult to ascertain whether it will be shaped normally or abnormally. Either the parent or the pediatrician will notice a persistent abnormal shape in early infancy, and the child is referred to either a craniofacial surgeon or a neurosurgeon.

Because the characteristic calvarial deformation is secondary to sutural fusion, clinical diagnosis is made more easily in the later perinatal months. Clinical history should include the primary birth events (length of term, birth weight, and complications during and after birth). A detailed history of the perinatal sleeping position is also critical in differentiating craniosynostosis from plagiocephaly without synostosis (vide infra). Syndromic craniosynostosis involves multiple systems (cardiac, genitourinary, and musculoskeletal), and a family history of abnormal head shape is critical in its diagnosis.

Clinical examination should include an examination not only of the head and neck to rule out torticollis, but it also should include the digits, toes, and spine. Measuring the head circumference aids in ruling out microcephaly and macrocephaly with associated hydrocephalus. The calvarial shape is characteristic for each type of sutural synostosis. An increase in anteroposterior length, or scaphocephaly, is associated with sagittal synostosis (Fig. 1). Transverse narrowing of the anterior calvaria, or trigonocephaly, is associated with metopic synostosis (Fig. 2). The distance between the medial canthi of the eyes (medial intermedial canthus distance) is decreased, whereas the distance between the centroids (center of each ocular globe) is normal. Metopic synostosis is therefore not associated with true orbital hypotelorism, but the orbital rims may appear hypoplastic. Bicoronal synostosis results in shortening of the skull in the anteroposterior direction and widening at the bitemporal region. The fronto-orbital rim is recessed bilaterally, and depending on the severity of the case and age of the child, this may lead to exorbitism with its associated complication of exposure keratitis (Fig. 3). Unilateral asymmetry, or plagiocephaly, can result from coronal or lambdoid synostosis. Unilateral coronal synostosis results in a recessed forehead and fronto-orbital rim and contralateral bossing of the forehead and fronto-orbital rim. The eyebrow on the ipsilateral side is raised, and the palpebral fissure is increased in its vertical height. The ocular globe is sometimes raised, along with the pupil, resulting in vertical dystopia (Fig. 4). An older
child with uncorrected unicoronal synostosis demonstrates asymmetry of the facial skeleton. This is associated with a reduction in height of the ipsilateral maxilla and mandible, resulting in an oblique occlusal cant. Lambdoid synostosis is rare and must be differentiated from deformational plagiocephaly, or plagiocephaly without synostosis.

**PLAGIOCEPHALY WITHOUT SYNOSTOSIS**
(DEFORMATIONAL PLAGIOCEPHALY)

Plagiocephaly without synostosis, or deformational plagiocephaly, is a very common cause for an asymmetrical head associated with an ipsilateral occipital flattening. The increase in this abnormality has been associated with the “Back to Sleep” campaign to reduce the incidence of sudden infant death syndrome (SIDS). If left in the same position for a prolonged duration of time, the skull of a newborn child can easily become deformed because it is so soft. In addition, infants have limited ability to move their head in the first few months of life because of their weak neck muscles. Torti-
collis, the tightening of neck muscles, may further complicate the situation by preventing the infant or parent from repositioning the head.

The deforming force flattens the occipital skull, and like a deflated ball, it preferentially tends to lie in that position. The continuing force flattens the occipital region and pushes the forehead and ear forward. In extreme cases, the cheekbones, jaw joints, and mandible may be involved. This leads to a typical parallelogram-shaped head, which differs from unicoronal or unilambdoid synostosis (Table I). This abnormality is known as deformational plagiocephaly, or plagiocephaly without synostosis. Other factors, such as prolonged labor, abnormal fetal position, and multiple births have also been incriminated in the origin of plagiocephaly without synostosis. Intentional change in positioning or the use of positioning devices, however, can improve head shape during this period. Keeping the child in the prone position while awake also contributes to improving head shape. The use of a customized molding helmet for 23 hours/day until the child is 1 year of age is also very successful in treating this condition when conservative measures have failed (Fig. 5). An early referral, preferably before the age of 6 months, is critical because molding helmet therapy is not very effective after 1 year of age. It is also extremely important to differentiate plagiocephaly without synostosis from craniosynostosis (Table I), because treatment of the former includes using either positional therapy or molding helmet therapy, whereas the latter requires surgical intervention.

**Investigations**

The clinical diagnosis of craniosynostosis or plagiocephaly without synostosis can be substantiated with radiologic investigations.

**Radiography of the Skull**

Skull radiography is a preliminary examination to visualize whether or not the sutures are patent. Although the test is simple, accuracy is

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**TABLE I**

Characteristics of Craniofacial Anomalies*

<table>
<thead>
<tr>
<th></th>
<th>Right Deformational Plagiocephaly</th>
<th>Right Unicoronal Synostosis</th>
<th>Right Unilambdoid Synostosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Frontal</td>
<td>Ipsilateral R bossing</td>
<td>Ipsilateral R flattening</td>
<td>No change</td>
</tr>
<tr>
<td></td>
<td>Contralateral L flattening</td>
<td>Contralateral L bossing</td>
<td></td>
</tr>
<tr>
<td>Occipital</td>
<td>Ipsilateral R flattening</td>
<td>No change</td>
<td>Ipsilateral R flattening</td>
</tr>
<tr>
<td></td>
<td>Contralateral L bossing</td>
<td></td>
<td>Contralateral L bossing</td>
</tr>
<tr>
<td>Palpebral fissure</td>
<td>Ipsilateral R narrowing or no change</td>
<td>Ipsilateral R widening</td>
<td></td>
</tr>
<tr>
<td>Ears</td>
<td>Ipsilateral R ventral translocation</td>
<td>Ipsilateral R ventral translocation</td>
<td>Ipsilateral R dorsal translocation</td>
</tr>
<tr>
<td>Mastoid process</td>
<td>No change</td>
<td>No change</td>
<td>No change</td>
</tr>
<tr>
<td>Face</td>
<td>Ipsilateral R ventral translocation</td>
<td>Ipsilateral R dorsal translocation</td>
<td>No change</td>
</tr>
<tr>
<td>Nose</td>
<td>Deviated to contralateral L side</td>
<td>Deviated to ipsilateral R side</td>
<td>No change</td>
</tr>
<tr>
<td>Chin</td>
<td>Deviated to contralateral L side</td>
<td>Deviated to ipsilateral R side</td>
<td>No change</td>
</tr>
</tbody>
</table>

* R, right; L, left.
not perfect and differentiating between a lambdoidal synostosis and plagiocephaly without synostosis is sometimes difficult. Special views are needed to better visualize and appreciate all sutures.

**Ultrasonography**

Ultrasonography, a noninvasive, nonradiologic procedure, has recently been studied to determine its utility in diagnosing a patent suture. Ultrasound is more accurate than a radiographic examination, but it requires a sonographer who is willing to specialize in interpreting the results.

**Computed Tomographic Scan**

A computed tomographic scan is the definitive standard for determining whether a suture is patent or fused. Two-dimensional views allow for the direct visualization of each suture, but grasping the extent of the associated deformity remains challenging. This ultimately makes preoperative planning very difficult and affects outcome reliability. In contrast, three-dimensional computed tomographic scans allow complete visualization of the skull and clearly document the extent of the deformity (Figs. 6 and 7). Scanning specific views (i.e., frontal, posterior [occipital], right and left profile, superior, and inferior ectocranial and endocranial) can be used as a protocol to allow comparison between preoperative, perioperative, and postoperative computed tomographic scans. A radiographic scale should also be placed beside the images to make approximate measurements of the various bony landmarks. This aids in preoperative planning and assists in performing a meaningful outcome analysis. Sophisticated software has also played a role in determining the best option by allowing manipulation of the osteotomies on a graphic workstation.36

**Sagittal Synostosis**

When the sagittal suture is fused, transverse growth of the skull becomes restricted (Fig. 8), resulting in a reduction of the transverse diameter at the parietal eminencies (biparietal diameter) and an increase in the anteroposterior length of the skull. This explains the frontal bossing and occipital protuberance associated with sagittal synostosis. The cephalic index (maximum transverse width/maximum anteroposterior length) is also reduced. Nomograms for cephalic indices are available and are one way to evaluate the severity of the scaphocephaly.37 Unfortunately, the cephalic index measures only two dimensions of the skull and does not allow assessment of either frontal bossing or occipital protuberance. Yet, it is the only readily available method for quantitative assessment.

**Metopic Synostosis**

The fused metopic suture restricts the transverse growth of the frontal bones, which

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**Fig. 6.** Three-dimensional computed tomographic scan of a child with metopic craniosynostosis. Note the ridging at the site of the fused metopic suture.

**Fig. 7.** Three-dimensional computed tomographic scan of a child with right unicoronal synostosis demonstrating asymmetry of the orbits. The right orbit is narrower and taller as compared with the contralateral side, and the facial axis is mildly deviated.
results in narrowing of the anterior cranial fossa and trigonocephaly (Fig. 6). The restriction in the transverse direction also results in reduction in the intermedial canthal distance (distance between medial canthi) and interdacryon distance (distance between lacrimal crests), but true hypotelorism (distance between the centroids of the globe) does not exist. The orbital rims may also be hypoplastic, depending on the severity of the metopic craniosynostosis. The metopic suture is the first suture to close physiologically. Previous studies documented this closure to be in the first and second year of postnatal life. However, computed tomographic scans taken in the neonatal period show that the metopic suture closes physiologically at a much earlier age (between 3 and 9 months postnatally). A physiologic closure, however, is not associated with the secondary deformity of narrowing of the anterior cranial fossa and hypoplastic orbital rims. The medial intercanthal distance should also be measured preoperatively and compared with a nomogram.

Bicoronal Synostosis

The bicoronal suture is fused, thus restricting the anteroposterior growth of the anterior cranial fossa. This results in an increased bi-temporal diameter and a decreased anteroposterior dimension of the anterior cranial fossa. In addition, the fronto-orbital bar is recessed bilaterally. The supraorbitale (the most prominent point of the fronto-orbital rim) is normally 2 mm ventral to the plane of the cornea. However, in a child with bicoronal synostosis, the fronto-orbital bar is recessed and the supraorbitale is more dorsal (posterior) to the corneal plane. In most circumstances, this leads to exorbitism with its associated complications. Therefore, the objective of surgery is to place the fronto-orbital bar 2 mm ventral to the corneal plane.

 unicoronal Synostosis

Unicoronal synostosis is characterized by a fused coronal suture. The ipsilateral fronto-orbital bar is recessed, whereas the contralateral fronto-orbital bar and frontal bone are more ventral. These conditions are the result of secondary deforming changes that occur because of the closed suture. When anteroposterior growth is restricted across the closed coronal suture, compensatory changes take place along the contralateral suture. This critical event must be understood when correcting unicoronal synostosis, because correcting the contralateral bones will require surgery. The fused suture also restricts the growth of the sphenoid bone, resulting in a reduction of the anteroposterior dimension of the anterior cranial fossa. Consequently, the anterior endocranial base deviates toward the ipsilateral side. Similarly, the ectocranium also deviates to the ipsilateral side. If uncorrected, deviation of the face and an occlusal cant may result. The posterior cranial base, however, remains unaffected. The roof of the orbit, formed by the greater wing of the sphenoid, is raised such that the shape of the ipsilateral orbit becomes more vertical (Fig. 7) compared with the more horizontal orbit on the contralateral side.

Lambdoid Synostosis

Lambdoid synostosis is characterized by an obliterated ipsilateral lambdoid suture that reduces the anteroposterior dimension of the posterior cranial fossa. The protruding contralateral suture and bulging ipsilateral mas-
toid process are distinguishing characteristics of lambdoid synostosis. The posterior cranial base will deviate to the ipsilateral side, whereas the anterior base is unaffected. The greatest difficulty lies in differentiating between deformational plagiocephaly (plagiocephaly without synostosis) and lambdoid synostosis. Synostosis simply restricts growth on the ipsilateral side, but deformational plagiocephaly deforms by exerting force in a ventral direction. Thus, lambdoid synostosis involves the petrous portion of the temporal bone being pulled toward the closed lambdoid suture. Correspondingly, the external auditory canal is also pulled toward the fused suture. In contrast, both the petrous portion of the temporal bone and the external auditory canal are pushed anteriorly in deformational plagiocephaly. Ipsilateral frontal bossing occurs regularly in deformational plagiocephaly but almost never in lambdoid synostosis. Thus, diagnosing deformational plagiocephaly, unlike in lambdoid synostosis, is made by assessing secondary changes instead of the suture itself. This eliminates diagnosis of partial suture closures and lambdoid suture sclerosis or thickening.

MAGNETIC RESONANCE IMAGING

A magnetic resonance imaging scan helps to delineate the pattern of cortical gyri and sulci underneath the fused suture. Whether the abnormal pattern is secondary to the pressure from the overlying fused suture or is a primary event leading to a fused suture is often debated. A simple follow-up study comparing the preoperative magnetic resonance imaging scans with the 1-year postoperative scans would offer some insight. However, it has been very difficult to objectively measure the extent of gyri and sulci flattening, despite having used sophisticated software from the National Institutes of Health. Lack of a centralized database of magnetic resonance imaging scans from normal children of a specific age further complicates the study. The major limitation of the magnetic resonance imaging scan is that it only allows for anatomical delineation of the cortex and provides no information about cortical function.

NEURODEVELOPMENTAL ANALYSIS

The purpose of neurodevelopmental testing is to obtain information about the cortical functioning before and after cranial vault remodeling. The Bayley Scales of Infant Development–II are often used to procure this information. The greatest difficulty with neurodevelopmental testing is the lack of accuracy in measuring cortical function in an infant 3 to 6 months of age. Therefore, a longitudinal assessment is essential in a thorough analysis. These tests compare the child’s mental and psychomotor scales with a nomogram, thus helping to quantify whether the child is developmentally delayed and if surgery will help reduce the severity of that delay. Children with syndromic craniosynostosis, characterized by multiple system involvement, are often developmentally delayed. This delay can be severe and the child could be classified as mentally retarded. Although children with single-suture craniosynostosis often do not seem to have significant developmental delays, a more detailed assessment with sophisticated analyses has shown otherwise. Children with metopic and sagittal craniosynostosis also have minor delays in learning and speech, a problem not previously recognized in this population. A study showed that children with developmental delays at 12 months also exhibited cognitive and motor deficiencies during their preschool years. Understandably, a 3-month-old infant cannot be tested for language delays, but the Bayley Scales of Infant Development–II is a comprehensive test that includes aspects of the child’s developmental skills. For these reasons, this test is a reliable tool for assessing infants.

NEED FOR SURGERY

The aim of surgical intervention is to excise the prematurely fused suture and correct the associated deformities of the calvaria. Uncorrected synostosis quite frequently is associated with an increase in intracranial pressure. This has been documented in the animal model and in humans. If the synostosis goes uncorrected, the deformity progresses to involve the facial skeleton, which is associated with asymmetry of the face and malocclusion. Asymmetry of the orbit leads to ocular dystopia and consequent strabismus. Therefore, the surgical goal is to increase the intracranial volume, especially under the fused suture, and prevent any long-term complications. Normalization of the calvarial shape successfully achieves this goal.

Surgery is best performed in early infancy because:
• Most brain growth occurs in the first year of life. The deforming vectors of the continually growing brain result in progression of the deformity with increasing age.
• Few studies have demonstrated an increase in intracranial pressure in nonsyndromic single-suture craniosynostosis.²⁹,⁴⁹
• Osseous defects following surgery undergo re-osseification more completely before 1 year of age as compared with later.
• A delay in surgery beyond the first 9 to 12 months of life leads to progressive deformity of the cranial base, resulting in abnormal facial growth and asymmetry of the maxilla and mandible.⁵⁰
• The calvaria in a child 3 to 9 months of age is still malleable and, therefore, quite easy to shape.

For these reasons, there is now an increasing consensus among craniofacial surgeons and neurosurgeons that surgery should be performed in early infancy.

**Sagittal Craniosynostosis**

Sagittal craniosynostosis is characterized by an increased growth in the anteroposterior direction and a reduction in the transverse direction (Fig. 8). This results in frontal bossing and occipital protrusion. Consequently, the objective of surgery is to correct the secondary deformational changes in addition to excising the fused sagittal suture. Traditionally, the aim has been to excise only the fused suture in the hope that the secondary changes would automatically correct themselves as the brain grows in the first year of postnatal life. This simple objective was achieved by removing the fused suture with a procedure called a strip craniectomy. Initial strip craniectomies were associated with refusion, and various agents were used to keep the sutures patent.⁵¹ Numerous modifications have been made to strip craniectomies; currently, as many as 16 types of procedures have been collated in an attempt to elucidate the rationale of performing surgery for sagittal craniosynostosis.⁵² Over the past decade, craniofacial surgeons have become more aggressive in trying to correct the suture and the associated deformities of frontal bossing and occipital protrusion, or bathrocephaly. The procedure of cranial vault remodeling involves excision of the frontal, parietal, and occipital bones. These are trimmed, reshaped, and then relocated and affixed with absorbable plates. The primary surgical objective is to release the synostotic constraint and promote normal calvarial growth. Reducing the anteroposterior length and increasing the transverse width also improves calvarial contour.

A retrospective quantitative analysis (using the cephalic index) of 40 infants who underwent surgery for sagittal craniosynostosis was conducted to determine whether any difference in outcome could be associated with either the child’s age at surgery or the extent of the operation. Despite the extended strip craniectomies performed before 4 months of age, the study concluded that cranial vault remodeling was more effective in normalizing the cranial shape (Fig. 9).⁵³

Recently, craniofacial surgeons and neurosurgeons have been using an endoscope to perform extended strip craniectomies, followed by molding helmet therapy for 6 to 8 months postoperatively.⁵⁴ Although this technique reduces intraoperative time, length of hospital stay, and blood loss, it does not allow the surgeon to intraoperatively alter the calvarial shape or cephalic index. After all, the procedure remains a strip craniectomy despite its being performed with an endoscope. Those authors recently reported their results using the cephalic index for nine patients who had undergone endoscopic strip craniectomy. Two patients had cephalic indices that were below normal, and seven had values within the normal range. Unfortunately, the mean follow-up was only 10 months after surgery. Also, no comparison was made between the improvements of cephalic index with either technique. This ultimately makes it difficult for the reader to determine whether the combined endoscopic strip craniectomy and molding helmet therapy is superior to cranial vault remodeling.⁵⁵

**Unicoronal Synostosis**

**Preoperative Planning**

**Ventral advancement of the supraorbital bar.** In unicoronal synostosis, the ipsilateral supraorbital rim is recessed and dorsal to the plane of the cornea.⁵⁶ A two-dimensional long axis view along the apex of the orbit and the center of the cornea demonstrates the extent of the recession. Nomograms have revealed that the supraorbital rim is approximately 2 to 3 mm ventral to the vertical plane of the cornea. The movement of the supraorbital rim is such that
it is 3 mm ventral to the vertical plane of the cornea. The extent of this movement varies between 7 to 15 mm, depending on the severity of the unicoronal synostosis. However, additional approaches to advancing the supraorbital bar have been offered by Marchac and Renier\textsuperscript{57} and Posnick.\textsuperscript{58}

Correcting orbital asymmetry. The ipsilateral roof of the orbit is higher than the contralateral orbit. The contralateral orbit, in contrast, is also wider than normal. The extent of the correction is determined by using nomograms. Excising bone at the frontozygomatic and nasomaxillary sutures reduces the height of the ipsilateral orbit. In addition, inserting a cranial bone graft into the supraorbital rim increases the width of the ipsilateral orbit. Similarly, the height of the contralateral orbit is increased by elevating the supraorbital bar, and its width is reduced by excising a bone segment from the supraorbital bar (Figs. 10 through 12).
BICORONAL SYNOSTOSIS

In treating bicoronal synostosis, the surgical objective is to increase the anteroposterior dimension of the calvaria by ventrally advancing the frontoparietal bones and the fronto-orbital bar. Advancement of the latter would increase the projection of the supraorbital rim 2 mm beyond the corneal plane. In addition, other techniques for treating bicoronal synostosis have been proposed by Marchac and Renier.57

For treatment planning, appropriate software allowing delineation of the bone segments and their advancements can be used. Most of these software programs are cumbersome and require prolonged duration of manipulation before the appropriate position is obtained. Despite the most accurate preoperative planning, the current software fails to provide intraoperative confirmation that the desired position is being reached. Intraoperative confirmation of the desired position will probably be available in the future because of the recent surge in frameless stereotactic neurosurgery.

A two-dimensional sagittal image is used for preoperative calculations. This image traverses through the centroid (center of the ocular globe) and the center of the orbital apex. The position of the supraorbitale is confirmed on this view. The corneal plane is developed by passing a ray connecting the infraorbitale to the most prominent point of the cornea. The ray is extended so that it passes 2 mm ventral to the cornea. This provides an estimate for ventral advancement of the fronto-orbital bar.

FIG. 10. Graphical representation of the method of correction of the orbital asymmetry. (Above) The right orbit is wider and shorter as compared with the left orbit, which is narrower and taller. (Center) The right orbit is decreased in width by reducing the width of the frontal orbital bar on the right. The left frontal orbital bar is widened by 5 mm. The height of the left orbit is reduced by 3 mm. (Below) The corrected position of the right and the left orbit, held in position by Lactosorb plates and screws. The 5-mm-wide bone graft on the left orbit is also fixed by using Lactosorb plates and screws.

METOPIC SYNOSTOSIS

In treating metopic synostosis, the surgical goal is to increase the volume of the anterior cranial fossa by performing a ventral advancement of the fronto-orbital bar. The reduced interdacyon distance must be normalized by increasing the transverse dimension between the pterions to correct the abnormality. This is done by placing a calvarial bone graft between the two halves to correct the reduced interdacyon distance. The width of the bone graft should be equal to the extent of correction needed to normalize the interdacyon distance. If the bar is thick at the midline, a tenon mortise bone joint is used to inherently increase the stability of the bone graft. A T-shaped absorbable plate is then used to fix the graft at the junction of the fronto-orbital bar and the nasion.59 Cohen et al.60 provided alternative techniques in treating metopic synostosis.

FIG. 11. Fronto-orbital bar demonstrating the correction described in Figure 10. Note the presence of an absorbable plate at the lateral aspect of the frontal orbital bar, which will allow rigid fixation and advancement of the left frontal orbital bar at the pterion.
After extubation, the child is transferred to the pediatric intensive care unit for 24 to 48 hours so that hemodynamic stability and level of consciousness can be monitored. Parents are informed about the considerable amount of swelling that occurs around the scalp and periorbital areas, and they are reassured that the swelling will subside after a few days. It is not unusual for the child to develop pyrexia of 38°C for the first 72 hours. Continuing pyrexia, persistent swelling, and cellulitis should be further investigated. Another important point is that the need for blood transfusions is based on the hemodynamic stability of the child rather than on a specific hematocrit level.

The senior author performs a perioperative three-dimensional computed tomographic scan on the fifth postoperative day. This allows an accurate method of documenting the osteotomy sites. In addition, the surgeon can confirm postoperatively whether the appropriate symmetry and ventral advancement were achieved intraoperatively. The child is discharged after completion of the computed tomographic scans.

Further clinical follow-up is done at 3 weeks, 6 weeks, 3 months, 6 months, and 1 year postoperatively. A three-dimensional computed tomographic scan and magnetic resonance imaging scan are also performed 1 year after surgery. These tests are valuable for documenting whether the ventral movement and symmetry of the orbits has been maintained. Thereafter, annual visits are required until age 6 years. Longitudinal follow-up by a craniofacial team is also recommended to assess the child’s neuropsychologic development and craniofacial growth.

**Complications**

To reduce mortality and morbidity, this surgery should be performed in a children’s hospital equipped with a pediatric anesthesiologist and pediatric intensive care facilities. Recent series have demonstrated only isolated cases of mortality, which often involved insufficient blood replacement during the intraoperative period. Therefore, blood loss should be minimized and replaced quickly when necessary.

The incidence of wound infection and dehiscence is low. Cranial bone graft infection can be minimized significantly by administering a 48-hour prophylactic antibiotic regimen. The current authors frequently use nafcillin and gentamicin. Continuing cerebrospinal fluid leakage is also extremely rare if adequate precautions are taken to ensure proper closure of dural tears intraoperatively.

Long-term follow-up may reveal minor asymmetries of the supraorbital bar and forehead; these can be addressed by elevating the scalp flap and applying a hydroxyapatite paste to the surface of the calvaria. Ossification within the orbit takes place quite rapidly and is complete within 1 year after surgery. Persistent areas of incomplete ossification, those larger than 2 cm in diameter, are also similarly treated with hydroxyapatite paste. Major asymmetries may require a revision osteotomy and re-advancement of the supraorbital bar. This, however, is rare under current surgical standards.
ABSORBABLE PLATES

Similar to the impact of titanium plating devices for maxillofacial fixation, absorbable plating systems have revolutionized pediatric craniofacial surgery by ensuring rigid fixation and a better outcome. The initial enthusiasm of using titanium-plating devices on a child’s growing calvaria was quickly followed by complications. Resorption of inner table and deposition of new bone on the outer table led to a gradual movement of the titanium plates and screws over the calvaria. This movement then progressed onto the dura, sometimes coming in direct contact with the brain.\textsuperscript{3,67–70}

At the same time, absorbable plating technology is becoming more dependable.\textsuperscript{71,72} Absorbable plates are manufactured from a combination of polyglycolic and polylactic acids. The addition of polylactic acid increases the resorption period of the plate. The resorption time, therefore, varies from 12 to 36 months, and resorption typically occurs by hydrolysis, which does not incite an inflammatory reaction. Studies have documented that the rigidity offered by absorbable plating devices is equivalent to that of titanium devices.\textsuperscript{73,74} The only drawback to this system is that absorbable screws must be tapped. This process adds time to the surgery, but with increasing practice, this is negligible.

USE OF HYDROXYAPATITE BONE SUBSTITUTE

Although hydroxyapatite has been used quite extensively by orthopedic surgeons for joint implants and by maxillofacial surgeons for chin augmentation, the compound has been available only as blocks. Recently, the material has been produced as a powder that is

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**Fig. 13.** Three-dimensional computed tomographic scans demonstrating (above, left) the osseous defect left in the calvaria after compound comminuted fracture; (above, right) abnormality in the contour and osseous defect over the left temporoparietal skull; (below, left) postapplication of hydroxyapatite into the osseous defect, which is completely obliterated; and (below, right) postoperative superior view demonstrating perfect contour between the right and left temporoparietal regions following application of hydroxyapatite.
mixed with sodium phosphate. The resultant paste is used to fill osseous defects and to augment frontal or temporal bones. Although the first products did not solidify for 24 hours, more recent products solidify in 10 minutes. These newer products give more reliable results even in the presence of moisture and blood. Craniofacial surgeons currently use hydroxyapatite quite extensively following cranial vault remodeling. For example, hydroxyapatite is used for osseous defects that have failed to ossify or to augment the temporal hollowing and minor asymmetry that may occur after cranial vault remodeling (Fig. 13). 75,76

CRANIOFACIAL SYNDROMES

Apert syndrome, a rare craniofacial disorder, occurs sporadically in most cases but can be transmitted by an autosomal dominant trait in some families. The syndrome is also known as acrocephalosyndactyly because of the cranial and extremity involvement. It is characterized by a bicoronal synostosis, turricephaly, severe exorbitism, midface hypoplasia, anterior open bite, and bilateral symmetrical complex syndactyly of the digits and toes (Figs. 14 and 15). In addition to developmental delays and hyperactivity, these children quite often suffer from obstructive sleep apnea because of airway restriction caused by the midface retrusion.

Crouzon syndrome, or craniofacial dysostosis, is a rare disorder with a prevalence of one in 25,000 in the general population. The syndrome, transmitted as an autosomal dominant trait, is characterized by exorbitism, midface retrusion, and brachycephaly because of the bicoronal synostosis. The midface is characteristicly box-shaped and may be associated with hypertelorism. The mandible has normal growth but may be secondarily deformed by maxillary retrusion and malposition (Fig. 16).

In 1964, Pfeifer described a syndrome consisting of craniosynostosis, broad thumbs, broad great toes, and, occasionally, partial soft-tissue syndactyly of the hand. The syndrome is inherited as an autosomal dominant trait with complete penetrance.

Carpenter syndrome is characterized by craniosynostosis in association with preaxial polysyndactyly of the feet. Short fingers with clinodactyly and variable soft-tissue syndactyly may also be present. Carpenter syndrome is transmitted as an autosomal recessive trait.

Unlike Carpenter syndrome, Saethre-Chotzen syndrome is inherited in an autosomal dominant manner. Characteristics of Saethre-Chotzen include craniosynostosis, low-set hairline, ptosis of the upper eyelids, facial asymmetry, brachydactyly, partial cutaneous syndactyly, and other skeletal anomalies.

The management of craniofacial syndromes can be summarized as follows:

- Step I: Correction of craniosynostosis between the ages of 3 and 6 months.
• Step II: Correction of syndactyly between the ages of 1 and 2 years.
• Step III: Correction of midface retrusion with distraction techniques by the age of 4 to 5 years. Timing and progress of the distraction may vary, depending on the severity of obstructive sleep apnea, malocclusion, and psychological disturbance.
• Step IV: Correction of hypertelorism and turricephaly, if present, at age 4 to 6 years. This may be done in conjunction with, or separately from, step III.
• Step V: Await full maturity and perform Le Fort I or Le Fort III procedure in conjunction with mandibular osteotomy to normalize appearance and correct malocclusion.

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REFERENCES


Management of Craniosynostosis
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1. WHICH OF THE FOLLOWING STATEMENTS REGARDING DEFORMATIONAL PLAGIOCEPHALY IS CORRECT?
   A) It is often due to an infant sleeping in the prone position in the early perinatal period.
   B) It is associated with occipital flattening, ipsilateral frontal bossing, and ventral translocation of the ipsilateral ear.
   C) It is associated with occipital flattening, contralateral frontal bossing, and ventral translocation of the contralateral ear.
   D) It can be treated successfully with molding helmet therapy as late as 24 months of age.
   E) It is familial in most cases.

2. WHICH OF THE FOLLOWING FINDINGS IS MOST CONSISTENT WITH CROUZON SYNDROME?
   A) Exorbitism
   B) Unicoronal synostosis
   C) Complex syndactyly
   D) Hypotelorism
   E) Mandibular hypoplasia

3. THE PRIMARY OBJECTIVE IN SURGICAL TREATMENT OF SAGITTAL CRANIOSYNOSTOSIS IS TO:
   A) Reduce the cephalic index
   B) Alter the shape of the calvaria to attain a normocephalic shape
   C) Decrease the transverse dimensions of the calvaria
   D) Resect the abnormal suture line
   E) Increase cranial volume

4. SURGICAL TREATMENT FOR CRANIOSYNOSTOSIS IS BEST PERFORMED AT WHICH AGE?
   A) 1 to 6 weeks
   B) 1 to 6 months
   C) 1 to 2 years
   D) 2 to 4 years
   E) Later than 4 years

5. RADIOGRAPHIC FINDINGS ON COMPUTED TOMOGRAPHY IN A CHILD WITH UNICORONAL CRANIOSYNOSTOSIS ARE BEST DESCRIBED BY WHICH OF THE FOLLOWING?
   A) Deviation of the cranial base to the contralateral side
   B) Ipsilateral roof of the orbit lower than the contralateral side
   C) Increased height of the orbit on the contralateral side
   D) Deviation of the cranial base toward the ipsilateral side
   E) Frontal bossing on the ipsilateral side

To complete the examination for CME credit, turn to page 2132 for instructions and the response form.