CHAPTER

Nonsyndromic Craniosynostosis

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INDICATIONS

Craniosynostosis refers to the premature fusion of one of the six major sutures of the craniofacial vault. Functionally, craniosynostosis may be defined as the premature conversion of a dynamic region of growth and resorption between two adjacent bones of the cranial vault into a static region of bony union. The final result is the formation of a single bony plate from two smaller segments. The term craniosynostosis is used interchangeably but actually describes the consequences of craniosynostosis. The first description of the morphologic changes created by premature fusion was recorded by Hippocrates. Galen also described a patient with craniosynostosis and coined the term oxycephaly. Sommerring first recognized that skull growth occurred at the sutures and fusion of these “growth areas” would create a deformity. Subsequently, Virchow initiated the use of the word craniosynostosis to describe the premature suture fusion and further established what is known as Virchow’s law for compensatory cranial vault growth after suture fusion.

CLASSIFICATION

Premature suture fusion may be characterized as described by Cohen. Nonsyndromic, or isolated, craniosynostosis predominates and is defined as suture fusion that creates functional impairments related to local effects of the fusion, that is, intracranial hypertension or ophthalmoplegia. Occurrences are usually sporadic, but rare familial tendencies have been reported. Furthermore, craniosynostosis in two members of the immediate family will increase the chance the next child will develop a premature suture fusion. Craniosynostosis associated with craniofacial syndromes (e.g., Apert’s syndrome, Crouzon’s disease, Pfeiffer’s syndrome) may be autosomal dominant or autosomal recessive and have second-
ary anomalies not directly associated with the suture fusion. These may include the cardiovascular, genitourinary, or vertebral organs. Craniosynostosis may also be classified as simple (one suture) or complex (two or more sutures) and primary or secondary as a reflection of the underlying cause.

The most commonly affected nonsyndromic suture fusion usually involves the sagittal suture. It is characterized by an increased anteroposterior position and decreased biparietal width (brachiocephaly). Presentation may be variable, but generally anterior sagittal fusion will present with significant frontal bossing; posterior sagittal fusion is characterized by an occipital bulge.

Coronal sutures may have unilateral (anterior plagiocephaly) or bilateral (brachiocephaly) involvement. The primary dysmorphology involves the forehead and the supraorbital region, which includes the zygomatic process of the frontal bone (lateral orbital rims) and the temporal area. In unilateral coronal synostosis, the forehead is flattened and there is retrusion of the ipsilateral superior orbital rim. If both coronal sutures are involved, the lateral dimensions of the skull are widened and the superior orbital rim is displaced bilaterally. There may also be an associated increase in the height of the forehead (turricephaly).

Trigonocephaly from metopic suture fusion is probably the most obvious deformity of the craniosynostoses. The occurrence is uncommon (7.9% to 10.0%), and the presentation may range from a simple midline ridge to full expression, including a prominent keel-shaped forehead, bitemporal narrowing, and hypotelorism. Associated intracranial midline anomalies may also be seen in these patients.

Lambdoid sutures are paired and may be involved unilaterally or bilaterally. The subsequent deformity is often referred to as an occipital or posterior plagiocephaly and posterior brachiocephaly, respectively. The deformity usually occurs on the right side and may have compensatory bossing of the contralateral anterior skull. Bilateral involvement is characterized by a widened biparietal width and occipital flattening.
Vertex elongation is present in both unilateral and bilateral suture fusions. Lambdoid suture fusion must be differentiated from deforming craniosynostosis, which also presents with a flattened occiput. True lambdoid synostosis is extremely rare, with reports being 1% to 2% of all craniosynostoses.

**Plagiocephaly without Synostosis**
Craniostenosis should be differentiated from plagiocephaly resulting from external forces on an otherwise normal cranial vault complex. Often called *deformational plagiocephaly*, or *plagiocephaly without synostosis* (PWS), shaping of the skull may come from intrauterine constraint and/or postnatal positioning. The incidence has been cited as being as low as one in 300 births to as high as 48% of otherwise healthy newborns. Postnatal forces may come from supine positioning favoring one side or mild flattening of the occiput during birth, which is accentuated in the supine position from head turning by force of gravity. Muscular torticollis, a fixed head position from vertebral malformations and extraocular motor dysfunction have also been related to plagiocephaly.

Deformational plagiocephaly is uniquely different from craniosynostosis-induced plagiocephaly and may be determined by the physical examination. Frontal examination of the patient with deformational plagiocephaly reveals retraction of the ipsilateral frontal bone and superior orbit with a narrow palpebral fissure, lower eyebrows, angulation of the nasal root, and a slightly inferior position of the ipsilateral ear. There is bossing of the contralateral frontal area. The vertex view demonstrates a "pushed" posterior position of the ipsilateral chin and ear associated with a parallelogram shape of the skull.

Unilateral coronal craniosynostosis is characterized by a widened palpebral fissure and a superimposed eyebrow and supraorbital rim. The ipsilateral ear may be higher, and the nasal root is deviated to the flattened side. The vertex view demonstrates a trapezoid shape to the skull and an anterior displacement of the chin and ear.

**NORMAL DEVELOPMENT AND ANATOMY**
Sites of suture formation in the neurocranium are thought to be determined by dural reflections. Cranial bone expands from intramembranous ossification centers within a fibrous membrane called the *ectomeninx*. The leading edge of these bone plates, referred to as osteogenic fronts, contains a wedge-shaped proliferation of osteogenic cells. A syndesmosis (no interposing cartilage) is formed through apposition of these bony plates. In contrast, a synchondrosis, which contains cartilage interposition, is seen in cranial base sutures.

A suture contains the leading edge of the bony plates and the intervening radiolucent fibrous tissue. Five distinct layers of the cranial suture were identified by Pritchard and include two cambial and two capsular layers of the periosteum with a middle vascular layer. The interposed fibrous tissue between the bony fronts has been shown to contain collagen types I, III, and V, fibronectin, osteoprogenitor cells, and osteoectin. The bony edges may approximate in an end-to-end relationship, as seen in the midline sagittal and metopic sutures, or overlap, as seen in the coronal, lambdoid, sphenotympanic, and squamosal sutures. Initially all the bone edges are smooth, but as the time of suture patency increases, the number of interdigitations also increases. Studies have not demonstrated an association between the number of interdigitations and the onset of suture fusion. Facial sutures formed in the absence of dura have different developmental and growth patterns from those seen in sutures of the cranial vault.

Cranial vault sutures usually close in early adult life, with the exception of the metopic suture, which begins to close at age 2. Initial bone bridging is usually seen on the endocranial surface, although it can also begin on the ectocranium. There is a single focus of suture fusion that may occur anywhere along the course of the suture and is especially true for the sagittal suture. In contrast, metopic suture fusion progresses from inferior to superior. During suture fusion, there is a zone of osseous obliteration characterized by nonlamellar bone across the preexisting suture. As one progresses away from the site of fusion, an area of both connective tissue and osseous union exists, followed by areas of thinned connective tissue (impending suture fusion) and, eventually, an area of uninvolved sutures with normal-appearing connective tissue.

**PATHOGENESIS**
The specific cause of premature suture fusion is unclear and may be multifactorial. Nonsyndromic suture fusion may be related to extrinsic causes, such as metabolic disorders (e.g., vitamin D deficiency, hyperthyroidism) or brain malformations (e.g., microcephaly, encephalocele, corrected hydrocephalus). Chromosomal abnormalities and exposure to teratogens (e.g., aminopterin, diphenylhydantoin, retinoic acid, valproic acid) are usually associated with syndromic fusion (Box 47-1).

Three general theories of the pathogenesis of craniosynostosis have been described. Virchow suggested that the primary abnormality was localized to the affected suture and translated to the cranial base. Moss theorized that the cranial base was the source of the pathogenesis. Tension translated from the cranial base to the cranial vault sutures (presumably by the dura) cause a premature fusion of the suture. Finally, Park and Powers suggested that the defect was secondary to an abnormality in the local mesenchymal blastema. It is important to understand that the latter theory of mesenchymal cell dysfunction does not stand in conflict with the initial two theories. The pathiology, as proposed by Moss and Virchow, reflects the initial site of abnormal growth, not the primary cause of the abnormality.

The biochemical activities at the cellular and molecular levels are not well understood, but some possible mechanisms have been suggested. The predominant cell type in fused sutures has been found to be osteoprogenitor cells. The dura in proximity to the suture is necessary to maintain pa-
known causes of craniosynostosis

monogenic conditions
chromosomal syndromes
metabolic disorders
hyperthyroidism
rickets
mucopolysaccharidoses
hurler's syndrome
morquio's syndrome
β-glucuronidase deficiency
mucolipidoses
mucolipidosis iii
hematologic disorders
thalassemias
sickle cell anemia
congenital hemolytic icterus
polycythemia vera
teratogens
aminopterin
diphenylhydantoin
retinoic acid
valproic acid
malformations
microcephaly
encephalocele
shunted hydrocephalus
holoprosencephaly

from cohen mm: craniosynostosis: diagnosis, evaluation and management, new york, 1986, raven press.

vault growth that more fully explain morphologic findings seen clinically.

with the assumption that sutural edges may have asymmetric growth activities, they proposed the following:

1. cranial vault bones that are prematurely fused act as a single bone plate with decreased growth potential.
2. abnormal asymmetric bone deposition occurs at perimeter sutures with increased bone deposition directed away from the bone plate.
3. perimeter sutures adjacent to the prematurely fused suture compensate in growth more than perimeter sutures distant to the sutural stenosis.
4. a nonperimeter suture that is contiguous to the prematurely fused suture undergoes enhanced symmetric bone deposition along both edges.

these four principles of cranial vault restriction and compensation have been supported by clinical findings in nonsyndromic craniosynostosis.

Potential functional complications

functional disability is an ill-fitting term in plastic surgery, especially in the area of craniofacial abnormalities. it not only includes anatomically related disabilities but should also incorporate psychologic and developmental issues. in isolated craniosynostosis, concerns related to exorbitism; speech; and associated neurologic anomalies, such as hydrocephalus and chiari's malformations, may be minimal.

intracranial hypertension

the primary concern with premature suture fusion relates to brain growth. the brain volume of the infant increases twofold by age 1 and threefold by age 3. this rapid brain growth is paralleled by an equally rapid accommodating increase in the size of the cranial vault (table 47-1). the major functional problem associated with restrictive craniosynostosis is the development of increased intracranial pressure (icp). elevated icp may manifest in two forms. the first is the well-recognized global increase in icp subsequent to a restrictive cranium. late radiographic signs may include "fingerprinting" or "copper beating" of the endocranial surface or loss of the cisternae on two-dimensional ct scans. if severe and untreated, intracranial hypertension can translate to the optic nerve with development of papilledema; nerve ischemia; and, eventually, optic atrophy.

elevated icp may also occur transiently and be limited to a region of the brain near the fused suture. focal regions of pressure and ischemia associated with areas of suture fusion have been identified using technetium 99 cerebral flow studies. focal hypertension probably has less dramatic consequences than are seen with overt elevations in icp, but the effects may correlate more with long-term brain function, such as mental development, learning disabilities, and intelligence quotients.

aside from the potential for optic atrophy, some forms of craniosynostosis may lead to other ocular disturbances. a

Morphogenesis

In 1851, virchow described the principle that growth is restricted in the plane parallel to the prematurely fused suture. This explanation was challenged by moss based on observations that abnormal skull shapes occur in the absence of suture fusion and that cranial vault suture fusion was often associated with a cranial base deformity. He concluded that the initiating event for premature suture fusion was an abnormality in the cranial base. He also expanded the concept that the approximating soft tissue plays an active role in the shape and size of the associated bone. In light of these principles, delashaw et al outlined four components of compensatory cranial...
Table 47-1.
Craniand and Brain Growth during the First 20 Years of Life

<table>
<thead>
<tr>
<th>AGE</th>
<th>VOLUME OF BRAIN (cm³)</th>
<th>CRANIAL CAPACITY (cm³)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Newborn</td>
<td>330</td>
<td>350</td>
</tr>
<tr>
<td>3 mo</td>
<td>550</td>
<td>600</td>
</tr>
<tr>
<td>6 mo</td>
<td>575</td>
<td>775</td>
</tr>
<tr>
<td>9 mo</td>
<td>675</td>
<td>925</td>
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<tr>
<td>1 yr</td>
<td>750</td>
<td>1000</td>
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<tr>
<td>2 yr</td>
<td>900</td>
<td>1100</td>
</tr>
<tr>
<td>3 yr</td>
<td>960</td>
<td>1225</td>
</tr>
<tr>
<td>4 yr</td>
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<td>1300</td>
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<td>6 yr</td>
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<td>9 yr</td>
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<tr>
<td>12 yr</td>
<td>1150</td>
<td>1450</td>
</tr>
<tr>
<td>20 yr</td>
<td>1200</td>
<td>1500</td>
</tr>
</tbody>
</table>


significant decrease in the volume of the orbit may cause exorbitism and subsequent corneal abrasions from exposure. Likewise, suture fusion associated with orbital hypertelorism may cause restricted binocular vision. The most common intrinsic ophthalmoplegia associated with coronal synostosis is either a divergent or convergent nonparalytic strabismus or exotropia. This is related to the misshapen orbital roof and subsequent malalignment of the extraocular muscles.

CONSENT

The importance of informed consent for any procedure has been well established (Figure 47-1). It does not relieve any physician of responsibility, but it does provide a format for discussing aspects of the procedure, including potential complications and the magnitude of the surgery. Specific complications related to corrective surgery are discussed in this chapter. The parents or legal guardians should be aware of the potential for blood transfusions and provide donor-directed blood, if possible. Complications associated with the intracranial components of the procedure should also be discussed in conjunction with the neurosurgeon. A lumbar drain may be necessary if there is potential for dural compromise (for instance, during a reoperation) or evidence of elevated ICP and should be included in the preoperative review. Finally, the expected scar, postoperative course (including recovery period in the intensive care unit), and potential donor sites should be discussed.

OPERATIONS

The first recorded surgical approach for craniosynostosis was performed by Lannelongue in 1890 and Lane in 1892, who completed strip craniectomies of fused sutures. The classic neurosurgical techniques developed over the ensuing decades were geared toward resecting the synostotic suture. It was thought that a new suture line would be created that would permit normalization of the cranial vault as further growth occurred. With the realization that this goal was rarely achieved, attempts were made to further fragment the cranial vault surgically, replacing the bone as autogenous grafts that would improve preoperative cranial shape. Uncontrolled postoperative skull molding during the healing process often resulted in skull distortions. Skull reossification by the technique of calvariectomy and morcellation was found to be unpredictable and associated with substantial residual cranial vault deformity. In 1967, Tessier described a new approach to the management of Crouzon's disease and Apert's syndrome. His landmark presentation and publications were the beginning of modern craniofacial surgery. Tessier combines an intracranial-extracranial approach with the use of a coronal incision, extensive periorbital subperiosteal dissection, autogenous bone grafting, and ingenious osteotomies. The concept of calvarial suture resection combined with skull reshaping in infancy was later pioneered by Hoffman, Whitaker, and Marchac. Hoffman reported lateral canthal advancement of the supraorbital margin as a new corrective technique in the treatment of coronal synostosis in 1976. This heralded reports by Whitaker in 1977 and the classic article by Marchac and Renier, which presented the floating forehead technique combined with frontoorbital advancement.

It has become accepted clinical practice with patients with nonsyndromic and single-suture craniosynostosis for clinicians to perform the primary operative procedure—frontoorbital advancement with cranial vault remodeling—at an early age to improve craniofacial form and function and lead to satisfactory long-term growth and development of the calvaria. Our current approach to single-suture and nonsyndromic craniosynostosis varies with the underlying suture fusion. In general, once a diagnosis has been established by physical examination and appropriate radiologic studies, a surgical treatment plan is recommended. It is critical that children undergo multidisciplinary evaluation by a craniofacial team. The team geneticist rules out associated abnormalities that can occur to a greater or lesser proportion in a number of craniosynostosis patients. In children with bicoronal synostosis, it is especially important to rule out syndromic involvement because this will not always be
Do not sign this form until you have read it and fully understand its contents.

Patient’s Name ___________________________ Date ___________________________

The following has been explained to me in general terms, and I understand that:

1. The diagnosis requiring this procedure is:

2. The nature of the procedure is:

3. The purpose of the procedure is:

4. Material risks of the procedure: As a result of this procedure being performed, there may be material risks of infection, allergic reaction, disfiguring scar, severe loss of blood, loss or loss of function of any limb or organ, paralysis, paraplegia or quadriplegia, brain damage, cardiac arrest, or death.

5. In addition to these material risks, there may be other possible risks involved in this procedure, including, but not limited to:

6. The likelihood of success of the above procedure is:
   ( ) Good   ( ) Fair   ( ) Poor

7. Practical alternatives to this procedure include:

8. If I choose not to have the above procedure, my prognosis (future medical condition) is:

9. I understand that the physician, medical personnel, and other assistants will rely on statements about the patient, the patient’s medical history, and other information in determining whether to perform the procedure or the course of treatment for the patient’s condition and in recommending the procedure that has been explained.

10. I understand that the practice of medicine is not an exact science and that no guarantees or assurances have been made to me concerning the results of this procedure and that sometimes a patient’s expectations may be greater than the reality of the treatment.

11. I understand that during the course of the procedure described above it may be necessary or appropriate to perform additional procedures that are unforeseen or not known to be needed at the time this consent is given. I consent to and authorize the persons described herein to make the decision concerning such procedures. I also consent to and authorize the performance of such additional procedures as they may deem necessary or appropriate.

12. I consent to diagnostic studies, tests, local and/or general anesthesia, x-ray examinations, and any other treatment or courses of treatment relating to the diagnosis or procedures described herein as may be deemed advisable.

13. I consent that any tissues, specimens, organs, or limbs removed from the patient’s body in the course of any procedure may be tested or retained for scientific or teaching purposes and then disposed of within the discretion of the physician, facility, or other health care provider.

14. I consent to the taking of photographs before and after this operation or treatment, as well as in the course of this operation or treatment. I understand and give my permission for the photographs to be used for the purpose of medical or instructional purposes, including lectures and/or publications.

15. By signing this form, I acknowledge that I have read or had this form read and/or explained to me, that I fully understand its contents, that I have been given ample opportunity to ask questions, and that any questions have been answered satisfactorily. All blanks or statements requiring completion were filled in and all statements I do not approve of were stricken before I signed this form. I also have received additional information, including, but not limited to, the materials listed below, related to the procedures described herein.

16. Additional materials used, if any, during the informed consent process for this procedure include:

17. I voluntarily allow
   along with any physician designated or selected by him or her and all medical personnel under him or her and all medical personnel under the direct supervision and control of such physician and all other personnel who may otherwise be involved in performing such procedures to perform the procedures described or otherwise referred to herein.

Signature of person giving consent: ___________________________

Relationship to patient if not the patient: ___________________________

Patient unable to sign because of: ___________________________

Date: ______________ Time: ______________ Witness: ___________________________

Figure 47-1. Informed consent.
apparent on preliminary examination yet has important
prognostic implications for the family and patient. An
ophthalmologist routinely sees each patient preoperatively
to determine whether associated ocular adnexal problems exist
and to rule out papilledema. The neurosurgeon ensures that
associated brain parenchymal abnormalities are not present
and evaluates the patient for the possibility of subclinical or
 overt intracranial hypertension.

The decision to operate on cases of single-suture synostosis
should not be based solely on aesthetic considerations, but
concerns regarding local or regional increases in ICP on brain
function should also be raised with parents. By the same token,
families with children in whom more minor degrees of skull
deformity exist, such as mild metopic synostosis, should not be
frightened into surgery because of the prospect of brain
damage secondary to undetected elevations of ICP. These
children can be followed in a craniofacial team setting to
optimize the chances for normal development and outcome.

The diagnosis of elevated ICP in children with single-suture
craniosynostosis must be made from a constellation of clinical
findings supported by the measurement of ICP in selected
cases. Children with a history of developmental delay,
headaches, and other neurologic symptoms of elevated ICP
should be referred to a pediatric ophthalmologist for fundus-
scopic evaluation. The presence of papilledema and other
findings consistent with elevated ICP should be noted. It is
important, however, to remember that increased ICP may be
present in the absence of papilledema, which is often a later
finding. Two- and three-dimensional computed tomography
(CT) scans should be routinely obtained. We rarely order plain
films of the skull, but occasionally these are brought by patients
from outside settings and should be reviewed for evidence of
craniosynostosis and digital printing or copper-beaten appear-
ances of the calvaria. In children less than 1 year of age with
isolated synostosis of the sagittal, metopic, or lambdoid suture
whose parents already wish to proceed to normalize skull
shape, ICP monitoring is unlikely to contribute to clinical
management. For children in these categories whose parents do
not desire surgical intervention, the measurement of ICP, if
indicated, may provide a means of excluding potentially
damaging consequences of the synostosis. In children with
uniconiral synostosis of any age, surgical intervention is
generally recommended because of facial growth problems
accompanying this type of sutureal fusion. Children presenting
with craniosynostosis later than 1 year of age, particularly those
with developmental delay or other signs of ICP, may be
referred for ICP monitoring. It is important that all factors be
taken into account before proceeding with surgical interven-
tions. The family should be apprised as best as possible and
given realistic expectations regarding improvement in develop-
mental delays.

Eppley and Sadove28 paved the way for the use of
biodegradable plates and screws in infant craniofacial surgery.
Our own center’s experience with these has been extremely
positive. They do require more patience than conventional
metallic plates and screws and, once positioned, if changes
are necessary, the biodegradables must usually be discarded
and replaced. They are also more expensive
than metallic plates and screws; however, the decreased
reoperation rate for removal will more than likely make up for
the added initial expense. Metallic plates and screws are still
useful, however, in infant craniofacial surgery, provided that
the parents are apprised of their need. Consideration can also
be given to removing them at a later date on a routine basis,
although we do not advocate this in most patients. Provided
that the plate lengths are kept small and the configuration
simple, migration may be less likely. Also, it is preferable to
avoid using metallic plates in the midline and especially the
nasofrontal region because this is where most clinical cases
associated with migration have been found.

PATIENT PREPARATION

In children undergoing simple sagittal or lambdoid synosto-
comies, two intravenous lines are used, but a Foley catheter and
intraarterial line are not routinely inserted. Our preference is to
perform synostectomies from 2 to 4 months of age, if at all
possible. When performing simple synostectomies for cases of
sagittal and true lambdoid synostosis, earlier surgery results in
the best chance for normalization of calvarial shape. Simple
sagittal and lambdoid synostectomies are performed with the
patient in prone position on a horseshoe headrest. Attention is
taken to ensure that no pressure on the eyes occurs because this
is a known cause of blindness. For the smaller infants, typically
chest rolls are not used because they may impede ventilation.
In patients undergoing either a Pi procedure41 or major cranial
vault reconstruction for sagittal synostosis, the modified prone
position with the neck hyperextended is used. In these cases, a
cervical spine x-ray is obtained before putting the child in this
position. In addition, because of the potential for increased
bleeding secondary to jugular venous hypertension, an intra-
arterial line is inserted, as is a Foley catheter in those children
undergoing major cranial vault reconstruction. For patients
with metopic, unicoronal, and bicoronal synostosis, the patient
is operated on in the supine position. In addition to two
intravenous lines, an intraarterial line is routinely used, as is a
Foley catheter. In the standard team approach, the neurosur-
gen begins the operation. The child undergoes a 10-minute
head scrub and a povidone-iodine (Betadine) preparation. We
do not routinely shave the hair. The incision is marked with a
circle and generally performed in a wavy S-shaped fashion
behind the hairline. A Shaw scalpel is used with the skin
incision made at 110 degrees and the deeper dissection
performed at 260 degrees. Michel clips are used to attach
sponges to the wound edges for hemostasis. Continuing with
the Shaw scalpel, the dissection is performed in the subgaleal
plane. This is carried out down to the level of the supraorbital
rims, and then the periosteum is incised along the insertions of
the temporal muscles and then horizontally at the level of the
anterior fontanelle. This permits subperiosteal elevation of a
generous tongue of peristeum and galea. Subperiosteal
dissection is then performed to expose the supraorbital rim and
lateral orbital rim down to the body of the zygoma, and
intrabosally to the level of the inferior orbital foramen. In
patients undergoing correction of metopic, unicoronal, and
bicornal synostosis, a bifrontal craniotomy is always carried out. The anterior cranial fossa is then exposed to perform the frontoorbital osteotomies. All children undergoing correction of craniosynostosis at our institution are typed and crossed for either donor-directed or banked blood. Before beginning the surgical procedure, the blood is transported to the refrigerator in the operating room, where it is immediately available at the commencement of the case.

SAGITTAL SYNOSTOSIS

Depending on the degree of deformity and the age of the patient, a variety of operations have been used for correction of sagittal synostosis. In young infants with mild scaphocephaly, a simple sagittal synostectomy is carried out. With the skull exposed, an incision is made with the Shaw scalpel 2.5 cm off the midline and carried from the coronal suture posteriorly through the lambdoid suture. This pericranial incision then continues well past the lambdoid sutures into the occiput. The extent of the posterior bone removal will depend on the severity of the posterior deformity. A Burr hole is made in the pericranial incision at the level of the vertex. With the Midas Rex drill (Midas Rex, Fort Worth, Tex.), it is unnecessary to strip the pericranium. A second pericranial incision is made in front of the lambdoid suture on either side 2.5 cm off the midline. The bone is removed from side to side, just in front of the lambdoid suture using either a fine Rongeur or the M-8 attachment of the Midas Rex. The pericranium is then incised along the posterior border of the anterior fontanelle. A curette opens the interval between the bone and dura. The B-5 attachment, which is the pediatric craniotome of the Midas Rex, is used to make a linear osteotomy. The parasagittal osteotomy is made from the coronal suture to the lambdoid suture on both sides. Typically the bone is stuck to the dura, especially over the sagittal sinus. A sharp periosteal elevator is used to dissect the remaining midline suture away from the sinus. Bleeding from the pacchionian granulations and draining veins are common, and the dissection must be performed quickly but carefully. Once the bone is removed, the dura is immediately covered with a wet sponge to stop significant bleeding. The bipolar cautery is then used to coagulate significant bleeds remaining on the dura. Now the bone has been removed from the coronal suture to just in front of the lambdoid. Dissecting underneath the lambdoid sutures and across the midline with periosteal elevator, the Midas Rex drill is then used to remove any significant occipital deformity. The bipolar is turned to 45, and the dura directly over the sagittal sinus is coagulated swiftly in order not to thrombose the sinus but rather to retard the dural osteoclast in the midline. The wound is then copiously irrigated with bacitracin, and the skin is closed with 4-0 Vicryl and staples. We have gotten away from using head wraps and prefer to place mupirocin (Bactroban) over the incision. The child is then carefully rolled back into supine position and taken to the recovery room.

This synostectomy procedure allows for significant improvement in head shape. It works especially well on large occipital knobs and in correcting the biparietal narrowing. The downside to the technique is that it fails to address significant forehead bossing. For children who present with moderate frontal prominence without saddling deformities or significant occipital abnormalities, a Pi procedure is used. This procedure is carried out in the modified prone position with the neck hyperextended. A cervical spine x-ray is obtained before putting the child in this position. Bilateral parasagittal strips of bone are removed between the coronal and lambdoid sutures. An osteotomy is then performed the length of the coronal suture on each side. This completes the formation of the Pi. A 3-0 silk is threaded through a twist drill hole between the midline construct and the forehead. When this is tied down, it foreshortens the forehead. Before tightening the suture, vertical osteotomies in the parietal bone are carried into the temporal fossa. Thus when the forehead is pulled back, the dura bulges laterally, correcting the biparietal narrowing. The procedure takes longer than a sagittal synostectomy and has a larger blood loss. Children who undergo strip craniectomy typically go home the following morning, and only 5% or less require a blood transfusion. Children undergoing the Pi procedure generally stay longer simply because they are sicker in the first day or so. Typically a higher percentage require blood transfusions, as well.

A major cranial vault reconstruction in an infant is reserved for children with severe skull deformities. In children under 7 months of age with severe sagittal synostosis, conventional strip craniectomy and even the Pi procedure fail to correct late deformities characteristic of severe scaphocephaly. The components of the deformity that require correction include extreme elongation, frontal and occipital bossing, temporal pinching, and angulatory apical skull deformations. Several investigators have reported unsatisfactory results in children of any age when the scaphocephalic deformity is severe, particularly when associated with large occipital shelves and marked frontal bossing. To improve surgical results, we prefer total cranial vault reconstruction (Figure 47-2) in these children. After
general anesthesia is induced, the endotracheal tube is secured to the mandible with a circummandibular wire. Two large intravenous catheters are placed, as is a Foley, to monitor urine output. An arterial line is used. The patient is placed in prone position with the head position maintained using an Olympic Vac positioning system (Sizall, Olympic Medical, Seattle) chin support. The neurosurgical and craniofacial portions of the procedure are carried out in sequential fashion. First, a frontal craniotomy is performed with a supraorbital osteotomy below the frontal boss. The posterior osteotomy is placed in front of the coronal suture, and the frontal bone is carefully removed. The two lateral temporoparietal bone segments are removed, staying just lateral to the sagittal suture. The occiput is removed, keeping the posterior osteotomy below the occipital shelf. A burr hole is placed adjacent to the midline of the occiput to facilitate stripping of the dura mater. Barrel-Stave osteotomies are performed in a vertical fashion along the remaining temporal bone and occipital bone and interconnected to permit lateral expansion of the cranium. Plication sutures are placed diagonally in two rows in the bulging frontal dura mater. The frontal bone is split into halves and expands in the transverse plane with radial osteotomies and interposition bone grafts. A wedge of bone is removed from the frontal bone at its junction with a supraorbital osteotomy to allow posterior rotation of the frontal bone. Biodegradable plates, titanium or Vitallium microplates, or wire osteosynthesis are used to link the two halves of the frontal bone in the supraorbital osteotomy suture. The occiput is split in half, allowing for lateral expansion in the same fashion as the frontal bone.

While the craniofacial surgeon reconstructs the frontal and occipital bones on a side table, the neurosurgeon inserts an ICP monitor through the right side of the large sagittal bone strip, which has been left between the osteotomized frontoparietal and occipital bone segments. This central strip serves as an anchor for the anteroposterior correction. Two small drill holes are placed in the central portion of the occipital and frontal bone plates. The central bone segment covering the sagittal sinus is then dissected free of the dura and sinus. A section of the central strip is removed according to the amount of anteroposterior correction deemed necessary, which averages 2.5 cm. A corresponding strip of the lateral temporoparietal bone segment is also removed. Small drill holes are placed through the central sagittal bone strip. The frontal bone segment is replaced using biodegradable plates, metallic plates, or wires along the supraorbital rim. Posteriorly, the occipital bone is replaced in a similar manner. Then 28-gauge wire is passed through the osteotomized occipital and frontal bone segments and attached to the central sagittal bone strip. While carefully monitoring the ICP, the right and left wires are slowly twisted on the frontal segment until bone contact occurs between the frontal bone and the central sagittal strip. As anteroposterior correction proceeds, bulging of the dura mater laterally is noted. The occipital correction is carried out by slowly twisting the wires, advancing the occipital bone segment toward the central sagittal strip until bone contact is achieved. ICP is maintained at 15 mm Hg with normocapnia. The anteroposterior correction is carried out slowly, often over 30 to 45 minutes in some cases. This permits equilibration of the ICP. Two small holes are drilled into the lateral temporoparietal bone segments, and these are then sutured directly to the dura mater with a horizontal mattress suture.

After all the osteotomized segments have been replaced, attention is then turned to the anterior temporal fossa region. Preoperatively this area is often pinched. To correct these depressions, excess bone that has been trimmed from the calvaria is cut and contoured to fit the temporal fossa. These may be attached with biodegradable screws or microscrews to the lateral orbital rim. The temporalis muscle is then advanced and resuspended to the lateral orbital rim. The large pericranial flaps, which had been elevated at the time of craniotomy, are replaced, covering the frontal bone and lateral orbital regions. In cases of total cranial vault reconstruction, we routinely insert a closed-system drain through a separate stab incision in the posterior occipital flap. The galea is closed using absorbable sutures in the skin with staples. Postoperatively, the patient is extubated in the recovery room and sequential neurologic examinations are performed. Patients undergoing total cranial vault reconstruction are placed in the intensive care unit for 24 to 48 hours. Careful monitoring of serum sodium is carried out over the first 72 hours. Transfusion of donor-directed blood, usually from parents, is begun intraoperatively and continues postoperatively for the first 24 hours as required. The drain is removed when its output has been reduced to less than 10 mm per day. The skin staples are removed from 7 to 10 days postoperatively.

Patients presenting after 2 years of age with significant scaphocephaly and sagittal synostosis undergo either three-quarters calvarial vault reconstruction or total calvarial vault reconstruction (Figure 47-3), depending on the degree of deformity. The main difference in technique in these patients...
is that calvarial defects are not left. When performing total cranial vault reconstruction, the calvaria between the frontal and occipital bones is usually removed in three transverse bands of approximately equal width. The lateral osteotomies for these bands are made just above the squamosal suture. The osteotomy across the midline is accomplished via three slightly off-center central drill holes to protect the sagittal sinus. In contrast to the early cases in which the bone over the majority of the sagittal sinus is left intact, in the later cases the bone is removed in its entirety. Each one of the three central bony segments is taken to the side table, where multiple partial-thickness osteotomies are made with a cutting burr on the inner surface of the bone. Next, gentle greenstick fractures are made along each of the partial-thickness osteotomies with a bone-contouring forceps. To expand the transverse dimensions of the skull, frequently these plates are split down the center, and an interposition bone graft is inserted. Biodegradable plates and/or microplates are used for stabilization of the interpositional bone. An average of 1.5 cm of bone is removed from the anterior and posterior skeletal bands to allow appropriate anteroposterior correction. The occipital and frontal bones, which have been typically split down the midline and enlarged transversely with placement of an interpositional bone graft, are reattached to the supraorbital and posterior occipital ledges with biodegradable plates or microplates. The central bone segment is then secured with biodegradable plates or microplates to the lateral temporal bone to allow it to act as a central post. Next, the anterior and posterior bands are replaced. The frontal bone is connected to the anterior strut either with 28-gauge wire or biodegradable plates or microplates, while careful attention is paid to ICP. The occipital bone is treated in a similar fashion. Lateral dural bolting into the Barrel-Style osteotomies that have been placed along the lateral temporal and parietal bones permit increased transverse dimensions (Figure 47-4).

**METOPIC SYNOSTOSIS** (Figures 47-5 and 47-6)

Although some variation of technique is used depending on the degree of deformity, in general the surgical approach is fairly routine for correction of metopic synostosis. After frontal craniotomy, bilateral supraorbital rim osteotomies are performed without a temporal tenon or Z-plasty unless wires are being used for stabilization. The supraorbital bar is split vertically with a reciprocating saw, and the deformed metopic suture is completed resected. A bone graft is routinely used in the midline to increase the transverse dimensions of the supraorbital bar. If hypotelorism is present, the nose is split in the midline and bone grafted as first described by Marchac and Renier and later by Sadove et al. In patients with severe hypotelorism, correction may be incomplete with the above technique. In such cases, we have used a modified medial orbital osteotomy (Figure 47-7) to increase the degree of interorbital separation. In these cases, the osteotomies are performed in situ. A Smith separator is placed in the osteotomy gap along the zygomatic temporal junction and gently spread to separate and advance the superior and lateral orbital rim to correct the triangular-shaped deformity of the skull. Partial-

![Figure 47-4. A, Eight-month-old with sagittal synostosis and severe scaphocephaly. B, Postoperative result after total cranial vault reconstruction.](image1)

![Figure 47-5. Technique used for correction of metopic synostosis. A and B, Deformity and osteotomies. C, Osteotomies. D, Reconstruction.](image2)
Figure 47-6. A, Lines of osteotomy for resection of synostosed metopic suture. Note that the osteotomy extends down to the nasofrontal region and is then continued along the midline of the nose. Segments are then differentially advanced laterally and transversely expanded. B, Artist’s version of the craniofacial reconstruction for metopic synostosis. Note the two-hole microplate on the lateral orbital rim to temporal bone region. This is fixed anteriorly only and left floating posteriorly. Also note the bone graft wedged in the midline after the nasal osteotomy and transverse expansion, plus anterior advancement of the lateral supraorbital bar. Posteriorly, the forehead is left floating. C, Preoperative frontal and lateral photograph of an 8-month-old girl with metopic synostosis. D, Postoperative frontal photograph 6 months after frontoorbital remodeling. E, At 16 months after frontoorbital remodeling, grade I surgical result with minimal aesthetic irregularities.

Figure 47-7. Modified medial orbital osteotomy used for patients with severe hypotelorism and metopic synostosis. A, Osteotomies. B, Bone grafts. Supraorbital rims are advanced in situ.
frontal bone may be onlaid over the advanced supraorbital rim and held in place with positional microscrews or biodegradable screws—the advancement-onlay technique. Typically stabilization with plates and screws is sufficient such that in children under 2 the frontal bone graft is left floating posteriorly. In older patients (>18 to 24 months), craniectomy defects are filled with bone grafts. The temporalis muscles are advanced and resuspended to the lateral orbital rim with Vicryl sutures.

UNICORONAL SYNOSTOSIS
(Figures 47-8 to 47-10)

The surgical approach to unicoronal synostosis again depends on the degree of deformity. Again, after a bifrontal craniotomy, an osteotomy of both supraorbital rims is carried out. The entire complex is then removed. In children with malar recession, the osteotomy along the lateral orbital rim may be carried into the zygoma and even include the inferior orbital rim. In cases in which the nasal deviation is severe, the osteotomy may be performed low across the nasal dorsum. We have not used nasal maxillary osteotomies to correct deviation in this region in this age-group after this deviation improves over time. It is not easy to predict which patients will go on to develop facial scoliosis secondary to synostosis along the cranial base. Current surgical techniques do little to prevent this outcome. When compensatory changes of the contralateral forehead are severe, the contralateral "unaffected orbit" will be lowered, in addition to the peaked harlequin orbit of the affected side. In such cases, it is important to reposition the entire supraorbital bar in a manner that normalizes the horizontal position of the orbital roofs. In milder cases, the transverse deficiency of the affected orbit can be corrected by carving out the area along the nasal junction. However, in more severe cases, a vertical osteotomy is usually performed through the affected supraorbital rim and an interpositional bone graft placed. The supraorbital bar in unicoronal synostosis must be inspected carefully because the curvature is frequently abnormal when viewed from the undersurface. Rather than the normal convexity of the curve at the midportion of the orbital roof, a concavity in this area is present. Partial-thickness wedge osteotomies will allow proper recontouring with the bone-contouring forceps. When an interposition bone graft is used, it is rigidly fixed with biodegradable plates or microplates into position. Once the supraorbital bar is recontoured, it is stabilized along the inferior portion of the lateral orbital rims. The temporal region is taken off as a separate strut rather than as a tenon so that it may be segmentally reconfigured. This allows for maximum flexibility in correcting the temporal region, which is often the site of residual deformity. Once the supraorbital rim has been adequately stabilized and the temporal region corrected and stabilized to the advanced supraorbital rim, the frontal bone is recontoured. Often by rotating the frontal bone 180 degrees, a more acceptable contour will be achieved. However, the region that had developed compensatory bossing will require multiple wedge osteotomies to allow proper recontouring. Generally, the frontal bone can be recontoured by the combination of wedge osteotomies and bone remodeling with the bone-bending forceps. Occasionally, however, it will need to be osteotomized into segments, which are then rigidly fixated to each other with biodegradable plates or microplates. The forehead is then reattached to the supraorbital rim, either flush or, if additional projection is necessary, as an advancement-onlay technique. Occasionally, additional bone grafts will be necessary to augment the affected supraorbital rim. Once the frontal bone has been restabilized, the peaked lateral aspect of the orbital roof is corrected with a contoured bone graft, as well. In cases of large advancements, the bone graft is actually wedged into the osteotomy gap created at the lateral superior aspect of the rim. The supraorbital rim is also stabilized at the nasal bridge either with a biodegradable plate or occasionally with Vicryl sutures affixed directly to the periosteum. This is done to prevent superior rotation of the rim. Occasionally a bone graft will need to be inserted along the nasal bridge to ease the transition at the nasofrontal junction. A lateral
canthopexy may be necessary, depending on the final position of the lateral canthus with respect to the contralateral side. The temporalis muscle is then resuspended. Once in a while, additional contoured bone grafts are inserted under the temporalis muscle to add further augmentation to the temporal region. Generally the forehead is left floating; however, in older children or in cases in which significant advancement has been performed, bone struts may be used for posterior stabilization, as well.

**BICORONAL SYNOSTOSIS**

(Figures 47-11 to 47-13)

Again, a frontal craniotomy that includes the fused coronal sutures is performed. The supraorbital bar is removed with a reciprocating saw. Some degree of turritbrachycephaly may be present, even in non-syndromic patients. In such cases, a craniotomy is carried posteriorly along the inferior portion of the parietal bone on both sides. Just proximal to the

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**Figure 47-10.** A, Preoperative photographs of a 6-month-old boy with unicoronal synostosis. B, Postoperative photograph following technique illustrated in Figure 47-11.

**Figure 47-11.** A to D, Technique used to correct bicoronal synostosis without turriccephaly.

**Figure 47-12.** A to C, Technique used when turriccephaly is present. (From Cohen SR, de Chalain TM, Burstein FD, et al: Ann Plast Surg 35:627-630, 1995.)
lambdoids, the craniotomy proceeds superiorly toward the sagittal sinus. The bone across the midline is carefully removed with a ronguer. With the maneuver repeated on the opposite site, a visor of bone still attached at the midline over the sinus may be gently compressed, overlapping the superior portion of the parietal bones over the inferior cranium. This lowers the vault, pushing the brain and dural coverings forward to fill out the forehead and supraorbital rims.\(^{17}\)

Stabilization of the supraorbital rims and forehead is carried out with biodegradable plates and screws placed at the lateral orbital rims and nasofrontal junction. The frontal bone is reattached to the advanced supraorbital rim with biodegradable plates and screws. Often a cranial bone graft is placed along the nose to ease the stepoff at the nasofrontal junction.

**LAMBDOID SYNOSTOSIS AND POSITION-RELATED HEAD DEFORMITIES**

The diagnosis and treatment of posterior plagiocephaly and true lambdoid synostosis is one of the most controversial aspects of craniofacial surgery.\(^{3,12,38,67,89}\) The features of true lambdoidal synostosis versus those of positional or deformational plagiocephaly are inadequately described in the literature. This has resulted in many infants across the United States undergoing major intracranial surgery to treat nonsynostotic plagiocephaly.

Patients with true lambdoid synostosis have a thick ridge over the fused suture with compensatory contralateral parietal and frontal bossing, as well as an ipsilateral occipitomastoid bulge (Figure 47-14). The skull base has an ipsilateral inferior tilt, with a corresponding inferior and posterior displacement of the ipsilateral ear. The skull, when viewed from above, takes on a trapezoidal appearance. These characteristics are opposite to findings in patients with positional molding and open lambdoid sutures.\(^{38}\) In positional molding, the head takes on a parallelogram shape with flattening of the occiput and contralateral frontal bossing.

Treatment of true lambdoid synostosis is surgical. In younger infants, a broad strip of craniectomy is performed. In older children (<2 years) or those with severe deformities, skull vault remodeling is carried out. The treatment of mild

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**Figure 47-13.** A and B, Preoperative photographs of a 6-week-old with bicornoral synostosis and turriccephaly. C and D, At 1 year postoperatively. (From Cohen SR, de Chalain TM, Burstein FD, et al: *Ann Plast Surg* 35:627-630, 1995.)

**Figure 47-14.** Vertex views. A, Infant with positional molding. Observe right occipitoparietal flattening, left occipital bossing, anterior displacement of right ear, and lack of right occipitomastoid bulge. B, Infant with left lambdoid synostosis. Observe right parietal bossing, left occipitomastoid bossing, and posterior displacement of left ear.
Positional molding is repositioning. Moderately severe deformities are treated as early as possible with helmet therapy or dynamic orthotic cranioplasty.

**POSTOPERATIVE MANAGEMENT**

The vast majority of children undergoing correction of nonsyndromic single-suture craniosynostosis are admitted first to the recovery room and then transferred to the surgical floor or the intensive care unit. At our institution, only those patients undergoing major cranial vault reconstruction and/or those patients with associated congenital disorders (e.g., cardiac disease) are admitted to the intensive care unit. Approximately 70% of the children are transferred directly to the surgical floor. When a head dressing is used, it is typically removed on the third postoperative day. Patients are given intravenous antibiotics until the time of discharge and begun on a clear liquid, progressing to an age-appropriate regular diet once awake and alert. A complete blood count (CBC) and electrolytes are routinely drawn in the recovery room and on the first postoperative morning. In children undergoing major cranial vault reconstruction, serum electrolytes are followed for the first 72 hours. The majority of children leave the hospital by the fourth postoperative day. The family is instructed to care for the sutures or staple lines. Typically, the staples are removed 7 to 10 days after operation. Children return to 4 to 6 weeks later, after which time routine follow-up is established at the craniofacial center. Children are typically followed every 4 to 6 months until their third birthday, when annual follow-up begins. Children are monitored for normal neurologic development, and symptoms and signs of elevated ICP are closely monitored.

**SECONDARY PROCEDURES**

In the Outcomes section of this chapter, the frequency and types of secondary procedures are listed. In patients undergoing correction of metopic synostosis, secondary procedures may be necessary to treat residual or recurrent temporal depressions. In such cases, any metallic hardware is removed, and hydroxyapatite or paste (bone source) is used for reconstruction. We have been pleased with the outcomes after simple recontouring using hydroxyapatite and recently reported our results in a small group of patients. Occasionally, as noted in the next section on outcomes, total reoperation is necessary. When total reoperation is carried out, generally the cases are longer and associated with higher blood loss. The same basic principles, however, are adhered to, as described above. Rarely, a patient will present with residual calvarial defects. Typically these children are given until around age 2 to develop new bone. If residual defects are present by this time, bone grafting is carried out. Although some authors have noted improvement in hypotelorism associated with metopic synostosis over time, others have found residual hypotelorism. In cases in which redundant epicantal folds are associated with intercanthal narrowing, a simple cranial bone graft to the nose may take up the extra tissue, and medial osteotomies are not usually necessary.

In patients with sagittal synostosis treated by strip craniectomy, a small proportion may ultimately require total or subtotal cranial vault remodeling. Patients who have undergone total calvarial reconstruction may require secondary cranial reconstruction for (1) hardware removal; (2) hardware removal and recontouring; and (3) rarely, redoing total cranial vault reconstruction. We have recently had good experience with bone source, a hydroxyapatite paste with which it is easy to work.

Patients with unicoronal synostosis may require secondary surgery for minor temporal depressions and supratrochlear rim asymmetry. A small percentage, who may have more extensive involvement of cranial base sutures, will go on to develop a clinical picture that has been referred to as *facial scoliosis*. Significant nasal deviation and sygomatic-maxillary deformities are often associated with occlusal cants and even mandibular asymmetry. Correction requires a complex craniofacial approach combined with orthognathic procedures to realign the jaws.

Patients undergoing correction of bilateral coronal synostosis usually do well. It is likely, however, that a percentage of children with "nonsyndromic" craniosynostosis have an undiagnosed craniofacial syndrome leading to a higher frequency of recurrent deformity, which may require partial or total reoperation.

Secondary procedures to expand the cranial vault may be necessary in a few patients with nonsyndromic craniosynostosis who develop increased ICP after frontoorbital advancement. We follow all children annually in the Center for Craniofacial Disorders to monitor skull growth and neuropsychiatric development.

**OUTCOMES**

Outcome analysis functions to determine the efficacy and reasonability of a service or product. This model is complicated in health care by the balance of cost containment to patient satisfaction and quality. Even so, application of outcome analysis provides the medical community a method of comparing and adjusting practice modalities to meet specific demands. It can also provide a means of establishing and communicating standard of care levels to those involved in the health care industry.

**COMPLICATIONS**

Cranial vault remodeling is usually completed within the first 12 to 15 months of age. The magnitude of the surgery is dictated by the sutures involved. The age of the patient and the intracranial component of the procedure carry inherent risks. Anesthetic risks are not significantly increased in the patient population compared with adults. But the introduction of
venous and arterial lines, as well as central venous monitoring ports, may be difficult in some cases. Because of the diminished size of the airway and the small oxygen reserve in these patients, potential airway problems should always be considered. Finally, the increased surface area of the pediatric patient can cause a quick decrease in the core body temperature if active steps to maintain body temperature are not taken.

Specific complications associated with craniosynostosis may be classified as either perioperative or postoperative. In a review of 204 patients treated for nonsyndromic craniosynostosis at Scottish Rite Children’s Medical Center, the overall perioperative complication rate was 9.8% (20/204) (Box 47-2). Major complications were seen in total cranial vault remodeling for sagittal synostosis in which four experienced hypovolemic shock. One patient had a transection of the sagittal sinus, and one death was recorded. This was the only death in our series. Seven patients were found to have syndrome of inappropriate antidiuretic hormone (SIADH) in the immediate postoperative period (four in the total cranial vault remodeling group). Other perioperative or immediate postoperative complications included severe chemosis (one case), wound infection (one case), and urethritis (one case). Perioperative complications, including air embolism after a venous tear, infarction, and damage to the unprotected brain, have also been reported.

Blood loss may be acute, but transfusions are usually necessary because of insidious losses throughout the case. Blood transfusions are required in almost all total cranial vault remodeling procedures. Transfusion requirements have been reported from 15% to 90% of the patient’s estimated red blood cell volume. Variability is seen based on the type of synostosis (Table 47-2). Acute complications of blood transfusion are well known and include hypocalcemia, hyperkalemia, coagulopathies, and transfusion incompatibility. Delayed complications of blood transfusions usually involve the potential for transmission of viral infections.

REOPERATION RATES

The rate of reoperation is an important outcome variable in the surgical treatment of craniosynostosis. Although quantitative changes in craniofacial remodeling are critical to understanding operative results, the decision to reoperate on a particular child is determined primarily from subjective measures of outcome, most commonly aesthetic appearance. Longitudinal studies of reoperation rates from a variety of centers are beginning to appear in the literature.

A prospective statistical study of reoperation rates was reviewed in the treatment of 167 consecutive children with nonsyndromic and syndromic craniosynostosis over a 6-year period.

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Table 47-2. Mean (SD) Percentage of Patient’s Estimated Red Cell Volume Lost and Type of Skull Deformities

<table>
<thead>
<tr>
<th>TYPE OF SYNOSTOSIS</th>
<th>DURING OPERATION</th>
<th>AFTER OPERATION</th>
<th>TOTAL</th>
</tr>
</thead>
<tbody>
<tr>
<td>Oxycephaly</td>
<td>49.1 (22.9)</td>
<td>-3.7 (16.8)*</td>
<td>45.4 (36.9)</td>
</tr>
<tr>
<td>Plagiocephaly</td>
<td>59 (37.4)</td>
<td>27.7 (41.7)</td>
<td>86.7 (56.2)</td>
</tr>
<tr>
<td>Trigonocephaly</td>
<td>92.4 (49.7)</td>
<td>11.7 (66.6)</td>
<td>104.1 (49.2)</td>
</tr>
<tr>
<td>Brachycephaly</td>
<td>105.3 (48.45)</td>
<td>25.5 (56.5)</td>
<td>130.9 (69.1)</td>
</tr>
<tr>
<td>Scaphocephaly</td>
<td>92.1 (65.2)</td>
<td>35.9 (38.4)</td>
<td>121.7 (78.2)</td>
</tr>
<tr>
<td>Complex</td>
<td>198.5 (165)</td>
<td>44.5 (97.6)</td>
<td>243.1 (259.4)</td>
</tr>
</tbody>
</table>

*Negative value indicates postoperative overtransfusion.

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period at Scottish Rite Children's Medical Center (Figure 47-15). Mean length of follow-up was 2.8 years. Frontoorbital remodeling with a floating forehead was completed at 4 to 6 months of age for nonsyndromic synostosis other than sagittal synostosis. This approach is similar to treatment of isolated synostosis in several centers.\(^{71,95,97}\) Bilateral frontoorbital remodeling has been shown to be comparable with or better than unilateral remodeling and was done in all cases.\(^{92,97}\) Strip craniectomies were limited to sagittal synostosis with mild-to-moderate deformities. Total cranial vault remodeling was completed for severe deformities if the patient was greater than 7 weeks old. Patients with syndromic craniosynostosis underwent frontoorbital advancement and cranial reshaping in 4 to 6 months unless increased cranial pressures required decompression.

Reoperation equal to or exceeding the magnitude of the original procedure occurred in 7% of cases (Figure 47-16). Total reoperation rates for syndromic and nonsyndromic synostoses were 27.3% and 5.9%, respectively. Five of the 12 reoperative cases (41.6%) were completed for significant relapse, as demonstrated clinically and radiographically. Neither early nor late sagittal strip craniectomies required reoperations; only one patient in the late cranial vault remodeling group for sagittal synostosis (>7 months) demonstrated relapse and required reoperation. Relapse was seen in two patients with bicoronal synostosis, requiring reoperation, and two more patients in this group underwent a second procedure for suboptimal cranial contouring. The rates of total reoperation significantly differed by gender (Fisher exact test \(p = .029\)). Of the female patients, 13.8% required reoperation compared with 3.8% of males.

The highest reoperative rate in nonsyndromic children was found in bicoronal synostosis. Of children with single-suture synostosis, reoperative rates were highest in those with sagittal suture fusion requiring total vault remodeling (6.45%). Reoperative rates in metopic synostosis were 2.86% with an average follow-up of 42 months. No statistical significance was found in reoperative rates for the various single-suture synostoses.

Multiple regression analysis revealed female patients and children with syndromic synostoses were more likely to require reoperation (Table 47-3). The reason for the increased odds ratio associated with female patients is unknown. Age did not appear to have an effect on reoperation rates. A 1-year increase in age at operation was not statistically significant after controlling for diagnostic group. The effect on reoperation rate did not change. In estimated blood loss was not significant after controlling for diagnostic group. Similarly, length of hospital stay, length of surgery, intensive care unit admission, and the amount of transfusion were not statistically significant after controlling for diagnostic category.

Whitaker's classification\(^{97}\) of clinical results after a craniofacial procedure includes Category III (C-III), requiring major bone grafting or other osteotomies, and Category IV (C-IV), requiring duplication of the previous craniofacial procedure. Total reoperations in our review were classified into the latter group. The increase in reoperation rates of syndromic were consistent with previous reviews. Whitaker\(^{96-97}\) showed a C-IV reoperation rate of 3% for asymmetric lesions (isolated synostosis) and 64% for the symmetric lesions (95% in Apert's syndrome). Excluding strip craniectomies, McCarthy had a 6.7% reoperation rate for isolated synostosis\(^{93}\) and 28.3% for syndromic deformities.\(^{96}\) Surgical approaches similar to ours were used in each of the aforementioned studies.

In most studies, no differences were seen in reoperative rates for treatment of single-suture synostosis as related to age (Table 47-4).\(^{24,73}\) recommendations for primary intervention ranged from 2 to 18 months. Wall et al\(^{95}\) showed an increased reoperation rate of 20% in nonsyndromic synostosis when primary treatment was in patients less than 6 months of age compared with 5.6% in patients greater than 6 months old. In syndromic disorders, patients less than 6 months old had a 30.2% reoperation rate compared with 9.1% when older than
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Table 47-3. Multivariable Analysis of Total Reoperation Rates

<table>
<thead>
<tr>
<th>CONTROL VARIABLE</th>
<th>SYNDROMIC VERSUS NONSYNDROMIC</th>
<th>SAGITTAL VAULT VERSUS OTHER</th>
<th>ONLY CONTROL VARIABLE (p value)</th>
</tr>
</thead>
<tbody>
<tr>
<td>None</td>
<td>6.0</td>
<td>1.1</td>
<td>-</td>
</tr>
<tr>
<td>Female gender</td>
<td>5.9</td>
<td>1.3</td>
<td>3.9 (0.04)</td>
</tr>
<tr>
<td>Age at surgery (1 yr)</td>
<td>9.7</td>
<td>0.7</td>
<td>0.6 (0.19)</td>
</tr>
<tr>
<td>Blood loss (each 100 units)</td>
<td>6.4</td>
<td>0.4</td>
<td>1.1 (0.31)</td>
</tr>
<tr>
<td>Length of stay (days)</td>
<td>1.3</td>
<td>0.5</td>
<td>1.2 (0.07)</td>
</tr>
<tr>
<td>Length of surgery (each 100 minutes)</td>
<td>5.2</td>
<td>0.5</td>
<td>1.3 (0.53)</td>
</tr>
<tr>
<td>Months of follow-up</td>
<td>5.7</td>
<td>1.1</td>
<td>1.1 (0.74)</td>
</tr>
<tr>
<td>Any ICU stay</td>
<td>9.1</td>
<td>1.7</td>
<td>0.6 (0.59)</td>
</tr>
<tr>
<td>Any transfusion</td>
<td>6.3</td>
<td>0.5</td>
<td>2.3 (0.46)</td>
</tr>
<tr>
<td>Any complications</td>
<td>4.2</td>
<td>0.9</td>
<td>5.7</td>
</tr>
<tr>
<td>Early complications</td>
<td>4.4</td>
<td>0.8</td>
<td>2.4</td>
</tr>
<tr>
<td>Late complications</td>
<td>5.0</td>
<td>1.4</td>
<td>6.2</td>
</tr>
</tbody>
</table>


Table 47-4. Age-Related Reoperation

<table>
<thead>
<tr>
<th>TOTAL REOPERATIONS</th>
<th>YES</th>
<th>NO</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of patients</td>
<td>11</td>
<td>144</td>
</tr>
<tr>
<td>Mean age of initial operation</td>
<td>0.57 yr</td>
<td>1.0 yr</td>
</tr>
<tr>
<td>Standard deviation</td>
<td>0.52 yr</td>
<td>1.27 yr</td>
</tr>
<tr>
<td>Standard error</td>
<td>0.16 yr</td>
<td>0.11 yr</td>
</tr>
</tbody>
</table>


6 months (40.9-month follow-up). Other reviews have shown no relationship of age to reoperative rates.99

FUNCTIONAL OUTCOME

Increased Intracranial Pressures

The first major study attempting to measure ICP in children with craniosynostosis was by Renier et al.77 Defining normal ICP as below 10 mm Hg and elevated as above 15 mm Hg, ICP were recorded preoperatively by an epidural sensor for 12 to 24 hours. Of 92 patients, %14% with nonsyndromic suture fusion had evidence of elevated ICP. After surgery, the ICP decreased to within normal limits in all but 7% of patients (Table 47-5). The review eventually consisted of 358 patients.76 Using Renier's criteria for normal and abnormal levels of ICP, Thompson44 also found 12.9% of patients with single-suture fusion, and 57% of patients with nonsyndromic multiple suture fusion, to have elevated ICP (Table 47-6). The latter study used subdural monitoring devices, which are thought to be more accurate.

The association between increased ICP and craniosynostosis is well established, but a direct corollary between the two is not clear in every patient.44,100 Recent advances in imaging techniques have provided a method of intracranial volume analysis that demonstrates a more complicated relationship between craniosynostosis and ICP. Several studies have demonstrated that suture fusion does not always cause a decrease in the volume of the cranial vault in nonsyndromic suture fusion.31,34,72 Furthermore, studies using intracranial-invasive monitoring did not find a consistent correlation between intracranial hypertension and decreased cranial volume. In one study, 13 of 66 patients were found to have elevated ICP, of whom 12 had decreased intracranial volume. In contrast, only 5 of 13 patients with the most severe reduction in volume demonstrated intracranial hypertension (above 15 mm Hg).30

Late postoperative increases in ICP were seen in 2% of patients at our institution. One patient developed elevated ICP
Table 47-5.
Baseline Intracranial Pressure in Patients Subdivided according to Skull Shape and Syndrome, Used in This Study

<table>
<thead>
<tr>
<th>TYPE</th>
<th>(\leq 10) mm Hg</th>
<th>10-15 mm Hg</th>
<th>(\geq 15) mm Hg</th>
</tr>
</thead>
<tbody>
<tr>
<td>31 Trigono</td>
<td>21 (28%)</td>
<td>8 (26%)</td>
<td>2 (6%)</td>
</tr>
<tr>
<td>118 Scapho</td>
<td>76 (64%)</td>
<td>33 (28%)</td>
<td>9 (8%)</td>
</tr>
<tr>
<td>65 Plagio</td>
<td>40 (62%)</td>
<td>17 (26%)</td>
<td>8 (12%)</td>
</tr>
<tr>
<td>34 Brachy</td>
<td>17 (50%)</td>
<td>8 (24%)</td>
<td>9 (26%)</td>
</tr>
<tr>
<td>66 Oxy</td>
<td>23 (35%)</td>
<td>7 (11%)</td>
<td>36 (54%)</td>
</tr>
<tr>
<td>9 Crouzon</td>
<td>3</td>
<td>0</td>
<td>6</td>
</tr>
<tr>
<td>16 Apert</td>
<td>3</td>
<td>6</td>
<td>7</td>
</tr>
</tbody>
</table>


Table 47-6.
Elevated Intracranial Pressures in Craniosynostosis*

<table>
<thead>
<tr>
<th>STUDY</th>
<th>SINGLE SUTURE FUSION % (n)</th>
<th>MULTIPLE SUTURE FUSION % (n)</th>
<th>CROUZON'S DISEASE % (n)</th>
<th>APERT'S SYNDROME % (n)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Thompson (1995)*</td>
<td>12.9 (8/62)</td>
<td>57.0 (12/21)</td>
<td>65.0 (13/20)</td>
<td>38.5 (5/13)</td>
</tr>
<tr>
<td>Gault (1992)*</td>
<td>3.9 (2/51)</td>
<td>71.4 (8/11)</td>
<td>67.0 (2/3)</td>
<td>100.0 (1/1)</td>
</tr>
<tr>
<td>Renier (1989)*</td>
<td>8.9 (19/24)</td>
<td>45.0 (45/100)</td>
<td>66.7 (6/9)</td>
<td>43.8 (7/16)</td>
</tr>
<tr>
<td>Renier (1982)*</td>
<td>13.5 (5/37)</td>
<td>43.3 (13/30)</td>
<td>100 (2/2)</td>
<td>50.0 (3/6)</td>
</tr>
</tbody>
</table>

*All studies used 15 mm Hg as upper limit of normal pressure.

Requiring decompression after a cranial vault remodeling. Recurrent intracranial hypertension after previous corrective surgery has been reported and requires a secondary cranial vault remodeling. 18,77

Mental Development

Studies on the effects of isolated craniosynostosis on mental development are difficult to interpret because of differences in the definitions of mental delay, variability of inclusion criteria, and differences in testing methods. Two questions exist: (1) does nonsyndromic synostosis cause a decrease in mental development? and if so, (2) does surgical intervention prevent the delay in mental development? Earlier studies have reported a prevalence of mental delay ranging from 0% to 20% in metopic synostosis, 2,4,5,8,10% to 60% in sagittal synostosis, 1,10,25,58,69 and 0% to 27% in coronal synostosis. 4,10,59,62

Recent studies elucidating the relationship of craniosynostosis and mental development have been conflicting. Recognizing the problems with the definitions of normal and abnormal ICP, Renier et al 76 obtained ICP (using an epidural sensor) in 358 patients with various types of craniosynostosis. Some 232 patients with nonsyndromic suture fusion underwent intelligence testing. Excluding trigonocephaly, more than 90% of the patients with single-suture fusion tested within the normal range. The proportion was lower in trigonocephaly (83%) because of the presence of syndromic anomalies. Patients with multiple-suture fusion had a lower number of patients in the normal range (brachycephaly 78%, oxycephaly 69%). Also interesting was the overall decrease in the number of normal patients over 1 year old in the normal range compared with patients less than 1 year old. When stratified for ICP, patients with scaphocephaly did not show a decrease in the proportion
of normal intelligence associated with elevated ICPs. In unicoronal plagiocephaly the percentage of patients testing within the normal range decreased from 97% to 86%, and the decrease was even more dramatic in patients with multisuture fusion.

A longitudinal study by Kapp-Simon et al.44 following mental development in patients with nonsyndromic suture fusion failed to confirm these findings. A review of 45 patients demonstrated no differences in terms of mental development between patients with synostosis and a normal control group. No decrease in mental development when analyzed for suture type, single-suture versus multiple sutures, timing of repair, or severity of deformity (4-point severity scores from patient and radiographic review) were found. Furthermore, no differences were noted between patients in the operative group and the nonoperative group. Mental developmental scores were actually higher in patients with multiple-suture synostosis compared with single-suture fusion. The major criticism of this study was the low number of patients who were assessed.75

Improvement in behavior has been reported with resolution of papilledema as an indicator of decreased ICP.10 Using "monitor-proven" decreases in ICP after surgery, Renier et al did not conclusively demonstrate an improvement in intelligence compared with nonoperated controls.76 It was concluded that the effective decrease in ICP after the operative intervention could possibly arrest the deterioration in intelligence. Therefore surgical intervention should be within the first year of life (during the maximal growth conflict stage). In contrast, Kapp-Simon found no improvement in mental development of patients with surgical intervention compared with the nonoperative group.44

Aesthetics

Recognizing the potential functional complications associated with nonsyndromic synostosis, operative intervention is usually undertaken because of aesthetic concerns. The acquired deformity of the various synostoses has been discussed previously. The importance of appearance in social interactions and the social advantages associated with attractiveness have been established. Few studies have attempted to clarify the impact of congenital malformation on psychosocial well-being. Pereshuk and Whitaker29 evaluated the psychosocial impact of congenital craniofacial malformations in two groups of children between ages 6 and 13, categorized by having early surgery (before age 4) or later surgery (after age 4). The earlier group demonstrated "no differences" to matched healthy subjects regarding psychosocial adjustment. In contrast, the later group expressed poorer self-concept, greater anxiety, more problematic behaviors, and more introversion. The psychosocial adjustments in this group appeared "limited." A final group of adolescent adult patients with uncorrected craniofacial malformations underwent similar analysis and was found to have obvious disturbances in social adjustment and self-concept. The analysis was not stratified for specific disorders, but general conclusions regarding the importance of improving appearance for psychosocial adjustment were demonstrated.

Standardized clinical assessments of aesthetic outcome in cranial vault remodeling are limited. Wagner et al.93 reviewed 22 patients with bicornal synostosis who underwent cranial vault remodeling. Using the grading system proposed by Whitaker et al, 45% (10) were judged to have satisfactory results, 14% were classified as grade II requiring minor revisions, and 41% required major reoperation rates (Tables 47-7 and 47-8). In a similar study,55 17 patients with trigonocephaly requiring cranial vault remodeling underwent review using a similar grading scale. Some 53% of the patients were found to have none or mild deformities (grade I), 35% had moderate deformities (grade II), and 12% had severe deformities requiring major reoperation (grade III).

**Table 47-7.**

<table>
<thead>
<tr>
<th>GRADE</th>
<th>NO. PATIENTS</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>10</td>
<td>45</td>
</tr>
<tr>
<td>II</td>
<td>3</td>
<td>14</td>
</tr>
<tr>
<td>III</td>
<td>9</td>
<td>41</td>
</tr>
<tr>
<td>TOTAL</td>
<td></td>
<td>100</td>
</tr>
</tbody>
</table>

FOA, Frontal orbital advancement; CVR, calvarial vault remodeling.


**Table 47-8.**

<table>
<thead>
<tr>
<th>AGE AT INITIAL FOA-CVR</th>
<th>GRADE I</th>
<th>GRADE II</th>
<th>GRADE III</th>
<th>TOTAL</th>
</tr>
</thead>
<tbody>
<tr>
<td>≤5 mo</td>
<td>5</td>
<td>0</td>
<td>8</td>
<td>13</td>
</tr>
<tr>
<td>≥6 mo</td>
<td>5</td>
<td>3</td>
<td>1</td>
<td>9</td>
</tr>
</tbody>
</table>

FOA, Frontal orbital advancement; CVR, calvarial vault remodeling.


Risk Stratification for Reoperation—Quantitative CT Scan Analysis

Current diagnosis and surgical correction of craniofacial anomalies benefited from accurate quantitative CT analysis as described by Waitzman and Posnick.94 Clinically useful growth information may be obtained, and the treating clinician may calculate preferred osteotomy movements to be achieved at operation. The intraoperative execution of the calculated changes at operation can be verified by analysis of postoperative CT scans.
Note: This pathway is a tool to be used in progressing the patient through a hospitalization. This document is maintained as a part of the chart during hospitalization only. It is not to be considered as a prescription for services, but rather as a guide. **THE PATHWAY IS NOT TO BE A PART OF THE PERMANENT RECORD.**

**Instructions:** Record the appropriate variance type (coded as: A = Practitioner, B = Patient, C = System, D = Other) in the blanks provided. Check marks are not acceptable. For each variance, document the cause of the variance in the table at the bottom of each page with the date, day, variance type (again), and your signature. **NOTE:** Blanks are provided for documentation of variances only. If there are no variances, check the box for no variances. In addition, when the review for the day is complete, check the box for review completed and sign as reviewer.

<table>
<thead>
<tr>
<th>Hospital Day</th>
<th>Dx/Tx Measures</th>
<th>Medications</th>
<th>Nursing Services</th>
<th>Anesthesia/Resp. Care</th>
<th>Nutrition/Diet</th>
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<td>CBC</td>
<td></td>
<td>Anesthesia preop</td>
<td>Anesthesia Assessment:</td>
<td>NPO as instructed</td>
</tr>
<tr>
<td></td>
<td>Type and cross-</td>
<td></td>
<td></td>
<td>History</td>
<td></td>
</tr>
<tr>
<td></td>
<td>match one unit</td>
<td></td>
<td></td>
<td>Physical exam</td>
<td></td>
</tr>
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<td></td>
<td>of PRBC</td>
<td></td>
<td></td>
<td>Respiratory/cardiac</td>
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<td></td>
<td>UA</td>
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<td></td>
<td>assessment</td>
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</table>

☐ No variances
☐ Review completed
Reviewed By:

☐ No variances
☐ Review completed
Reviewed By:

☐ No variances
☐ Review completed
Reviewed By:

☐ No variances
☐ Review completed
Reviewed By:

**Variation Types:**  A = Practitioner  B = Patient  C = System  D = Other

<table>
<thead>
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<th>DATE</th>
<th>DAY</th>
<th>VARIANCE TYPE</th>
<th>VARIANCE/CAUSE</th>
<th>SIGNATURE</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
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</tr>
</tbody>
</table>

**Figure 47-17.** Sagittal synostosis critical pathway. Strip craniectomy patients only.
We recently performed a quantitative CT scan analysis in patients with metopic synostosis to determine if preoperative findings are predictive of the need for reoperation. In 35 children with metopic synostosis, measurements of the cranial length, cranial width, anterior intercoronal distance, anterior interorbital distance (intercanthal distance), lateral orbital distance, and interzygomatic buttress distance were taken from the preoperative CT scans and normalized relative to each child's age. Abnormal measurements were defined as those that fell outside of the age-appropriate mean by greater than or equal to 5% of the mean. To separate overall facial hypoplasia from regional hypoplasia, a ratio of intercanthal distance to interzygomatic buttress distance was determined. These prognostic factors were analyzed using chi-squared or Fisher's exact test with respect to length of hospital stay, transfusion requirement, postoperative complications, and reoperation rate. Some 29% of the children underwent reoperation (5% total reoperation and 24% contouring) for correction of residual contour deformities. All of the reoperations occurred in children with abnormally small intercanthal distance ($p = 0.16$). The ratio of intercanthal distance to midfacial width was significantly related to reoperation rate with those children who had a ratio of less than or equal to 0.80 having a reoperation rate of 43.8% ($p = 0.05$). This relationship suggests that the preoperative CT scan measurements can be used as means of risk stratification in outcome analyses of the surgical treatment of craniosynostosis. In children treated for metopic synostosis, a foreshortened intercanthal distance compared with interzygomatic buttress distance was significantly related to reoperation rate, particularly in those children who were under 12 months of age when treated.

Economic Issues

Critical pathways are case management mechanisms. From 1994 to 1995, Scottish Rite Children's Medical Center adopted a variety of critical pathways. Patients undergoing sagittal synostectomies were included in one such critical path. In an environment in which multiple clinical disciplines provide patient care services, case management through the use of critical paths is one of the more efficient ways to ensure continuity of care. Critical pathways are class project-management and industrial-engineering tools that are simple, visual, and two-dimensional. They plot time along one axis and staff actions-tasks, interventions, and orders along another to form a "schedule of events within time periods." The pathway development model Scottish Rite adopted is an integrated model developed by interdisciplinary teams and supported by the quality improvement structure. The critical path is a clinical tool for achieving better quality and cost outcomes by outlining and sequencing the usual and/or desired care for particular groups of patients. At its core, the critical path is a communication tool, facilitating coordination among a wide variety of clinicians and departments. As Bruce Campbell, Director of Medical Liaison at Scripps Memorial Hospital in San Diego, observed, "Care is so complex now, and variable from patient to patient, that if the essential components of care are not "blueprinted," they are either forgotten or not done on time." As a result, patient outcomes are jeopardized. Critical paths create a matrix that profiles interventions on one axis and time on another. Second-generation critical paths, which are called care maps, incorporate usual patient problems, desired clinical outcomes, and intermediate goals into the matrix. Critical paths are generally used in high-volume, high-cost, high-risk, or high-interest patient populations. They involve physicians in all stages of critical path development. The variances for critical pathways are used to stimulate discussion among practitioners and to identify opportunities to improve organizational systems and care practices.

Economic issues are discussed in terms of hospitalization costs rather than surgical fees. Every effort has been made with the individual surgeon's fees to remain competitive. At our center, discussions are underway to develop package pricing for craniosynostosis services. The cost of hospitalization for nonsyndromic craniosynostosis patients was assessed through a biopsy of the hospital bills of 197 patients operated on from 1988 to 1996 at our institution. The costs range from $5130.30 to $120,121. The mean costs range from $52,381 in 1990 to $14,225 in 1996. In 1990 and 1994, costs exceeded $100,000 in two patients who had suffered complications. The patients' diagnoses included sagittal, bicoronal, uniconoral, and metopic synostoses. After surgery the presence or absence of an early complication was noted. When an early complication occurred, the mean hospital charge was $75,995 compared with $22,293 ($p = 0.03$).

In 1995 a critical pathway was introduced for patients undergoing sagittal synostectomies. Figure 47-17 shows an example of the sagittal synostectomy critical pathway. Variances in variation types are recorded. Variation types may be related to the practitioner, patient, system, or other. Although the number of observations is small, the mean cost of hospitalization dropped substantially, but numbers are still too small for meaningful statistical analysis.

REFERENCES


