Upper Facial Asymmetries Resulting from Unilateral Coronal Synostosis

Diagnosis and Surgical Reconstruction

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Virchow coined the term craniosynostosis and formulated the classic theory known as Virchow's Law.22 This states that premature fusion of a cranial vault suture (craniosynostosis) inhibits normal skull growth perpendicular to the fused suture and leads to a compensatory growth at the open sutures.

The birth prevalence of craniosynostosis is about 343:1 million.3 Most cases of simple (isolated) craniosynostosis are sporadic. Syndromic craniosynostosis usually is genetic and may be autosomal dominant or autosomal recessive. The term craniofacial dysostosis is used in a general way to describe familial forms of craniosynostosis involving not only the cranial vault but also various cranial-base and midface sutures (i.e., Crouzon syndrome, Apert syndrome, Saethre-Chotzen syndrome, Pfeiffer syndrome, and Kleeblattschädel anomaly).11,21

The treatment of craniosynostosis and the craniofacial dysostosis syndromes is surgical, but the specific indications for surgery and the timing, type, and effectiveness of reconstruction have not always been well evaluated.1,5,7,9,10,12,17,18,21,23-25 Unilateral coronal synostosis results in extensive upper face asymmetry. This article describes a philosophy and rationale for the treatment of this frequent craniofacial malformation.

CLASSIFICATION SYSTEMS

The classification used most commonly for craniosynostosis is based on the shape of the skull, which in general reflects the underlying prematurely closed suture(s).3 The major cranial vault sutures that are involved include left and right coronal (ante-
rior plagiocephaly); metopic (trigonocephaly); sagittal (scaphycephaly); left and right lambdoidal (posterior plagiocephaly); bilateral coronal (anterior brachycephaly); and bilateral lambdoidal (posterior brachycephaly). Craniosynostosis should be documented both clinically and radiographically and not be confused with either external postural or positional skull molding problems that frequently result in nonsynostotic anterior or posterior plagiocephaly.2,10

FUNCTIONAL CONSIDERATIONS

Elevated Intracranial Pressure, Brain Growth, and Hydrocephalus

Intracranial volume in the normal child almost triples within the first year of life. By 2 years the cranial capacity is four times that at birth. At 5 years of age the cranial capacity approaches 90% of its adult size. For this rapid brain growth to proceed unhindered the open cranial vault and base sutures must spread during phases of rapid growth, resulting in marginal ossification.

Elevated intracranial pressure is the most worrisome functional problem associated with premature suture fusion. Its late and devastating effect is documented indirectly on plain radiographs as the "copper beaten" or "finger printing" appearance along the inner table of the cranial vault and base. When raised intracranial pressure goes untreated brain function is affected.

Intracranial hypertension can be documented invasively by means of a craniotomy used to place an epidural pressure sensor or by lumbar puncture monitoring. When using standard measurements as indications of significantly elevated intracranial pressure (greater than 15 mm of mercury), Renier et al15 determined that 14% of untreated children with just one prematurely fused suture fell into this category. The corollary is that 86% of their study group did not demonstrate increased intracranial pressure despite documentation of craniosynostosis. It therefore must be acknowledged that a specific head shape correlated with radiographic findings of synostosis is not definitive for decreased intracranial volume or increased intracranial pressure. A clear understanding of the relationship between intracranial volume and intracranial pressure in patients with craniosynostosis continues to elude clinicians.16

Hydrocephalus affects a small percentage of patients with craniosynostosis. The cause is not always clear but a high level of suspicion for hydrocephalus should be maintained. Early diagnosis and consequent ventriculoperitoneal shunt placement is indicated.

Effects on Vision

Divergent or convergent nonparalytic strabismus or exotropia occurs frequently in unilateral coronal synostosis. This may be the result of anomalies of the extraocular muscles themselves or of orbital dystopia.11

If increased intracranial pressure does occur, and is left untreated, papilloedema and eventual optic atrophy develops, resulting in partial or complete blindness. An accurate fundoscopy examination is useful in the documentation of these findings.

Morphologic Pattern in Unilateral Coronal Synostosis

Unilateral coronal synostosis results in an anterior plagiocephalic shape to the cranial vault.3 With premature closure of the coronal suture a frontoparietal bone plate is formed with restricted growth potential.4 Open adjacent cranial vault sutures compensate in growth more than those sutures not contiguous with the synostotic one. En-
hanced symmetric bone deposition occurs along both sides of the contralateral (open) coronal suture.

In anterior plagiocephaly resulting from a unilateral coronal synostosis, the ipsilateral palpebral fissure is widened, the supraorbital rim and eye are displaced superiority and posteriorly, the ipsilateral ear is higher and anteriorly located, and the root of the nose is deviated toward the flattened side. The anterior fontanelle and chin are both displaced to the contralateral side.

When viewing an anteroposterior plain cranial radiograph in a unilateral coronal synostosis patient characteristic features include a "harlequin" deformity owing to ipsilateral superior displacement of the lesser wing of the sphenoid; deviation of the nasal root toward the synostotic side; and absence of the normal coronal sutural radiolucency. A three-dimensional cranial-base CT scan view documents angulation of the midlines of the anterior and posterior cranial bases toward the side of unilateral coronal synostosis.

**RECONSTRUCTIVE OBJECTIVES**

The achievement of symmetry, proportionality, and balance with the restoration of the normal orbital aesthetic units is essential to forming an unobtrusive face in the child born with unilateral coronal synostosis. The frontoforehead region is the central area of dysmorphology in this malformation. The establishment of a normally positioned forehead is critical to overall facial symmetry and balance. The forehead may be divided into two separate aesthetic aspects: (1) the supraorbital ridge and (2) the superior forehead. The supraorbital ridge includes the glabella region and supraorbital ridge, and extends laterally and inferiorly down each frontozygomatic suture and posteriorly along each temporoparietal region. In the normal forehead, at the level of the frontonasal suture, an angle ranging from 90 to 110 degrees is formed by the supraorbital ridge and the nasal bones when viewed in profile. The eyebrows, overlying the supraorbital ridge, should be anterior to the cornea. The supraorbital rim should arc posteriorly to achieve a gentle 90-degree angle at the temporal fossa with a center point of the arc at the level of each frontozygomatic suture. The superior forehead begins approximately 1 cm above the supraorbital rim. It has a gentle posterior curve of about 60 degrees and levels off at the coronal suture region when seen in profile.

**SURGICAL PERSPECTIVES**

The first recorded surgical approach to craniosynostosis was by Lannelongue in 1890. He completed a strip craniectomy of the involved suture. His aim was to limit brain compression (intracranial hypertension) within a constricted cranial vault. For the treatment of unilateral coronal synostosis, this strip craniectomy approach was favored until the mid-1970s.

In 1976, Hoffman and Mohr proposed the "lateral canthal advancement" procedure for this form of craniosynostosis. In addition to a frontal craniectomy, they completed a limited unilateral superior and lateral upper orbital advancement. The "lateral canthal advancement" procedure gave superior results compared with the strip craniectomy, but anatomic-symmetric correction rarely was achieved. In 1977, Whitaker et al proposed a more formal cranial vault and orbital reshaping procedure for unilateral coronal synostosis. They completed a more extensive orbital osteotomy, stabilizing the advanced supraorbital bar with a tongue-in-groove technique and reshaping the forehead bone and stabilizing with interosseous wire fixation. McCarthy et al further modified the unilateral fronto-orbital advancement by removing the sphenozygomatic suture at the cranial base. In 1979, Marchac and Renier suggested that, although the craniosynostosis malformation was unilateral, a bilateral deformity resulted that required a bilateral operation. They removed the supraorbital bar bilaterally, reshaping the segment for symmetry and replacing it loosely to the residual or-
bits. A segment of cranial vault bone was reshaped and secured to the newly repositioned supraorbital bar with interosseous wires. The “free-floating” supraorbital ridge and forehead unit was to allow the now unrestricted brain to push the forehead and upper orbits forward with the filling in of lateral bony defects in the temporal and upper forehead regions. Unfortunately, the large open areas often reossified with temporal and upper forehead constrictions or regions of prominent bulging of the cranial vault.

**THE AUTHOR’S APPROACH AND SURGICAL TECHNIQUES**

When a newborn is noted to have cranial vault dysmorphology, early referral to a craniofacial surgeon should be carried out. If the craniofacial surgeon’s examination is suspicious for craniosynostosis then the child also should undergo an evaluation by a pediatric neurosurgeon, pediatric ophthalmologist, and neuroradiologist including a complete two- and three-dimensional CT scan of the craniofacial complex.

If an isolated form of unilateral coronal synostosis is documented both clinically and radiographically, the degree of dysmorphology is significant. Without evidence of increased intracranial pressure, however, my recommendation for the timing of surgical intervention generally is when the child is 10 to 12 months of age. Whereas some surgeons feel that early surgical intervention (between 2 and 9 months of age) is an advantage because “rapid ongoing brain and cranial vault growth will improve on the surgical results initially achieved,” this hope has not been borne out through scientific analysis. By postponing surgical intervention until 10 to 12 months of age, a greater percentage of the child’s overall brain and cranial vault will have occurred and the surgical improvements achieved are less dependent on ongoing growth that potentially distorts the initial satisfactory results.

Although some technical aspects of the reconstruction are dependent on individual morphologic variation, in general I use a standardized surgical approach for the reconstruction of unilateral coronal synostosis (Fig. 1). It involves suture release as well as simultaneous anterior cranial vault and bilateral three-quarter orbital osteotomies with reshaping and repositioning. The reshaped orbital and forehead units are replaced and stabilized with microbone plates and screws or interosseous wires. Lateral canthopexies are completed and the wounds are closed in two layers over suction drains (Fig. 2).
Figure 2. An 8-month-old girl born with right unilateral coronal synostosis resulting in anterior plagiocephaly. 
A. Preoperative frontal view. B. Frontal view 2 years later. C. Preoperative bird's eye view. D. Worm's eye view 2 years later.

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Figure 2 (Continued). E, Bird’s-eye view of removed orbital osteotomy unit prior to reshaping. F, Bird’s-eye view of orbita osteotomy unit after reshaping. G, Bird’s-eye view of anterior cranial vault prior to reshaping. H, Bird’s-eye view of anterior cranial vault after osteotomies, reshaping, and fixation of bone segments. Illustration continued on following page.
Figure 2 (Continued). I. Comparison of standard three-dimensional CT scan view of cranial base before and after procedure. (From Posnick JC: Unilateral coronal synostosis (anterior plagiocephaly): Current clinical perspectives. Ann Plast Surg, in press; with permission.)

COMPLICATIONS

The complications associated with unilateral coronal synostosis result either from neglect or surgical intervention. The problems that result from lack of intervention relate to increased intracranial pressure, impairment of vision, and poor self-esteem.

Potential morbidity associated with the surgical procedures include neurologic injuries, ophthalmologic injuries, cardiopulmonary injuries, infection, and inadequate early or late morphologic improvements. When the procedures are carried out by an experienced craniofacial surgeon and team the incidence of operative complications is low.17,11,25

MANAGEMENT OF SECONDARY DEFORMITIES AND NEGLECTED PATIENTS

Despite initial primary intracranial procedures, secondary deformities may present that require further surgical intervention (Fig. 3). When a second intracranial procedure is required to further reshape the cranio-orbital region additional morbidity relates to brittle cortical bone, previously placed and imbedded fixation devices, dural invaginations within the inner table of the cranial vault, and soft-tissue scarring.15 Approximately 5% to 10% of infants undergoing their initial cranio-orbital reshaping will benefit from a second cranio-orbital procedure carried out later in childhood.

Some affected children occasionally are allowed to grow into adulthood without undergoing surgical intervention (Fig. 4). Evaluation of several untreated adult patients born with unilateral coronal synostosis has resulted in a personal observation that there is little if any worsening of the deformity over time. I believe that unilateral coronal synostosis represents a nonprogressive craniofacial asymmetry rather than a worsening skeletal malformation.
Figure 3. A 4-year-old boy born with a right unilateral coronal synostosis underwent a "strip" craniectomy procedure when he was 10 weeks of age. His anterior plagiocephalic morphology remained and he arrived for management of his residual cranio-orbital deformities. 
A. Frontal view prior to cranio-orbital procedure. 
B. Frontal view after undergoing anterior cranial vault and three-quarter orbital osteotomies with reshaping.  
C. Bird's-eye view just prior to cranio-orbital procedure.  

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Figure 3 (Continued). D, Three-dimensional bird's eye CT scan views indicating residual anterior plagiocephaly after strip craniectomy procedure earlier in life. E, F. Bird's-eye view of cranial vault just prior to and after osteotomies, reshaping, and fixation of bone segments.
Figure 3 (Continued). G, Comparison of standard axial-sliced CT scan images through cranial vault prior to and after cranio-orbital procedure. H, Comparison of standard three-dimensional CT scan view of cranial vault prior to and after cranio-orbital procedure. (From Posnick JC: Unilateral coronal synostosis (anterior plagiocephaly): Current clinical perspectives. Ann Plast Surg, in press; with permission.)
When reconstructing the neglected, older, unilateral coronal synostosis patient the procedure carried out is similar to that undertaken in the young child. Differences relate to the necessity of closing all surgically created skull defects, the greater reliance on stable fixation techniques, and the management of the fully developed frontal sinus.

CONCLUSION

Unilateral coronal synostosis results in a consistent pattern of upper facial asymmetry. The craniofacial malformation affects both sides of the upper face, necessitating a bilateral reconstruction. Confusion may arise when postural or skull molding deformations are confused with unilateral coronal synostosis. The morphologic differences between the two are obvious to the astute clinician and important because craniostenosis requires surgery, whereas skull molding is treated with parental education and observation.

Details of the preferred timing and techniques for the morphologic correction of unilateral coronal synostosis are suggested. The author has found them to give consistent results.

References


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