

Pitfalls for the Pediatrician: Positional Molding or Craniosynostosis?

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As many as one-quarter of single births and more than half of twins are born with abnormal head morphology.³ A variety of causes, including intrauterine crowding, passage through the birth canal and vacuum- or forceps-assisted delivery, have been implicated in these cranial deformations.⁴ The majority of these infants, however, will have complete resolution by age 2, with normal development of the craniofacial skeleton.⁵

Craniosynostosis, on the other hand, does not resolve, and early recognition and surgical correction is essential for a satisfactory outcome. Beginning in the early 1990s with the American Academy of Pediatrics recommendation to place infants on their backs to sleep to decrease the risk of sudden infant death syndrome, there has been a dramatic increase in the incidence of occipital plagiocephaly, or positional molding. In the majority of cases, the skull deformity is mild, a diagnosis of positional molding is made easily, and appropriate treatment is initiated rapidly. On occasion, however, pediatricians may encounter difficult situations that are resistant to traditional treatments, leading them to question the diagnosis and seek subspecialty opinions.

This article provides an overview of the nomenclature, epidemiology, and etiology of craniosynostosis, including a review of the literature in these areas. The most common types of craniosynostosis are discussed, as well as appropriate treatment strategies for each. Finally,

diagnostic criteria distinguishing craniosynostosis from positional molding and the use of helmet therapy are reviewed in detail.

DEFINITION AND HISTORY

Craniosynostosis is the premature fusion of one or more sutures of the skull. It may occur as an isolated primary abnormality, as part of a syndrome, or secondary to a systemic disorder. Craniosynostosis has many different classification systems. Perhaps the most useful is to distinguish simple, or nonsyndromic, craniosynostosis from complex, or syndromic, craniosynostosis. More than 150 different syndromes described by geneticists involve craniosynostosis, but most are esoteric and rarely seen by pediatricians. Therefore, this report will focus on simple craniosynostosis, which is the most common type. Syndromic and secondary forms of craniosynostosis (eg, from metabolic disorders, microcephaly, and others) will not be discussed.

As early as 1791, Sommering¹ recognized the role of premature calvarial suture fusion in the pathogenesis of primary craniosynostosis. Several years later, Virchow² further advanced the concept and characterized the dysmorphic sequelae of individual suture fusion. Cranial growth occurs as a direct result of the growing brain. An infant's brain doubles in volume in the first 6 months and again by age 2. If all cranial sutures are open, the skull allows the brain to grow normally. Once a suture becomes fused, however, growth perpendicular

to that suture becomes restricted and compensatory growth occurs at the remaining open sutures. Premature suture fusion may involve the sagittal, metopic, coronal (unilateral or bilateral), or lambdoid (unilateral or bilateral) suture (Figure 1, see page xxx).

There are six principal clinical manifestations of craniosynostosis, categorized predominately by the resulting head shape. As noted by Virchow, characteristic head shapes (named with Greek or Latin derivatives) will depend on which sutures are involved. Sagittal craniosynostosis results in scaphocephaly or a "boat shaped" head (also known as dolicocephaly), with increased anteroposterior length and decreased biparietal diameter. Metopic craniosynostosis presents as trigonocephaly or a "triangular" head, with a narrowed anterior cranial fossa. Unilateral coronal craniosynostosis causes anterior plagiocephaly or a "twisted head," with flattened ipsilateral forehead and ipsilateral fronto-orbital bar recession. Analogously, unilateral lambdoid craniosynostosis causes posterior plagiocephaly, with ipsilateral flattening of the occiput and contralateral occipital bossing. Bilateral coronal craniosynostosis results in anterior brachycephaly or a "short head" due to reduced anteroposterior length of the anterior cranial fossa. Multiple suture involvement results in more complex head morphologies such as Kleeblattschadel (cloverleaf skull) or oxycephaly (towering and cone-like cranium).

EPIDEMIOLOGY

The incidence of primary craniosynostosis has been reported to range from 0.3 to 1.4 per 1,000 births.¹¹⁻¹⁵ Much of the variability stems from differences in the diagnostic criteria used at individual centers, although an average incidence of 0.5 per 1,000 births is quoted widely. Multiple-suture craniosynostosis accounts for about 14% of all cases, while between 9% and 15% of cases are associated with craniofacial syndromes.⁸⁻¹⁰ There is no racial predilection for craniosynostosis, but individual suture involvement often presents with a strong gender bias. Sagittal craniosynostosis, for instance, is more frequent in males, who account for between 70% and 90% of cases. On the other hand, coronal craniosynostosis has a slightly higher incidence among females, who account for

between 55% and 70%.^{9,10} A series by Sloan et al.¹⁰ indicates that bilateral involvement of the coronal suture accounts for much of this female predilection, with 76% of bicoronal craniosynostosis occurring in females, as compared with the more modest 54% female majority in unicoronal craniosynostosis.

Primary involvement of the sagittal suture is the most frequently encountered single-suture craniosynostosis. Most reports indicate an occurrence of between 40% and 60% of all cases.¹⁴ Coronal craniosynostosis accounts for between 17% and 24% of cases.¹⁴ The percentage of craniosynostosis cases reported to involve the metopic suture is roughly 10%,^{16,17} although numbers as low as 4% and as high as 50% have been reported in the literature.^{8-10,14,18} Lambdoid suture involvement is seen in 0.9% to 4% of cases.^{19,20} One must keep in mind, however, that premature closure of a lambdoid suture accounts for less than 5% of posterior plagiocephaly cases, which is overwhelmingly more often positional in nature.^{19,20}

While the majority of craniosynostosis is sporadic, genetic patterns have been demonstrated in 8% of patients with coronal involvement and in 2% to 5% of those with sagittal involvement.²¹ Given the sporadic nature of single-suture craniosynostosis, it is difficult to predict accurately the subsequent risk, although it appears to double for siblings. In addition, a number of discrete genetic syndromes, namely Crouzon, Pfeiffer, and Apert syndromes, produce complex craniosynostosis. These variants, however, are much less common and account for less than 15% of all patients with craniosynostosis.

ETIOLOGY AND PATHOGENESIS

While a complete understanding of the etiology of craniosynostosis is unavailable, current theories implicate both environmental and genetic influences. The heterogeneous nature of cra-

niosynostosis is reflected, in part, by the diverse environmental factors that have been reported to be associated with its development. A small sample of these environmental factors includes maternal smoking, increased antenatal maternal altitude, and certain paternal occupations, such as agricultural and mechanic work.²²⁻²⁵ Most of these associations have not been confirmed with large-scale studies, and the mechanism by which these environmental factors predispose the fetus to develop craniosynostosis is unknown. The intrauterine environment also has been implicated in the development of craniosynostosis. The current thinking is that mechanical stress on the developing cranium due to intrauterine head constraint is causative in at least some cases.^{26,27}

At a cellular level, the neural crest-derived precursor cells of the cranial bones form discrete condensations by the middle of the first trimester.²⁸ As these condensations expand and ossify, sutures form at the leading edges of adjacent calvarial bones. A paradigmatic shift in our understanding of the pathogenesis of craniosynostosis emerged as it became appreciated that diffusible factors from the underlying dura are necessary for this initial formation and maintenance of sutures.²⁹ Expansion of the cerebral hemispheres likewise is essential to the maintenance of suture patency. Thus, microcephaly predictably leads to early suture closure whereas hydrocephalus prolongs suture patency. Studies have shown that sutures are sites of bony adaptation that normally allow for cranial expansion, compliance, and adaptability well into the third decade of life.²⁸ Premature suture fusion, generally occurring before birth, severely disrupts this developmental program.

Recent advances in molecular medicine have allowed a better understanding of craniosynostosis via identification of candidate genes and their mutations. Mutations in the genes encoding the fi-

CME EDUCATIONAL OBJECTIVES

1. Describe the nomenclature, epidemiology and etiology of craniosynostosis.
2. Recognize the infant with an abnormally shaped head and explain the difference between positional plagiocephaly and craniosynostosis.
3. Discuss when to refer a child with a skull deformity.

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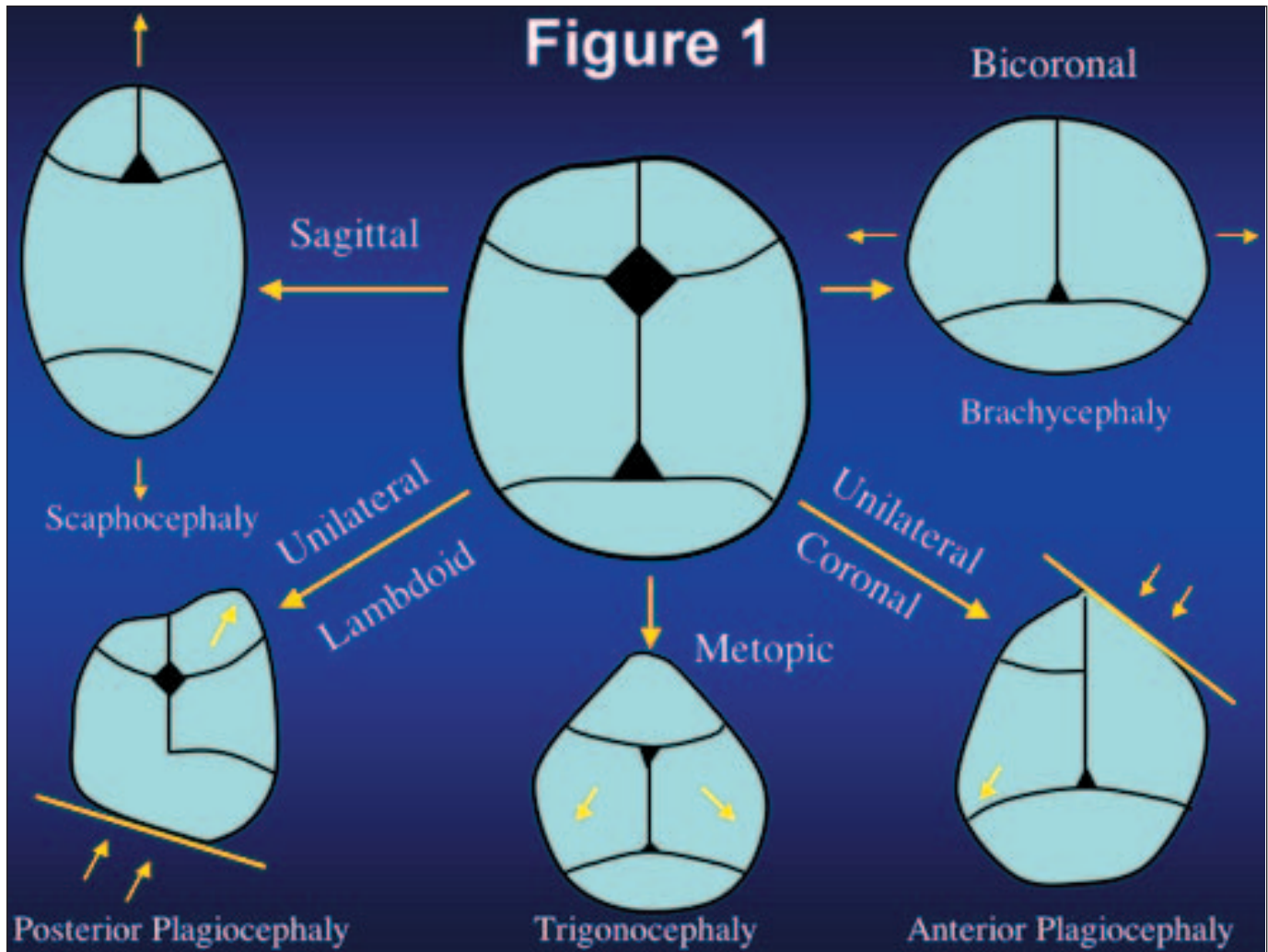


Figure 1. Schematic overview of the most common types of craniosynostosis. Restriction of growth at a specific suture will prevent growth perpendicular to the involved suture and lead to compensatory growth at unfused sutures.

broblast growth factor receptors-1, -2, and -3 (FGFR-1,-2,-3) are known to be associated with specific craniosynostotic syndromes, including Apert, Crouzon, and Pfeiffer syndromes.^{30,31} Nonsyndromal sporadic and familial forms of craniosynostosis likewise have been associated with FGFR mutations. Because FGF receptors have been shown to promote skeletogenic differentiation of neural crest cells, it is likely that there is a causal link between mutations in FGFR genes and altered development of the craniofacial skeleton. In addition, upregulation of transforming growth factor-beta expression has been demonstrated in the dura underlying prematurely

fused sutures.³² Although research efforts in this area have focused primarily on FGFR and transforming growth factor, a number of other factors have also been implicated, including MSX2 and TWIST [SPELL OUT OR DEFINE].^{33,34} Alterations in these growth factors have been observed in patients with nonsyndromic craniosynostosis, and it has been suggested that at least a subset of these patients carry cytogenetically undetected mutations in the aforementioned candidate genes.³⁵

The intimate interaction between genetic and environmental factors in the pathogenesis of nonsyndromal craniosynostosis has been elucidated by a

number of recent studies. In these studies, intrauterine fetal head constraint was found to be necessary and sufficient to cause craniosynostosis in the genetic background of a novel FGFR-2 gene mutation.³⁶ Hunenko et al.³⁷ also demonstrated a potential link between genes and environment in the pathogenesis of craniosynostosis, demonstrating upregulation of transforming growth factor receptors type I and II and FGFR-2 in mouse sutures subjected to in utero head constraint. The prevalence of environmentally induced craniosynostosis in the setting of predisposing mutations, however, remains unknown. The factors involved in the development of the cra-

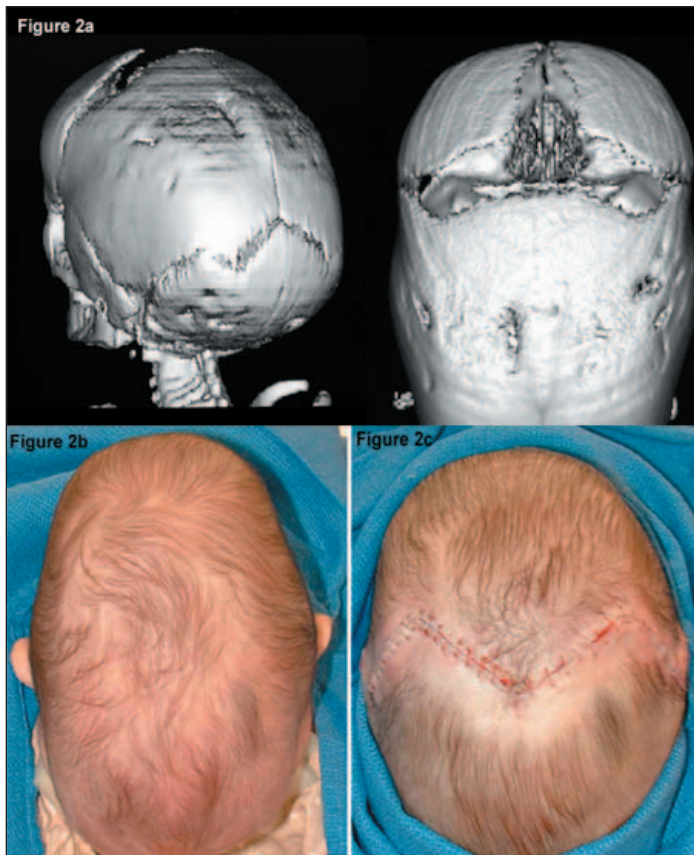


Figure 2. (A) Three-dimensional reconstruction of a CT scan demonstrating sagittal craniosynostosis. (B) Preoperative and (C) postoperative photographs of surgically corrected sagittal craniosynostosis.

niofacial skeleton are manifold, but deciphering the complex processes involved will undoubtedly expand our therapeutic options for the treatment of craniosynostosis in the future.³⁸

DIAGNOSTIC CONSIDERATIONS

Craniosynostosis that develops in utero can be diagnosed by routine prenatal ultrasonography in the second and third trimesters.^{39,40} Specific syndromic and familial cases of craniosynostosis also may be detected by fetal genetic testing. In most cases, however, a diagnosis of craniosynostosis will be made in early infancy during well-baby checkups or when obvious defects in head morphology become a source of concern for the parents. A definitive diagnosis of nonsyndromal single-suture craniosynostosis usually can be made after a thorough history and physical examination, and

further testing is most often unnecessary.

At a minimum, the elicited history should include information concerning significant prenatal events (ie, breech presentation), the method of delivery, the preferred sleeping position of the infant, and the existence of a family history of craniosynostosis or abnormal head shape. The clinical examination should assess for asymmetries and malformations in all anatomical regions of the

infant. As previously outlined, characteristic head morphologies are predictive of specific premature single suture fusion. A careful viewing of the infant's head from all angles should be done, as should palpation of the calvarium, which may reveal bony ridging at the fused suture(s). Palpation of the anterior fontanelle may reveal a triangular rather than diamond shape in infants with unilateral coronal, sagittal, or metopic craniosynostosis (Figure 1). Head and facial deformity, torticollis, scoliosis, congenital hip dislocation, pelvic obliquity, rib cage molding, and other signs of generalized asymmetry in infancy often present together and should be noted.⁴¹ Extremities and digits should be examined as particular syndromic forms of craniosynostosis present with syndactyly (eg, Apert syndrome). Finally, passive and active neck range of motion

should be evaluated to evaluate possible torticollis.

Plain skull radiography often is used to confirm a clinical diagnosis of craniosynostosis. This modality can be unreliable, however, as it often fails to demonstrate any sutural fusion in the presence of craniosynostosis, and it can be interpreted as being abnormal when no true pathology exists. Radiographic findings indicative of craniosynostosis include bone bridges across the suture, suture sclerosis, suture straightening and narrowing, or suture obliteration.^{42,43}

Computed tomography (CT) scanning with bone windows offers superior views of sutural pathology, and is warranted when skull radiography is equivocal. Brain windows may also reveal associated intracranial pathology such as evidence of increased intracranial pressure or agenesis of the corpus callosum, although magnetic resonance imaging should be obtained if there is clinical suspicion of such pathology. Three-dimensional CT scanning has emerged as a useful tool in surgical planning and evaluation of complicated suture pathology, but is not essential for making a diagnosis of craniosynostosis.

SURGICAL TREATMENT

Most children with craniosynostosis will not manifest neurological complications or have an adverse long-term cognitive outcome secondary to limitations in calvarial growth. Thus, for the majority of children with simple craniosynostosis, surgical intervention is undertaken for cosmetic and psychosocial considerations. However, it is important to keep in mind that as many as 60% of syndromic craniosynostosis patients and up to 20% of nonsyndromic single-suture craniosynostosis patients have been reported to develop increased intracranial pressure (ICP).^{8,59} Infants diagnosed with craniosynostosis and presenting with papilledema, irritability, vomiting, or fontanelle bulging may have elevated

ICP. CT evidence of expanded subarachnoid spaces is seen frequently in craniosynostosis but should not be the sole indicator of elevated ICP, as it is highly nonspecific in this population.

While expedited surgical intervention is indicated in suspected cases of elevated ICP secondary to craniosynostosis, the insidious nature of elevated ICP development in infants with craniosynostosis generally permits physiological adaptation such that the usual clinical signs are not present. Elevated ICP, whether clinical or subclinical, is the presumed mediator of cognitive deficits in the majority of children with nonsyndromal craniosynostosis.⁶⁰ Because it is impossible to determine with certainty the children who are likely to develop increased ICP, surgical intervention often is performed.

When children present with clear signs of elevated ICP or a severe calvarial deformity, spontaneous resolution is very unlikely, and these children are best served by early surgical intervention.⁴⁷⁻⁵⁴ When children present with mild to moderate degrees of cranial deformity, the decision to operate is more difficult, and both the age and severity of the calvarial defect will guide decision making. If the deformity is mild and the child is approaching age 1, it is unlikely surgery will be needed, as a mild deformity at 12 months is unlikely to become significantly more pronounced. In these instances, the decision to operate depends on the family's perception of the asymmetry (which is likely to remain unchanged), and consideration of psychosocial issues that may be encountered during the child's life.

While there continues to be substantial debate as to the timing of surgical correction for craniosynostosis, the general principle is that early surgical release produces the best long-term results and minimizes secondary craniofacial abnormalities. When surgery is performed at an early age, however, the sta-

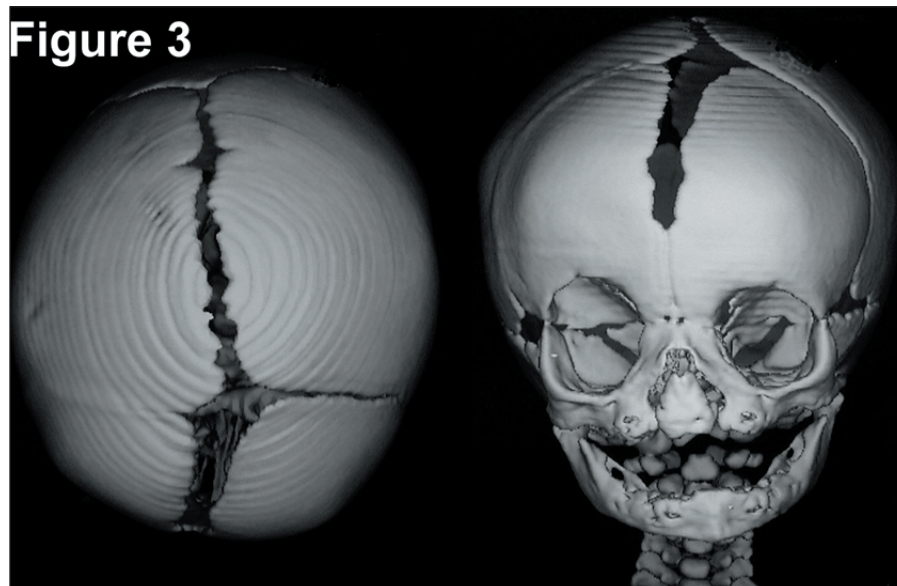


Figure 3. Three-dimensional reconstruction of a CT scan demonstrating right unilateral coronal craniosynostosis. Note the "harlequin eye" deformity, or the upward and outward deviation of the orbit.

bility of cranial re-shaping and advancements can be compromised secondary to the softness of the bone. In addition, regrowth of the removed bone can occur and occasionally necessitate repeat surgery. Most surgeons, therefore, balance these concerns and operate between ages 3 and 12 months.

Myriad surgical procedures, ranging from removal of the suture only to extensive cranial vault remodeling, have been used in the treatment of craniosynostosis. Current surgical therapy includes resection of the involved suture and correction of secondary facial and calvarial abnormalities. Improvements in techniques now allow for corrective surgery with minimal morbidity (below 10%) and mortality (below 1%).⁷⁻¹⁰ In addition to the standard open techniques, endoscopic techniques are being used increasingly with success in young patients.^{52,55-57} Jimenez et al.^{18,58} recently reported a series of 100 patients treated with endoscopic techniques and demonstrated safe and efficacious results. The decreased blood loss, reduced need for transfusions, and shorter length of stay make this procedure a promising alternative to other more extensive pro-

cedures.

Sagittal Craniosynostosis

Sagittal craniosynostosis is the most common type of single suture craniosynostosis. With an incidence of between 0.2 and 1 per 1,000 live births, it represents 40% to 60% of reported craniosynostosis.^[REFERENCE] Between 75% and 85% of cases occur in boys, and the majority are sporadic. Secondary sagittal suture closure also may be seen in infants following shunting procedures for hydrocephalus (approximately 1%).^{16,69} Children with sagittal craniosynostosis present with an elongated and narrow skull. Depending on the degree of fusion, frontal bossing and occipital protrusion also may be seen (Figure 2, see page xxx).

Surgical management of sagittal craniosynostosis has changed considerably during the past 50 years. As early as 1892, approaches advocating the simple removal of the pathologic suture were proposed. Approaches have evolved from minimal strip craniectomies to extensive total calvarectomies, and now minimally invasive endoscopic and distraction techniques.⁷² At the Children's

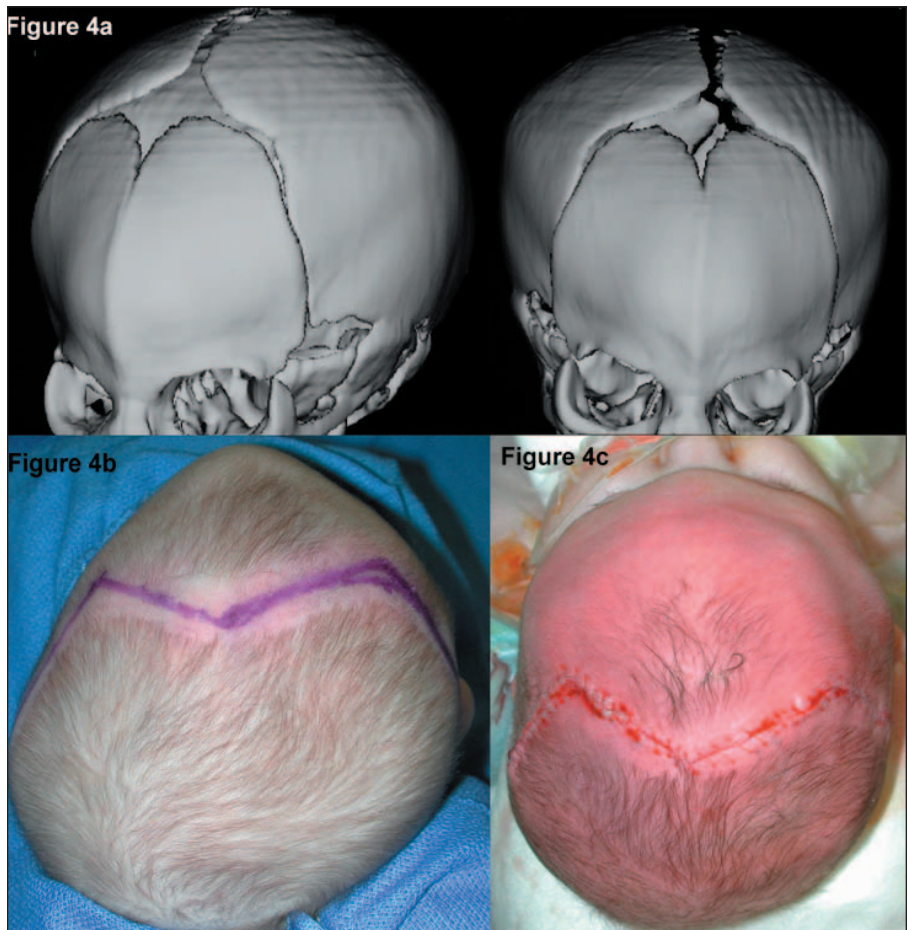


Figure 4. (A) Three-dimensional reconstruction of a CT scan demonstrating metopic craniosynostosis. (B) Preoperative and (C) postoperative photographs of surgically corrected metopic craniosynostosis.

Hospital of New York, we perform an extended vertex craniectomy (ie, removal of the sagittal suture and a strip of bone 3 to 6 cm wide from the coronal to lambdoid sutures) combined with bilateral parietal osteotomies for patients younger than 6 months. In infants older than 6 months, significant pathology at the skull base usually dictates a larger procedure to be undertaken to provide optimal correction. This is the rationale for total calvarectomy and reconstruction in the setting of severe or late presentation scaphocephaly.^{56,76-80}

Coronal Craniosynostosis

Premature closure of a single coronal suture will lead to anterior plagiocephaly, whereas bicoronal craniosynostosis will lead to brachycephaly. Unilateral

coronal craniosynostosis occurs in approximately 1 in 10,000 live births and accounts for 20% to 25% of all craniosynostosis.⁵⁶ In addition, deformational anterior plagiocephaly, similar to posterior positional plagiocephaly, is seen occasionally and has been reported to occur in 17% to 24% of live births secondary to in utero molding. This deformation is frequently associated with other congenital conditions thought to be the result of inhibited fetal growth such as congenital hip dysplasia, micrognathia, and torticollis.²⁶

Patients presenting with unilateral coronal craniosynostosis have flattening of the ipsilateral forehead with a recessed fronto-orbital bar. When anteroposterior growth is restricted across the closed coronal suture, compensatory changes

take place along the contralateral suture. A slight female predominance exists (approximately 60%), and both sutures are affected equally. The presence of a “harlequin” appearance on skull radiographs and reconstructed CT scans is caused by superior elevation of the sphenoid wing and is considered pathognomonic for coronal craniosynostosis (Figure 3, see page xxx). Strabismus also is common (50% to 60%) and results from a shortened anterior skull base displacing the orbital roof and trochlea, which contributes to an imbalance of the superior oblique muscle.^{81,82}

As with sagittal craniosynostosis, primary indications for surgical correction most often are cosmetic. However, concerns regarding orbital symmetry and strabismus are important when dealing with coronal craniosynostosis. The goals of surgical correction with unilateral coronal craniosynostosis are to restore the anterior fossa intracranial volume, repair orbital deformities, and correct growth restriction at the coronal and anterior skull base sutures.⁴⁶

The surgical approach for both unilateral and bilateral coronal craniosynostosis involves releasing of both coronal sutures, reshaping bilateral frontal bones, and advancing the orbital bar. Although the proper timing of surgery is still debated, surgical intervention commonly is performed between ages 3 and 10 months.⁵⁶ The majority of patients with unilateral coronal craniosynostosis have excellent outcomes after surgery, whereas those treated for bicoronal craniosynostosis may require secondary surgery due to the frequent syndromic association.⁸³⁻⁸⁵

Metopic Craniosynostosis

Premature fusion of the metopic suture restricts the transverse growth of the frontal bones, which results in narrowing of the anterior cranial fossa and trigonocephaly (Figure 4, see page xxx).⁸⁶ Metopic craniosynostosis represents ap-

proximately 10% of all craniosynostosis and shows a male predominance of between 75% and 85%.^{16,87} Mild variants of metopic craniosynostosis seem to represent familial inheritance and have been associated with CNS anomalies and abnormalities of chromosomes 3, 9, and 11.^{17,51,88-90} For example, Tubbs et al.⁹¹ found a 30% incidence of Chiari I malformation in the evaluation of patients with simple metopic ridges.

While the majority of patients have simple metopic craniosynostosis, a significant number (10% to 22%) may manifest syndromal features and are at additional risk for cognitive dysfunction.^{16,56,87} Although single-suture craniosynostosis has a low incidence of mental retardation, metopic suture craniosynostosis has the highest rate among the subtypes of craniosynostosis.^{9,92} For these reasons, an MRI usually is recommended in patients with metopic craniosynostosis.

Although under normal circumstances the metopic suture fuses around age 2, recent reports suggest that it may close earlier without causing deformity. Premature fusion of the metopic suture is necessary but not sufficient for the development of trigonocephaly. The degree of trigonocephaly varies considerably and will direct the need for surgical intervention. Similar to coronal craniosynostosis, surgical intervention is commonly done between ages 3 and 10 months.⁵⁶

The goals of surgery are to increase the volume of the anterior cranial fossa, reshape the forehead, and advance the superior orbital bar. Patients with a simple midline keel may best be served by simple contouring of the frontal bone, as opposed to those with significant trigonocephaly and hypotelorism, who often need orbital reconstruction and lateral expansion.⁵⁶ More recent modifications propose radical treatment of the involved sphenoid bone and simultaneous correction of hypotelorism.^{93,94} For more than 90% of children, the long-term outcome

TABLE.

Clinical Features of Lambdoid Synostosis Versus Positional Molding

Abnormal Feature	Lambdoid Synostosis	Positional Molding
Persistent positioning on one side	Rare	Invariable
Torticollis, tight SCM [SPELL OUT], or delay	Absent	Present
Bony riding along suture line	Present	Absent
Occipitomastoid bony protrusion	Present	Absent
Frontal bossing	Contralateral	Ipsilateral
Ipsilateral ear displacement	Posterior	Anterior
Geometric shape from vertex	Trapezoid	Parallelogram

is excellent.⁹⁵⁻⁹⁷

LAMBDOID CRANIOSYNOSTOSIS VERSUS POSTITIONAL MOLDING

The majority of posterior plagiocephaly seen today represents positional molding or flattening without craniosynostosis. True craniosynostosis of the lambdoid suture is a rare event with an incidence ranging from 0.9% to 4% in patients presenting with occipital flattening.^{18,20,56,61} Positional molding, on the other hand, is very common and has been found to have an incidence of nearly 10% in the general population.⁶² Positional plagiocephaly is seen more frequently in boys than girls (ratios between 3:120 and 3:263), is more common on the right (70%), and is associated with torticollis. The increased incidence of positional plagiocephaly in recent years is attributable to parents following the American Academy of Pediatrics “Back to Sleep” positioning recommendations aimed at decreasing the risk of sudden infant death syndrome instituted in the early 1990s.^{3,20,64-66} Infants placed in a supine sleeping position appear to have increased predilection for persistent flattening over one side of the occiput, representing transient calvarial deformation.

The differentiation between positional plagiocephaly and “true” lambdoid craniosynostosis is important when deciding the appropriate treatment course.

In the majority of cases, deformational plagiocephaly can be differentiated from lambdoid craniosynostosis on the basis of history and physical examination. Imaging studies are unnecessary to make a diagnosis in most cases (Table, see page xxx). Children with positional molding usually are born without significant flattening but develop it over the first several months with persistent positioning on one side, especially if any torticollis exists. Children with lambdoid craniosynostosis, on the other hand, often are born with the occipital flattening and do not have a history of unilateral positioning or torticollis.

On physical exam, children with lambdoid craniosynostosis usually have distinctive features including a trapezoid-shaped head in association with a posteriorly displaced ear when viewed from the top of the head, a palpable ridge over the suture line, and contralateral occipital bossing. This is in contrast to the parallelogram-shaped head, ipsilateral frontal bossing, and anterior displacement of the ipsilateral ear seen in positional plagiocephaly (Figure 5, see page xxx).

The management of deformational plagiocephaly is conservative, with preventive counseling of parents, positional maneuvers, and exercises. Preventive counseling should include early discussion with parents, encouraging them to provide infants with time in the prone

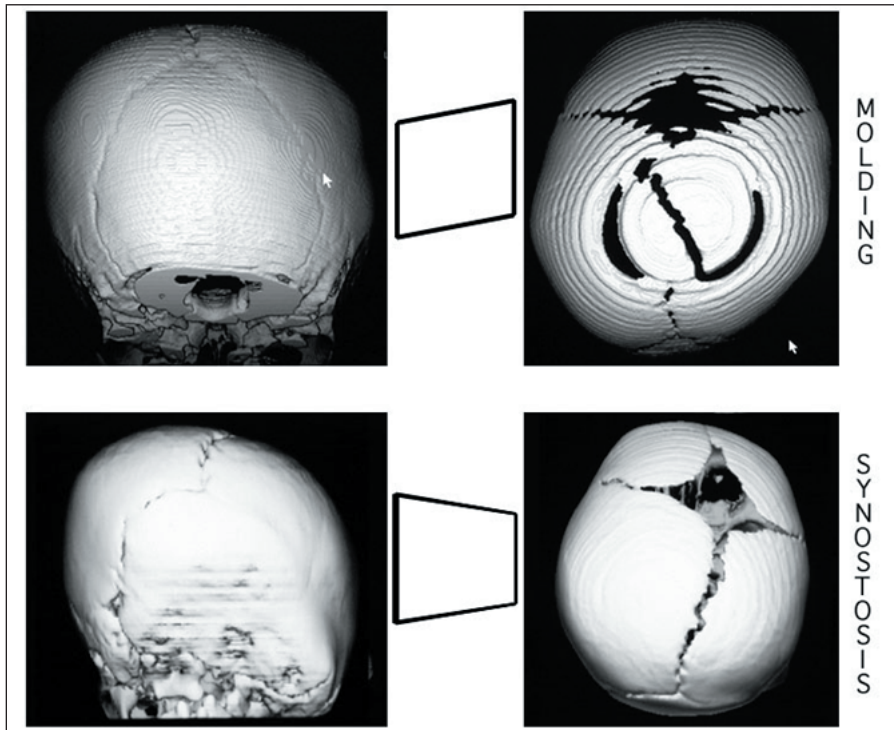


Figure 5. Three-dimensional reconstruction of a CT scan demonstrating the difference in geometrical shape (parallelogram versus trapezoid) between positional molding and lambdoid craniosynostosis.

position while awake and supervised. Beginning at birth, most deformational plagiocephaly can be prevented by nightly alternating the supine head position (left and right occiput) during sleep.⁴⁴ If positional molding already exists, parents should be counseled to change the infant's sleeping position with the goal of placing the opposite side of the head against the mattress. In addition, supervised "tummy time" while awake and neck motion exercises should be encouraged if torticollis is present.

The majority of affected infants will improve in 3 to 6 months, particularly after they begin to sit up and spend less time lying on the back of the head. If there is progression of the deformation or lack of improvement after a trial of positional modification, then referral to a subspecialist is appropriate.⁴⁴ A referral both enables diagnostic confirmation and can expedite the initiation of definitive management if required.

Cranial molding helmets or bands are an option for children with severe

deformity that is refractory to positional changes. The role of orthotic molding, however, remains controversial and may be of no additional benefit over repositioning.⁴⁵ There are no randomized or prospective clinical trials that demonstrate convincingly that helmet therapy offers superior cosmetic results to positional maneuvering in the long term. Furthermore, helmet therapy is costly and may not be well tolerated. If helmet therapy is used, the best response occurs between ages 4 and 12 months.⁴⁴ At the Children's Hospital of New York, we tend to recommend helmet therapy only for infants with extreme and unresolving cases.

In contrast to the success of conservative treatment in children with deformational plagiocephaly, those with lambdoid craniosynostosis most often require surgical intervention. While craniotomy for expansion of the posterior cranial vault is satisfactory in greater than 90% of patients, some children with lambdoid craniosynostosis have significant pari-

etal and frontal compensatory changes that may require a more extensive reconstruction⁵⁶ (Figure 6, see pages xxx). The greatest improvement has been seen in children operated on at early ages (younger than 18 months).^{67,68}

SUMMARY

Abnormal head morphology is common among infants and often leads parents to ask their pediatricians for guidance. While the vast majority of children will have positional molding, early diagnosis and referral for surgical evaluation are important in obtaining the best possible results for children who are affected by craniosynostosis. While the primary goal of such treatment is enhancement of craniofacial cosmesis, surgical correction may also promote normal psychosocial development and prevent adverse neurobehavioral sequelae secondary to increased intracranial pressure or impaired cerebral growth. The coordination of a multidisciplinary team in the setting of a center experienced in the management of craniosynostosis will assure the best possible results for each patient.

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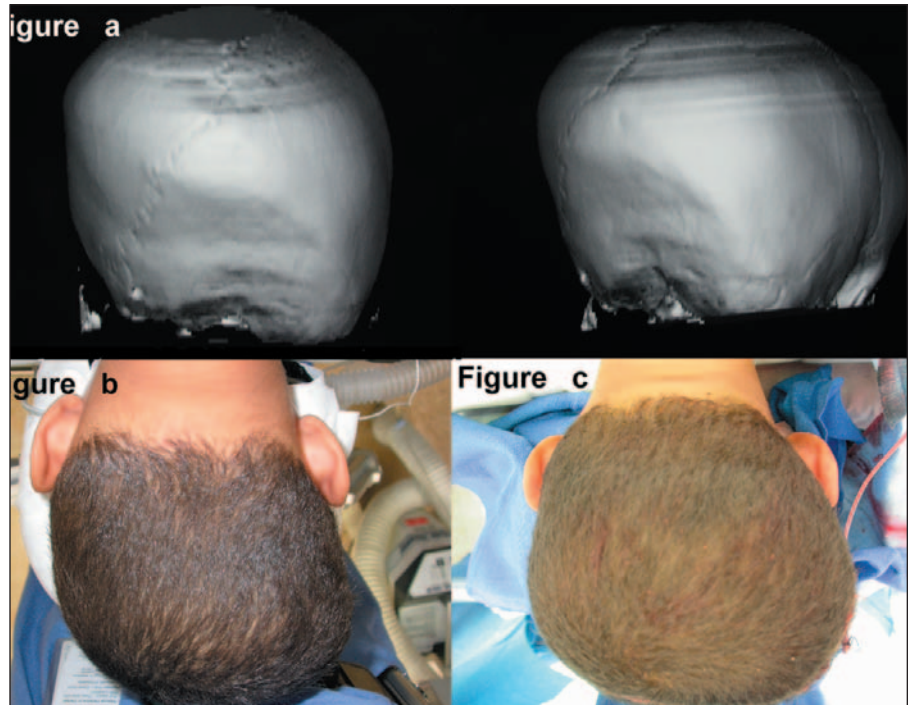


Figure 6. (A) Three-dimensional reconstruction of a CT scan demonstrating unilateral lambdoid craniosynostosis. (B) Preoperative and (C) postoperative photographs of surgically corrected lambdoid craniosynostosis.

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