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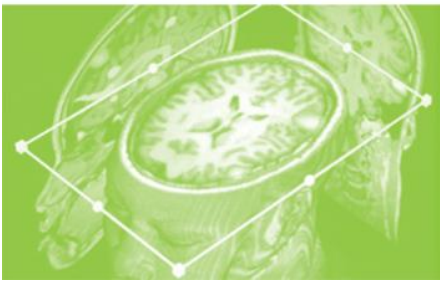

Neuro Update℠

A HEADS UP ON CRANIOSYNOSTOSIS

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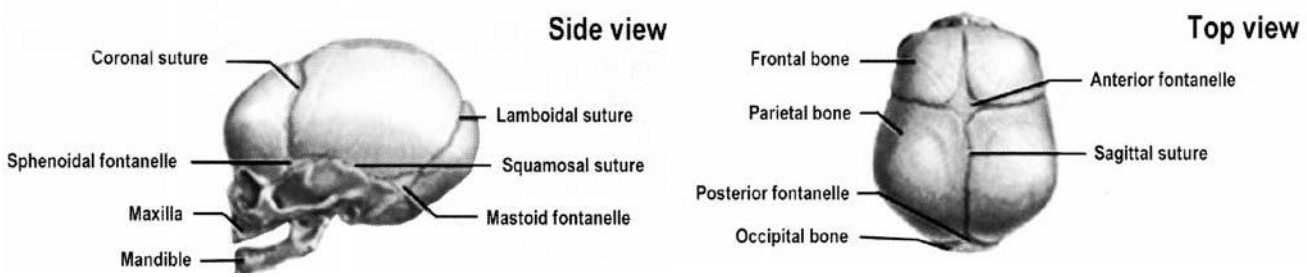
Craniosynostosis, an early closure of the growth plates of the skull, results in a skull deformity and may result in neurologic compromise. Craniosynostosis is surprisingly common, occurring in one in 2,100 children. It may occur as an isolated abnormality, as a part of a syndrome or secondary to a systemic disorder. Typically, premature closure of a suture results in a characteristic cranial deformity easily recognized by a trained observer. it is usually the misshapen head that brings the child to receive medical attention and mandates treatment.

This issue of Neuro Update will present the diagnostic features of the common types of craniosynostosis to facilitate recognition by the primary care provider. The distinguishing features of other conditions commonly confused with craniosynostosis, such as plagiocephaly, benign subdural hygromas of infancy and microcephaly, will be discussed. Treatment options for craniosynostosis will be reserved for a later issue of Neuro Update.

Types of Craniosynostosis

Sagittal synostosis
The sagittal suture runs from the anterior to the posterior fontanella (Figure 1). Like the other sutures, it allows the cranial bones to overlap during birth, facilitating delivery. Subsequently, this suture is the separation between the parietal bones, allowing lateral growth of the midportion of the skull. Early closure of the sagittal suture restricts lateral growth and creates a head that is narrow from ear to ear. However, a single suture synostosis causes deformity of the entire calvarium. With restriction of lateral bone growth, brain growth is similarly restricted in that direction. As a consequence, the brain and skull grow excessively from anterior to posterior, creating the football-shaped head characteristic of sagittal synostosis (Figure 2). The forehead bulges forward, usually with a prominent knob of bone at the occiput. often, there is another knob of bone at the anterior fontanella and a palpable ridge of bone along the sagittal suture.

Figure 1



The head deformity associated with sagittal synostosis is termed scaphocephaly, which can be confused with molding caused by vaginal delivery. The two can be differentiated with time—cranial molding diminishes after a few days to a few weeks, but the head deformity of sagittal synostosis worsens. This worsening is caused by continued brain growth, which accentuates the deformity.

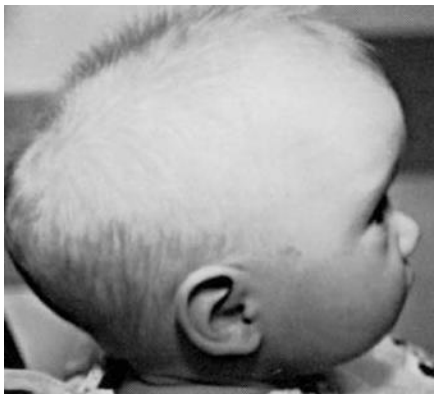


Figure 2

Sagittal synostosis is the most common form of craniosynostosis and occurs much more often in boys. it is not clear whether brain damage results if sagittal synostosis is untreated; however, it is known that intracranial pressure is elevated in 8 percent to 13 percent of children, which can restrict regional cerebral blood flow. it is clear that children with uncorrected sagittal synostosis have an increased incidence of emotional problems and difficulty with peer interactions. This probably relates to the reaction of the latter to the child's abnormally shaped head.

When sagittal synostosis arises, the child should be referred for pediatric neurosurgical evaluation as soon as possible. To be discussed in future issues of *Neuro Update,* the surgery required to correct sagittal synostosis becomes more complex and risky as the child gets older.

Coronal synostosis
The coronal suture separates the frontal from the parietal bone (Figure 1 above). Unilateral coronal synostosis presents with a flattening of the forehead on the affected side (Figure 3). Also, the supraorbital ridge is swept backward creating frontal plagiocephaly. As with sagittal synostosis, brain growth is restricted parallel to the involved suture. Compensatory overgrowth occurs, causing a blowing outward of the other side of the head—accentuating the deformity.

Unilateral coronal synostosis must be differentiated from contralateral lambdoid synostosis and contralateral occipital plagiocephaly caused by positional flattening. Both of these entities cause forehead bowing, mimicking the asymmetry of unilateral coronal synostosis. Major points of difference include:

1. The back of the head is not flat with unilateral coronal synostosis.
2. The supraorbital ridges are only affected in unilateral coronal synostosis.
3. The root of the nose is deviated toward the flat side of the head in coronal synostosis—the tip of the nose deviates the opposite way.

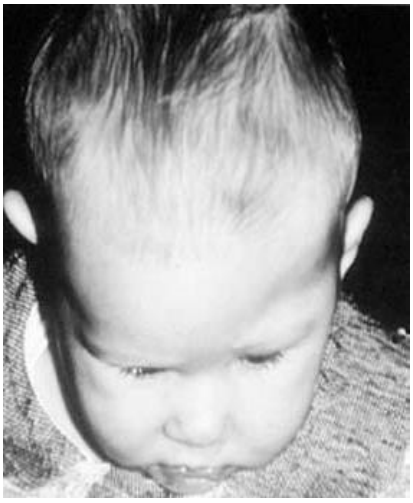


Figure 3

Bilateral coronal synostosis creates a head deformity termed brachycephaly characterized by a foreshortened, extreme high forehead (Figure 4). This is created by the lack of forward growth of the frontal bone, associated with a small anterior cranial fossa. This is a symmetric synostosis, and the nose, accordingly, remains with a chromosomal abnormality. The more common syndromes are Crouzon's (midface hypoplasia), pfeiffer's and Apert's. A thorough physical examination and chromosomal analysis is mandatory for every child with bicoronal synostosis.

With two sutures closed, intracranial pressure is frequently elevated, especially in syndromic bicoronal synostosis. due to the shallow orbits, severe proptosis can occasionally occur. Blindness is a risk from corneal exposure, necessitating an early intervention to allow normal brain development and prevent loss of sight.

Infants with benign subdural hygromas can be misdiagnosed with bicoronal synostosis due to their prominent foreheads. Clinically, these children can be differentiated by the retruded, high forehead of bicoronal synostosis versus the forward-jutting, squared forehead of benign subdural hygromas. Children with benign subdural hygromas are usually macrocephalic with none of the other stigmata of the syndromes associated with bicoronal synostosis. Finally, if physical criteria do not differentiate these entities, then a computed tomography (CT) scan will show whether the coronal sutures are patent.

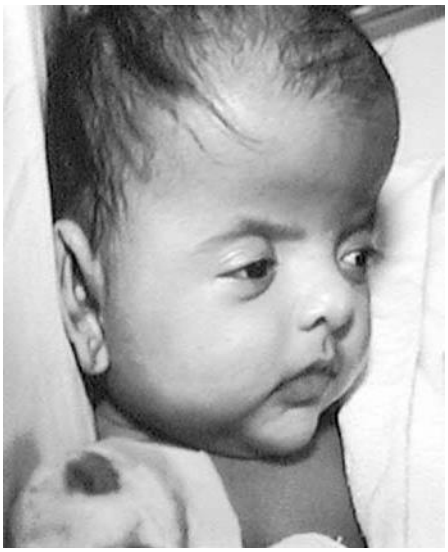


Figure 4

Metopic synostosis

The metopic suture separates the two halves of the frontal bone. Early closure results in a triangular appearance to the forehead, termed trigonocephaly (Figure 5). The supraorbital ridges are swept back laterally, and there is a prominent ridge of bone in the midline from the anterior fontanella to the glabella (root of the nose). Metopic synostosis can be associated with midline brain anomalies, such as holoprosencephaly. All children with severe metopic synostosis need brain imaging with either CT scan or magnetic resonance imaging (Mri) scan.

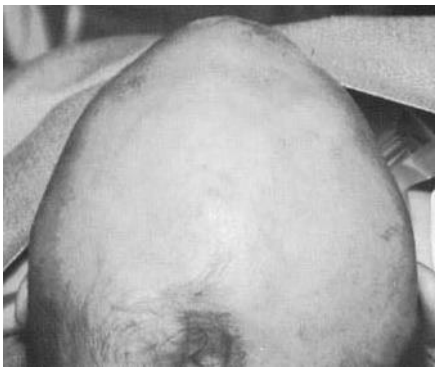


Figure 5

The metopic suture is the earliest suture to close normally, usually between ages 1 to 2—the other sutures usually do not close until late teens or early 20s. Accordingly, many children present with the above mentioned midline forehead ridge, exhibiting an otherwise normal appearance. These children do not require operative intervention, with time, the ridge regresses and the forehead shape remains normal.

Lambdoid synostosis
The lambdoid suture separates the parietal from the occipital bone. The results of synostosis include the flattening of the ipsilateral side and the compensatory bowing of the opposite side. Bilateral lambdoid synostosis produces symmetric flattening of the back of the head. Both of these conditions are quite uncommon (less than 2 percent of all craniosynostosis). Unilateral occipital flattening is now much more commonly caused by positional flattening.

Positional plagiocephaly is now the most common diagnosis for new patients to our practice. While the national institute of Child Health and Human development's Back to Sleep campaign has effectively reduced the incidence of sudden infant death syndrome (SidS), an unintended consequence has been an explosion in the number of infants with flat occiputs. Although either side may be involved, the right side is most commonly flattened. There may be associated muscular torticollis keeping the infant's head turned toward the affected side. The ear on the ipsilateral side is frequently rotated forward, and the forehead and cheek may also be asymmetric, creating a trapezoid-like appearance to the head when viewed from above. Some have claimed positional plagiocephaly left untreated may be associated with an increased incidence of ear infections and later temporomandibular joint disorder problems. However, there is no credible medical evidence to suggest this.

The primary treatment for positional plagiocephaly is repositioning. With simple repositioning, the abnormal head shape will resolve in most children. For those children with significant facial involvement, a molding band or helmet may be appropriate.

We usually do not place infants in cranial orthotic devices until after the age of 6 months, preferring an emphasis on repositioning for younger children.

Multisuture synostosis
Fortunately, this condition is rare, generally associated with one of the above syndromes. The head shape is markedly deformed, usually a cloverleaf pattern. The deformity is due to the only open suture being a minor suture in the temporal area—the squamosal suture (Figure 6). Thus, the head juts out laterally and also at the anterior fontanel. These infants need decompressive surgery within the first few days of life to allow for brain growth.

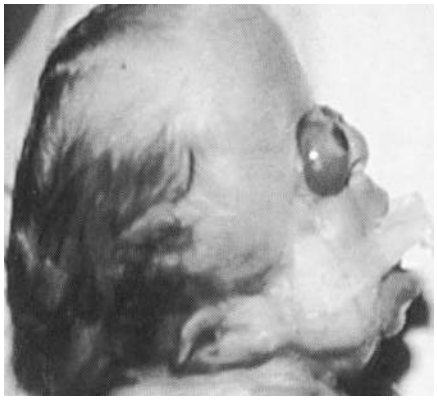


Figure 6

Microcephaly with a normal shaped head is almost never due to craniosynostosis. Usual causes include a normal variant (just a small head) or inadequate brain growth. A CT or MRI scan may be necessary to rule out neurologic abnormality.

Diagnosis of Craniosynostosis
The diagnosis of craniosynostosis is primarily clinical: recognition of the characteristic head shape abnormality for each type of synostosis. A CT scan can be obtained to assess the patency of the sutures if the diagnosis is not obvious, or other pathology needs to be ruled out, including brain abnormalities in metopic synostosis and hydrocephalus. Skull films are of minimal value and should not be obtained.

Obviously, the more craniosynostosis you have seen, the better you are at recognizing it. As pediatric neurosurgeons, we are trained, and have much experience, in the recognition of the various types of synostosis. We are always happy to see any of your patients if there are questions, and we can frequently obviate the need for a CT scan, lowering the cost and avoiding radiation exposure to the child.