CHAPTER 47

Nonsyndromic Craniosynostosis

J. Kerwin Williams
Steven R. Cohen
Fernando D. Burstein
Roger J. Hudgins
William R. Boydston

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 *craniostenosis* 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.[60] Galen[91] also described a patient with craniosynostosis and coined the term *oxycephaly*. Sommering[86] first recognized that skull growth occurred at the sutures and fusion of these "growth areas" would create a deformity. Subsequently, Virchow[92] 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.[14] 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.[32] Furthermore, craniosynostosis in two members of the immediate family will increase the chance the next child will develop a premature suture fusion.[70] 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 secondary anomalies not directly associated with the suture fusion.[15] 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.[63] It is characterized by an increased anteroposterior position and decreased biparietal width (scaphocephaly). 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.[70] 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%),[28,84] 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.