Treatment of Craniosynostosis

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Premature closure of the sutures of the cranial vault is thought to be a mesenchymatous defect of unknown cause; it appears more often than by chance in the same family, with a higher incidence of some types in males. It is frequently recognized at birth but may become evident and progress during early childhood. By its nature, craniosynostosis causes distortions of the cranium and constrictions of growth of the brain if multiple sutures are involved. Other deformities may coexist, particularly syndactylism and distortions of the maxilla in children who have synostosis of the coronal suture.

Recognition, timing, and treatment are interdependent factors of equal importance in proper care.

Recognition

Appearance. Craniosynostosis should be suspected in the infant with an abnormally shaped head or premature closure of the fontanel. It may be obvious at birth or appear during the time of active growth of the head.

In infancy, growth and shape of the skull are determined by mechanical forces exerted by the growing brain on the cranial case. When the skull is not able to expand in certain directions, because of premature synostotic changes in sutures, abnormalities of shape are the result. Thus, the patient with sagittal synostosis has a long narrow head and often a visible or palpable ridge along the sagittal suture line (Fig. 1 A, B, C); the patient with coronal synostosis has a foreshortened broad head and bulging of the temporal regions (Fig. 2 A, B, C), and he may also have prominent eyes and deformities of the face. The patient with premature closure of the metopic suture has a prow-shaped forehead, and the one with unilateral coronal or lambdoid closure has an asymmetrical head. The infant with multiple closed sutures has a small head, often with prominences in the temporal regions and in the region of the anterior fontanel, if this was the last area to close.

The diagnosis may frequently be made on clinical observation, but radiologic studies will confirm the diagnosis and differentiate between craniosynostosis, microcephaly, or molding caused by sick or quiet babies lying in one position for long periods.

Roentgenography. In radiologic diagnosis, it is necessary to view the entire extent of all the sutures since at the outset only a small segment may be fused; fusion of only a portion of a suture is sufficient to retard expansion of the skull and is the precursor to advancing obliteration of that suture. In addition, it must be remembered that fusion of one suture may be followed by obliteration of other sutures causing more serious compression of the brain.

In sagittal synostosis, outward “beaking” is seen, and often a straight line is demonstrated rather than a serrated appearance of the outer portion of the abnormal suture. “Beaking” inward and increased density of the lower end of the obliterated suture blending into the wing of the sphenoid bone can be seen in coronal synostosis. Shortening of the base of the skull and sometimes a relative degree of platybasia can be recognized in coronal synostosis. The skull of plagiocephaly, in which one coronal suture is closed, has obliteration of that suture with associated flattening of this region of the skull and a characteristically elevated lateral displacement of the superolateral margin of the orbit on the side involved.

If multiple sutures are closed, in addition to the absence of suture lines, signs of increased intracranial pressure are present manifested by exaggerated digital markings, bulging of the thin temporal bone, and sometimes changes in the sella turcica. The sutures in microcephaly are patent in early