Review Article

Cranial vault growth in craniosynostosis

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Skull growth after single suture closure was described in 1851 by Virchow, who noted that growth in the
plane perpendicular to a fused suture was restricted. However, this observation failed to predict compensatory
growth patterns that produce many of the deformities recognized as features of individual syndromes. The
deformities resulting from premature closure of a coronal, sagittal, metopic, or lambdoid suture can be
predicted on the basis of the following observations: 1) cranial vault bones that are prematurely fused secondary
to single suture closure act as a single bone plate with decreased growth potential; 2) asymmetrical bone
deposition occurs mainly at perimeter sutures, with increased bone deposition directed away from the bone
plate; 3) sutures adjacent to the prematurely fused suture compensate in growth more than those sutures not
contiguous with the closed suture; and 4) enhanced symmetrical bone deposition occurs along both sides of a
non-perimeter suture that is a continuation of the prematurely closed suture. These observations regarding
growth in craniosynostosis are illustrated with clinical material in this report.

KEY WORDS · craniosynostosis · skull growth · restricted bone plate · compensatory skull growth · skull deformity

DESPITE many advances in the treatment of craniosynostosis, a normal skull shape is difficult to achieve by surgical measures. Surgical treatment is hampered because of an incomplete understanding of the pathophysiology of craniosynostosis. The modern perception of craniosynostosis began in 1791 with Sömmerring's observations that skull growth occurred along calvarial sutures and that failure of growth at a particular suture resulted in cranial deformity. In 1831, Otto described craniosynostosis in man and animals as secondary to microcephaly or premature suture closure. In 1851, Rudolf Virchow expanded upon Otto's observations, noting that growth was restricted in a plane perpendicular to the fused suture, and enhanced in the corresponding parallel plane. These observations have served as the principal guide in understanding craniosynostosis.

The role of calvarial sutures in determining skull shape was questioned by Moss in 1959. His skepticism was based on several observations: that cranial vault deformities can occur in the absence of a fused suture and that skull growth in the rat was not altered by experimental removal of a normal cranial vault suture; moreover, in cephalometric data, he found cranial base abnormalities associated with craniosynostosis. He described abnormalities in the planum, orbital, and foramino-clival angles in patients with sagittal or bicornal synostosis. Moss, therefore, proposed that cranial base abnormalities were primarily responsible for the characteristic forms of craniosynostosis rather than the result of premature closure of a vault suture. He emphasized that the principal influence for growth of the skull was brain development.

Although consistent cranial base abnormalities have not been described in sagittal and lambdoid synostosis, multiple deformities are seen in coronal synostosis. For example, in bilateral coronal synostosis several abnormalities have been described, including: protrusion of the greater wing of the sphenoid bone; a short anterior cranial base and clivus; an enlarged sella turcica; frontalization of the orbital roof; and deficient nasion-sella-basion angle. The cranial base is markedly asymmetrical in unilateral coronal synostosis. The anterior cranial base is short and the slope of the lesser
wing of the sphenoid bone is pointed upward and laterally ipsilateral to the stenotic coronal suture. The cranial base angle is also increased and the sphenoidal-petrosal angle is narrowed due to ipsilateral anterior displacement of the petrous ridge.

Despite cranial base involvement, premature closure of a cranial vault suture alone can be responsible for the characteristic deformities seen in each form of craniosynostosis. Isolated restriction of growth in a normal cranial vault suture of a growing animal can cause permanent cranial vault, cranial base, and facial skeleton abnormalities. Clinical observations show that these skull base changes are similar to cranial base maldevelopment in humans with craniosynostosis. Calvarial growth is a result of a complex interaction between the cranial vault, skull base, and facial skeleton. Thus, there is a theoretical basis for predicting the clinical form of each single suture synostosis by making reference to the vault suture alone.

To begin with, the observed calvarial deformities cannot be totally explained by restricted growth perpendicular to the fused suture. For example, multiple stereotyped deformities are seen in unilateral craniosynostosis (Fig. 1). Virchow's guidelines predict a deficit in growth in ipsilateral frontal and parietal bones perpendicular to the fused suture. He also implied that compensatory growth must occur at the remaining sutures to accommodate a growing brain. Virchow, however, did not establish a specific compensatory growth pattern that fully predicts the observed calvarial deformities. If the normal sutures could effectively accommodate for growth restriction caused by a prematurely fused suture, then the skull could conform to a shape similar to the underlying round brain. Indeed, the twofold purpose of cranial sutures is to allow molding during birth and to facilitate growth of a basically round head to accommodate the human round brain. It is of considerable interest that closure of a single suture should result in such a profoundly abnormal skull shape that secondarily changes the shape of the brain itself. In unilateral coronal synostosis, a normal calvarial shape could still develop if bone deposition along the metopic suture would increase and compensate for growth restriction at the coronal suture. Instead, contrary to the basic notion that brain growth primarily influences skull shape, enhanced compensatory growth results in an ipsilateral temporal and contralateral frontal boss. Although many of the characteristics of this compensation have been described, no unified theory is available to predict the outcome of compensatory growth. A new understanding is needed to explain how skull growth patterns change in craniosynostosis. By understanding these growth patterns, we might also understand the pathological mechanism of premature sutural stenosis.

A Working Hypothesis for Calvarial Growth

Each form of craniosynostosis involving premature closure of a single suture was analyzed to define the exact patterns of abnormal skull growth. This analysis found that all the deformities resulting from premature closure of a coronal, sagittal, metopic, or lambdoid suture can be predicted on the basis of four rules. Using unilateral coronal synostosis as an example, the four rules are described and illustrated: these rules are modified from a previous formulation described by Jane and Persing.

1. Cranial vault bones that are prematurely fused act as a single bone plate with decreased growth potential. In unilateral coronal synostosis, the fusion of the coronal suture results in a single bone plate of restricted growth potential that comprises the frontal and parietal bone (Fig. 2A).

2. Abnormal asymmetrical bone deposition occurs at perimeter sutures with increased bone deposition directed away from the bone plate. Under normal conditions, the growth pattern is equal at a suture bordering bones of similar form, while bone deposition is unequal along a suture separating skull bones of different form. In sutural synostosis, however, we postulate that abnormal bone deposition occurs at all perimeter sutures with additional growth directed away from the restricted bone plate. The asymmetrical bone deposition at perimeter sutures in unilateral coronal synostosis results in the characteristic expansion of the contralateral frontal and parietal bones and of the ipsilateral squamosal region (Fig. 2B).

3. Perimeter sutures adjacent to the prematurely fused suture compensate in growth more than pe-
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Fig. 2. Patterns of abnormal skull growth in craniosynostosis. A: Premature fusion of the left coronal suture results in a frontoparietal bone plate with restricted growth potential. B: Asymmetrical bone deposition occurs at the perimeter sutures with increased bone deposition directed away from the frontoparietal bone plate. C: Sutures adjacent to the fused suture (black arrows) compensate in growth more than those sutures not contiguous with the closed suture (white arrow). D: Enhanced symmetrical bone deposition occurs along both sides of the right coronal suture (arrows) which is a continuation of the prematurely fused left coronal suture.

rimeter sutures distant to the sutural stenosis. The ipsilateral lambdoid suture of unilateral coronal synostosis is a distant perimeter suture of low growth potential (Fig. 2C). Therefore, growth compensation is not appreciated and occipital symmetry appears to be maintained. Subtle asymmetry may be present as predicted by these rules, but examination of occipital bone growth in patients with unilateral coronal synostosis has not been conclusive. Contralateral occipital and temporal deformities are not usually seen with frontal plagiocephaly because the contralateral lambdoid and squamosal sutures are not perimeter sutures, and are not significantly involved in the compensatory growth process.

4. A non-perimeter suture that is contiguous to the prematurely fused suture undergoes enhanced symmetrical bone deposition along both edges. The patent contralateral coronal suture of plagiocephaly compensates with symmetrical growth along both sides (Fig. 2D). This growth combined with the asymmetrical bone deposition of the metopic and sagittal sutures produces the characteristic contralateral frontal and parietal prominence (Fig. 3).

Other Forms of Craniosynostosis

Metopic Synostosis

Premature closure of the metopic suture results in the well-recognized trigonocephaly because of deficient growth in both frontal bones. Symmetrical bone deposition occurs at the sagittal suture, but growth at the coronal suture is asymmetrical. Bone deposition is directed away from the restricted frontal bones. This enhanced growth of the parietal bones produces the characteristic biparietal pear shape of metopic synostosis (Fig. 4). The lambdoid and squamosal sutures are distant from the bifrontal restricted bone plate and do not participate in the compensatory growth process.
FIG. 3. *Left:* Photograph of a patient with left unilateral coronal synostosis demonstrating a flat left forehead and right frontal bulge. The *arrow* points to her prominent right parietal area which is contralateral to the fused suture. *Right:* Following surgical correction of the frontal abnormalities, her mild parietal prominence persists (*arrow*).

Bilateral Coronal Synostosis

The same observed rules for calvarial growth can be applied to both frontal plagiocephaly and the symmetrical deformities of bilateral coronal synostosis (Fig. 5). Two symmetrical frontoparietal bone plates are restricted in growth especially at the fused suture. The metopic and sagittal sutures are perimeter sutures of both bone plates. Asymmetrical enhanced bone deposition directed away from the restricted bone plate affects each side of the suture equally, resulting in symmetrical growth compensation. This symmetrical growth produces an abnormally tall head. However, unequal bone deposition occurs along the squamosal sutures producing the characteristic bilateral temporal bulge seen in turriencephaly. The lambdoid sutures do not participate significantly in the compensatory process because they have limited growth potential and are not adjacent to the coronal suture.

Sagittal Synostosis

With sagittal synostosis the principles of growth restriction and compensation are readily seen (Fig. 6). Premature closure of the sagittal suture results in a biparietal bone plate of restricted growth. Reduced growth in this bone plate results in a narrow biparietal dimension (the Virchow deformity). The coronal and lambdoid sutures, which are adjacent to this restricted bone plate, compensate by enhanced bone deposition at the edge of the frontal and occipital bones, respectively. The metopic suture, which is a contiguous non-perimeter suture, also compensates by symmetrical bone expansion along its suture line. As predicted by the rules of calvarial growth, the compensatory growth along the coronal, metopic, and lambdoid sutures produces the characteristic frontal and occipital prominence seen in sagittal synostosis. Since the squamosal sutures are distant from the fused suture and of limited growth potential, they do not participate significantly in the compensatory growth process. Therefore, the deformities of scaphocephaly do not include prominent bitemporal regions.
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The deformities resulting from only the anterior or posterior half of the sagittal suture being prematurely fused can also be predicted by these rules. The parietal bones remain restricted in growth. The characteristic frontal bulge of anterior sagittal synostosis is produced by the compensatory growth process of the adjacent coronal and metopic sutures (Fig. 7). Enhanced bone deposition of the occipital bones at the lambdoid suture creates the occipital prominence of posterior sagittal sutural synostosis without an associated significant anterior cranial vault deformity (Fig. 8).

**Lambdoid Synostosis**

Since bone deposition along the lambdoid suture is normally small compared to that along other calvarial sutures, lambdoid synostosis may produce only modest changes in skull shape. Clinical presentation of unilateral lambdoid synostosis includes a flattening of the ipsilateral parieto-occipital region as a result of a restricted parieto-occipital bone plate (Fig. 9 left). Significant compensatory growth occurs at the sagittal suture, contralateral lambdoid suture, and ipsilateral squamosal suture. Since the occipital bone has no discernible midline suture, it is joined to the left parietal bone to form a parieto-occipital bone plate with reduced growth potential. Therefore, the contralateral lambdoid and sagittal sutures act as perimeter sutures, with more bone deposited along the parietal border of the lambdoid suture and the contralateral border of the sagittal suture. Asymmetrical growth also occurs along the ipsilateral squamosal suture, with bone deposition directed away from the fused suture. The clinical result is an ipsilateral temporal bulge with anterior and inferior displacement of the pinna (Fig. 9 right).

With bilateral lambdoid synostosis, a similar pattern develops. Growth is restricted bilaterally in the parieto-occipital regions, and as predicted, enhanced asymmetrical deposition of bone along both squamosal sutures produces the characteristic bilateral temporal prominence with anterior and inferior displacement of the pinnae.

**Discussion**

The bones of the cranial vault originate from multiple ossification centers within the fibrous desmocranium that surrounds the embryonic brain. The calvarial bones continue to enlarge as the brain increases in size. Eventually, the margins of adjacent cranial bones achieve close approximation and sutural morphogenesis is initiated. Unlike facial bones, the fibrous desmocranium develops into the outer ectoperiosteal layer and inner dural layer, but does not envelope the margins of the cranial bones. At the margin of each cranial bone three distinct morphological zones develop: an inner mineralized zone, a midzone of osteoid accumulation, and an outer zone of cells undergoing differentiation. As the calvaria grows, the suture becomes a site for continuous bone deposition and resorption. An expanding, growing brain causes widening of these sutures.
FIG. 9. Left: Drawings showing the observed rules predicting the abnormalities of lambdoid synostosis. Direction of bone growth is indicated by the arrows. Right: Vertex view of a patient with left lambdoid synostosis demonstrating a mild left frontal prominence and anterior displacement of the left pinna (arrow). The flattening of the left occiput is not seen in this view. The left parietal scalp deformity is a capillary hemangioma.

and facilitates deposition of a layer of osteoid along the sutural bone margin. The osteoid layer becomes mineralized and transformed into bone. The calvarial bones continue to grow along their margins in response to an enlarging brain by reduplicating this process. Tensile forces generated from the growing brain and basocranium may be transmitted to the dura and influence sutural bone deposition. Moss demonstrated in the newborn rat that normal interfrontal (metopic) suture fusion could be inhibited by incision of the falx cerebri. Unlike Moss, Babler, et al., did not find a difference between accelerated growth in rabbits with coronal suturectomy and those that also had their dura and falk excised at the coronal suture. However, the adjacent frontonasal suture that is oriented parallel to the coronal suture demonstrated reduced growth approaching statistical significance. This suggests that sutural growth can be influenced by both changing tensile forces and alterations in growth at other sutures. Similarly, premature fusion of a cranial suture restricts growth of the calvarial bones involved and requires that other sutures compensate for the growing brain.

As proposed by Otto and Virchow, the observed rules for calvarial growth reiterate the hypothesis that the primary pathology of isolated vault craniosynostosis is premature fusion of a cranial vault suture. Virchow emphasized that growth is restricted in a plane perpendicular to the prematurely fused suture. Through his description of the various calvarial deformities produced in each type of craniosynostosis, he implied that perpendicularly oriented sutures have the capacity to compensate for the growing brain. However, he failed to describe how growth at compensating sutures is altered when a suture is prematurely fused.

Guidelines for predicting compensatory calvarial growth were developed after careful analysis of craniosynostosis deformities. Sutures adjacent to the fused suture appear to participate significantly in the compensatory growth process. Premature sutural fusion produces a bone plate of restricted growth potential. Sutures along the perimeter of this bone plate compensate by directing growth away from the sutural synostosis. Non-perimeter but adjacent sutures are also involved with enhanced symmetrical bone deposition along both edges.

These rules for calvarial growth are based primarily upon clinical material. Craniosynostosis case material consistently fits the patterns of abnormality predicted by these four rules. The rules assume that compensating cranial sutural edges have the capacity to grow asymmetically. Selman and Sarnat have also demonstrated unequal growth patterns in the normal frontonasal suture of rabbits. Normally, two-thirds of the growth that takes place at this suture occurs in the nasal bone while the remaining one-third involves the frontal bone. This asymmetrical growth may be attributed to the differing influences of the cranial base and facial skeletons. However, unequal bone deposition has also been demonstrated by Baer along the parieto-interparietal suture in albino rats. Using vital staining, he demonstrated that the growth increment along the posterior margin of the parietal bone is approximately twice as large as that measured from the anterior margin of the interparietal bone. Studies in animal models and humans to measure growth along each edge of compensating sutures are needed to determine if asymmetrical bone deposition occurs in craniosynostosis.

It is unclear whether these rules of calvarial growth apply to the cranial base abnormalities of craniosynostosis. Despite general descriptions of deformities asso-
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associated with coronal synostosis, detailed knowledge of cranial base abnormalities does not exist. Closer scrutiny of skull base morphology associated with craniosynostosis may disclose a pattern of skull growth that also predicts characteristic cranial base abnormalities not yet described.

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References


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