Cranial Base Changes Following Surgical Treatment of Craniosynostosis

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Three-dimensional osseous surface images from CT scans have been used to study the endo- and exocranial bases of 87 patients with a variety of craniosynostoses. Patients were studied prior to cranial surgery in infancy, perioperatively, and 1 year postoperatively. The dysmorphology of the endocranial base is diagnostically specific for synostosis of the metopic, sagittal, unicoronal, and bicoronal sutures. Cranio-orbital surgery in infancy for nonsyndromal solitary and bicoronal synostosis seems to induce normalization of endocranial symmetry in the first postoperative year. This normalization occurs to a lesser degree in patients with multiple synostoses. These findings suggest that the cranial base dysmorphology of craniosynostosis is a secondary manifestation of an undefined primary disorder. Furthermore, persistent postoperative dysmorphology may reflect abnormal neural rather than osseous growth.

Study of the dysmorphology of premature closure of cranial sutures generally has focused upon the cranial vault. Characteristic vault shapes have been recognized for single and some multiple sutural synostoses for over a century (Virchow, 1851). Description of these shapes has produced the standard nomenclature for synostoses, e.g., trigonencephaly, scaphocephaly (Marsh and Vannier, 1985). Furthermore, until the past decade, treatment for craniosynostosis was surgery of the cranial vault (Anderson and Geiger, 1965; Shillito and Matson, 1968). Although clinical attention has focused upon the cranial vault, the search for etiologic mechanisms has been directed toward the cranial base since the 1960s (Moss and Young, 1960; Kreiborg et al, 1976; Osterhout and Melsen, 1982; Burdi et al, 1986). The dysmorphology of the cranial base in craniosynostosis, however, is not well described. The limited in vivo data available have been restricted to the midsagittal plane by the use of cephalometric radiographs for analysis (Friede et al, 1983). The recent advent of new medical imaging technology now makes available for anatomic study both the endo- and exocranial bases (Marsh, Gado et al, 1986). Therefore, using this technology our investigation was undertaken to answer the following questions: (1) What is the endocranial base dysmorphology of craniosynostosis? (2) Is this dysmorphology suture specific? (3) Does cranial vault surgery affect this dysmorphology? and (4) What are the implications of the findings, if any, upon hypotheses regarding the pathogenesis of craniosynostosis?

MATERIALS AND METHODS

Three-dimensional osseous surface reformatations from CT scans, which included images of the endocranial base, became available at Washington University Medical Center in 1983 (Marsh and Vannier, 1983). The details of this technology have been reported previously (Vannier et al, 1983; Marsh et al, 1985; Marsh, Vannier et al, 1986). Since that time, 87 patients with craniosynostosis have had osseous surface processing. Some or all of the scans for 16 of these patients were processed retrospectively from archived CT data obtained prior to 1983. The remainder were processed synchronously with scan acquisition. Forty-seven percent of patients underwent CT scanning preoperatively and 1 year postoperatively. Perioperative postopera-
TABLE 1 Sequence of 3-D Images by Synostosis

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<tr>
<td>Bicoronal (N=10)</td>
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<tr>
<td>Multiple (N=26)</td>
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Results

Metopic Synostosis

The endocranial bases of all patients with isolated metopic synostosis had a tear drop configuration with a ventral mid saggital keel or prow (trigonencephaly). There was symmetric constriction of the anterior fossae and widening of the posterior fossae compared to control skulls (Fig. 2A). The magnitude of anterior fossa constriction varied from patient to patient. Deformity of the sphenoid ridge, consisting of acute angulation with respect to the anterior clinoids and absence of the lateral horizontal portion, was noted to varying degrees. The midlines of the anterior and posterior fossae were colinear in all cases with right-left symmetry of the hemicrania about the midsagittal plane. The three cranial fossae were right-left symmetric as well. The maximum width of the posterior fossae was more dorsal than in the normal skulls.

The ventral keel and anterior fossa constriction were absent on all perioperative images. The brow was flat and the lateral anterior fossae expanded compared to the preoperative images. The surgery acutely did not effect the size, shape, or symmetry of the middle or posterior fossae.

One year postoperatively (Fig. 2B), the cranial perimeter was more normal appearing for the two patients with longitudinal images. The posterior fossa no longer dominated the cranial width. The dimensions of the anterior fossa essentially were those achieved surgically and had not changed from those recorded perioperatively. In contrast, the endocranial base of a 3-year-old boy with untreated metopic synostosis shows persistent tear drop configuration with maximum width in the posterior fossa (Fig. 2C).

Sagittal Synostosis

The endocranial bases of all patients with isolated sagittal synostosis had apparent increased anteroposterior length and decreased bitemporal width (scaphocephaly) compared to control skulls. The actual length-to-width ratios were equal to normal skulls in half of the patients and either greater or lesser than normal, divided equally, in the remainder. A constriction, or excessive narrowing, was present at the pterion (junction of the greater wing of the sphenoid with the calvaria) in half of the cases (Fig. 3A). In the remaining cases, the cranial perimeter was a smooth curve resembling an elongated ellipse with expansion of the anterior fossae as compared...
FIGURE 1  Normal skull endocranial bases. 3-D osseous surface reformattions from CT scans of museum dry specimens. The neonate's endocranial base is dominated by the posterior fossa. As the infant grows, the anterior fossa enlarges in length and width. All fossae are relatively right-left symmetric, regardless of age, bisected by the colinear midsagittal planes of the anterior and posterior fossae. A, Neonate, B, 15- to 18-month-old infant, C, 8-year-old child. (Figs. 1A, B, and C courtesy of Marsh JL, Vannier MW. Comprehensive care for craniofacial deformities. St. Louis: CV Mosby, 1985:105).
FIGURE 2  

A, Endocranial base 3-D images of a 2-month-old girl with untreated metopic synostosis. The perimeter configuration is trigonocephalic. The anterior fossae are compressed, and the cribriform plate is depressed caudally. The horizontal portions of the greater wings of the sphenoid are absent. 

B, Same patient 1 year after frontal bone recontouring, bilateral superolateral orbital advancement and recontouring, and extended bicoronal craniectomies performed at 3 months of age. The endocranial configuration is normalized with respect to shape and location of maximum width. The anterior fossae remain anomalous in spite of their expansion. 

C, Endocranial base 3-D image of a 3-year-old boy with untreated metopic synostosis. Although the trigonencephaly is not as marked as for the patient in Figure 2A, the endocranial configuration remains more anomalous than that seen in Figure 2B.
to normal skulls (Fig. 4A). The midlines of the anterior and posterior fossae were colinear in all cases with right-left symmetry of the hemicrania about the midsagittal plane. The three cranial fossae were right-left symmetric as well.

The operation which we perform for sagittal synostosis does not involve the cranial base.

One year postoperatively (Figs. 3B and 4B), the cranial shape was more normal appearing in a majority of cases. Pterion constriction persisted in those cases in which it was present preoperatively. Actual length-to-width ratios were normal for half of the patients and greater than normal for the remainder.

**Unicoronal Synostosis**

The endocranial bases of all patients with isolated unicoronal synostosis were asymmetric without a flat midsagittal plane (plagiocephaly) compared to normal skulls (Fig. 5A). The brow ipsilateral to the synostosis was recessed, and the contralateral brow was protrusive, reflecting the compression of the ipsilateral and expansion of the contralateral anterior fossae respectively. Reciprocal changes were present in the posterior fossae with expansion ipsilateral and compression contralateral to the synostosis. The posterior fossae findings were not as consistent as those of the anterior fossae. Thus, the perimeters of the hemicrania were asymmetric. The midlines of the anterior and posterior fossae were not colinear; they were angulated toward the side of the synostosis an average of 9 degrees (Marsh, Gado et al, 1986). None of the three cranial fossae were right-left symmetric and the anterior fossa was the most asymmetric.

The anterior fossa had become essentially symmetric on all perioperative images. The brow was flat, and the lateral anterior fossa ipsilateral to the synostosis was expanded compared to the preoperative images. The surgery acutely affected neither the angulation of the anterior and posterior fossa midlines nor the size, shape, or asymmetry of the middle and posterior fossae.

One year postoperatively, the cranial perimeter was more symmetric and normal appearing in all cases (Fig. 5B). The improved symmetry of the

![FIGURE 3 A, Endocranial base 3-D image of an 8-month-old girl with sagittal synostosis. There is indentation or constriction of the cranial perimeter at the pterion bilaterally. There is apparent and absolute scaphocephaly (length/width = 1:0.55 vs 1:64 for age-matched control). The anterior cranial fossa is large and the posterior fossa narrow for an infant this age. B, Same patient 1 year after extended sagittal, bicoronal and bilambdoid craniectomies performed at 10 months of age. The cranial configuration is normalized with respect to shape and length-to-width ratio. The anterior fossa remains relatively large for age; however, the posterior fossa has increased in width. The pterion constriction persists. (Figs. 3A and 3B are courtesy of Marsh JL, Vannier MW. The anatomy of cranioorbital deformities of craniosynostosis: insights from 3-D images of CT scans. Clin Plast Surg 1987; 14: )](image-url)
The asymmetry of the middle and posterior fossae was less apparent. Two populations were discernible based upon angulation of the midlines of the anterior and posterior fossae. The first had normalization of the angulation with the average angle reduction of nine degrees. The angulation essentially remained unchanged in the second population. The significance of these two populations is unknown at this time. The postoperative normalization of cranial perimeter configuration and anterior-posterior fossa angulation are not seen on an adult museum skull with untreated unicoronal synostosis (Fig. 5C).

Bicoronal Synostosis

The endocranial bases of all patients with isolated bicoronal synostosis had both apparent and absolute decreased anteroposterior length and increased bitemporal width (brachycephaly) compared to normal skulls (Fig. 6A). The maximum skull width was ventral to that of the normal skull. The midlines of the anterior and posterior fossae were colinear with symmetric hemicrania in all cases. Although each of the three cranial fossae were right-left symmetric as well, the anterior fossae were constricted and the middle fossae expanded compared to normal skulls.

The anterior fossa was expanded ventrally and laterally on all perioperative images. The surgery acutely did not affect the size, shape, or symmetry of either the middle or posterior fossae. One year postoperatively, the cranial perimeter was more normal appearing (Fig. 6B). The maximum skull width had moved dorsally from the region of the pterion to that of the petrous ridges. While the surgically created expansion of the anterior fossae persisted, these fossae still appeared somewhat compressed compared to normal skulls. In contrast, the endocranial base of a 6-year-old boy with untreated bicoronal synostosis has persistent brachycephaly with marked middle fossae widening and anterior fossa compression (Fig. 6C).

Lambdoid Synostosis

There was inadequate material and follow-up of patients with lambdoid synostosis for definition of the endocranial dysmorphology or the effect of cranial vault surgery.
FIGURE 5  A, Endocranial base 3-D image of a 6-week-old girl with right unicoronal synostosis. All cranial fossae are asymmetric. The midline of the anterior fossa is angulated 14 degrees toward the side of the synostosis with respect to the midline of the posterior fossa. The right forehead is retracted and the right anterior fossa compressed; the left is protruded ("bossed") and the fossa expanded. The arcs of the wings of the sphenoid are asymmetric. B, Same patient 1 year after frontal bone recontouring, right superolateral orbital advancement and recontouring, and extended bicoronal craniectomies performed at 3 months of age. The symmetries of the cranial perimeter and the respective fossae are improved. The angulation of the anterior and posterior fossae midlines has decreased to four degrees. (There is a three-degree angulation in the 15 to 18 month normal skull, see Fig. 1B). (Figs. 5A and 5B are courtesy of Marsh JL, Vannier MW. The anatomy of cranioorbital deformities of craniosynostosis: insights from 3-D images of CT scans. Clin Plast Surg 1987, 14:Jan.). C, Endocranial base photograph of an adult museum specimen with untreated left unicoronal synostosis. (The image has been right-left reversed to facilitate comparison with the other figures). The angulation of the fossa midlines and asymmetries of the cranial perimeter and fossae as seen in our affected infant (Fig. 5A) are persistent in this untreated adult (skull courtesy of Timothy Turvey, D.M.D.).
FIGURE 6  

A, Endocranial base 3-D image of a 4-month-old girl with sporadic bicoronal synostosis without craniofacial dysostosis syndrome. There is apparent and absolute brachycephaly (length-to-width ratio of 1:0.86 versus 1:0.64 for normal). The maximum skull width is displaced ventrally to the pterions. The anterior fossae are compressed, the middle fossae expanded, and the posterior fossae compressed compared to the normal. 

B, Same patient 1 year after frontal bone recontouring, bilateral superolateral orbital advancement and recontouring, and extended bicoronal craniectomies performed at 4 months of age. The cranial perimeter configuration is normalized. While the length-to-width ratio is improved, it still exceeds normal (0.75 versus 0.60 for age). The proportions of the anterior, middle, and posterior fossae also are normalized. 

C, Endocranial base 3-D image of a 6-year-old boy with untreated sporadic nonsyndromal bicoronal synostosis. There is marked brachycephaly. The middle fossae are widened excessively. The anterior fossae are compressed and the brow is retruded. The cranial configuration is an exaggeration of the dysmorphism seen in our untreated infant (Fig. 6A). (Courtesy of Marsh JL, Vannier MW. Comprehensive care for craniofacial deformities, St. Louis: CV Mosby, 1985:150).
Multiple Synostoses

The endocranial bases of patients with multiple synostoses were similar in having decreased length-to-width ratios (brachycephaly), compressed-to-almost-absent anterior fossae and markedly enlarged middle fossae as compared to normal skulls (Fig. 7A). When the anterior fossae were absent, the most ventral portion of the cranial base consisted of the sphenoid wings and the glabella. The cribriform plate, the sole remainder of the anterior fossae in such cases, was displaced caudally. The amount of anterior fossa did not correlate with either the specific sutural synostoses or the presence and type of associated syndrome. The sutures and fontanelles about the occipital and sphenoid bones were widely patent in most cases.

The anterior fossa was expanded ventrally and laterally on all perioperative images. The surgery acutely did not affect the size, shape or symmetry of either the middle or posterior fossae.

One year postoperatively, the cranial perimeters were rounder with brachycephalic proportions (decreased length-to-width ratio) (Fig. 7B). However, unlike the brachycephaly of bicoronal synostosis, the maximum skull width was at the petrous ridges rather than the more ventral pterions. The surgical ventral and lateral enlargement of the anterior fossae persisted unchanged. The expansion of the anterior fossae, which normally occurs over the first year of life, was absent in these cases of multiple synostoses. The endocranial base of a 23-month-old girl with untreated sporadic metopic, bicoronal, and bilambdoid synostoses had excessive width and depth of the middle and posterior fossae (Fig. 7C).

FIGURE 7 A, Endocranial base 3-D image of a 2-month-old boy with metopic and sagittal synostosis in association with Carpenter syndrome. The configuration is brachycephalic. The cribriform plate is the only vestige of the anterior fossa. The middle and posterior fossae are widened. The sutures and fontanelle are widely patent about the sphenoid and occipital bones. B, Same patient 2 years after frontal bone revision, bilateral superolateral orbital advancements, extended bicoronal, sagittal, and squamosal craniotomies, and partial skull reconstruction with biparietal floating bone grafts, performed at 3 months of age. Although an anterior fossa has been created, it has not enlarged since the operative procedure. There has been no significant change in the cranial configuration from that achieved perioperatively. C, Endocranial base of a 23-month-old girl with untreated sporadic metopic, bicoronal and bilambdoid synostoses in association with osteogenesis imperfecta and camptomelic dwarfism. While the cranial perimeter is brachycephalic (reduced length-to-width ratio), the configuration differs from the brachycephaly of isolated bicoronal synostosis in that the excessive width is in the posterior third rather than the anterior third of the skull. The middle and posterior fossae are both enlarged and excessively deep. (Figures 7A and 7C are courtesy of Marsh JL, Vannier MW. Comprehensive care for craniofacial deformities. St. Louis: CV Mosby, 1985:163-168).
CONCLUSIONS

Three-dimensional osseous surface reformat- tions from CT scans are a useful tool to study the dysmorphology of the endocranial base. These images document a dysmorphology in patients with craniosynostosis that is diagnostically specific for premature closure of the metopic, sagittal, unicoronal, and bicoronal sutures. Although there is a dysmorphology characteristic for the set of patients with multiple suture synostoses, this dysmorphology does not seem to be diagnostic for specific combinations of synostoses. Cranio-orbital surgery within the first 6 months of life for patients with nonsyndromal solitary and bicoronal synostoses seems to result in normalization of endocranial symmetry. The improvement in symmetry affects the calvarial perimeter, the hemicrania, and the individual cranial base fossae. There is a lesser degree of normalization of the anterior fossa than of the middle and posterior fossae, especially in patients with metopic, bicoronal and sagittal synostoses. While syndromal patients with multiple synostoses have more normally configured endocranial bases after cranio-orbital procedures, compared to similar untreated individuals, their endocranial bases remain quite dysmorphic. These findings suggest that the dysmorphology of the cranial base is a secondary manifestation of an undefined primary disorder in craniosynostosis. If the brain has normal growth vectors, surgical release of the vault constriction caused by the synostosis will allow remodelling of the endocranial base as well as the vault. The minimal postoperative anterior fossa expansion in metopic bicoronal and multiple synostoses may reflect impaired growth of the frontal lobes rather than osseous impediment within the skull.

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REFERENCES