The surgical correction of coronal and metopic craniosynostoses

JEFFREY L. MARSH, M.D., AND HENRY G. SCHWARTZ, M.D.

Department of Surgery, Plastic and Reconstructive, Department of Pediatrics, and Department of Neurology and Neurological Surgery, Cleft Palate and Craniofacial Deformities Institute, Washington University School of Medicine, St. Louis, Missouri

Procedures for relief of coronal synostosis and metopic synostosis have resulted in some undesirable sequelae. The authors present combined neurological and plastic surgical modifications to prevent additional synostoses, forehead ridging, and lateral orbital wall step-off. They recommend bifrontal craniotomy with lateral wall osteotomy into the body of the zygoma and self-retaining grafts.

KEY WORDS: craniosynostosis □9 craniofacial deformities □9 skull □9 skull deformity □9 orbit □9 synostosis

VIRCHOW recognized that premature stenosis of a cranial suture stops bone growth perpendicular to that suture and impairs radial growth of the calvaria. The resultant physical deformity may involve the orbits and face as well as the cranium, thereby producing subsequent psychosocial difficulties. The traditional surgical treatment for craniosynostosis, namely strip craniectomy, has produced inconsistent results with respect to suture patency, correction of cranial deformity, and prevention of orbitofacial deformity. Our experience with these procedures, however, has revealed new problems: induction of an additional synostosis, midline forehead ridging, lateral orbital wall step-off, and palpable fixation wires. We report our modifications to overcome the untoward sequelae we have experienced with the Hoffman procedure for unilateral coronal synostosis and the Marchac procedure for metopic synostosis.

Summary of Cases

This series includes 21 consecutive cases of infants who underwent surgical treatment for craniosynostosis within the first 19 months of life (Table 1). All patients underwent pre- and postoperative multidisciplinary evaluations at the Cleft Palate and Craniofacial Deformities Institute, St. Louis Children's Hospital, Washington University Medical Center. All operations were performed by a single plastic surgeon in conjunction with one of four associated neurosurgeons. None of the patients had intra- or perioperative complications. All patients were discharged from the hospital within 1 week following surgery.

The first seven patients (Cases 1 to 7) underwent the procedures of either Hoffman and Mohr or Marchac as described in the literature for coronal or metopic synostosis, respectively. Of the patients with coronal synostosis, the fusion was unilateral in one and bilateral in two, one of whom also had midface retrusion. The average age for surgery in these three patients was 8.7 months. One of the remaining four infants with metopic synostosis also had sagittal synostosis and underwent sagittal strip craniectomy as well as the Marchac procedure. The average age at surgery in these four patients was 6.0 months. The remaining 14 patients (Cases 8 to 21) underwent the modified operations reported herein. Five of these infants had unilateral coronal synostosis. An additional five had bilateral coronal synostosis. Of these, two also had midface retrusion. The average age for surgery for coronal synostosis for these 10 infants was 5.4 months. Two patients had isolated metopic synostosis. One had metopic and sagittal synostosis. Their average age at surgery was 4.9 months. While both the standard coronal and metopic groups were older at the time of surgery than the groups with modified techniques, the differences of 3.3 and 1.1 months, respectively, are unlikely to account for the untoward
results observed in the standard groups. The follow-up period in all patients was at least 18 months.

**Modified Operations for Coronal and Metopic Synostosis**

**Evolution of Surgical Procedures**

Linear craniectomy was introduced by both Lannelongue and Lane in the 1890's to decompress the presumed increased intracranial pressure (ICP) thought to be the necessary consequence of premature cranial suture closure. The procedure was abandoned shortly thereafter because of poor results and high mortality. Today it is believed that their results reflect a failure to separate microcephaly from craniosynostosis. Faber and Town revived strip craniectomy in 1927, and later recommended surgery within the first 3 months of life to minimize the risk of blindness and other sequelae, and to optimize the results. The inclusive term "craniosynostosis" was introduced by Sear in 1937 to describe all varieties of premature closure of the cranial sutures in lieu of Virchow's restrictive term "craniosstenosis," which meant narrowed or strictured skull. Technical modifications in strip craniectomy, from its revival in the 1930's until the development of craniofacial surgery in the 1970's, have been characterized by the attempt to prevent rapid reossification of the resected suture. Metallic foil, acid chemicals, and plastic sheeting have been proposed, popularized, and then abandoned in turn as the solution to rapid osseous bridging. Furthermore, in the early 1960's, Hemple, et al., and Freeman and Borkowf began to question the actuality of increased ICP in patients with craniosynostosis, especially isolated affected sutures. In turn, they questioned the indication for strip craniectomy.

In the 1970's, attention was directed to individualization for operative correction of specific craniosynostoses. Seeger and Gabrielsen, in a radiological report published in 1971, drew attention to the absence of hard evidence of increased ICP, such as erosion of the sella turcica or clinical signs, when accentuated digital skull markings were observed in patients with isolated coronal synostoses. Moreover, they reiterated Bertelsen's observation of the continuity of the frontosphenoidal, sphenethmoidal, and coronal sutures; they suggested that release of this entire suture ring would be necessary to allow normal anterior fossa expansion. In fact, this approach had been advocated by Anderson and Geiger in 1965 to improve the poor results in patients aged 3 months or older who underwent unilateral coronal synostosis treated with simple coronal craniectomy and sphenoid wing resection. Hoffman and Mohr extended this technique to include advancement of the supralateral orbit in a further attempt to achieve adequate anterior fossa expansion. Their procedure has become the standard in contemporary craniofacial centers, as it most consistently achieves normalization of the frontal and orbital deformities on the side of the coronal synostosis.

The flattening of the nasofrontal angle in bicoronal synostosis was addressed by Stricker, et al., who advocated restoration of the angle by rocking the supraborital ridge forward upon the nasion. This technique was subsequently modified and popularized by Marchac. In the same paper, Marchac described recontouring of the supralateral orbit and forehead to correct the de-
Surgery for coronal and metopic craniosynostoses

FIG. 2. Case 11. Computerized tomography scan analysis of unilateral coronal synostosis and its surgical correction in a 13-month-old girl (same patient as shown in Fig. 1). Comparable scans at the level of the floor of the anterior cranial fossa (left), the superior orbit (center), and the midorbit (right) are shown. Upper Scans: Preoperative study demonstrating left frontal protrusion (bossing), right frontal recession, and asymmetry of the greater wings of the sphenoid and orbits. Lower Scans: Scans taken 2 weeks after extended bicoronal craniectomies show bilateral frontal bone revision and right supralateral orbital advancement. The self-retaining tenon-in-mortise bone grafts are seen on the center and right images (double arrows), lateral to the osteotomy of the right orbit lateral wall (arrowheads).

formities of metopic synostosis. This technique has become the accepted procedure for patients with trigonocephaly because it addresses the orbital deformity as well as that of the cranium. While the need for orbital correction has recently been questioned by Shillito, our computerized tomography (CT) studies of patients with metopic synostosis consistently demonstrate posterior displacement of the lateral orbital wall with post- lateral lateral rotation of the orbit itself as compared to unaffected individuals.

Unilateral Coronal Synostosis

Unilateral coronal synostosis produces bilateral deformity (Fig. 1). The asymmetry of the resultant distortions of the skull, orbit, and midface reflects two different mechanisms of deformity. On the side of the coronal fusion, impairment of bone growth thwarts the forward expansion of the anterior cranial fossa, resulting in a recessed brow with a flattened forehead (Figs. 2 and 3). Ipsilateral frontal lobe growth vectors are redirected superiorly, inferiorly, and contralaterally. The superior pressure elongates the forehead. The inferior pressure alters the underlying middle cranial fossa with forward bowing of the greater wing of the sphenoid (Figs. 2 and 3). This results in effacement of the adjacent temporal fossa with shortening of the lateral wall of the orbit as well as direct encroachment upon the orbital volume by the middle cranial fossa, thereby producing proptosis. The floor of the anterior fossa remains elevated, creating the “harlequin” orbit. On the side opposite to the coronal synostosis, redirected growth vectors produce the contralateral deformity. Clinical manifestations of this secondary deformity vary a good deal among patients, presumably reflecting different timing in the occurrence of the synostosis in utero. The “normal” forehead is usually pushed forward (bossed) (Figs. 2 and 3). The orbit may appear malpositioned (dysopia) with inferolateral displacement. This asymmetry in orbital position presumably reflects obstruction to the normal embryonic migration of the orbit from lateral to medial by the pressure of the frontal lobe. Finally, there is often asymmetry of the midface as well, which seems to correlate with torque in the clivus.

The major limitation of the Hoffman and Mohr procedure for the treatment of unilateral coronal synostosis is that it does not correct the contralateral deformity. The recommended midline osteotomy to mobilize the unilateral frontal bone flap has resulted in a visible forehead ridge with forehead asymmetry in one of our patients so managed (Case 8). Consequently, we have adopted the procedure of bifrontal craniotomy to recontour both frontal bones. The six patients who have undergone this modification over the past 2 years all have smooth symmetrical foreheads (Fig. 1). The recommended medial greenstick fracture of the brow segment is at times uncontrolled, thereby producing an undesirable fracture. The medial “hinge” is now cut with a saw to insure a correct position. A more subtle limitation of the Hoffman procedure is its failure to correct the recession of the lower lateral orbital wall. Although the zygomatic process of the frontal bone (the superior third of the lateral orbital rim) is advanced in continuity with the superior orbital rim in the Hoffman
technique, the frontal process of the zygoma (the inferior two-thirds of the lateral orbital rim) is unaltered. This may result in a lateral rim step-off irregularity and fails to affect the frequently associated malar retrusion. In addition, the lateral canthus, which is attached to a tubercle on the frontal process of the zygoma, is in fact not advanced as its insertion is unaltered. McCarthy, et al., extended the lateral wall osteotomy inferiorly to the level of the inferior orbital fissure to remedy this problem. Similarly, we have extended the lateral orbital wall osteotomy through the zygomaticofrontal suture into the body of the zygoma. A greenstick fracture, rather than an osteotomy, is then made at the malar block allowing the entire lateral orbital rim and underlying soft tissue, such as the lateral canthus, to advance with the superior rim. Finally, based upon recently created three-dimensional osseous surface CT images, we now lower the supralateral orbit on the affected side as well by removing bone from the nasion and lateral orbital wall (Fig. 3). Absorbable sutures rather than wire are used to fix the mobilized orbit and recontoured frontal bone. The early results with respect to rim configuration have been satisfactory over the past 2 years; however, the ultimate effect upon the midface asymmetry will require study of these infants into adolescence. Claims regarding beneficial, or for

**FIG. 3.** *Upper:* Preoperative three-dimensional osseous surface reconstructions of a 5-month-old girl with right unilateral coronal synostosis. The periorbital osteotomies are indicated by a solid line; the bone resections with stippling. The images were produced from standard axial computerized tomography scans by the method of Marsh and Vannier. *Upper Left:* Frontal projection. Verticalization of the orbital long axis and temporal bulging are seen on the right. While the superior and inferior orbital fissures are visible in the left orbit (as is usual in normal individuals in the frontal view), the fissures are obscured on the right by the incomplete mesial migration of the orbit due to the synostosis. *Upper Center:* Right lateral projection. *Upper Right:* Top view projection. The top of the calvaria has been made transparent to allow inspection of the cranial base. The recession of the right frontal bone, protrusion of the left frontal bone, compression of the right anterior cranial fossa, and ballooning of the right temporal fossa are seen. *Lower:* Two weeks after extended bicoronal craniectomies, bilateral frontal recontouring, and right supralateral orbital advancement (7 mm for the midpoint of the orbital roof) with caudal displacement (5 mm). *Lower Left:* Frontal projection. The bicoronal craniectomies are seen between the triangular recontoured “frontal” plate and the unaltered midcranial vault. Note the equalization of the orbital rim configuration. *Lower Center:* Right lateral projection. The tenon-in-mortise self-retaining bone graft (dashed line) in the lateral orbital wall osteotomy maintains the orbital advancement. The osteotomy extends to the body of the zygoma. The brow contour has been augmented with a calvarial onlay graft. *Lower Right:* Top view projection showing expansion of the right anterior cranial fossa, equalization of the brow contour, and autogenous calvarial bone grafts (dashed lines) in the orbital roof and lateral orbital wall.
that matter detrimental, effects on facial growth for certain craniofacial procedures performed in infancy must wait until 10 to 15 years of follow-up data are available.

A final modification has been made to prevent rapid resynostosis. Hoffman and Mohr 9 described placing a calvarial bone graft strut between the supralateral orbital rim and the parietal bone to maintain their advancement. The placement of Silastic sheeting on the parietal bone edge was recommended to prevent fusion of the graft and subsequent tethering. Early suture re-stenosis requiring repeat craniectomies occurred bilaterally in one of our patients with bicoronal synostosis who had such grafts (Case 1). We have had similar problems with the temporal bone-slot procedure of Tessier. 22 Nonetheless, in our experience some buttressing of the supralateral orbital advancement is necessary in all cases to prevent relapse from the tension of scalp closure. This is in contradistinction to the recommendation of Anderson, 1 who stated that struts are necessary only in patients over 6 months of age or with severe

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**Fig. 4.** Case 18. Left: Photograph of a 1-month-old baby girl with non-familial bicoronal synostosis. Note the proptosis on the lateral view. The maxillary-mandibular relationship is normal. Right: The patient 2 weeks following bilateral extended coronal craniectomies, bilateral frontal bone revision, and bilateral supralateral orbital advancement. Note the change in forehead profile and the brow-globe relationship.

**Fig. 5.** Case 18. Computerized tomography scan analysis of bilateral coronal synostosis and its surgical correction in a 3-month-old baby girl (same patient as shown in Fig. 4). Comparable scans are shown at the level of the upper anterior cranial fossa (left), orbital roofs (center), and midorbit (right). Upper: Preoperative studies demonstrating the asymmetry in frontal recession and proptosis. Lower: Scans taken 2 weeks after bilateral extended coronal craniectomies, bilateral frontal bone revision, and bilateral supralateral orbital advancement with self-retaining tenon-in-mortise bone grafts (double arrow). The orbital roof defects, documenting the increase in anteroposterior orbital dimension, are seen on the center scan (arrowheads).
A wide craniectomy, of at least 2 cm, is performed between the advanced supralateral orbit and the temporal cranium to avoid rapid reossification with restenosis.

**Bilateral Coronal Synostosis**

Careful study of the appearance and CT scans of individuals with bicoronal synostosis documents varying degrees of asymmetric deformity in most cases (Figs. 4 and 5). The greater part of the deformities observed can be explained by the pathological mechanism presented above for the primarily affected side in unilateral coronal synostosis. We hypothesize that the asymmetries reflect temporal variance from side to side in the occurrence of the synostosis in utero, and therefore differing degrees of secondary contralateral effects.

We currently begin the correction of bilateral coronal synostosis with a bifrontal craniotomy. The two frontal bone plates are usually separated to achieve the best frontal configuration after rotation and modeling of the plates. The right and left supralateral orbits are mobilized as described previously for unilateral coronal synostosis. A greenstick fracture of the frontal process of the zygoma is made at the malar block, and the advanced orbit is self-retained with a tenon-in-mortise...
bone graft. When the degree of orbital deformity is asymmetrical, differential advancement is performed to achieve osseous symmetry.

Metopic Synostosis

We perform the procedure of Marchac for the correction of trigonocephaly, with two modifications. We extend the lateral orbital wall osteotomy into the body of the zygoma, and place the self-retaining tenon-in-mortise low lateral orbital wall calvarial buttress graft (Figs. 6 and 7). We believe it is important to keep the buttress graft below the transition between the coronal (frontotemporal) and the sphenozygomatic (lateral orbital wall) sutures to prevent the inadvertent induction of secondary coronal synostosis. We have treated one patient with trigonocephaly who developed late acrocephaly in which the buttress ran from the suprolateral orbit to the squamosal temporal bone (Case 5). Our current technique has been easily extended to include sagittal craniectomy when there is concurrent scaphocephaly, as in Case 8.

Comment

The surgical correction of craniosynostosis has evolved from simple removal of the suture, purportedly to prevent brain damage, to cranio-orbital recontouring for an optimal aesthetic result. The most recent surgical advances reflect a union of neurological and plastic surgery through the format of multidisciplinary teams to provide care for craniofacial deformities. The procedures of Hoffman for unilateral coronal synostosis and Marchac for metopic synostosis exemplify such procedures. In our experience, however, there have been untoward sequelae from these operations as described: induction of additional synostosis, midline forehead ridging, lateral orbital wall step-off, and palpable fixation wires. We report 14 patients in whom our surgical modifications have prevented these sequelae. We emphasize avoidance of osseous struts between the pterion and temporal or parietal bones, bifrontal craniotomies, extension of the lateral wall osteotomy into the body of the zygoma, and use of self-retaining grafts and absorbable sutures in lieu of wire fixation. The combined neurological and plastic surgical approach to craniosynostosis can release radial brain and calvarial growth vectors, resulting in a more normal-appearing craniofacial configuration.

References

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