Surgical Correction of Severe Scaphocephalic Deformities

Fernando D. Burstein, MD*
Roger J. Hudgins, MD†
Steven R. Cohen, MD*
William R. Boydston, MD, PhD†

Atlanta, Georgia

Sagittal synostosis may result in severe skull deformities. Characteristic components of the deformity include extreme elongation, frontal and occipital bossing, temporal pinching, and angulatory apical skull deformation. Conventional strip craniectomy often fails to correct these complex problems completely in severe early or late cases of sagittal synostosis. Techniques for total calvarial vault reconstruction have previously been reported, but a single large series has not been presented. Eighteen consecutive patients ranging in age from 3 months to 5 years (mean = 12 months) with severe early and late scaphocephalic skull deformities underwent total calvarial vault reshaping. All children required transfusions ranging from 250 to 1,100 mL. Operative times averaged 6 hours, and hospital stay ranged from 4 to 7 days. There was no perioperative mortality. Two patients experienced transient syndrome of inappropriate secretion of antidiuretic hormone, which responded to fluid restriction. One patient was noted to have a 2-cm parietal craniectomy defect 9 months after operation. Microscrews, which were used in all 18 patients, had to be removed in 2 patients when they became palpable. Excellent aesthetic results were noted in all 18 patients up to 36 months of follow-up.

Key Words: Sagittal synostosis, scaphocephalic deformities

Sagittal synostosis, with its characteristic oblong calvarial shape (scaphocephaly), is the most common type of craniosynostosis [1–3]. The severity of the calvarial deformity varies from slight cranial elongation with sagittal ridging to extreme elongation, with a large occipital shelf and pronounced frontal bone bossing. Persing, Jane, and Edgerton [4] suggested that the deformity is not limited to the sagittal suture but also involves the base of the skull, which in part contributes to the degree of the cranial dysmorphology. Many neurosurgeons and craniofacial surgeons consider strip craniectomy of the sagittal suture to be the standard of surgical treatment for early sagittal synostosis [5]. Some investigators reported excellent results when the initial deformity is mild and surgery is performed within the first few months of life [5,6]. Others, however, reported unsatisfactory results in children of any age when the scaphocephalic deformity is severe, particularly when associated with large occipital shelves and marked frontal bone bossing [7–9].

At Scottish Rite Children's Medical Center, we have found strip craniectomy to provide unsatisfactory results, even when performed within the first few months of life in infants with marked scaphocephalic deformities (Fig 1). Because of its high failure rate, strip craniectomy in children with scaphocephaly who are diagnosed after 18 months of age is generally not indicated. Children presenting with late scaphocephalic deformities often have marked frontal bossing, large occipital shelves, temporal pinching, and severe angulatory deformities at the apex of the skull (Fig 2). Because the majority of the brain growth has already taken place and the deformity is fixed, strip craniectomy has little or no effect on outcome [10]. To improve surgical results, Marchac performed near-total calvarial vault reconstruction using a transposition technique in older patients with scaphocephaly [11]. Persing, Jane, and Edgerton described a technique for complete one-stage correction of severe scaphocephalic deformities in 1989 but did not report a large series or an analysis of their results [4]. Marsh and Vannier [12] discussed a group of patients with scaphocephaly and quantitated their radiographic results but did not focus on perioperative problems or technical nuances of surgical management. We recently reported our initial results using a modification of their technique in patients older than 18 months [13]. Herein, we report the results of total calvarial vault reconstruction in 18 consecutive patients over a 3-year period.

From the Departments of *Plastic Surgery (Craniofacial Surgery) and † Pediatric Neurosurgery, Scottish Rite Children's Medical Center, Atlanta, GA.

Address correspondence to Dr Burstein, Department of Plastic Surgery, Scottish Rite Children's Medical Center, 5455 Meridian Mark Rd, Suite 200, Atlanta, GA 30342.

A modified surgical approach is recommended to both maximize protection to the frontal branch of the facial nerve and minimize dissection of the fat pad (Fig 4). As described previously, elevation of the coronal flap in the temporal region proceeds in the subaponeurotic plane to a level approximately 2 cm above the zygomatic arch. The middle temporal artery and vein can be seen in some patients, coursing within the fat pad deep to the superficial layer of the temporal fascia. The superficial layer of the fascia is incised below this level, and dissection proceeds inferiorly immediately on the undersurface of the fascia to the periosteum of the zygomatic arch. The superficial temporal fat pad is maintained in its normal location, and dissection within the fat is minimized. At the conclusion of the reconstructive procedure, the transected superficial layer of the deep temporal fascia is repaired.

Conclusion

We presented 6 patients with obvious temporal contour deformities after bicoronal flap elevation and exposure of the lateral craniofacial skeleton. The deformity results from atrophy or prolapse of the superficial temporal fat pad. In the future, the surgical approach to the lateral craniofacial skeleton will have to be done not only with the frontal branch of the facial nerve in mind but also the superficial temporal fat pad. Meticulous dissection of the fascial layers, preservation of the fat pad in its normal anatomical location, and avoidance of the middle temporal artery are critical.

Presented at the 3rd Congress of the International Society of Craniofacial Surgery, Santiago de Compostella, Spain, June 1991.

References

1. Khosla VM. Pseudoankylosis of the mandible produced by temporal muscle fibrosis: report of a case. J Oral Surg 1970;28:521
2. Summers L. False ankylosis of the temporomandibular joint following craniotomy. Br J Oral Surg 1985;18:138
3. Coonan TJ. Ankylosis of the temporomandibular joint after temporal craniotomy: a cause of difficult intubation. Can Anaesth Soc J 1985;32:158
4. Mitz V, Peyronie M. The superficial musculoaponeurotic system (SMAS) in the parotid and cheek area. Plast Reconstr Surg 1976;58:80
5. Abul-Hassan HS, von Drasek Asched G, Acland RD. Surgical anatomy and blood supply of the fascial layers of the temporal region. Plast Reconstr Surg 1986;77:17
6. Stock AL, Collins HP, Davidson TM. Anatomy of the superficial temporal artery. Head Neck Surg 1980;2:466
7. Baker DC, Conley JC. Avoiding facial nerve injuries in rhytidectomy: anatomic variations and pitfalls. Plast Reconstr Surg 1979;64:781
8. Stuzin JM, Wagstrom L, Kawamato HK, Wolfe SA. Anatomy of the frontal branch of the facial nerve: the significance of the temporal fat pad. Plast Reconstr Surg 1989;83:265