hydroxyapatite grafts, Proplast, and silicone have been used frequently, but all have some disadvantages, such as resorption and extrusion.[10–12] Maxillofacial contour could also be corrected by autologous tissues, such as dermofat graft or bone. However, in our patient soft tissue augmentation was not considered to be a solution because the main problem was the underlying bony contour deformity. Moreover, bone grafting has several disadvantages, such as increased operation time, donor site morbidity, and resorption to some extent.[12,13] Therefore, alloplastic implant was preferred. Medpor implant integrates with surrounding soft tissue and does not undergo resorption. It is a good alternative for restoration of maxillofacial contour deformities.[14–17] The long-term follow-up of the patient reported here revealed good results with symmetric facial appearance.

In conclusion, differential diagnosis between ACS and Goldenhar syndrome should be taken under consideration, and when the diagnosis is confirmed, both the auricular and the mandibular defects should be corrected. Here, we presented two simple solutions that offer satisfactory results for the patient.

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The Bowstring Canthal Advancement: A New Technique to Correct the Flattened Supraorbital Rim in Unilateral Coronal Synostosis

Bianca I. Knoll, MD, Joseph Shin, MD, John A. Persing, MD

New Haven, Connecticut

Abstract: Patients with unilateral coronal synostosis characteristically have a recessed and flattened supraorbital rim ipsilateral to the fused suture. Despite lateral canthal advancement procedures to correct these anomalies, patients often have a persistent flattened and recessed supraorbital rim after surgery. Current procedures address the pathologic features of the orbital rim only partially by advancing forward a deformed supraorbital rim without correcting the abnormal flattening of the normal rim curvature. The authors describe a technique modification of the supraorbital rim advancement procedure that addresses not only the lack of anterior positioning of it, but also its flattened contour: the bowstring canthal advancement.

A reduced projection of the ipsilateral supraorbital rim is one of the characteristic features of unilateral coronal synostosis. However, this anteroposterior malposition of the rim is only one abnormal feature; there are deficiencies in contour of the medial

From the Section of Plastic Surgery, Department of Surgery, Yale University School of Medicine, New Haven, Connecticut.

Address correspondence to Dr. John A. Persing, Section of Plastic Surgery, Yale University School of Medicine, PO Box 208041, New Haven, CT 06520; e-mail: john.persing@yale.edu

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rim, mediolateral shortening and superoinferior elongation of the axes of the orbit ipsilateral to the fused suture. Since the introduction of the simple strip craniectomy, more complex unilateral and bilateral techniques have evolved to correct the pathologic features of unilateral coronal synostosis.[1–4] Despite many advances in understanding the morphology,[5–9] pathophysiology,[10–12] and treatment of craniosynostosis, operative techniques[1,4,6,13–19] have developed that only partially address the existing orbital deformities. The plethora of different techniques reflects the difficulty in achieving pleasing long-term symmetrical results[20–27] in size, shape, and projection of the orbital rim.

The characteristic features of unilateral coronal synostosis are caused by restricted bone growth perpendicular to the fused coronal suture. The frontoparietal bone plate and the anterior cranial fossa are shortened ipsilateral to the fused suture. Compensatory expansion of the skull caused by the growing brain occurs in the contralateral frontal bone, and in the squamous temporal bone in the ipsilateral middle cranial fossa. The sphenoid bone is positioned anterosuperiorly ipsilateral to the fused suture, with the greater wing of sphenoid becoming more obliquely oriented, upward and laterally, when compared with the contralateral sphenoid. Orbital height is increased, and the mediolateral span of the orbit is reduced in comparison to the contralateral orbit. Compensatory frontal "bossing" depresses the contralateral supraorbital rim, reducing the vertical and increasing the mediolateral dimension of the orbit. Thus, the ipsilateral supraorbital rim is elevated, recessed, flattened, and associated with an elevated eyebrow and widened palpebral fissure. With a narrowed sphenopetrosal angle, the zygoma, ear, and the glenoid fossa are anteriorly displaced, resulting in a prominent malar eminence, often anterior to the orbital rim and deviation of the chin point to the contralateral side. The nasal root is deviated toward the fused suture while the tip points away from it. The feature, not as well recognized, is that the medial and lateral portion of the supraorbital rim do not have a "convex outward" profile like the normal orbital rim. Our approach is to provide a technique that offers the potential for a long-lasting correction of this deformity.

Materials and Methods

Operative Technique

A bifrontal parietal cranioplasty and a unilateral orbital rim advancement is performed. A coronal incision and a subgaleal supraperiosteal dissection is carried out to approximately 1 cm above the orbital rim bilaterally. An incision is placed in the periosteum, and subperiosteal dissection is carried down to the frontozygomatic suture, but the frontozygomatic ligamenteous attachments are maintained. An osteotomy is performed in the temporal region, extending anteriorly to the midline frontal region, using a combination of the Midas Rex and a lateral burr hole. A bifrontal bone graft is developed. An osteotomy of the orbital roof is performed extending into the greater wing of the sphenoid. A separate anterior parietal bone graft is harvested on the side contralateral to the fused suture to effect a "bowstring" advancement of the orbital rim. The bow is created by bending the supraorbital rim and is aided by intracranial kerfs (Fig 1). The harvested bone graft is secured with absorbable screws to just beyond the midline in the right supraorbital region and the temporal bone to maintain convexity. The supraorbital rim is then projected outward with the inferior frontal bone pivoting on, but still attached at, the frontozygomatic suture. This stabilizes the advancement inferiorly and allows correct anterior inclination of both the orbital rim and the inferior frontal bone. The space between the advanced orbital rim and the anterior portion of the squamous temporal bone is filled with multiple small bone grafts. The hollow temporal bone is then kerfed and reshaped with the Tessier rib bender. The bifrontal bone graft undergoes kerfs and

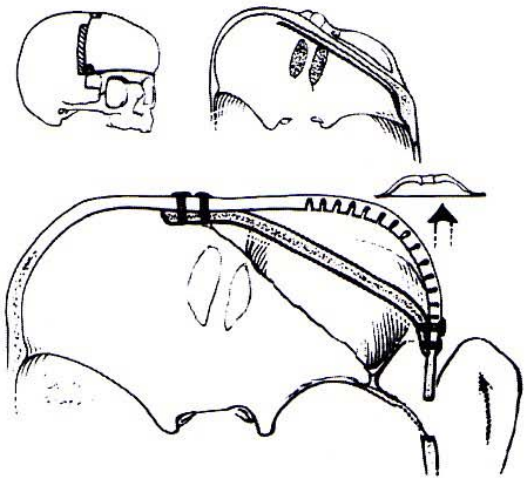

Fig 1 Depicting osteotomy performed and schematic principle of the bowstring canthal advancement technique (horizontal view).

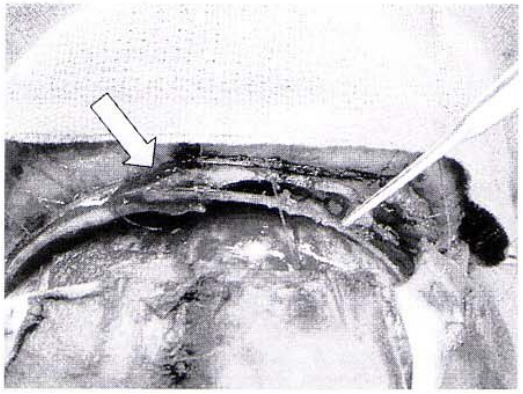

Fig 2 Intraoperative view of the bowstring canthal advancement: fixation with absorbable screw just beyond midline to the nonfused side (arrow).

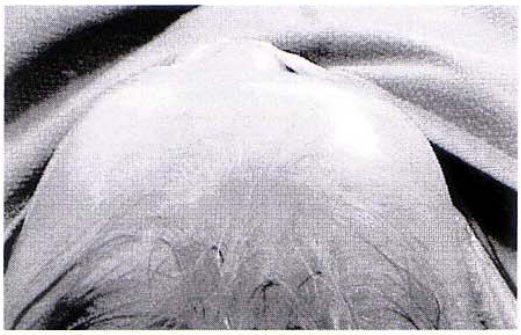

Fig 4 The bowstring canthal advancement: immediate intraoperative result.

remodeling and is attached to the advanced supraorbital rims bilaterally to achieve symmetrical projection (Fig 2).

Case Report

A 2-month-old infant received a diagnosis of right unilateral coronal synostosis presenting with typical morphologic features. The right frontal area was recessed, the supraorbital rim was flattened, and the left frontal bone showed compensatory bossing. The right palpebral fissure was expanded compared with the left, and the left orbital roof was displaced inferiorly. The right ear was malpositioned anteriorly and the chin deviated to the left. The nasal root was deviated to the right (Fig 3A–C). No ridge of the right coronal suture was palpable. The anterior fontanelle was open and flat, and the child had no signs or symptoms of neurologic dysfunction. Ophthalmologic exam revealed pseudostrabismus but no papilledema. Genetic testing revealed a P250R mutation in FGFR3, a mutation associated with Muenke syndrome or FGFR3-associated coronal synostosis, whose phenotype is variable.[28,29] The described bowstring canthal advancement was performed (Fig 4,5), maintaining the convexity of the frontal bone and supraorbital projection of the orbital rim 24 months after surgery (Fig 6A–C).

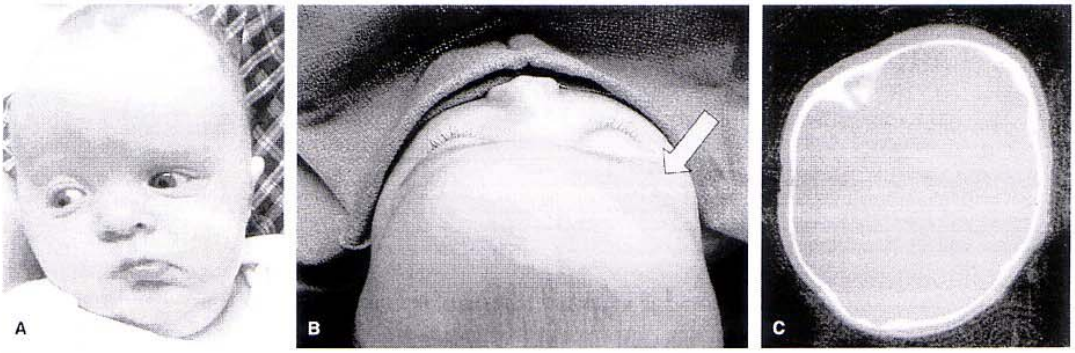

Fig 3 (A) Patient with right unilateral coronal synostosis before surgery. Note flattened, recessed, and elevated supraorbital rim. (B) Preoperative view: note the flattened and recessed right supraorbital rim. (C) CT scan with right unilateral coronal synostosis before surgery.

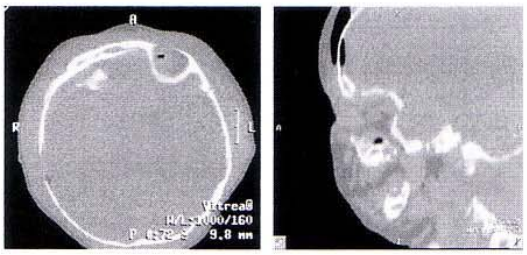

Fig 5 Computed tomography scan 1 month after surgery depicting the bowstring in a horizontal and lateral view.

Discussion

Surgical correction and satisfying long-term results of the orbital deformities associated with unilateral coronal synostosis are challenging. Postoperatively, a common feature in the long-term evaluation of patients with unilateral coronal synostosis is a recession and flattening of the supraorbital rim ipsilateral to the fused suture, regardless of the surgical technique used.[30,31] Some of this residual flattening and diminished projection is related to a hypoplastic ipsilateral cranial base. Other possibilities include incomplete correction of the intrinsic deformity of the orbital rim.

The orbit and its contents develop between 3½ and 10 weeks of gestation. Orbital wall and globe development are two different processes. By the 4th week, the orbital walls are visible. Centers of ossification appear 2 weeks later. The bones encompassing the orbit are delineated with periosteum by 6 to 7 months of gestation. The lateral wall and the orbital roof revealing the typical pathognomonic feature in unilateral craniosynostosis (harlequin deformity) have different origins. The lateral wall and the orbital floor are formed by the maxillary process, whereas the roof develops from the paraxial mesenchyme, which forms the capsule of the forebrain. The medial orbital wall arises from the nasal process. The zygomatic and maxillary bones, with the exception of the frontal process of the maxilla, are derived from the maxillary process. The lateral nasal process forms the frontal process of the maxilla, the nasal bone, lacrimal bone, and the lateral portion of the ethmoids. Because the globe size and extraocular muscles are not affected in most forms of developmental exorbitism, the orbital deformity in unilateral coronal synostosis is secondary, and a result of orbital displacement from abnormal positions of adjacent brain and

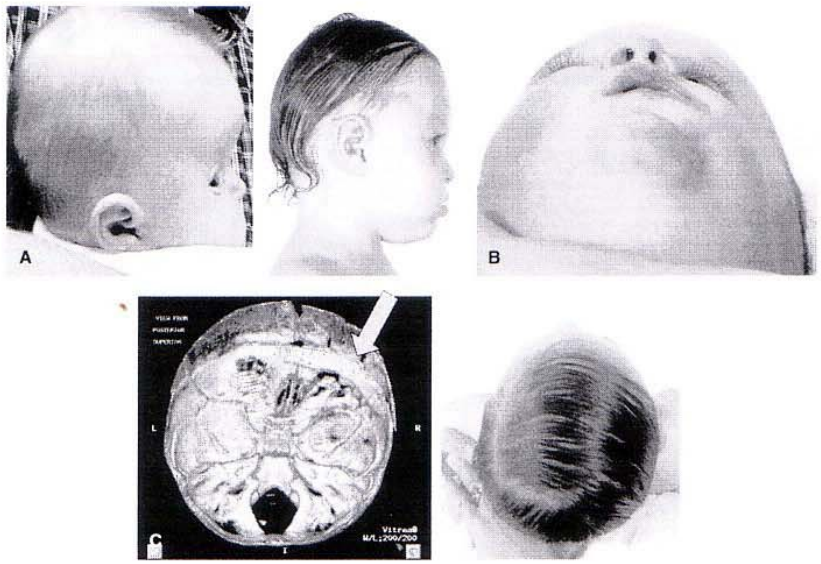

Fig 6 (A, B) Patient with right unilateral coronal synostosis 24 months after surgery in which the bowstring canthal advancement was performed, maintaining the symmetric projection and convexity of the supraorbital rim. (C) Cranial-caudal view depicting three-dimensional reconstruction of the bowstring 1 month after surgery and patient 24 months after surgery.

orbital bones is due to the premature fused cranial suture (ie, paraxial mesoderm).[32–34]

Commonly used techniques to correct periorbital deformities associated with unilateral coronal synostosis, such as the lateral canthal advancement[1] or tongue-in-groove technique,[4] take the recession of the supraorbital rim into account but do not address the pathologic deficient convexity of the anterior surface of the supraorbital rim. Recession of an advanced orbital rim may be caused by fixation instability. Improved stabilization of the supraorbital rim superiorly is achieved with the tongue-in-groove technique. Frontal bone remodeling to match the contour of the orbital rim and securing to the advanced supraorbital rim further contribute to support supraorbital rim projection.[4] Inferiorly, the fibrious attachment at the frontozygomatic suture allows for inferiorly located stability but, more importantly, an anterior angulation of the rim pivoting on the facial skeleton. Hardesty et al,[14] who used a combination of techniques, showed postoperative improvement of the orbital height and mediolateral width to near normal. Although an increased width of the orbital rim was seen after reshaping (and advancement), bone thickness may limit the amount of the reshaping possible at the zygomatic process of the frontal bone, and frequently it remains narrow on the fused side, compared with the contralateral (nonfused) orbit. Although both methods advance the supraorbital rim, the deficient convexity of the affected supraorbital rim is still not corrected.

Attentive to this issue, we tried to correct the lacking convexity of the supraorbital rim by placing vertical oriented kerfs on the internal surface of the orbital rim. This allowed creation of a convex shape of the supraorbital rim, as Marchac[35] described; however, thorough evaluation of these patients after surgery showed improved, but still flattened, ipsilateral orbital rim and frontal bone projection. This suggests that relapse of the shaped supraorbital rim occurs because of a lack of support of the anterior curvature superiorly and inferiorly. The bowstring technique keeps the curved supraorbital rim taut in its anterior position with resorbable screws and a tightened band of bone. We believe this allows the postoperative result to be more predictable and long lasting. It also gives the opportunity to create the desired convexity of the supraorbital rim without restriction of the expanding developing brain. Postoperative computed tomography scan 1 week after surgery demonstrated brain mass occupying the space immediately posterior to the advanced orbital rim. The sculpting influence of the developing brain and the enlargement of the globe during the first year of life to the upper face and orbital growth is still possible.[17]

Most importantly, it gives the opportunity to create the desired convexity of the supraorbital region and continues to influence the developing brain, and enlargement of the globe on orbital rim shape during the first year of life continues.[17] Likewise, the anterior cranial base position may influence the degree of orbital rim and frontal bone projection. This technique does not address these concerns directly, but it is able to ameliorate and even correct intrinsic orbital rim deformities.

With this technique, the improved rim contour has persisted for at least 24 months after surgery; however, additional follow-up monitoring is needed to determine whether this improved effect will last throughout childhood into adulthood.

Conclusion

The bowstring canthal advancement presents a promising technique that addresses typical postoperative periorbital stigmas of unilateral coronal synostosis. The correction of the flattening and recession of the supraorbital rim enables a more symmetric and normal appearance that might enhance the quality of the results of surgery.

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