Unicoronal Craniosynostosis and Plagiocephaly Correction with Fronto-orbital Bone Remodeling and Advancement

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Abstract

The standard surgical treatment for unilateral coronal synostosis is fronoto-orbital advancement. The technique is known for its high recurrence rate and established plagiocephaly remains a challenge. In this case report, the management of a 5-year-old with unicoronal plagiocephaly correction with fronto-orbital bone remodeling and advancement is presented. The previous surgery done at 6 months of age resulted in failure with establishment of hypertelorism and sudden progressive diminishing vision. Furthermore, temporal hallowing was evident in the imaging technique. As the child is in the active growing phase, only one end of the bandeau was secured with plates and screws while the other end was welded with ultrasonic waves. This ensured that the plate was held in position while bone remodeling will ensure normal healing and establish regular sutural growth. The child recovered well with regaining of vision. The report also discusses the possible reason for failures and the effectiveness of modification of the standard technique for growing child.

Keywords: Craniofacial deformities, craniosynostosis, fronto-orbital advancement biodegradable implants, plagiocephaly, sonic welding

Introduction

Unilateral coronal synostosis (UCS) is a rare, congenital craniofacial abnormality reported to be the second most common type of craniosynostosis with a birth prevalence of 66 in 1 million live birth.[1][3] The etiopathogenesis of this deformity has not been clearly defined and has been associated with mutation of FGFR2, FGFR3, TWIST, or EFNB1 genes in nearly one-third of the cases.[3][4] The UCS leads to formation of plagiocephaly and often at birth presents with clinical finding of retrusion of the forehead and superior orbital rim on the side of the fusion (involved side), contralateral side showing frontal bossing, nasal deviation, and orbital asymmetry.[3][5] The orbital asymmetry with growth may lead to aniso-astigmatism, ocular torticollis, and amblyopia. The condition poses a challenge in treating because the UCS by virtue of fusion of one coronal suture influences and alters the regular growth of adjacent cranial bone.[6][7] The effects of this extend beyond the fronto-orbital region to cause angulation of the cranial base, undergrowth of the ipsilateral cranium, anterior displacement of the ipsilateral ear, orbital dystopia, and contralateral rotation of the middle and lower side of the face.[6][7]

Fronto-orbital advancement (FOA) is the gold standard corrective procedure for UCS and is typically deferred until late infancy. The immediate goal of the FOA would be to expand cranial volume thereby minimizing damage to the brain. The surgery does not come without significant complications including prolonged hospitalization, bleeding, potential for relapse, and temporal hollowing. Furthermore, the procedure do not offer esthetical relief for middle and lower facial asymmetry as well as ocular anomalies that are other characteristics of this disorder.[1]

In limited-resource setting and lack of awareness often are the causes of delayed treatment-seeking behavior. Coupled with the fact of high degree of relapse, challenge of treating a patient in childhood (3–8-year-old) becomes manifold. The aim of this manuscript is to present Indian experience of treating a young child with previously mismanaged case of UCS with established plagiocephaly.

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A 5-year-old girl reported to center with a chief complaint of deformed head and wanted to correct it [Figure 1]. Patient’s parents gave a history of frontal and orbital bone reconstruction done for craniosynostosis at the age of 6-month elsewhere with unsatisfying surgical outcome. There was minimal documentation with a diagnosis of UCS with unknown etiology. On clinical examination, patient’s cranial vault was disproportionately large with frontal bossing, concave facial profile with increased intercanthal distance, flattened nasal dorsum, increased alar base width, and decreased malar prominence. She also had strabismus with laterally displaced left eyeball and decreased vision. No evidence of any other syndromic findings or other medical complications noted. Radiologically, computed tomography (CT) examination, there was absence of left coronal suture with incomplete ossification of frontal bone, broad nasal bridge, pneumatized perpendicular plate of ethmoid, and bilateral malar hypoplasia, and increased left orbital volume was seen. Furthermore, evidence of temporal hollowing observed [Figure 2]. Based on the findings, the patient was diagnosed as nonsyndromic unicoronal plagiocephaly owing to UCS and the treatment plan was to correct the cranial vault deformity by frontoorbital bone remodeling and advancement in relation to the left side of the skull. After adequate imaging studies and clinical examination, using the latest CT set of
images, a customized 3D-stereolithographic model was prepared [Figure 3] and the planned osteotomy cuts were prepared on the model before the surgery [Figure 4].

**Surgical procedure**

Using standard surgical protocols, oro-endotracheal intubation was done and general anesthesia was induced. Infiltration of local anesthesia with vasoconstrictor was injected along the previous incision line. Bicoronal incision was placed and the dissection proceeded in the subperiosteal plane to reach the supraorbital rim. Dissection was continued to expose the medial orbital walls and the frontonasal bone. The supraorbital nerve was separated from the supraorbital notch using osteotome. A craniotome was used and osteotome cuts were done to separate the frontal bone segment with careful dissection of the dural attachments below. Then, the osteotomy lines were extended along the lateral orbital wall to encompass wide bony resection in the region of the sphenozygomatic region in relation to the left side of the skull, across the roof of the orbit, and the frontonasal junction. The temporal muscles were bilaterally minimally mobilized to enable the preparation of the lateral struts of the orbital bandeau. The entire left fronto-orbital bandeau of bone was carefully separated from the adjacent structures of the skull and trimmed using burs and recontoured to match the contour of the right side of the orbit. The fronto-orbital bandeau and the frontal bone segment were significantly advanced anteriorly to correct the deformity of the skull as well as to increase the volume of the skull to accommodate the growing skull. Then, the fronto-orbital bandeau was fixed to the frontal bone flap using titanium plates and screws. Further fixation of the frontal bone to the rest of the calvarium was achieved using SonicWeld bioreabsorbable plates and screws. The lateral canthus was suspended using nylon sutures using holes drilled in the lateral orbital rims [Figures 5-7]. The dura was inspected for any tears and cerebrospinal fluid leak. Closure of the scalp was done and a full head compressive dressing was placed after the closure of the surgical wound. Appropriate antibiotics and nonsteroidal anti-inflammatory drugs were given and healing was uneventful. The patient is being regularly followed up [Figure 8].

**Discussion**

It has been documented that the surgical techniques for craniosynostosis still require clarity in simplifying and objectifying the cranial remodeling process. In UCS and FOA, the entire surgical procedure, esthetic and functional outcome relies on the operator’s experience and his/her ideologies in this regard. This subjective issue was observed by Marchac and developed a template-based solution for the same. His template aimed to assist in remodeling the fronto-orbital bandeau, called Marchac template. This was improved by Mommaerts et al., Pappa et al., Burstein et al., Salyer and Hall, and Saber et al. Since then, many templates including 3D-printed ones were suggested. Every subsequent method improved the level of customization and ease of applicability during operation, reducing operator’s error which would cause significant morbidity and/or poor outcomes.

The ability of this surgical invention to cause molding of the involved bone is only one of the many variables affecting the long-term cosmetic outcome in UCS. The other critical factors include but limited to extent of the cranial base deformity, (though compensated by FOA) and timing of surgical intervention have an impact on long-term outcome. As per the reported and documented experience of Foster et al. and Marchac and Renier, children who were operated between ages of 6 and 12 months have excellent outcome and lower recurrence rate. In the present case too, the child was operated at the age of 6 months with FAO which for reasons, unknown has failed and accentuated the problem. The probable causes might be due to compromised vascularity of the fronto-orbital bandeau either during the procedure or strangulation of vascular elements during the subsequent remodeling of bone or due to inherent genetic deficiencies. Another mechanism suggests the pathological fusion of sutures. Pathologic fusion of the coronal sutures in UCS usually begins in the middle portion and is reported to expand cephalad to the anterior fontanelle and caudad into the frontosphenoidal suture along the anterior cranial base. It has been further documented that pathologic fusion extension
into the frontosphenoidal suture is time-dependent, that is, the suture is patent up to age 3 months, but goes on to fuse by 6 months of age in all patients. The FOA surgery is positioned at 6–8 months of age, after the frontosphenoidal suture is fused. During the surgery, the fronto-orbital unit is advanced and this leads to formation of bony gaps that rapidly ossify; functional “sutures” never arise. Probably, the absence of functional sutures on the ipsilateral side in the face of functional sutures on the contralateral side may account for recurrent fronto-orbital asymmetry reported after FOA for UCS.[1]

Like in the present case, temporal hollowing is not an unusual finding after UCS repair. The exact cause of the hollowing remains still debated. The suggested causes include temporal muscle atrophy, temporal fat atrophy, and type of surgical technique in the causation of postsurgical temporal hollowing.[9] In the present case too, for unknown reasons, there has been a failure of earlier surgery as well as temporal hollowing necessitating an immediate surgery for two reasons – the eye being progressively involved causing damage to vision as well as possibility of increasing intracranial pressure.

In the present case, the developing loss of vision with uncoordinated cranial base growth necessitated the recorrection attempt. The overcorrection of the surgery was a choice available, but the age of the patient did not permit the same. In an attempt to accommodate the growing brain as well as the correction of diminishing of vision, the FOA was planned and performed. The template would not be an ideal solution here in as the growth at this phase is quite unpredictable and the cause of failure of previous surgery was not clear. Hence, the surgery was planned in such a fashion to perform the advancement.

- As maximum as possible to maintain symmetry with contralateral side
- To facilitate or accommodate growth of brain – for this, one side of the bandeau was stabilized with screws and plates while the other side was fixed with sonic welding of bone. This ensured that the fixation element was resorbed in due course of time, permitting natural remodeling to pursue, and aid the latter growth.

This dual approach helped achieve the dual objective of securing normal expansion of cranial vault with growth while firmly protecting the delicate internal structures.

**Conclusion**

A modified approach for late correction of recurrence in a case of UCS is presented with the role of probable mechanisms that lie behind the recurrence. The approach ensured the solving of diminishing vision as well as adapting to growing brain. The beneficaility of the modification has to be followed up using large sample size to understand the condition better and offer better solutions.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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