Technical Note

Modified facial bipartition

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ABSTRACT

Orbital hypertelorism is a craniofacial abnormality that arises on its own or as part of a number of syndromes. It is not uncommon to find the condition in association with maxillary hypoplasia. This manuscript reports an uncommon case of Tessier 2, 12 with orbital hypertelorism and dentoskeletal maxillary prognathism. To correct the condition, the first stage procedure was a modification of facial bipartition, according to the need of the case, followed by correction of alar cleft by Denonvilliers technique.

Keywords: Epicanthal fold, facial bipartition, hypertelorism

INTRODUCTION

Orbital hypertelorism refers to an abnormally increased distance between the two orbits. The condition is always congenital and never acquired. The characteristic feature of this condition is increased interpupillary distance in addition to increased intercanthal (inner canthi) distance. In contrast, telecanthus is a condition that is very often acquired, though sometimes congenital and is characterized by increased intercanthal distance with normal interpupillary distance.[1] Orbital hypertelorism is mostly a component of craniofacial anomalies, syndromes or facial clefts but have been documented as isolated entity as well. It is not uncommon to have a prominent epicanthal fold requiring an additional soft tissue correction.

Hypertelorism is a symptom manifested in a variety of syndromes, including 1q21.1 duplication syndrome, Basal Cell Nevus syndrome, DiGeorge syndrome, Loeys-Dietz syndrome, Apert syndrome, craniofrontonasal dysplasia, Noonan syndrome, Neurofibromatosis, LEOPARD syndrome, Crouzon syndrome, Wolf-Hirschhorn syndrome, Andersen-Tawil syndrome, Waardenburg syndrome, and a host of other conditions.[1]

Facial bipartition was primarily designed for "medial fasciotomy," performed by van der Meulen,[2] in which the median facial cleft was obliterated and the partial segments were rotated medially in conjunction with advancement. Since then, this technique had been modified by various surgeons numerous times and applied for hypertelorism correction.[3] Though the surgical technique is versatile it requires improvisation based on the clinical findings in the case to accommodate the other phenotypic abnormalities. One such modification has been discussed and presented.

CASE SCENARIO

An 8-year-old boy reported with a complaint of wide set eyes, flat facial appearance, and nasal deformity since birth. On

Figure 1: (a and b) Preoperative picture showing telecanthus (50 mm), orbital hypertelorism (interpupillary width = 65 mm) with prognathic maxilla

clinical examination, the child had an interpupillary width of 65 mm, intercanthal distance of 50 mm, inter eyebrow width of 35 mm, alarbase width of 32 mm, commissural width of 40 mm confirming orbital hypertelorism [Figure 1a-b]. In addition, the child also had Tessier's 2 alar clefts. He had a mongoloid slant with epicanthal folds. He had decreased malar projection with a wide nasal bridge. On CT examination, he had absent nasal bones replaced by a broad nasal bridge, pneumatized perpendicular plate of ethmoid, bilateral malar hypoplasia, bilateral Tessier 12 frontal defects, partial absence of parietal bones with prognathic maxilla. The coronal section showed evidence of a small encephalocele. The case was diagnosed as Tessier 2 and 12 with hypertelorism, encephalocele, epicanthal fold, posterior cranial defect, and malar hypoplasia.

SURGICAL TECHNIQUE

Under general anesthesia, lumbar puncture was performed and a CSF drain placed. Through a bicoronal approach, craniotomy was done 2 cm above the supraciliary arches, with the posterior cut just anterior to the coronal sutures. The squamous portion of the frontal bone was removed and preserved to be replaced at the end of surgery. The frontal lobes of the brain were exposed. CSF

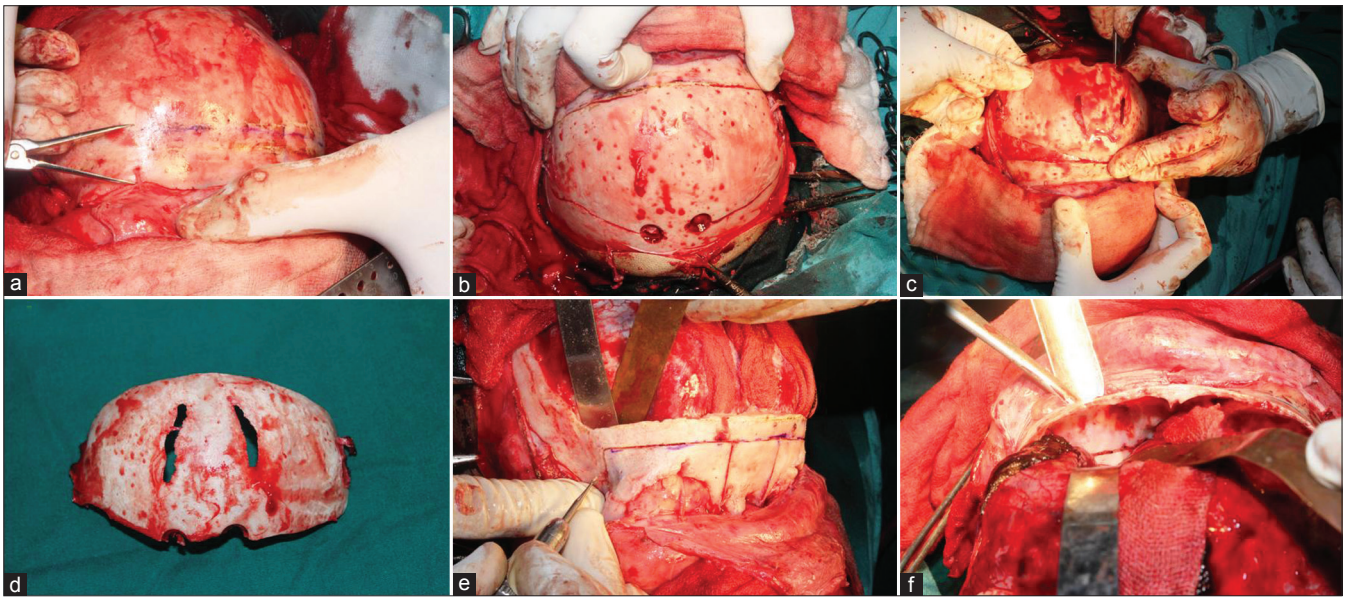

Figure 2: (a) Intraoperative picture showing frontal bar planned 15 mm above the supra orbital ridge, (b–d) craniotomy cuts placed and removal of the squamous portion of the frontal bone, (e, f) intraoperative picture showing the facial bipartition osteotomy cuts

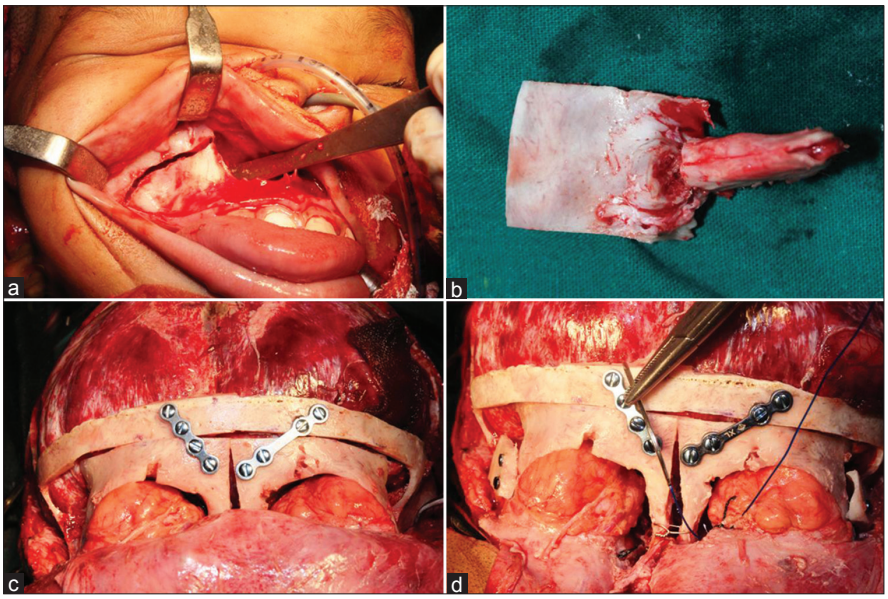

Figure 3: (a) Intraoral view showing the modified maxillary osteotomy, (b) exenterated nasal elements – pneutamized nasal septum, cribriform plate of ethmoid with the excess interorbital frontal bone, (c) medial mobilization of the orbits and fixation to superiorly placed frontal bar with mini plates, (d) MCL canthopexy

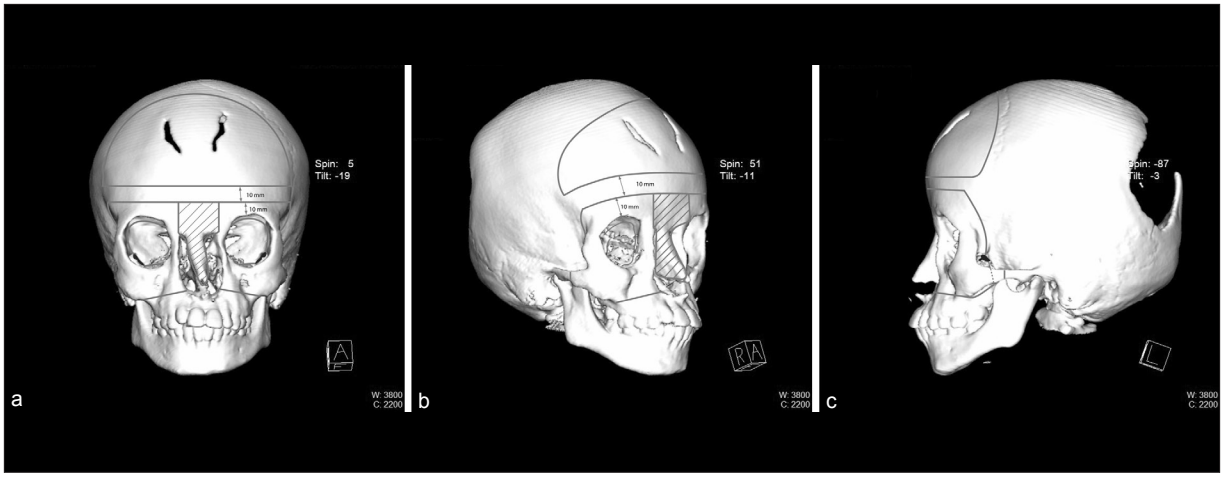

Figure 4: (a–c) 3D CT scan showing Tessier 2, 12 cleft and orbital hypertelorism with markings of the planned modified facial bipartition

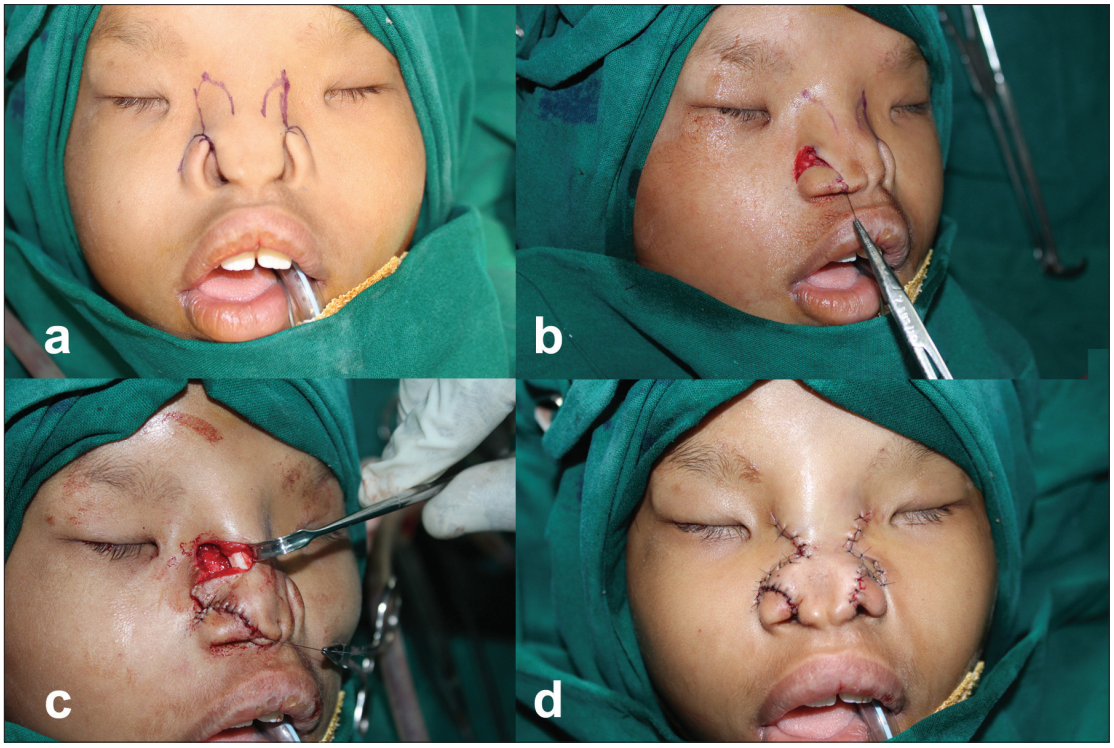

Figure 5: Intraoperative pictures of IInd stage surgery a) Marking for bilateral Denonvilliers procedure b) Down rotation of lateral lower crura and creation of alar rim c) Dorsal augmentation with rib graft d) Immediate postoperative after bilateral Denonvilliers procedure

was drained through the lumbar drain along with administration of I.V. mannitol facilitating the retraction of the frontal lobes from the floor of the anterior cranial fossa. An osteotomy was done 10 mm inferior and parallel to the craniotomy cut, 10 mm superior to the supraciliary arches in order to create a frontal bar. The temporalis muscle was retracted from the temporal fossa on either sides, exposing the inferior orbital fissure. Supraorbital osteotomy was continued laterally into the upper end of the inferior orbital fissure bilaterally. Osteotomy of the zygomatic arch was done bilaterally. A transverse osteotomy was done across the roof of the orbit, 10 mm anterior to the optic nerve. The osteotomy was continued to the opposite side, by-passing the cribriform plate. Medially the osteotomy is continued to the medial orbital wall bilaterally posterior to the posterior lacrimal crest. Though a vestibular incision, maxilla and zygoma were exposed till the infra orbital margins. Osteotomy was performed bilaterally with the cut extending from the lateral wall of the piriform aperture continued across the anterior maxillary wall, underneath the zygomatic arches reaching the inferior orbital fissure. Two vertical osteotomies were performed perpendicular to the frontal bar, reaching the superior wall of the piriform aperture, 3 mm medial to the medial orbital wall. The nasal elements comprising the cribriform plate of the ethmoid bone, pneumatised nasal septum, turbinates were exentrated. Medial

Figure 6: Postoperative picture showing reduced intercanthal distance, prominent nasal bridge with anatomic nostrils

wall osteotomy was continued into the orbital floor. The encephalocele was repaired. The orbits were mobilized medially with anterior advancement of the zygoma and superiorly fixed to the frontal bar using mini plates. The medial canthal ligament was identified and lassoed, transnasal canthopexy was performed. The bone exentrated from the nasal complex was used as graft at the lateral defects created by the medial orbital translocation. Pericranial flap was raised and used to line the floor of the anterior cranial fossa. Skin redraped [Figures 2, 3]. Later nose was corrected with Denonvilliers technique and dorsal augmentation with rib graft.

DISCUSSION

Facial bipartition aims at separating the mid-facial component of the skull from the anterior skull base. The squamous portion of frontal bone is split from the supraorbital rims initially. Then the orbits and the mid-face are released from the skull base using monoblock osteotomy along the mid-face. The most important part of this initial step is marking of facial midline and creating the guiding frontal bar [Figure 4]. The orbits are mobilized by doing the 180° continuous osteotomy including all walls of the orbits. The mid-facial mobilization is done conventionally followed by the resection of mid-facial skeleton including the ethmoid bone. After removing this segment it is possible to rotate the two halves of the mid-face toward each other, thus resulting in reduction of the distance between the orbits.

The modification in this technique has facilitated us to avoid the subciliary approach. Moreover since this case warranted the advancement of the zygoma along with the medial translocation of the orbit, it was achieved by combining the facial bipartition cuts with that of Lefort III osteotomy sparing the dentate segment. The modification ensured better result at postoperative period. Later as second stage the nasal alar cleft repair was done by Denonvilliers technique with dorsal augmentation using rib graft [Figures 5 and 6]. Further he is planned to undergo secondary rhinoplasty and epicanthal fold correction for cosmesis.

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