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A rare case of surgical correction of bifid nose in an Adult: A Case Report

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Abstract: *Tessier Pclassified the rare facial clefts which simplifies the management of these patients[1]. Still rarities keep appearing which cross the defined boundaries and cast a doubt on the completeness of these classifications. It is immensely difficult to perceive the morphogenesis of a rare cleft which spans a zigzag path through the embryological processes of the foetal head enlarge. This patient presented to us with a bizarre facial cleft which was classified as a combination of Tessier no. 0 and 1 cleft.*
Tessier craniofacial cleft may involve entire soft tissue skeletal element throughout the course of cleft, resulting in distorted craniofacial growth pattern and altered pattern for normal growth. The presentation of Tessier number: 0 cleft patient may vary from minimal changes on median facial structures such lip, vermilion and nose, and nose to wide clefts dividing all median craniofacial structures. The variability of expression of the unusual orofacial clefts can be challenging for the surgeon, while reconstructing affected facial structures.

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I. Introduction

Craniofacial clefts cause severe facial disfigurement even in minor forms. The surgical reconstruction is imperative to restore function and appearance of facial structures. Orofacial cleft is a failure in embryonic facial development during the first 8 weeks of life. The cause of craniofacial clefts is not clear. However, some theories about orofacial cleft have been submitted such as failure of fusion theory and the failure of mesodermal penetration theory[2].Various risk factors (radiation, infection, maternal metabolic imbalances, and drugs and chemicals) have been published[3]. Craniofacial clefts were classified by Tessier, according to their anatomical basis[4]. This classification system contains numbered clefts from 0 (midline cleft of the lip and nose) to 14 and 30 (mandibular). Presentation of median cleft may be varied from minimal changes on median facial structures such lip, vermilion, and nose to wide clefts dividing all median craniofacial structures[5]. The median craniofacial skeleton (crista galli, ethmoid, vomer, nasal, and premaxillary bones) and the cartilaginous septum can be affected in severe cases. Tessier number: 0-cleft may cause death if it is associated with holoprosencephaly[6]. Although the clinical presentation of patients has been almost always enough for a diagnosis of the craniofacial clefts, computed tomography (CT) scans help the surgeon to plan surgery. Minimal malformations on median facial structures can be corrected with local flap options such as z-plasties and v-y advancement flaps[7]. However, severe cases require reconstruction of midline of the craniofacial skeleton. Surgical correction of bifid nose can be challenging and requires multiple surgical procedures. A few articles related to surgical correction of this unusual malformation have been published[6,7,8].

II. Case report

A 17-year-old girlfrom poor socioeconomic class presented to our clinic for Aesthetic repair of her deformed nose presented with congenital midfacial disfigurement with hypertelorism diagnosed as Tessier's cleft number 0 (bifid nose) was posted for reconstruction surgery. She had no siblings with clefts.She had achieved expected intellectual and physical milestones commensurate with her age. The physical examination revealed a midfacial cleft with normal calvarial and lower facial structures. The patient had a flat nasal dorsum and a deep groove between the two alar domes. The nose was short and bifid. The width of nasal radix was increased. The nasal septum was thick and doubled. The nares were symmetric and separate. The phitral dimple was clefted and phitral is columned. Computed tomography scan (CT Scan) of the craniomaxillofacial skeleton was performed to rule out median cleft face syndrome and holoprosencephaly. CT scans revealed bony cleft over anterior nasal septal region. All the laboratory tests were in normal ranges. Systemic examination and routine blood investigations were within normal limits. The patient was prepared as per standard guidelines. Post-operative stay was uneventful and discharged from hospital on 15th post-operative day.

III. Surgical technique

Under general anaesthesia, the skin groove between the 2 alar domes was elevated. By transcolumelar incision, dermis and the soft tissue covering the upper two-third of the nose was raised as an inferiorly pedicled single flap. The upper lateral cartilages and the lower lateral cartilages were detached from the overlying skin and subcutaneous tissues with a combination of sharp and blunt dissection. The right and left alar parts of the bifid nose were transposed to the midline and sutured by interdomal sutures. The septal mucosa was detached from cartilaginous skeleton of septum. The nasal septum was thick. After the septoplasty, the nasal passage was open. The hypertrophic bone segment, lying at the base of the pyriform aperture, was removed. Autologus costal cartilage graft, 1.5 cm × 2.5 cm in diameter, was harvested from right 6^{th} rib. The cartilage graft was reshaped to form the new cartilaginous nasal vault [Figure 3]. The cartilaginous frame was sutured to each other with 5/0 polyamide sutures. A subcutaneous tunnel was created with a dissector from the base of the columella. The lateral osteotomies were completed percutaneously with a 2-mm osteotome. The lateral walls were fractured to close the roof and reshaped the nose in upper third. The inferiorly based flap was used to cover osteocartilage frame. The nostrils were packed with gauze with Vaseline. The nasal splint was applied for 3 weeks.

IV. Discussion

Although facial bipartition is essential for the reconstruction of the facial skeleton, sometimes patients prefer minimal surgery as our patient did. The clefts including upper lip structures (Cupid's bow, labial philtrum, vermillion, and buccal mucosa) can be reconstructed with local flaps[9].When the nasal structures were affected, more complex surgical correction is needed. The laterally displaced alar and upper nasal cartilages should be corrected to obtain more acceptable nasal shape. Moreover, optimal nasal length could be obtained. Costal cartilage graft was used to augment the nasal dorsum. Our technique resembles the embryologic movement of the nasal structures[10].We believe that correction of the lower one-third of the nose with native structures plays a key role in obtaining natural nasal appearance. Nasal septum can be duplicated in some cases[11].Our patient had one, thickened nasal septum. Lateral osteotomies are usually insufficient to prevent a saddle nose appearance. We think that the usage of cartilage graft works well for obtaining nasal projection in severe bifid nose cases. Skin excision from nasal dorsum is widely suggested in the literature. However, we did not perform skin excision.

In case of early repair of bifid nose more conservative, surgical approaches are described. The case presented here was corrected during adulthood. We are not sure whether our technique has an adverse effect on nasal development when performed in childhood. The only disadvantage of our technique is that it needs two stages. Second stage can be performed 3 months later.

IV. Conclusion

In case of severe bifid nose, cartilaginous and soft tissue structures of the nose must be restored separately. Besides to facial bipartition, we think that the lower one-third part of the nose, including the lower cartilages must be reconstructed by native nasal structures. Following reconstruction of the cartilaginous framework with nasal tissues or grafts, remaining nasal soft tissue and local flaps can be used to cover the soft tissue.

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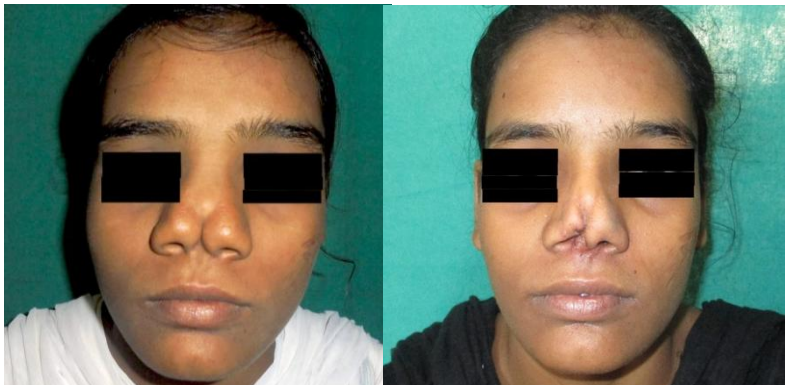
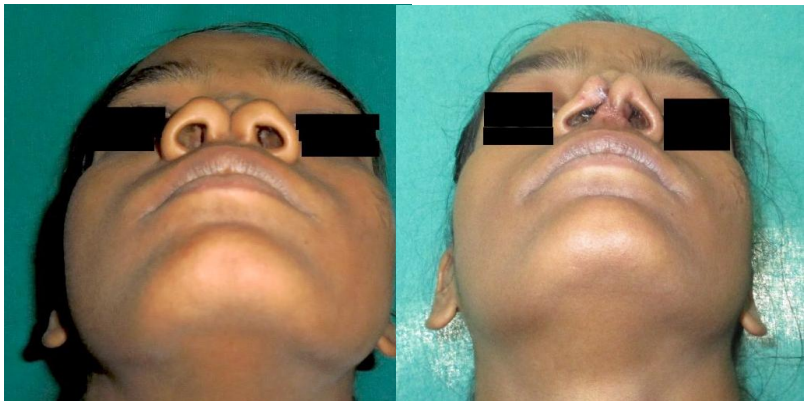
Photographs.



Before **After**



Before **After**

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