

Morpho functional cleft lip and palate repair in a binderoid syndrome patient - A case report

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Abstract

Maxillofacial region is quite complex, growth and developmental defect cause various anomalies. Cleft lip and/or palate is the most popular congenital anomaly and is seen in 0.2% of general population. An 8-month-old boy’s parents, attended the Oral and Maxillofacial Surgery department and was diagnosed with Binderoid unilateral left Cleft lip and palate with bilateral microtia and left anterior plagiocephaly. Morpho functional repair of the cleft lip and palate was done. This article outlines the major characteristics of Binderoid syndrome with cleft lip and palate and the surgical treatment methods used in the authors’ department and worldwide.

Keywords: Binderoid Syndrome, Cleft lip, Cleft Palate, Morpho functional repair

Introduction

The term Binderoid (binder like) cleft lip and palate team was suggested by Mulliken et al in the year 2003. It is used in the patients of unilateral or bilateral complete cleft lip and palate with Nasola bio maxillary hypoplasia and orbital hypotelorism, without evidence of holoprosencephaly. The term ‘Binderoid’ comes from its similarity with Binder syndrome (anomaly), which is characterized by Nasola bio maxillary hypoplasia [1–3].

These patients are characterized by hypo plastic nasal tip, conical columella, tinyprolabium, underdeveloped

lateral labial elements, and small/mobile premaxilla. The ipsilateral central and lateral incisors are absent in patients with unilateral cleft, and single-toothed premaxilla is typically found in bilateral patients. Early recognition of this entity is important for counselling parents and because alterations in standard operative methods and orthodontic protocols are necessary [4,5]

Case Report

An 8-month-old boy's parents, attended the Oral and Maxillofacial department with complain of defect in lip and palate along with difficulty in feeding.

On clinical examination, unilaterally cleft lip was present extending to the alveolus, hard and soft palate of left side. The mid-face profile was hypoplastic, the nose was flattened on left side, the upper lip was convex with a small philtrum, wide alar base not in proper ring shape, the nostrils were typically crescent in shape due to the short columella, total severance of upper lip and a deep fold between the upper lip and the nose, resulting in an acute nasolabial angle. There was a normal slope of the fossa prenasalis on the non-cleft side with an absent fossa on the cleft side. The deficient nostril sill areas with the convex contour of upper lip tend to give rise to an overall concave midface of C-shaped profile. The ala of nose tend to attach at right angles to the cheek tissues and when viewed from below, the lumens have a crescentic shape. The premaxillary segment was underdeveloped with absence of anterior nasal spine. Midface hypoplasia was evident with an absence of frontonasal angle reflected in a straight profile. He also had thin vermilion, and hypoplastic nasal septum. There was also reduced sagittal development of the nose. Bilaterally microtia was present along with left anterior plagiocephaly (Figure 1). It was impossible to use the dentofacial orthopaedic appliance since patient came

from a distant place and regular follow ups were not possible for the patient due to Covid pandemic.

The final diagnosis made was unilateral complete cleft lip, alveolus, hard and soft palate with bilateral microtia and left anterior plagiocephaly with Binder's Syndrome suggestive of Binderoid Unilateral Cleft lip and palate.

Patient was admitted under Department of Oral and Maxillofacial Surgery after physical fitness of Anaesthetist and Paediatrician.

Figure 1: Profiles



The treatment plan for the patient was as follows

- Correction of Anterior Plagiocephaly-Patient was not willing for surgery
- Sept cheiloplasty done at 8 months
- Palatoplasty done at the age of 1 yr.

Further treatment plan

- Speech Therapy at 3-4 yrs. of age
- Alveolar bone grafting at 7-8 yrs.
- Bilateral microtia repair at the age of 10 yrs.
- Orthodontic treatment at the age of 12-13 yrs.
- Maxillary advancement procedure at the age of 15 yrs.

• Secondary cleft rhinoplasty at 16 yrs. of age
Morpho functional cleft lip and palate repair

- Sept cheiloplasty (Afroze Incision)

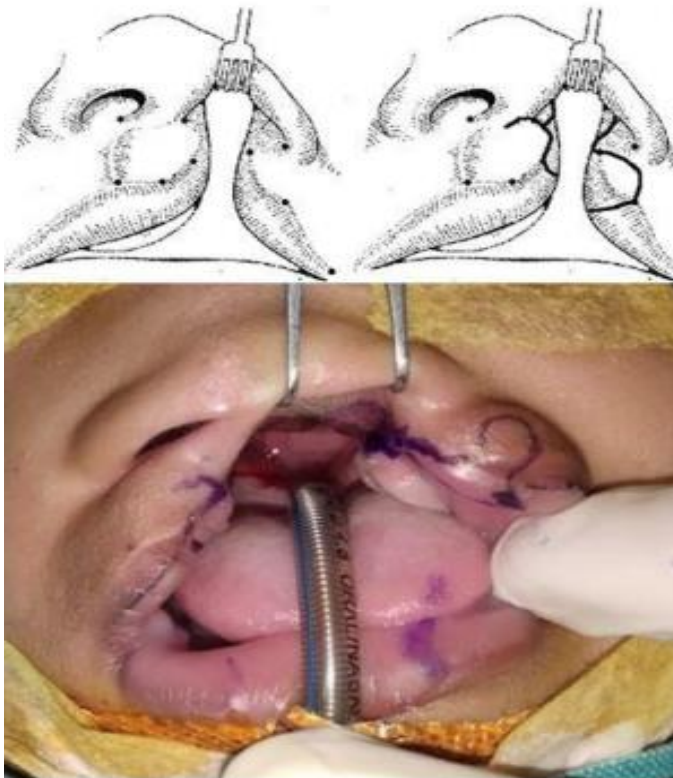
Incision marking was done according to Afroze incision for Sept cheiloplasty. Markings were made with gentian violet dye, using a sharpened wooden toothpick.

Following points were marked (Figure 2)

- Tip of the nose
- Base of columella
- Lateral points of columella

- Highest point of white roll on the non-cleft side [POINT A]
- Deepest point of white roll on the non-cleft side [POINT B]
- Point C is 2mm more than the distance between point A & point B
- Point D is on the highest point of the white roll fading on the cleft side.

Figure 2. Sept cheiloplasty Markings



Afroze incision is a combination of Millard incision (on non-cleft side) and Pfiefer wavy incision (on cleft side). (Reddy et al., 2009a) On the cleft side, a frozen incision was made, followed by muscle dissection, with the ala nasalis muscle being dissected first. On the cleft side, a 5 mm back cut was made in the vestibule, followed by a large sub-periosteal dissection to lift the complete face mask from the piriform rim, fronto-nasal area, infra orbital, and maxillary buttress region. It's known as facial mask elevation. The aberrant attachments from the anterior nasal spine and the columella were relieved with

minimum dissection on the non-cleft side. Septal cartilage was discovered on the non-cleft side during the same incision. To achieve septum straightening and columellar centralization, the perichondrium was entirely removed from the septal cartilage on both sides, as well as from the anterior nasal spine, septo-vomer junction, and septo-spinal ligaments (Agarwal and Chandra, 2007). All aberrant attachments to the nasal septum were removed, and the septum was restored to its morphological position. Suturing the alveolar flap from both sides posteriorly, which later forms the nasal floor in the anterior region, was followed by peri alveolo plasty. A suture was passed through the alar head of the nasalis muscle on the cleft side to the contralateral muscle via the septum to fix the alar base to match that of the contralateral side. The nasal floor was rebuilt after this alar cinch suture was put. The septal cartilage was then sutured in three locations superiorly, intermediately, and inferiorly with the perichondrium on the non-cleft side to transfer it to a normal position. A single quilting suture was put in the nasal septal area to reduce dead space. The lip was then closed in layers, beginning with the mucosal layer, followed by muscle where the orbicularis orris was approximated and repaired to a normal position, and finally tension - free skin closure enabled by facial mask elevation and suturing of the transverse nasalis muscle.

Key steps in Morpho-Functional Sept cheilo plasty

- Facial mask elevation (wide sub-periosteal dissection)
- Peri alveolo plasty
- Septoplasty
- Ala nasalis suturing
- Cheiloplasty with triangular flaps on the vermillion

Patient then reported back at the age of 1yr for palatoplasty, the weight of 9.42 kg and Hb of 9.6 g/dl was noted and the patient was admitted for the same.

➤ Palatoplasty (Two flap with optimal muscle dissection)

Incisions were made bilaterally along the cleft edges, laterally near to the palato-gingival margins of maxillary teeth, and anteriorly as far as the defect's limit, including the palatine rugae in the flap.

The marking was done posteriorly from the cleft borders at the base of the bifid uvula and up to 10 mm bilaterally over the palate pharyngeal arches. Starting at the palato pharyngeal arches, incisions were made across the base of the bifid uvula, along the cleft edges, and then lateral releasing incisions. On either side, a muco periosteal flap was reflected to expose the posterior nasal spine medially and the pterygoid hamulus laterally. The muscular aponeurosis connected to the hamulus was released to allow for muscle mobilisation medially after the PNS was exposed medially and the pterygoid hamulus was exposed laterally.

The soft palate mucosa was dissected from the levator muscle bundle along the same plane as the aberrant insertion of the levator palatine muscle. The nasal layer was carefully detached from the levator muscle. The periosteal fibres across the pterygoid hamulus could be incised using a no. 15 BP blade to ensure a tension-free closure of the oral mucoperiosteum. On both sides, the nasal layer was approximated. Suture was used to approximate the levator muscle bundle by moving it transversely and posteriorly.

4-0 vicryl sutures were used to rebuild the uvula. The oral mucoperiosteum was sutured with 4-0 vicryl sutures that alternated between simple interrupted and vertical mattress sutures, with the vertical mattress sutures passing solely through the oral mucoperiosteum and the simple interrupted sutures passing through both the oral and nasal layers. 4-0 vicryl sutures were used to suture the oral layer's lateral release incisions to the palate-

gingival borders or the labial mucoperiosteum (Figure 3).

Figure 3: Palatoplasty



Discussion

Cleft lip and palate is linked to around 340 syndromes. There are currently few published data about patients with cleft lip and palate who have Binderoid syndrome. Although Binderoid syndrome is not life-threatening and does not affect any vital organs or systems, it causes substantial facial dysmorphia, which leads to a diminished social life and a significant reduction in quality of life.

As a result, patients who exhibit all or portion of the syndrome's symptoms require complex surgical procedures to disguise the syndrome's consequences^[6]

Mulliken et al. described a condition called "Binderoid complete cleft lip or palate" in 2002.

Their patients (15 cases) had cleft lip and/or palate, as well as Nasola bio maxillary parts that were underdeveloped. Patients with chondrodysplasia punctata, cervical spine deformities, foetal exposure to warfarin, and a cleft lip and palate may all have a Binderoid phenotype, according to the investigators.

Mulliken et al. reported that the patients they presented were mostly women (2:1), which matched the findings of the present authors. This ratio is the total opposite of what is seen in patients with isolated cleft lip and palate. The individuals in this study had a wide range of dysmorphic characteristics, ranging from minor nasal hypoplasia to severe Naso maxillary under development with malocclusion^[11].

The surgical protocol used at the institution is based on early closure of the cleft lip and palate in patients presenting this co-morbidity.

Cleft lip surgery can be done in a variety of ways. Depending on the surgeon's inclination and training, different procedures are used (Hetal et al., 2015). Millard's rotation-advancement approach, as well as its several adaptations such as Noord off (1984), Mohler (1987), and Randall-Tennison (Mohler, 1987), is still one of the most often utilised surgical repairs today. These changes were made to address observed flaws in rotation advancement technique, such as wide cleft application, limited medial element rotation, insufficient vermilion fullness, and philtral column aesthetics. Delaire (Delaire et al., 1988; Markus and Delaire, 1993), as well as Nakajima and Yoshimura, pioneered their own individual repairs (1993). Fisher's anatomical subunit approximation technique for unilateral cleft lip repair was also recently published (Fisher, 2005).

None of these methods account for the morphological changes present in unilateral complete cleft lip, particularly in patients with syndromic cleft lip. Not every unilateral full cleft lip is the same; some have a Simon art's band, some have a large discrepancy between the bigger and lesser segments, and still others are linked with significant nasal deformity, all of which affect surgery outcomes.

All of these variations were corrected using the surgical technique, "Morpho-Functional Sept cheiloplasty." The Afroze incision is a mixture of Millard and Pfeifer incisions on the non-cleft side and Afroze incision on the cleft side [6]. Because the incision is essentially horizontal in nature and the scar contractures horizontally rather than 9 vertically, there is little tension on the post operative scar using this approach. For the

same reason, there is no pressure on Cupid's bow (Reddy et al., 2009a)

The deviated septum must be corrected because it offers stability and precise alignment of the previously elevated alar crus of the cleft side and nasal tip, allowing the nose to expand in a balanced manner with equal muscle force on both sides. It also ensures a balanced airflow for both inspiration and expiration. In children, studies have shown that septum modification has no major deleterious consequences (Hans et al., 2008).

Unilateral Cleft Lip Defect is a 3-Dimensional Problem

- Oral -Discontinuity and mal insertion of Orbicularis oris muscle causing horizontal and vertical lip length discrepancy
- Nasal -Deformity of nasal form caused due to mal insertion of Nasalis and other oro-nasal muscles
- Displacement of septum
- Alveolar -Loss of bony support

Correction of the deviated septum provides stability and exact positioning of the previously lifted alar crus of the cleft side and nasal tip, and the nose can grow in a balanced way with equal muscular force being exerted on both sides.

Depending on the surgeon's preference and training, palatoplasty can be performed in a variety of ways. Palatoplasty faces challenges beyond only achieving tension-free closure of the cleft and preventing palatal fistulas, such as increasing palatal length, improving speech results, and having little to no impact on maxillofacial growth. Of the several surgical techniques available, we chose the levator myoplasty to repair the soft palate and the Bardach two-flap technique to repair the hard palate representing Morpho functional repair of cleft palate which helped overcoming the challenges^[8].

Figure 4: Pre and Post Surgery.



Summary

This article demonstrates that the Morpho functional approach, with its numerous combinations and alterations, is versatile enough to provide satisfactory surgical outcomes in patients with unilateral cleft lip, despite the significant variability in size and kind of defects present in patients with syndrome. This is due to the technique's comprehensive character and the balance that it produces among the impacted structures.

Thus, a rare and challenging presentation of Binderoid complete cleft lip and palate case was successfully repaired with intra op technical modifications in nasolabial area.

The operative changes in the nasolabial dimensions were made in anticipation of the rate of growth for these particular features and in consideration of those distortions that would be expected.

Long-term follow-up is needed to assess the nasomaxillary growth and nasolabial repair. Secondary procedures will be required after the growth of this patient and a long follow up.

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