REVIEW ARTICLE

Diagnosis and presurgical orthopedics in infants with cleft lip and palate

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ABSTRACT

Cleft lip and palate are one of the most common congenital craniofacial malformations. While preoperative treatment for infants with cleft lip and palate is still a scientific debate, patients with this malocclusion usually have to be treated from infancy to adulthood. Orthodontist plays an important role in the treatment of patients with cleft lip and palate. The purpose of this review is to give information about cleft lip and palate and presurgical nasoalveolar molding.

Key words
Cleft lip, cleft palate, nasoalveolar molding

INTRODUCTION

Clefts of the lip and/or palate (CLP) are complex craniofacial anomalies that require a multidisciplinary approach. Orofacial clefts involving mandibular-facial system can be classified as syndromic or nonsyndromic according to the association with other syndromes. Cleft palate alone is observed in 30%, whereas cleft lip is observed in association to cleft palate in 50% of CLP-related syndromes.[1-3] Nonsyndromic orofacial clefts include a variety of abnormality related to lip and oral cavity. These abnormalities can be observed at lip alone, palate alone or at both lip and palate [Figure 1]. CLP can negatively influence speech, hearing, perception, health and social life of individual in addition to esthetic issues.[4]

Unilateral cleft deformity is characterized by enlarged external naris and segmented lip at ipsilateral side. Lateral and inferior displacement of lower lateral nasal cartilage results in increased nasal alar rim, oblique columella and prolapsed external naris. If cleft palate is present in addition to cleft lip, nasal septum can deviate to the contralateral side with shifting of nasal base [Figure 2].[5,6]

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Bilateral cleft deformity is typically characterized by frontal localization and rotation of premaxilla. Width of nasal ala is significantly increased with wider segmentation of lip.[7] Flattened nose tip is directly attached to prolabium due to severe deficit or absence of columella. Lower lateral nasal cartilages are severely enlarged, and they are concave rather than being convex. Major problem in esthetic reconstructions is deficit or absence of columella [Figure 3].[8]

CLEFTS OF LIP AND/OR PALATE EPIDEMIOLOGY

Orofacial clefts can be observed in 1–2: 1000 live births. Cleft can be seen at either lip or palate alone, but it can be seen in both lip and palate with different genetic patterns. The highest incidence for cleft lip and palate association is observed in Asian and American populations while it is lowest in African population. There is no racial difference in the incidence of cleft palate alone.[9-11] There is a male preponderance in CLP while cleft palate is more prevalent among females.[11] The incidence was found as 1: 800 in the studies from Turkey.[12] In a study at Southeast Region of Turkey by Göyenç *et al.*,[13] it was suggested that infants with CLP were from rural regions; that they had low socioeconomic level; and that there was higher rates of consanguinity among parents.

CLEFTS OF LIP AND/OR PALATE ETIOLOGY

It has been thought that many factors play a role in the etiology of CLP; thus, genetic and environmental factors

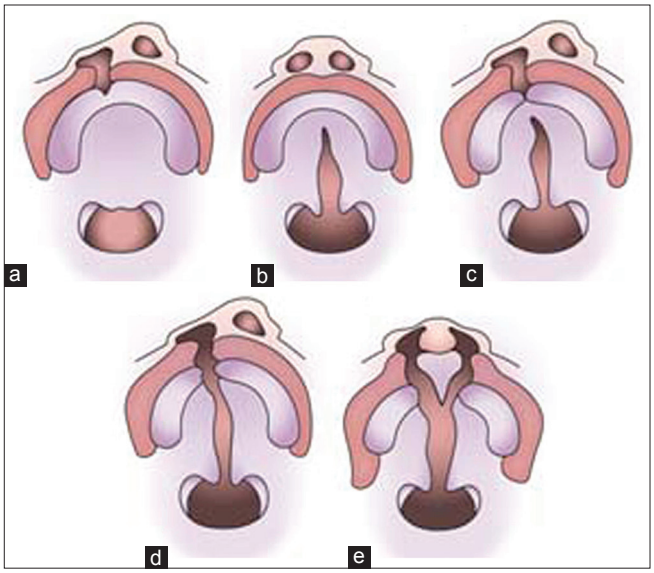

Figure 1: Nonsyndromic orofacial clefts, (a) cleft lip and alveoli (b) cleft palate (c) unilateral incomplete clefts of lip and/or palate (CLP) (d) unilateral complete CLP (e) bilateral complete CLP[5]

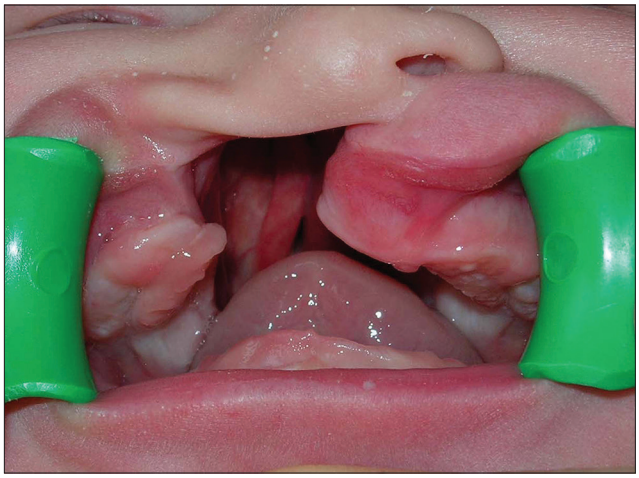

Figure 2: Unilateral cleft

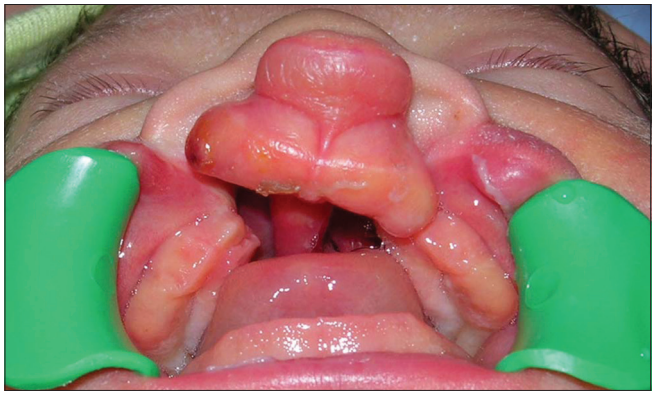

Figure 3: Bilateral cleft

have been investigated. Craniofacial morphogenesis is highly complex and involves a number of interplaying events. Any harmful factor influencing embryo within the first trimester of gestation disrupts normal growth and development, causing irreversible abnormalities.[14-16]

Genetic factors

Gene identification is a novel and rapidly evolving area in individuals with orofacial cleft. Genes thought to be responsible from the anomaly has been identified, and the likelihood of disease in individuals harboring these genes is found to be 25%. However, this rate doesn't necessarily indicate the occurrence of the anomaly.[17]

Common genes thought to be responsible from the syndromes characterized by orofacial clefts have been investigated. It was suggested that interferon regulating factor-6 gene mutation is involved in van der Woude syndrome while poliovirus related gene-1 mutation is involved in autosomal ectodermal dysplasia. In some nonsyndromic clefts, gene mutations related to a specific disease has not been encountered so far. There are ongoing studies on suspected genes.[18] CLP probability ranges from 25% to 45% in monozygotic twins while it ranges from 3% to 6% in dizygotic twins.[19,20] In ongoing human genome studies, it was found that a major gene located on chromosome 9 that causes CLP when mutated is commonly observed in individuals with CLP from several ethnic groups.[17]

Environmental factors

For environmental factors, smoking and alcohol consumption during pregnancy are considered as risk factors for CLP.[21,22] Moreover, it has been reported that several factors such as rubella infection, drug use (phenytoin, salicylate, steroids, aminopterin and busulfan), radiation exposure, excess or deficit of Vitamin A during pregnancy, maternal diabetes mellitus and stress causes CLP.[15,23]

In addition, it was suggested that folic acid and multivitamin supplementation during pregnancy reduces CLP risk.[24] These factors determine CLP risk but do not necessarily cause formation of the anomaly. It is thought that some genes are associated to environmental factors and alterations in these genes can play role in CLP.[25]

ORTHODONTIC TREATMENT OF INFANTS WITH CLEFTS OF LIP AND/OR PALATE BEFORE SURGERY

Individuals with CLP undergo several reconstructive surgeries during management of the disorder. The success of the surgeries depends on successful preoperative orthopedic treatment program. Treatment of infants with CPL requires a multidisciplinary work including a pediatrician, orthodontist, a plastic and reconstructive surgeon, a maxillofacial surgeon, an audiologist, otolaryngologist, a pedodontist, a child psychologist, and a speech therapist.[26,27]

The goals in preoperative orthopedic therapy include facilitation of nutrition, locating tongue at normal position, provision of psychological support to parents, correction of arc form, reduction of cleft width, ensure palatal bone development and soft tissue growth, prevention of risk of collapse in bone segments, prevention of auricular infection and guidance to tooth eruption.[28] It is believed that the alignment in accordance to alveolar anatomy increases chance of surgeon to perform successful lip reconstruction.[29] Prahl *et al.*[30] suggested that alveolar molding procedure has minor, but important effect on speech development.

In 1950s, McNeil[31] designed a plate that brings cleft segment closer. McNeil[31] performed paraffin wax set-up procedure in dental stone cast obtained from impression taken, which approximates cleft regions. Latham,[32] intended to approximate cleft segment using an apparatus with screws. Although there is an approximating spring between screws, plate is adapted into mouth by assistance of a pin. Gnoinski[33] ensured approximation of cleft segments through plates made from acrylic with hard outer and soft inner component.

In 1991, nasoalveolar molding (NAM) method was applied by Grayson *et al.*, Orthodontist and Plastic and Reconstructive Surgeon and MD Court Cutting, Plastic and Reconstructive Surgeon. This method made up shortages of previous methods.[26] By NAM, alveolar reshaping as well as nasal reshaping were emphasized. Maternal estrogen hormone is passing to infant after birth allows reshaping of cartilages by enhancing their plasticity. As estrogen hormone level that increases immediately after birth will show a gradual decrease, reshaping should have to be performed within first 3–4 months after birth. Success rate of nasal reshaping will decrease after postnatal week 12.[26,34]

Goals of nasal reshaping include:[35]

- To approximate alveolar segments located at discrete and asymmetric position by providing symmetry,
- To correct cartilage malposition, to erect columella and nasal dome, to narrow enlarged nasal base.

Although unilateral CLP is more frequently observed, bilateral CLP is more complex deformity with more challenging management compared to unilateral CLP [Figures 4 and 5].[13,35] In bilateral CLP cases, columella formation fails as lower lateral cartilages don't augment toward tip of nose and columella remains short due to failure of sufficient muscle tissue formation in prolabium. Premaxilla and prolabium are located at entirely out of the mouth while lateral alveolar segments are located within mouth independent of premaxilla. In these cases, there is a deformity at nasal region in addition to CLP and alveol [Figure 5].[36,37]

To increase success rate in individuals with bilateral CLP, preoperative procedures include orthopedic retraction of premaxilla, lengthening and erecting columella, correction of deformity in nasal cartilage, approximation of alveolar segment by aligning and approximations of discrete lip segments by narrowing enlarged nasal base and external nares.[36,38]

STAGES OF NASOALVEOLAR MOLDING

Nasoalveolar molding defines the reshaping procedure of nose together with alveolar segments in neonates with

Figure 4: An infant with unilateral clefts of the lip and/or palate

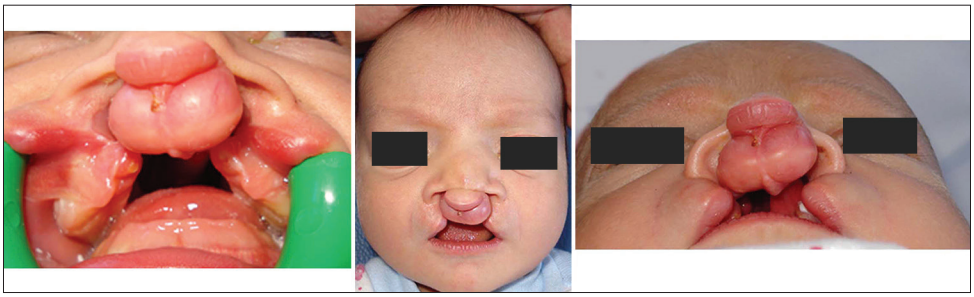

Figure 5: An infant with bilateral clefts of the lip and/or palate

CLP before surgery. In a neonate presented with CLP, initial impression is taken using silicon-based impression material [Figure 6]. It is important to hold an infant upside down or at upright position due to the risk of aspiration of impression material into airway. Then, traditional molding plate is prepared form cast obtained with dental stone using hard acrylic. In order to prevent removal of plate from mouth, adhesive tapes are applied from retention arms at plate to cheek for retention. One retention added is added to plate in infants with unilateral cleft, whereas 2 retention arms in those with bilateral cleft [Figure 7]. It should be taken care to position retention arms at an angle of 45° to occlusal plane. The dentist has 3–4 months after birth to correct alar cartilage distortion and asymmetry and to restore nasal tip projection considerably.[26]

Stages of unilateral nasoalveolar molding

In unilateral cleft lip, a major segment is located as being deviated to lateral at anterior and to medial at posterior while minor segment is located as being deviated to medial at anterior and to lateral at posterior. Thus, soft acrylic is added to the outer component of plate that encompasses major segment while hard acrylic molding was performed with same amount at contralateral side [Figure 8]. Soft acrylic should be added to plate fitting to internal surface of a minor segment while hard acrylic at same amount should be abraded at contralateral side. The amount of soft acrylic added should not excess 1–1.5 mm. Adjustment of alveolar segments should be performed by 1-week intervals over 3–4 weeks and nasal reshaping should not be performed until distance between segments is reduced below 5 mm. Otherwise; there is a risk for formation of mega nostril.

After alignment of alveolar segments, reshaping procedure of nasal cartilage is undertaken. Nasal reshaping procedure is achieved by a nasal stent, which extends from labial surface of plate within nose as passing middle of cleft line. There is no need to take a nasal impression when preparing nasal stent. Nasal stent is made up of 0.36 or 0.04 stainless steel wire by guidance of wax template prepared while plate is in the mouth of infant. Nasal stent is attached to the inferior surface using hard acrylic. The part of nasal stent that will be inserted to nose is coated by hard acrylic first with overlaying soft acrylic in order to prevent irritation of nasal mucosa. As adaptation of nasal stent changes, activation required can be obtained by adding soft acrylic [Figure 9].

Figure 6: Intraoral impression

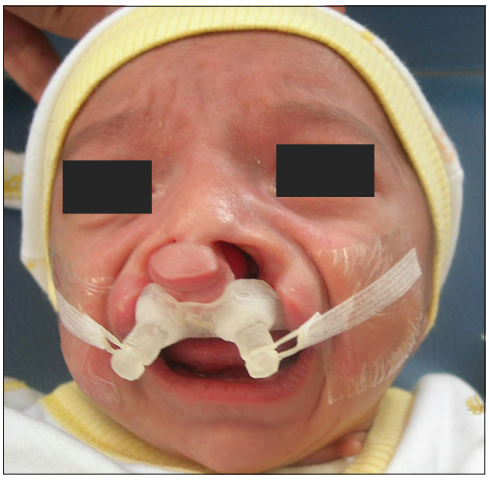

Figure 7: Adhesive tapes from retention arms to cheek

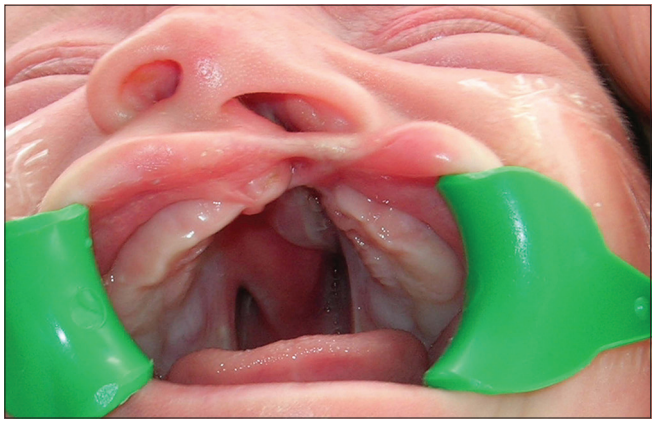

Figure 8: Deviation of alveolar segments in unilateral cleft lip

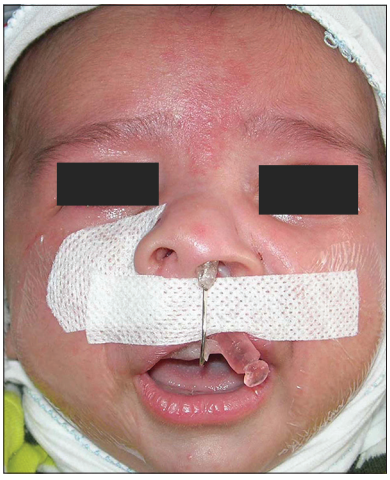

Figure 9: Application of horizontal and vertical adhesive tapes over Tegaderm

At this point, adhesive tapes applied to bilateral cheeks of infant that approximates lips will allow surgical procedure with less tension as it ensure lengthening of lip muscles. In addition, adhesive tapes applied vertically at lateral to nasal ala will help supporting nasal ala [Figure 9].[35]

Stages of bilateral nasoalveolar molding

After preparation of the plate on the cast obtained from intraoral impression, 2 retention arms are applied on cleft line. Plate retention is achieved by adhesive tapes that are looped on retention arms and secured to cheeks [Figure 5]. It should be assessed whether there is any deviation at premaxilla and whether there is enough space for meshing of premaxilla. If there is a deviation at premaxilla, this deviation should be corrected first, and premaxilla should be forced to its position between posterior segments. In the case of insufficient space between posterior segments, it is required to enlarge posterior segments. For this purpose, soft acrylic is added to internal part that covers posterior segments while hard acrylic is removed from contralateral side. When alignment is achieved, and distance between segments is reduced below 5 mm, the procedure proceeds with nasal reshaping after 3–4 weeks.

Since estrogen level is at peak levels within first 3–4 months after birth and decreases gradually, nasal reshaping should be completed within first 4–5 months of life.[26,34] In contrast to infants with unilateral CLP, 2 nasal stents are attached to plate in infants with bilateral CLP. Nasal stents are made up of 0.36 or 0.04 stainless steel wire which is coated with soft tissue in order to prevent irritation nasal mucosa. Soft tissue bridges from soft acrylic are added between nasal stents in accordance to columellar fold. Lengthening columella before surgery yields better aesthetic outcomes and reduces scar risk as it eliminate the need for an incision on the lip. Otherwise, scar tissue formed restricts development of columella and nasal projection, causing a flattened and large nasal appearance. This also reduces the risk for potential nasal surgical revision [Figure 10].[29,38]

Tegaderm™, commercially available protective tapes, can be applied to prevent irritation of cheek while applying adhesive tapes in cases with unilateral or bilateral CLP. These special tapes can be left in place over a week and allows application of retention and lip tapes over it [Figure 9].

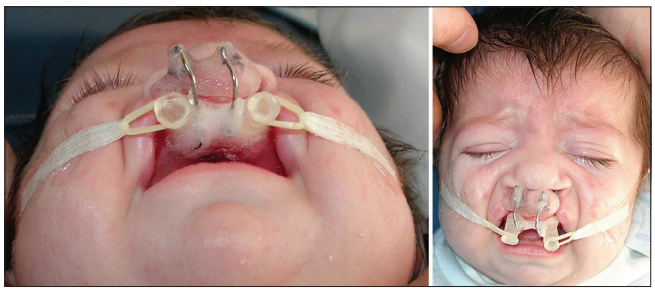

Figure 10: Reshaping columella with nasal stents

When NAM procedure is successfully performed, deformity at lip, nose alveolar region can be treated by a single surgery, eliminating need for additional gingivoperiosteoplasty. Need for secondary bone graft will be reduced by 60% when gingivoperiosteoplasty procedure is performed together with first surgical intervention where alveolar segments are approximated. Moreover, tension will be reduced with minimal risk for scar formation when distance between gingival tissues are closed or reduced to a minimal level.[29,35,38]

Cases without need for preoperative orthodontic treatment

If there is no cleft at hard palate and cleft is a secondary cleft palate involving soft tissues and uvula or if there is no nasal deformity with a cleft forming an alveolar notch without a condition that make nutrition of infant challenging, no preoperative orthodontic treatment is needed.[39]

However, a nutrition plate will be appropriate if there is an isolated cleft palate that makes nutrition of infant challenging.

SURGICAL PROCEDURE

The first operation is performed when child is 6 months old, and anterior nares are formed by suturing lip and nasal alae [Figure 11]. It is recommended to suture cleft in hard plate before speech development at about 12–15 months of age based on consensus of speech therapists, orthodontist and plastic and reconstructive surgeons.[17,29] The child should be followed until 5–6 years of age regarding speech development, oral hygiene, pediatric dentistry and skeletal development counseling.[29]

CONCLUSIONS

Individuals with cleft lip and palate should be evaluated very carefully. Enough knowledge regarding malocclusion and long-term orthodontic intervention is needed for successful management of patient with cleft lip and palate.

Advances in the correction of deformity by team approach in CLP management allow increasing success level markedly. Functional and esthetic improvement achieved in these patients has already reached to a level that ensures optimum adaptation to the social environment and comfort of these patients.

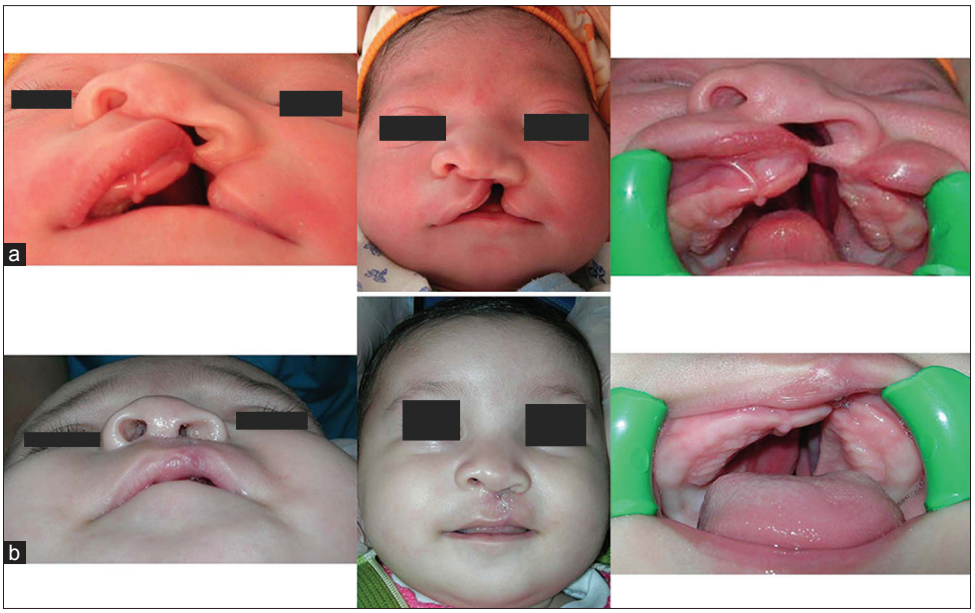

Figure 11: (a) Intraoral and extraoral photographs of the case at baseline. (b) Intraoral and extraoral photographs of the case later first surgery

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