

Cleft Lip and Palate: The Multidisciplinary Management

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ABSTRACT

Patients with cleft lip and palate usually face a multitude of problems, esthetic compromise being the most noticeable, malocclusion, missing teeth, oronasal fistula, speech and hearing pathology are also present in most cleft patients; this necessitates a multidisciplinary treatment across various medical and dental specialties that extends from birth to adulthood but in separate stages. A protocol for the treatment of cleft patients should followed by the healthcare providers, and coordination amongst them is a major contributor to success in cleft treatment.

KEYWORDS

Cleft patients, Treatment protocol, Teamwork management.

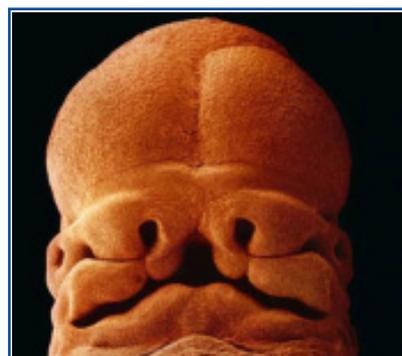
INTRODUCTION

Cleft lip and/or palate are the most common craniofacial anomalies, occurring disproportionately across the world. Cleft anomalies affect several organs and functions within the human body, necessitating multidisciplinary treatment across various specialties.

This article gives a brief comprehensive overview of cleft lip and palate, including the embryology and etiology of cleft formation, the incidence of clefting, and the contemporary multidisciplinary treatment approach of cleft lip and palate.

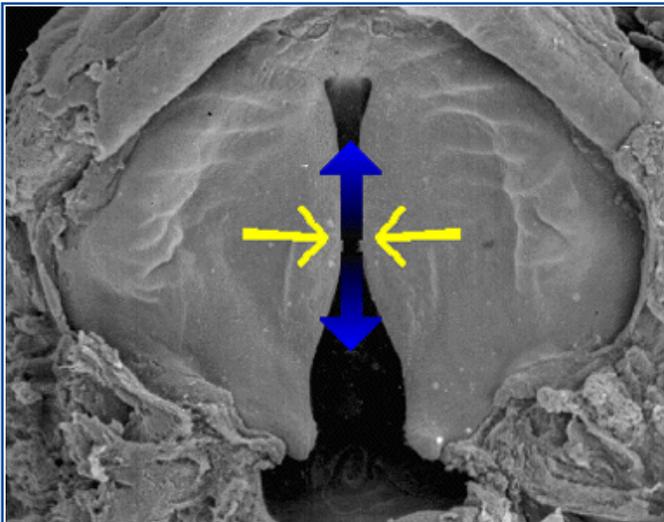
Embryology of cleft formation

The critical period for proper intrauterine development of the face is between the fifth and seventh weeks of gestation, with the sixth week being the most important. During this time, the morphodifferentiation and orientation of the unpaired frontonasal process, which includes the medial and lateral nasal processes, occur simultaneously with progressive medial migration and growth of the paired maxillary processes (**Fig. 1**).¹



(Fig. 1) Formation of lip and primary palate

The palate is formed in two stages: In the first stage, the primary median palatal triangle is formed, derived from the merging of the two mesial processes originated from the median frontonasal process. This is completed by the eighth intrauterine week. In the second stage, shelf-like outgrowths known as lateral palatine processes, derived from the maxillary processes, grow horizontally above the tongue to form the secondary palate.



(Fig. 2) Formation of secondary palate

The merging or fusion of these processes is completed by the twelfth intrauterine week **(Fig. 2)**.

Any insult to the fusion process between the fifth and seventh intrauterine weeks leads to the formation of clefts. This may lead to irritation of the nasal septum, causing repeated respiratory tract and middle ear infections. Additionally, the tongue may be postured in the cleft space during swallowing, which may further widen the cleft space.

Etiology of cleft Lip and Palate

The etiology of cleft remains unclear. It is presumed to be multifactorial, with various contributing environmental and genetic factors.²

Toxin exposure during the antenatal period is more likely to be present in developing countries due to poor sanitation, inadequate infrastructure, and political instability. For many of the same reasons, metabolic disorders and malnutrition are also more likely to be an issue for pregnant women in developing countries. Genetic factors and consanguinity are also major potential risk factors for birth defects.³

Problems Associated With Clefts

Patients with cleft lip and palate usually face a multitude of problems, esthetic compromise being the most noticeable.

The esthetic obstacle in children with cleft lip and palate may often lead to various types of psychosocial distress. For example, patients may feel that there is a stigma attached to their appearance. They often perceive themselves as unattractive, which may influence their psychological behavior and lead to problems with communication.⁴

Esthetic issues can also limit a child's life prospects. Some affected children may not attend school regularly, due to teasing, bullying, speech difficulty, and hearing problems.⁶

Additionally, adult patients with cleft anomalies may also suffer from marital problems. These and other scenarios all require psychosocial support for the cleft patient as well as his/her family.⁵

By improving a child's social skills, educational pursuit, both the child and his/her parents may be able to overcome the issues that stem from the child's physical appearance.⁵

Other associated abnormalities include oronasal fistula, speech and hearing pathology, malocclusion, which is always present in cleft lip and palate patients. Anterior and posterior crossbites due to anteroposterior and transverse deficiency of the maxilla, rotation of the incisors due to muscle pull, lateral incisors at the cleft site are frequently missing and supernumerary teeth may be present at the non-cleft side. Carious teeth and periodontal inflammation are often present due to dental neglect **(Figs. 3-5)**.^{6,7}



(Fig. 3) Skeletal and Dentoalveolar irregularities associate with cleft lip and palate



(Fig. 4) Oronasal fistula associated with cleft palate



(Fig. 5) Repaired oronasal fistula during alveolar bone graft

TREATMENT TIMING & PROTOCOL

The First Closure: Lip

The first procedure that cleft patients receive is surgical closure of the lip. Following the "rule of 10s," a patient admitted for lip closure should be 10 weeks of age, weigh at least 10 pounds, have a hemoglobin $\geq 10\text{mg}/100\text{ml}$, and have aWBC count $< 10.000\text{cu}/\text{mm}$ in order to withstand surgery and anesthesia (Figs. 6,7).^{8,9}

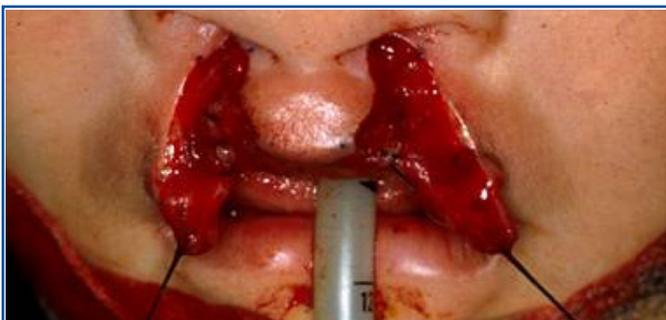


(Fig. 6) Bilateral Complete Cleft lip and palate before lip repair



(Fig. 7) Bilateral Complete Cleft lip and palate after lip repair

Neonatal orthopedic correction is still used in several cleft lip and palate centers around the world, despite several reports questioning its effectiveness. In patients with wide bilateral cleft lip and/or a severely protruding premaxilla, surgical taping as an alternative method for neonatal orthopedics has many benefits: it is easy to use, does not require several visits to pediatric dentists or orthodontists over a period of months, and the results are often successful. When used, this taping is worn for two weeks prior to the Von Langenbeck lip or Millard forked flap surgery (Fig. 8).⁹



(Fig. 8) Surgical lip repair

In this approach, physiological forces will enhance palatal molding and will bring the premaxilla backwards. This in turn shortens the time for lip closure. When surgical taping is used rather than lengthy neonatal orthopedics, lip

closure surgery can be performed 10 weeks later, thereby decreasing the psychological distress of the family as they witness the correction of their child's cleft.

Hard & Soft Palate Surgical Repair

Speech is learned by hearing, which can be aided by surgical repair of the hard and soft palate. This surgery is performed around 12-18 months of age, when children first begin to talk. The surgery is also performed at this age with airway considerations in mind, as it benefits dentition eruption.⁹

Palatal repair is done in a single stage using the Vomer flap for anterior palate closure, in addition to the Bardach two-flap palatoplasty with soft closure into both alveolar clefts (Fig. 9).



(Fig. 9) Surgical repair of the palate

Treatment in the Primary Dentition Stage

In the primary dentition stage, focus is on restoration of the primary dentition using mainly fillings, stainless steel crowns, and oral hygiene rather than orthodontics or surgery (Fig. 10).



(Fig. 10) Dental management in the primary dentition stage

Sometimes, expansion by a removable appliance is used in cross bites with functional shifts of the mandible upon closure. At this young age, cooperation of the patient remains a factor. This further necessitates delay of treatment to the mixed dentition stage.⁷

Treatment Principles in the Mixed Dentition Stage

The main objective during the mixed dentition stage is to prepare the cleft patient for a bone graft. The collapsed (overlapped) alveolar segments may be impeded in their growth. It is important that these segments be unlocked by expansion during the early stages of development when growth is most rapid. Orthodontic expansion of collapsed buccal segments will also facilitate the push-back of the premaxilla to restore a favorable arch form, which was initially interrupted by the lack of alveolar bone continuity on the cleft side.

Expansion is carried out successfully by a quadhelix expansion appliance (Fig. 11), giving the surgeon a more favorable surgical field to perform the bone graft. Expansion devices should be used for at least four months after the bone graft, as freshly grafted bone is unable to maintain the expansion (Fig. 12).⁷



(Fig. 11) A quadhelix expansion appliance



(Fig. 12) Collapsed buccal segments in unilateral cleft palate patient before expansion

Crowding is usually present on the non-cleft side in unilateral cleft patients, and should also be relieved during the mixed dentition stage. At the crowding site, serial extractions can sometimes be carried out to provide space for eruption of the canine, while lateral incisors are usually missing at the cleft side. Supernumerary teeth should be extracted during the bone graft surgery and primary teeth adjacent to cleft should be extracted at least two months prior to surgery (Fig. 13).¹⁰



(Fig. 13) Post orthodontic expansion of collapsed buccal segments

The central incisor is usually distolabially rotated and inclined due to muscular pull (Fig. 14). Care should be taken while leveling this incisor and other teeth, so as not to push the roots of these teeth into the cleft space. Following a successful graft placement, it is preferable to keep the incisor roots invested in bone at the pre-grafting stage while the correct tooth angulation is achieved at least 2 months after grafting (Figs. 15, 16).⁷



(Fig. 14) The central incisor is usually distolabially rotated and inclined in a cleft palate patient



(Fig. 15) Corrected position of the central incisor

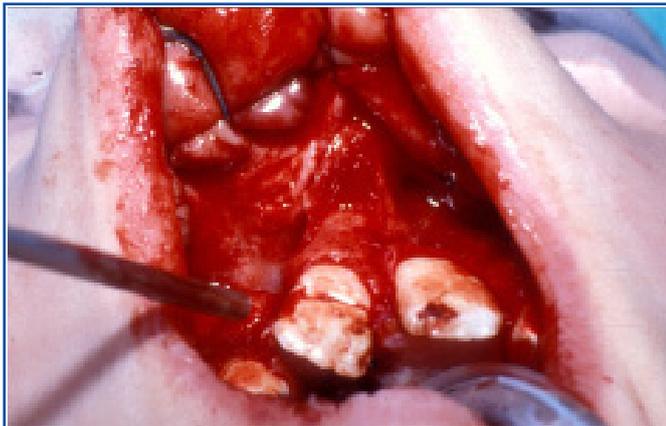


(Fig. 16) A successful bone graft placement appearing by a periapical X-ray

Bone Graft at the Cleft Side

Bone grafting has several advantages. It is performed to support the long-term expansion of the dental arch, maintaining arch continuity and form while also providing bone for the passage of the erupting canine through the graft.⁶

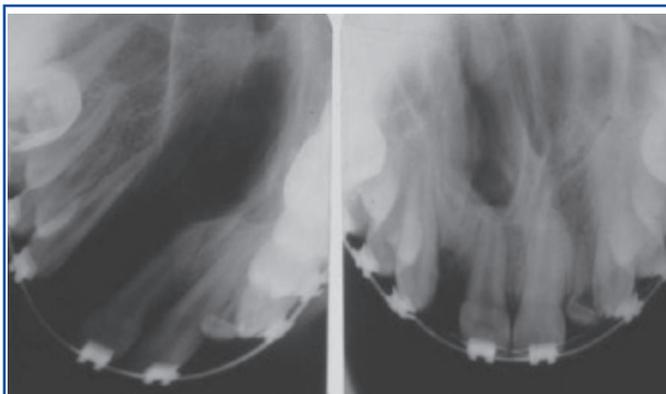
A bone graft will also support the teeth adjacent to the cleft site, ensuring orthodontic movement of these teeth without periodontally compromising them. The bone graft will also improve facial esthetics by providing support to the base of the nose, which is lacking due to the cleft space (Figs. 17-19).



(Fig. 17) Alveolar bone exposed at cleft side



(Fig. 18) Cancellous bone packed into cleft



(Fig. 19) Incisor roots invested in bone at the pregrafting stage while the correct tooth angulation can be achieved after successful graft placement

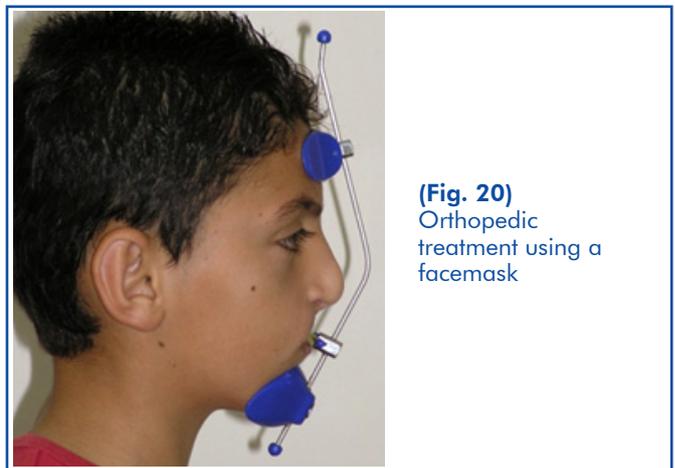
The bone graft is usually taken from the cancellous bone of the iliac crest. Other donor sites that provide cancellous bone include the mandibular symphysis and the skull.

Timing of alveolar bone graft relates to tooth development and is carried out between the ages of 6-9 years, using the stage of root formation of the maxillary canine as a guide for the proper timing for bone graft is better than just following chronological age, when one third of the canine is formed viewed by a periapical x-ray is the best time to consider bone graft. This may lead to spontaneous canine eruption through the graft and healthy gingiva surrounding the graft, with normal bone height.^{6,9}

Orthopedic Mid Face Growth Enhancement

Maxillary hypoplasia is a well-known feature of cleft lip and palate patients. It may be due to scarring from the surgical repair of the cleft lip and/or palate, which may cause lip tightness and palatal scarring that restricts the growth of the maxilla. Studies by Ross et al. showed that untreated cleft patients exhibited normal maxillary growth, further supporting this hypothesis.

Orthopedics applies interrupted forces which can aid maxillary sutures. However, the success of maxillary orthopedic advancement remains limited due to the tightness of the lip and palatal scarring. The decision to use a face mask or to undergo orthognathic surgery should be made early during treatment planning for patients with mild to moderate skeletal discrepancies (Fig. 20).^{11,12}



(Fig. 20) Orthopedic treatment using a facemask

Orthognathic surgery

Cleft patients will usually have jaw disharmony at the end of their facial growth period, manifested by mid-face deficiency. Growth of the maxilla is impaired in sagittal, transverse and vertical dimensions, evident in the form of anterior and posterior cross bites. Mandibular over-closure is also apparent due to maxillary deficiency in the vertical dimension.

Orthognathic surgery is required in about 25% of adult cleft patients according to the severity of skeletal features. The most commonly affected jaw in cleft patients is the upper jaw. Thus, in most cases,

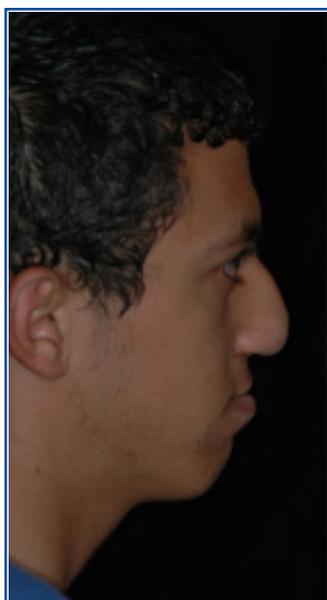
the orthognathic procedure performed involves the advancement of the upper jaw (Le Fort I) rather than setting the mandible back. When the reverse overjet is severe, a two-jaw surgery may be required, impacting breathing and esthetics (Figs. 21-24).⁷



(Fig. 21) Cephalometric X-ray for adult cleft patient before surgery



(Fig. 22) Surgical advancement of the upper jaw (Le Fort I) for the same patient in Figure 21



(Fig. 23) Lateral facial view of a cleft patient before orthognathic surgery



(Fig. 24) Lateral facial view of the same cleft patient in Figure 23 after maxillary advancement and mandibular set back

The proper timing of orthognathic surgery is when facial growth has ended. A hand and wrist radiograph can help determine the end of facial bone growth.

Orthodontic preparation for the orthognathic surgery aims to correct dental compensation due to the skeletal discrepancy. Other goals of pre-surgical orthodontics include arch coordination and removal of any occlusal prematurity.

Relapse in the upper jaw position is possible due to the scar tissue resulting from the upper lip and palate repair. Distraction osteogenesis enables the gradual advancement of the hypoplastic maxilla with corticotomy cuts that will allow expansion of the scar tissue (Figs. 21,22). This decreases the possibility of relapse and allows for a longer-term correction over orthognathic surgery.

Distraction osteogenesis has recently benefited from major advances in instrumentation, especially regarding the materials used to create instruments and the creation of bidirectional instruments. This has improved results and further decreased the need for more orthognathic surgery.¹²

SUMMARY

Patients with cleft lip and palate require a team approach for their treatment, comprised of several specialists. This multidisciplinary care starts from birth and continues into adulthood, and coordination amongst specialists is a major contributor to success in cleft treatment.

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