Review article

Cleft Lip and Palate: Overview of Causes, Surgical Repair, and Treatment Approaches: A Narrative Review

Amir Hossein Alaviyan[1*]

1. General Surgeon, Department of General Surgery, School of Medicine, Tehran University of Medical Sciences, Tehran, Iran.

ABSTRACT

Cleft lip and palate is one of the most common congenital malformations of the skull and face (craniofacial). The deformity is characterized by a loss of integrity of the lip muscles, alveolar bone, and hard and soft palate. The severity of the deformity can range from a small hole in the lip to a large fissure extending into the roof of the mouth and nose. Management and control of cleft patients is a challenge. Intervention in these patients begins very early in life, from the fetal period, and continues into adulthood, and the patient's relatives are always involved in the treatment process. Since there are multiple problems in these patients, the treatment process is managed in a group and multidisciplinary manner to achieve the desired result. The goal of all treatment measures for a person with a cleft lip and palate is to provide intelligible speech, hearing within a normal range, good facial appearance, proper lip and nose symmetry, an attractive smile, good occlusion, healthy oral structures, and ultimately appropriate self-confidence. In the current study, I reviewed the causes, pathophysiology, surgical repair, and treatment approaches in patients with cleft lip and palate.

KEYWORDS
Cleft lip; Cleft palate; Congenital anomalies; Surgery

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***Corresponding Author:**

Amir Hossein Alaviyan

General Surgeon, Department of General Surgery, School of Medicine, Tehran University of Medical Sciences, Tehran, Iran

Email:
amirhosseinalavian41@gmail.com
dralaviyan@gmail.com

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INTRODUCTION

Congenital anomalies include chromosomal defects, molecular and anatomical abnormalities that occur during the embryonic period and their effects are visible in the newborn after birth [1, 2].

Cleft lip and palate are the most common congenital defects in the jaw and face, which are observed in various forms of cleft lip, palate or cleft lip and palate [3]. These anomalies are observed in one of 3 forms: isolated, syndromic or associated with other anomalies but non-syndromic. The syndromic form of the disease is associated with syndromes such as Velo-cardio-facial, Stickler syndrome and Hemifacial Microsomia syndrome [4, 5].

Non-syndromic cleft lip with or without cleft palate is one of the most common birth defects, and its prevalence varies in different ethnic, social, and economic communities [6, 7]. Some studies have been conducted worldwide on the prevalence of cleft lip and palate, and

different incidence and prevalence rates have been reported [8]. Based on the results of a meta-analysis on the prevalence of cleft lip and palate in Iran from 1987 to 2009, this figure was calculated to be one in every thousand live births [9].

Genetic factors play an important role in causing these abnormalities, but environmental factors are also important. Maternal hormonal disorders, use of neuropsychiatric drugs, deficiency of some vitamins and folic acid, hypocalcemia, smoking, maternal obesity, and even the effect of the season on their prevalence have been reported [10, 11].

Cleft lip and palate can cause various problems for sufferers, such as changes in facial appearance, feeding problems, dental disorders and improper occlusion, as well as respiratory, hearing, speech and social interaction problems [12]. The treatment of these defects is usually long-term and multi-stage, requires various and multidisciplinary specialties, and imposes high costs on the family and healthcare institutions[11].

The study aimed to review the causes, pathophysiology, surgical repair, and treatment approaches in patients with cleft lip and palate.

Cleft lip

Failure of fusion of the fronto-oral and maxillary processes, resulting in varying degrees of cleft of the lip, alveoli, and nasal floor (an incomplete cleft does not cross the nasal floor, while a complete cleft means there is no connection between the alar base and the middle labial element)[13].

Cleft palate

Failure of fusion of the palatine shelves of the maxillary appendages, resulting in a cleft of the hard and/or soft palate. Clefts develop during the fourth stage of development [14]. The exact location of their appearance is determined by the locations where fusion of the various facial appendages has not occurred, and this in turn is influenced by the time in fetal life when some developmental interference has occurred [13].

Pathophysiology

These complications occur due to disruption in the process of connecting the facial prominences during the embryonic period. In cleft lip, it occurs due to disruption in the connection between the middle nasal processes and the maxillary prominences in the sixth week of embryonic development. Certainly, syndromic cases should be evaluated for abnormalities of other organs in the specific syndrome [13, 15]. The development of the primary and secondary palates occurs at different times and involves different developmental processes. The primary palate includes the medial part of the upper lip and the alveolar ridge that encloses the central and lateral teeth. The secondary palate includes the remainder of the hard palate and all of the soft palate. A cleft lip is the result of a defect in the fusion of the maxillary bone with the nasal protuberance on the frontal protuberance to form the primary palate, which occurs naturally at 6 weeks of gestation. Cleft palates vary from a bipartite uvula to a complete cleft palate that extends from the soft palate to the hard palate [13, 16]. Cleft palate is divided into two groups: complete, which extends into the nose (involving both the soft and hard palates), and incomplete, which maintains a midline connection. In addition, submucosal cleft palate, which is characterized by three features: a cleft uvula, a thin membranous portion, and a palpable pharyngeal depression, is also highly prevalent [17, 18].

Etiology

Many factors can lead to cleft lip and palate or both, which may be internal or external causes. Taking medications such as corticosteroids, antiepileptics, salicylates, aminopterin, and contracting diseases such as rubella and diabetes during pregnancy have an effect on the development of cleft lip and palate [19]. The incidence of associated congenital anomalies is increased in infants with oral clefts, especially in the case of a single cleft palate. Facial clefts can be sporadic or part of a syndrome [18].

Prenatal period: diagnosis

Today, with advances in ultrasound technology, it has become possible to diagnose oropharyngeal clefts before birth. Although cleft lip and alveolar bone clefts can be diagnosed with ultrasound imaging in the second trimester of pregnancy, the diagnosis of cleft hard and soft palates is still limited and difficult, even with 3D imaging [20, 21].

Dental problems in cleft lip and palate

Various abnormal dental conditions include:
1. Natal and neonatal teeth: The presence of neonatal teeth does not appear to affect the deciduous or secondary teeth in the clefts. Unlike in infants without clefts, most of the natal teeth in infants with clefts are located between the clefts on the lateral margins of the premaxilla and mandible [21,22].
2. Microdonti: Small teeth (microdonts) are often present in the impacted lip/teeth and are generally more prevalent in cases where the lateral incisors are absent. The lateral incisors of the upper jaw are generally seen as wedge-shaped [21].
3. Taurodontism: According to studies, taurodontism is associated with certain syndromes and dental growth disorders.
4. Ectopic eruption: The clefts contribute to the misalignment of the primary lateral incisors, which may erupt palatal adjacent to or within the cleft, while the permanent canines may erupt palatal on the side of the alveolar clefts. Delayed eruption of the permanent incisors may be observed [22,23].
5. Enamel hypoplasia: Enamel hypoplasia, especially in the maxillary central incisors, is more common in individuals with cleft lip/palate compared to non-cleft populations [22].
6. Delayed tooth maturation: Several growth factors are important during craniofacial development. These factors may be over- or under-expressed in cleft lip and palate defects. This abnormal expression causes altered odontogenesis as well as tooth blade abnormalities [22].

Newest treatment of cleft lip and palate
Early, Pre-Surgical Treatment

presurgical nasoalveolar molding is a pre-surgical procedure (PNAM, also known as the Figueroa NAM technique). This procedure was first introduced by Grayson in 2004[22]. This reduces the stigmata of the cleft, especially around the nose and lip area. It shows a decrease in the width of the columnella and an improvement in the height of the columnella. It also shows a decrease in the bilateral width and an increase in the bilateral height [23].
Preoperative preparation may require a NAM plate, an acrylic orthodontic appliance designed to stimulate maxillary growth and alter the growth pattern of a patient with cleft lip and palate. The new plates are 3D printed. The NAM plate helps rotate the premaxilla and reduce the size of the cleft lip and palate [24]. Moreover, changes in the width of the maxilla are also observed in the canine and molar regions [25].
It should be applied as fast as possible due to the possible problems with neonate adaptation. After the preparation, the plate should be worn daily with only a short time for cleaning the plate twice a day [26]. Preoperative preparation also changes the rotation of the incisal bone, so the alveolar bone shape becomes less triangular and more natural, and the cleft palate spontaneously decreases [23]. However, long-term follow-ups have shown that only the shape of the dental arch remains stable over time, other abnormalities persist [27]. Possible methods for mobilizing soft tissues include lip massage and lip taping to make the skin more elastic and close it over the alveolar bone [28].

Lip and/or Palate Closure

The primary surgical procedures for patients with cleft lip and palate are cosmetic surgery of the lip and palate as well as alveolar bone grafting. In most countries, lip and palate operations are performed separately and have different timings: 3–6 months for the lip and 6–18 months for the palate closure [29]. Recently, the C-flap technique was introduced by Jung et al [28].
This method results in lip elevation during surgery and, therefore, less philtrum and cupid's bow deformity than the traditional method. The selection of surgical repair method depends on the surgeon's experience and the type of cleft. Most surgeons combine the Fischer's technique with a modified Maillard rotational-progressive flap for lip repair [30, 31]. Among the many surgical procedures for closing the palate, non-radical intravelar veloplasty is one that may be used to repair it [32]. After lip surgery, post-operative care includes scar management, massage, and monitoring for wound potential dehiscence, and the use of silicone gel, as well as steroid administration[29]. The modern scar management methods require lasers to reduce scar tissue by softening and smoothing it[33].

Bone Grafting

Most patients with alveolar bone clefts require

bone grafting to restore bone shape for future tooth movement and prosthesis repair. The timing of bone grafting is controversial - several techniques are used for it. The most prevalent donor site is the iliac crest (due to the possibility of increasing cancellous bone up to 50 ml) or calvarial bone. Autogenous bone is the best material that can be used in any type of graft. Bone should be grafted slowly, as bone fragmentation increases its resorption, thereby reducing its volume and quantity [34, 35]. Bone grafting is usually performed in the mixed dentition stage and, in addition to repairing the resorbed nasal bone, it also becomes a foundation for the upper nasal bone. Reconstruction of the length of the alveolar bone allows for the restoration of missing teeth [36]. Now, bone grafting must be performed based on 3D planning, which allows for the production of realistic, true-to-size bone grafts. Additionally, 3D scans can be used to print bone graft scaffolds made of bio-glass [37].

Postoperative care: cleft lip

One of the most important things to do after surgery is to protect the surgical site. Children who undergo cleft lip surgery should not be breastfed or fed by syringe while the child is awake after surgery. Some surgeons recommend that the child be discharged after cleft lip repair on an outpatient basis or after an overnight stay in the hospital for monitoring [38]. Pain control continues at home, and parents are instructed in the use of appropriate doses of painkillers. A thin layer of antibiotic ointment may be prescribed for 6 days, followed by a gel for several weeks [39, 40].

CONCLUSION

Cleft lip and palate is one of the disorders that leads to physical and emotional disorders in the individual, family, and society after birth. Therefore, correcting the defect at an earlier age and reducing subsequent disorders associated with cleft lip and palate can be an effective health and treatment approach. Treatment of patients with cleft lip and palate is challenging due to the multiple problems they present which requires strong teamwork. A multidisciplinary approach to treatment of cleft patients, when combined with proper diagnosis and treatment planning, results in good occlusion, function, and aesthetics. Ultimately, the ideal treatment for a cleft patient is one that enables them to speak coherently, hear within normal limits, have a good facial profile, symmetrical lips and nose, an attractive smile, good occlusion, and healthy oral structure, and ultimately have the self-confidence to participate in society.

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CONFLICT OF INTERESTS

There is no conflict of interests.

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