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Cleft Lip and Palate - A Review Article

Shweta Chaudhary

Intern, SGT University, Gurugram, Haryana.

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ABSTRACT

Craniofacial anomalies, and in particular cleft lip and palate, are major human birth defects with a worldwide frequency of 1 in 700 and substantial clinical impact. The cause of clefts is multifactorial with both genetic and environmental input and intensive research efforts have yielded significant advances in recent years facilitated by molecular technologies in the genetic field. In 2008, the World Health Organisation (WHO) has recognised that non-communicable diseases, including birth defects cause significant infant mortality and childhood morbidity and have included cleft lip and palate in their Global Burden of Disease (GBD) initiative. This will further facilitate the birth defects registration and international efforts aimed at improving quality of care and ultimately prevention of non-syndromic clefts of the lip and palate.

Key words: Cleft lip, Cleft palate, Dentistry, Etiology, Management.

INTRODUCTION

A cleft is a congenital abnormal space or gap in the upper lip, alveolus or palate. Thus, a cleft lip and palate may be defined as:

Cleft lip: The failure of fusion of the fronto-nasal and maxillary processes, resulting in a cleft of varying extent through the lip, alveolus, and nasal floor (an incomplete cleft does not extend through the nasal floor, while a complete cleft implies lack of connection between the alar base and the medial labial element).[1]

Cleft palate: The failure of fusion of the palatal shelves of the maxillary processes, resulting in a cleft of the hard and/or soft palates.[1] Clefts arises during the fourth developmental stage. Exactly where they appears is determined by locations at which fusion of various facial processes failed to occur, this in turn is influenced by the time in embryologic life when some interference with development occurred.[2]

Cleft lip and palate is a congenital anomaly affecting the orofacial region. Orofacial clefts are the most frequently occurring craniofacial birth defects. It may present alone or in combination and/or with other congenital deformities such as congenital heart diseases. The condition is often associated with various syndromes such as, Pierre Robin syndrome, Apert's syndrome and more.

The defect may present itself as a result of interplay of various genetic and environmental factors (eg: Nutritional factors, such as vitamin A deficiency or excess., Maternal illness and drug usage., Infections such as rubella., and more).

Overall incidence of cleft lip and palate is approximately 1 in 600 to 800 live births (1.42 in 1000) and isolated cleft palate occurs approximately in 1 in 2000 live births. Thus, the typical distribution of cleft types are:[3]

1. Cleft lip alone – 15%
2. Cleft lip and palate – 45%
3. Isolated cleft palate – 40%.

This condition has a slight male predilection. Generally, left side of the face is more involved than the right side. A complete cleft may extend upwards and involve the nostril, which results in

deflection of nasal tip towards the non-cleft side and larger nares on the cleft side. The zones commonly affected are:

1. Upper lip
2. Alveolar ridge
3. Hard palate
4. Soft palate
5. Nose and eyes, in rare cases.

The condition has a significant effect on parents as well as the child due to the psychological and socio-economic implications associated with these deformities. Breastfeeding a child with oro-facial cleft becomes difficult since they're unable to generate sufficient suction along with the problem of nasal regurgitation of the milk.

Impairment of facial aesthetics, speech, mastication, deglutition and dental occlusion are potential problems associated with this condition. The clefting of lip and/or palate occurs at such a strategic place in the orofacial region, at such a crucial time (before birth) that it becomes a complex congenital deformity.[3]

Successful management cleft lip and palate requires a multidisciplinary approach, complex long-term treatment plan and a rehabilitation program designed specifically for individual case. The goals and objectives for the treatment are aimed at:

1. Normalized/Improved facial aesthetics
2. Increased survival rates
3. Improved speech, mastication and other associated functions
4. Normal psychosocial development of the child

EMBRYOLOGY

Embryology of Cleft Lip:

During the first 2 weeks of embryonic life, the human embryo resembles a flat circular plate. In the third week, as the cranial region expands and the neural tube elongates, its shape becomes pear-like. Specialized neural crest cells derived from the neuroectoderm appear as paired columns on the dorsolateral aspect of the neural tube. Despite their ectodermal origin, these neural crest cells make a major contribution to the mesenchyme of the head and neck (ectomesenchyme). The lengthening nervous system results in a flexing of the embryo, bringing the cranial and caudal ends into close proximity. Rapid neural crest cell growth also results in lateral folding.

During the third week of gestation the neural crest cells proliferate and migrate into the fronto-nasal and visceral arch region to form five facial structures or primordia (Early in week 4, the five facial primordia develop around the stomodeum (primitive mouth): the fronto-nasal prominence formed by mesenchyme ventral to the forebrain and paired maxillary and mandibular prominences derived from the first branchial arch mesenchyme. The fronto-nasal prominence will form the forehead, nose, and the top of the primitive mouth. The maxillary prominences will form the lateral sides of the stomodeum, and the mandibular prominences will constitute the caudal boundaries.

By the end of week 4, the embryo resembles a horseshoe- shaped cylinder.

Toward the end of the fourth week, two oval thickenings, the nasal placodes, develop from the ectoderm around the primitive mouth on the lower aspect of the frontonasal prominence. Proliferation of the mesenchymal tissue at the periphery of these ectodermal thickenings produces the medial and lateral nasal prominences. The placodes deepen and sink to form nasal pits which are the precursors of the nose and its structures. The medial nasal prominences and the area above the primitive mouth continue to grow and eventually merge with each other to form the middle part of the upper lip, known as the philtrum.[5, 16-19]

Rapid growth continues during the fifth and sixth week. By the end of the sixth and the beginning of the seventh week, rapid proliferation of the maxillary prominences results in the medial nasal prominences merging with each other and the lateral nasal prominences to form the lateral nose and the cheek regions. During the eighth

week, the maxillary processes on each side of the mouth grow for- ward and fuse with the lower edges of the lateral nasal prominences. They extend below the nasal pits to reach and merge with the upper lip's groove, producing a continuous ridge above the mouth that forms the upper lip. Mesodermal tissue migrates from the first branchial arch and rein- forces the fused tissues in the developing lip. Normally, this mesodermal tissue assumes a medial position, and the two masses formed by the maxillary prominence will assume lateral positions. If this process is delayed, or if one mass is absent, the branchial membrane will pull apart and a CL will ensue. If the maxillary prominence on the affected side fails to merge with the merged nasal prominence, a unilateral cleft will result. If tis- sues fail to merge on both sides, two grooves are formed, resulting in a bilateral CL.[5,16-19]

Embryology of Cleft Palate:

The palate begins to form during the fifth week and is not completed until the twelfth week of gestation. The most critical stage is between weeks 6 and 9. During this stage, the maxillary prominences merge with the medial nasal prominences beneath the nasal pits, forming a wedge-shaped mass of mesenchymal tissue. As this mass of tissue grows, it separates the future nostrils from the upper lip and becomes the median palatine process or primary palate. The primary palate is located immediately behind the gum and extends to the incisive foramen.[5,16-19]

The secondary palate develops from the paired lateral palatine processes. These shelf-like mesodermal projections arise from the medial aspect of the maxillary prominences and are initially oriented vertically on either side of the developing tongue. Development of the lower jaw results in a relatively smaller tongue, which moves inferiorly, allowing the palatine shelves to grow towards each other and rotate to a horizontal position during the seventh week of gestation. Once the shelves are elevated to the correct position, there is apoptotic programmed cell death of the medial edges, thinning the epithelium and allowing the tissue from each side to join on the midline in an anterior-to-posterior sequence. During the ninth week, the palatal shelves begin to merge with the free edges of the nasal septum posteriorly. By twelve weeks, fusion is complete and extends from the maxillary and palatine bones to the palatal shelves, forming the hard palate. The most posterior part that does not ossify becomes the soft palate and the uvula. A CP occurs when this fusion fails.[5,16-19]

INCIDENCE

Overall incidence of orofacial clefting is around 1.5 per 1000 live birth (about 220,000 new cases per year) with wide variation across geographic areas, ethnic group and nature of cleft itself.[6]

The incidence appears high among Asians (0.82 – 4.04 per live births) intermediate in Caucasian (0.9 – 2.69 per 1000 live birth) and low in Africans (0.18 – 1.67 per 1000 live births). Chinese showed 1.76 per 1000 live birth, while Japanese reported 0.85 to 2.68 per 1000 live birth of orofacial clefting.[7]

Isolated CL comprises about 25% of all clefts, while combined CL/P accounts for about 45%. CL/P occurs more frequent and more severe in boys than in girls. Unilateral clefts are more common than bilateral clefts with a ratio of 4:1, and for unilateral clefts, about 70% occur on the left side of the face. Cleft palate is seen more frequently in females than in males. CL/P is frequently associated with other developmental abnormalities and majority of cases are presented as part of a syndrome. Syndromic clefts account for about 50% of the total cases in some reports with about 300 syndromes described. Although the percentage of cases directly linked to genetic factors is estimated to be about 40%, all clefts appear to show a familial tendency.[6]

Various epidemiological studies show that, if one parent affected with a cleft has a 3.2% chance of having a child with cleft lip and palate and a 6.8% chance of having a child with isolated cleft palate (Grosen *et al*., 2010). Presence of a cleft in one parent and in one sibling is associated with a 15.8% chance that the next child will have a cleft lip or palate, and a 14.9% chance that the next child will have a cleft palate (Christensen *et al*., 1996). In case where parents with one is child affected with a cleft have a 4.4% chance of having another child with a cleft lip and palate and a 2.5% chance of having a child with isolated cleft palate.[8]

CLASSIFICATIONS OF CLEFT LIP AND PALATE

Cleft lip and palate may be classified on the basis of:

1. **Location:** Lip, alveolus, palate
2. **Completeness:** Complete/incomplete or microform
3. **Extent:** Unilateral/bilateral.

A. **Davis and Ritchie classification (1922)**, had anatomical basis:
   - GROUP I: Pre-alveolar clefts (unilateral, bilateral and median)
   - GROUP II: Post-alveolar clefts
   - GROUP III: Complete alveolar clefts (unilateral, bilateral and median)

B. **Veau (1931):**
   - GROUP I: Cleft of the soft palate only
   - GROUP II: Cleft of hard and soft palate
   - GROUP III: Complete unilateral cleft, extending from uvula to incisive foramen and then deviates to one side extending through the alveolus
   - GROUP IV: Complete bilateral alveolar cleft[9]

ETIOLOGY

While the exact etiology of cleft ip and palate is unknown, it is believed to be associated with multifactorial reasons. Chiefly, involving both hereditary and environmental factors.

Hereditary/Genetics-

Transmission is believed to be caused through a male, sex-linked recessive gene. With a family history of CLP, preponderance of occurrence is about 40%. Whereas, it is only 18-20% with cleft palate alone.[9]

Genetic causes include:

[1] Syndromic: Here cleft is associated with other malformation. Usually, it is due to a single gene (monogenic or Mendelian) disorder.[10] Association with various syndrome is unknown, namely-

- Pierre Robin syndrome
- Apert's syndrome
- Down syndrome
- Marfan syndrome
- Crouzon syndrome
- Nager syndrome

(2) **Non-syndromic:** Here the cleft is mostly an isolated feature and occurs in the vast majority of individuals having a cleft lip or palate (up to 70% cases). In this form, a cleft is neither a recognised pattern of malformation nor a known cause for the disorder can be identified.[10]

Environmental factors-

Besides genetic factors, environmental influences are also known to have a role in the etiology of CLP. These factors are-

a) Smoking: The relationship between maternal smoking and CLP is not strong, but it is significant. Several studies have consistently yielded a relative risk of about 1.3-1.5. When maternal smoking was considered together with a positive genetic background, the combined effect was more significant. Furthermore, Beaty *et al.* (2002) reported that maternal smoking and infant *MSX1* genotypes acted together to increase the risk for CLP by 7.16 times.[11]

b) Alcohol use: Heavy maternal drinking, apart from causing fetal alcohol syndrome, also increases the risk of CLP. Munger *et al*. (1996) showed that maternal drinking increased the risk for CLP by 1.5–4.7 times in a dose-dependent manner Low-level alcohol consumption, however, did not seem to increase the risk of orofacial clefts. The link between alcohol consumption and genotypes on the risk of CLP has yet to be demonstrated.[11]

c) Other factors: Various other factors may also contribute in the etiology of CLP, these are-

- Psychogenic, emotional or traumatic stress in pregnant mothers
- Relative ischemia to the area due to defective vascular supply
- High dose of steroid therapy during pregnancy
- Nutritional factors, such as deficiency of riboflavin and deficiency of or excess of Vitamin A
- Intrauterine exposure/influence of drugs- Excessive use of antibiotics, insulin and anti-epileptic drugs.
- Maternal age- Older the mother, greater the chance of incidence of congenital anomalies.9

CLINICAL FEATURES

The various clinical findings in patient with cleft lip and palate can be categorised under the following-

General features:

- These defects occur more commonly in the male population.
- Clefting involves left side of the face more than the right side.
- A complete cleft lip may extend upward and involve the nostril, thus resulting in deflection of nasal tip towards the nostril side and larger nares on cleft side.
- Cleft palate may involve both hard and soft palates or the soft palate alone.

Dental problems in cleft lip and palate:

Various abnormal dental conditions include:

1. Natal and neonatal teeth:

Presence of neonatal teeth does not appear to influence primary or secondary dentition in clefts. Most natal teeth among clefts are located in the lateral margin of the premaxillary and maxillary segments unlike in non-cleft neonates.[12,13]

2. Microdontia

Small teeth (microdonts) frequently are found with CL/P. This is usually more common in cases where lateral incisors are not missing (van der Wal, 1993; Stahl *et al*., 2006; Rawashdeh and Bakir, 2007). Generally, peg shaped upper lateral incisors are seen.[12]

3. Taurodontism

Taurodontism has been reported to be associated with certain syndromes and dental developmental disorders (Cichon and Pack, 1985).[13]

4. Ectopic eruption

Clefts also contribute to the ectopic eruption of primary lateral incisors which may erupt palatally adjacent to or within the cleft side while permanent canine on side of alveolar clefts may erupt palatally. Delayed eruption of permanent incisors may be seen.[13,14]

5. Enamel hypoplasia

Enamel hypoplasia was found to occur more frequently in CL/P subjects compared with non-cleft populations, especially involving the maxillary central incisors (Vichi and Franchi, 1995).[13]

6. Delayed tooth maturation

Several growth factors are of major importance during craniofacial development, and these factors may be over-expressed or under-expressed when a cleft defect occurs. This aberrant expression can modify odontogenesis and cause abnormalities of the dental lamina.[15]

Other associated conditions

1. **Speech difficulties:** Due to the dysfunction of m. levator veli palatini muscle phonation are affected. Retardation of consonant sound (p, b, t, d, k, g) is most common findings. Abnormal nasal resonance and difficulty in articulation are

another characteristic feature in most individuals with cleft lip and palate.

2. **Ear infection**: Due to improper function of m. tensor veli palatini muscle, which opens the Eustachian tube, otitis media is observed in these patients. In a case where infections frequently occur, results that can lead to hearing loss may occur. The incidence, however, increases sharply when there is associated submucous cleft palate.

3. **Feeding problems:** A child with a cleft palate can have difficulty sucking through a regular nipple due to the gap in the roof of the mouth. An infant's ability to suck is related to two factors: the ability of the external lips to perform the necessary sucking movements and the ability of the palate to allow the necessary build-up of pressure inside the mouth so that foodstuff can be propelled into the mouth. Most babies require a personalized or special nipple to properly feed. It may take a couple of days for the baby and parents to adjust to using the nipple before going home. Most babies learn to feed normally with a cleft palate nipple.[16]

MANAGEMENT OF CLEFT LIP AND PALATE

This correction involves surgically producing a face that does not attract attention, a vocal apparatus that permits intelligible speech and a dentition that allows optimal function and aesthetics. The cleft palate team concept has evolved from that need. Because optimal care is best achieved by multiple types of clinical expertise, the team may be composed of individual in:

(1) the dental specialties (orthodontics, oral surgery, pediatric dentistry, and prosthodontics)

(2) the medical specialties (genetics, otolaryngology, pediatrics, plastic surgery, and psychiatry), and

(3) allied health care fields (audiology, nursing, psychology, social work, and speech pathology).[20]

Surgical treatment

Unlike the artistic nature of the cleft lip repair, the cleft palate repair is very functional in nature. A team approach has decreased the morbidity and secondary deformities caused by the cleft and mostly focuses quality of speech.[21] Soft palate repair techniques may be used in isolation or combined with hard palate procedures, as necessary. Most surgeons today perform either some modification of an intravelar veloplasty, vs. a two flap palatoplasty with double opposing z-plasty to achieve levator muscular repositioning.[22] Maxillary distraction is increasingly used for the correction of severe maxillary retrusion in patients with cleft lip and palate.[23] Cleft lip and palate children benefit from team approach special treatment requirements. such a team lead by the plastic surgeon should include a speech therapist and orthodontist having ready access to pediatric, ENT and dental treatment facilities.[24] Esenlik *et al*. reviewed the literature on nasoalveolar molding (NAM) with an eye to both benefits and limitations. A review of the literature suggests that NAM Cleft lip and palate children benefit from team approach special treatment requirements. such a team lead by the plastic surgeon should include a speech therapist and orthodontist having ready access to pediatric, ENT and dental treatment facilities does not alter skeletal facial growth when compared with the samples that did not receive PSIO (Presurgical infant orthopedics). Nevertheless, the published studies on NAM show evidence of benefits to the patient, caregivers, the surgeon, and society. These benefits include documented reduction in severity of the cleft deformity prior to surgery and as a consequence improved surgical outcomes, reduced burden of care on the caregivers, reduction in the need for revision surgery and consequent reduced overall cost of care to the patient and society.[25] Robotic cleft surgery is a new and exciting field that holds numerous advantages to both patients and surgeons. Previous research in allied health

specialities has paved the way to the feasibility studies of robotic cleft surgery. Finally, the use of surgical robots at present introduces economic challenges to implementation because of increased operative time and high capital and operating costs and it is hoped that over time, costs will reduce and performance will increase as more systems are developed in the future.[26]

CONCLUSION

Cleft lip and palate are both birth defects that affect different structure and function such as speech difficulty, aesthetic, eating, nutrition etc. The primary aim in cleft lip and palate is to educate the parents as it facilitates the treatment to take place at the right age. This further helps to achieve functional and aesthetic wellbeing. While surgical treatment holds importance, it is also essential to consider the mental status of the patient so that adequate psychological rehabilitation may be considered. Extensive dental treatment is generally required in CLP patients to achieve reasonable oral health. The multidisciplinary approach towards this problem led to a steady improvement in its end results.

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