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Cleft Lip and Palate

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Cleft Lip and Palate

Introduction

- ☑ Cleft lip and palate (CLP) are the most common congenital anomalies of orofacial region. "A cleft is a congenital defect in the upper lip, alveolus, or in the palate.
- ☑ CLP can occur either isolated or together in various combinations or along with other congenital defects particularly congenital heart diseases". CLP are as associated features in >300 identified syndromes.

Cleft Lip

- ☑ "The failure of fusion of the frontonasal 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 no connection between the alar base and the medial labial element.)".
- ☑ Cleft lip can occur as one sided, i.e., unilateral or both sided as bilateral condition. Microform cleft, a mild form of cleft lip sometimes can appear as a small dent in red part of upper lip.

"The failure of fusion of the palatal shelves of the maxillary processes, resulting in a cleft of the hard and/or soft palates".

Cleft Palate

Definitions

Overall incidence of CLP is 1 in 1,000 live births and isolated cleft palate occurs in 1 in 2,000 live births.

The typical distribution of cleft types:

- ☑ Cleft lip alone: 15%
- ☑ Cleft lip and palate: 45%
- ☑ Isolated cleft palate: 40%

Many epidemiological studies reveal that, if one parent affected with cleft has a 3.2% chance of having a child with CLP and a 6.8% chance of having a child with isolated cleft palate (Grosen et al., 2010).

The etiological factors of CLP are classified as:

- ☑ *Nongenetic (environmental factors):*
 - Maternal smoking
 - Maternal consumption of alcohol
 - *Others:* Maternal disease, maternal hypoxia, stress during pregnancy, chemical exposure, decreased blood supply in nasomaxillary region, folic acid deficiency, increased parental age and exposed to retinoid drugs.
- ☑ *Genetic:*
 - *Syndromic:* CLP is associated in >300 identified syndromes and many among them follows classic Mendelian inheritance pattern (single gene disorder) (**Table 1**).

TABLE 1: Syndromic causes of cleft lip and palate.

Syndromes	Gene name (symbol)	Location on chromosome	Inheritance
Waardenburg syndrome, type II A	Microphthalmia-associated transcription factor (MITF)	3p14,1-12,3	AD
DiGeorge syndrome	DiGeorge syndrome chromosome region (CATCH 22)	22q11	AD
Treacher–Collins mandibulofacial dysostosis	Treacle (TCOF1)	5q32-q33,1	AD
Van der Woude syndrome	Interferon regulatory factor-6 (IRF 6)	1q32-q41	AD
CLP-Ectodermal dysplasia syndrome	Poliovirus receptor-related-1 (PVRL-1)	11q23,3	AD

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Syndromes	Gene name (symbol)	Location on chromosome	Inheritance
Ectrodactyly, ectodermal dysplasia orofacial cleft syndrome	P63	3q27	AD
Zollinger syndrome-3	Peroxisomal membrane protein-3 (PXMP3)	8q21,1	AD
Diastrophic dysplasia	Diastrophic dysplasia sulfate transporter (DTDST)	5q32-q33,1	AD
Gorlin syndrome (Basal cell nevus syndrome)	Patched (PTCH)	9q22,3	AD

- *Nonsyndromic*: It accounts for 70% of CLP cases. Many genes are identified for clefting whose mutation may lead to nonsyndromic CLP (**Table 2**).

TABLE 2: Non-Syndromic causes of cleft lip and palate.

Name of gene	Symbol	Chromosome location
Transforming growth factor-alpha	TGFA	2p13
Transforming growth factor-133	TGF 133	14q24
Methylenetetrahydrofolate reductase	MTHF3	1p36,3
Blood clotting factor XIII gene	ETI	6p24
Endothelin-1 gene	ETI	6p24
Proto-oncogene BCL3	BCL3	19q13,2
Retinoic acid receptor alpha gene	RARA	17(t15/17)
MSX-I	MSX-I	4q25

Cleft lip and palate may lead to problems with feeding, ear disease, speech, cognition, and socialization.

- ☑ Cosmetic deformity
- ☑ *Dental problems* seen as natal teeth, microdontia, taurodontism, ectopic eruption, enamel hypoplasia, and delayed tooth maturation. Delayed in tooth development can be proportionate to severity of CLP. Most problems arise after the eruption of permanent teeth. Other problems include missing teeth, fused teeth, and extra teeth erupting behind normal teeth; however, missing teeth or extra teeth both sometimes are normal occurrences. In CLP, there is an increased rate of dental caries, malocclusion, thereby resulting in an open bite or cross bite. This in turn can then affect the patient's speech too.

Clinical Features

- ☑ *Speech and language difficulties:* Dysfunction of levator veli palatini muscle affects phonation. The most common finding is retardation of consonant sound (p, b, t, d, k, and g). Abnormal nasal resonance and difficulty in articulation are seen.
Language issues develop since lips and palate are both used in pronunciation. They will have problems in expressive language acquisition as well as receptive language acquisition. Tentative evidence has suggested that those with clefts perform less well at language.
- ☑ *Feeding problems:* CLP patients have difficulty in suckling the breast due to gap in the roof of the mouth and lack of suction.
- ☑ Middle ear issues; CLP children often suffer with following issues:
 - Frequent ear infections eventually may lead to hearing loss
 - External ear canals and Eustachian tubes may be tortuous or angled, causing food or other contamination to develop a middle obstruction
 - Speech issues due to compromised hearing
- ☑ *Psychosocial issues:* Cosmetic issues due to visible deformities and language impediments are mainly responsible for following psychosocial issues:
 - Impact on social skills, behavior, and self-esteem
 - Social anxiety
 - Feeling of anger, fear, sadness, and alienation from peer group

- ☑ Fourth to twelfth embryonic week is the critical period for cleft development.
- ☑ Accurate evaluation of craniofacial malformations is usually possible with the ultrasound scan performed during pregnancy enabling cleft team to timely discuss the long-term treatment planning and educate the parents about diagnosis.
- ☑ This helps improve the quality of treatment received by the child and improves quality of life.

Prenatal Diagnosis

- ☑ The patients with orofacial cleft deformity should be treated at right time and at the right age to achieve functional and aesthetic well-being. Minimum age for cleft lip repair is after the third month and cleft palate after the ninth month. Minimum requirement to get fitness for surgery: hemoglobin 10 g/dL and weight should be 10 lbs (5 kg).
- ☑ The correction of the cleft lip and face involves surgically producing a face, making intelligible speech, optimal functioning of dentition, and aesthetic look. This is best achieved by involvement of multidisciplinary clinical experts at various stages of the child's development.
- ☑ The medical specialties (pediatric surgery, plastic surgery, pediatrics, genetics, otolaryngology, and psychiatry)
- ☑ The dental specialties (orthodontics, oral surgery, pediatric dentistry, and prosthodontics)
- ☑ Allied healthcare fields (audiology, nursing, psychology, social work, and speech pathology)

Surgical Treatment

Unlike the artistic closure of the cleft lip repair, cleft palate repair is very functional in nature. A team approach will decrease the morbidity and secondary deformities and mainly focus on quality of speech.

Soft palate repair techniques may be used in isolation or combined with hard palate procedures. Most of the surgeons perform either intravelar veloplasty or two flap palatoplasty with double opposing Z-plasty to achieve levator muscular repositioning.

Maxillary distraction is used for correction of severe maxillary retrusion in patients with CLP.

There are six procedures in repair of CLP:

1. Cleft lip repair—third month
2. Cleft palate repair—ninth month
3. Palatal shelf expansion—5–7 years
4. Alveolar bone grafting (ABG)—ninth year
5. Cleft lip rhinoplasty—after 13 years
6. Scar revision over lip—14–16 years

- ☑ Feeding is preferred in more upright position as gravity helps preventing milk from coming from baby's nose. Specialized instruments such as Haberman Feeder and customized bottle having a combination of nipples and bottle inserts help in achieving gravity feeding. A large hole or a slit in the nipple, a protruding nipple and rhythmical bottle squeezing can control the milk flow.
- ☑ The mental status of patients with CLP should be considered and supported by psychological rehabilitation and their morale should always be bolstered.

Management of Feeding Issues

Outcome Response

- ☑ It is very difficult to measure the outcome response of CLP child due to longitudinal nature of cleft case and complexity associated with its management.
- ☑ However, international consortium for health outcome measurement assesses standard set of measures outcomes in important domains such as hearing, breathing, eating/drinking, speech, oral health, appearance, and psychosocial well-being.

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Further Reading