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Cleft Lip and Cleft Palate

Abstract

Cleft lip (cheiloschisis) and cleft palate (palatoschisis) can occur individual or together. Individuals with CLP may experience problems with feeding, speaking, hearing and social integration that can be corrected to varying degrees by surgery, dental treatment, speech therapy and psychological intervention. In India over 35,000 babies per year are affected. This review signifies the etiological factors, signs and symptoms, classification and its comprehensive treatment.

Key Words

Cleft lip; cleft palate; chelioschisis; palatoschisis

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INTRODUCTION

Cleft lip (cheiloschisis) and cleft palate (palatoschisis) can occur individual or together. This is a congenital deformity caused by abnormal facial development during gestation i.e. 3rd and 9th week. Literally, cleft means a fissure or opening. Individuals with cleft lip & palate (CLP) may experience problems with feeding, speaking, hearing and social integration that can be corrected to varying degrees by surgery, dental treatment, speech therapy and psychological intervention.[1]

INCIDENCE

Approximately 1 in 100 child births are affected. So, for India, it is over 35,000 babies per year. Occurrence of orofacial clefts including nose, ears, eye, cheeks and forehead (called craniofacial clefts) is extremely rare. Predominance of cleft is seen on the left side. There is predilection for male child.

ETIOLOGICAL FACTORS

These can be hereditary or environmental.

Hereditary

Transmission is said to be caused through a male, sex linked recessive gene. With a family history of cleft lip and palate, preponderance of occurrence is about 40% where as it is 18-20% with cleft palate alone.

Environmental

Maternal hypoxia e.g. maternal smoking, maternal hypertension, pesticide exposure, malnutrition specially retinoids, members of vitamin A family, anticonvulsions, exposure to lead and illegal drugs such as heroin, cocaine etc, use of steroids, anorexia, anaemia, viral infections, excessive consumption of alchohol, consanguineous marriages, elder age group mothers.[1,2] Cleft might be linked to genetics as in mutations as in the gene PHF8 is said to cause cleft lip / palate. Current research continues to investigate the extent to which Folic acid can reduce the incidence of clefting.[3]

SIGNS AND SYMPTOMS

Cleft affecting the lip only are classified as microform or partial cleft and complete cleft (Fig. 1 & Fig. 2) which extends into the nose. These can be unilateral or bilateral. It occurs due to the failure of fusion of the maxillary and median nasal processes which are responsible for formation of the primary palate.[2]

CLEFT PALATE

It is a condition in which the two palates of the skull which form the hard palate are unable to fuse. The critical problem is failure of Lateral nasal processes to make contact with median nasal process. Palatal cleft may be complete (soft and hard palate) or incomplete (Fig. 3 & Fig. 4).[3] Uvula may or may not be involved. It occurs due failure of fusion of the lateral palatine processes the nasal septum and/or the median palatine processes. In cleft palate, the oral cavity directly overlooks the nasal cavity. Because of this, velopharyngeal incompetence happens. There is leakage of air into the nasal cavity resulting in a hypernasal voice resonance and nasal emissions.[4]

ASSOCIATED PROBLEMS

The upper lip cleft entails loss of the important orbicularis oris muscle complex premaxilla and prolabium are deviated away from the cleft. In

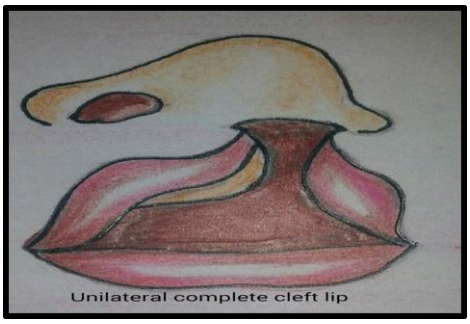
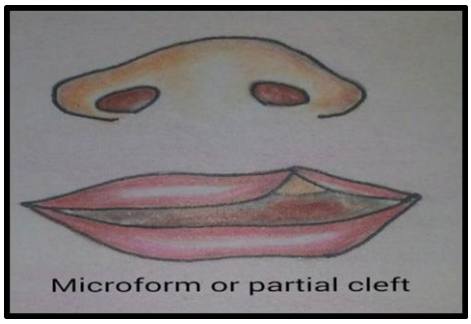


Fig. 1



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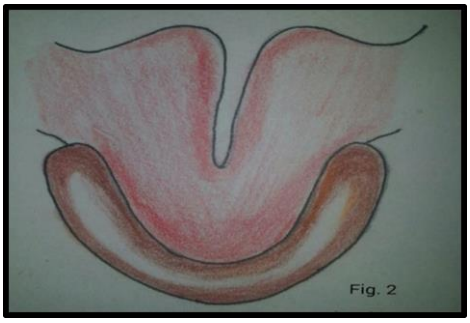

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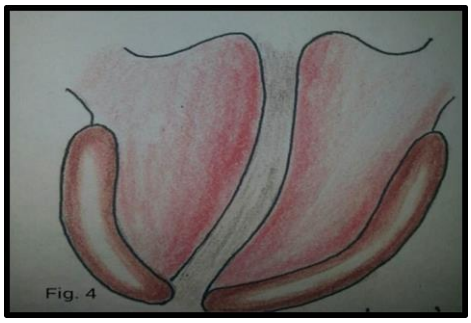

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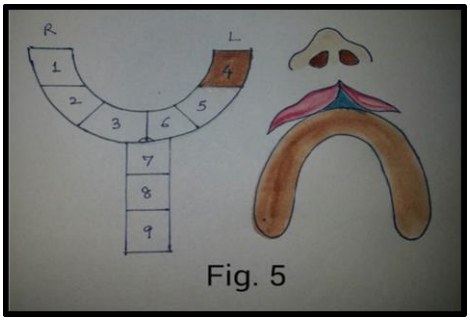

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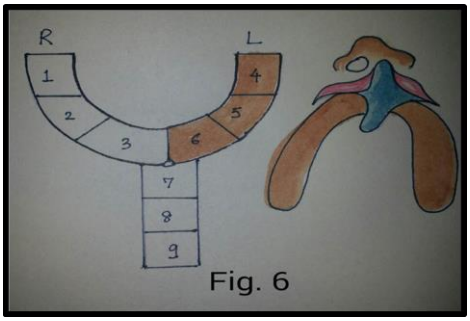

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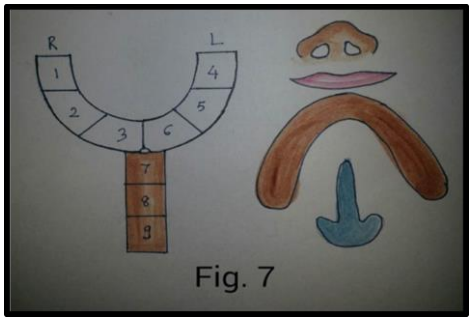

Fig. 7

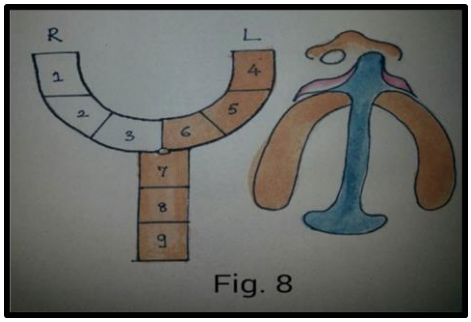

Fig. 8

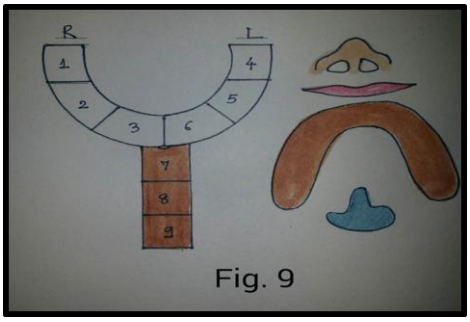

Fig. 9

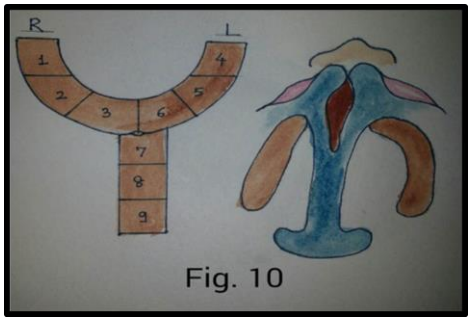

Fig. 10

bilateral cases, premaxilla and prolabium project anteriorly and columella is extremely short. Feeding is a major problem since structural defects of cleft lips and palate prevent negative oral pressure

required for sucking.[5] Other effects include speech articulation errors, like distortions, substitutions and omissions, compensatory misarticulations and mispronunciations (e.g. glottal soaps) and posterior nasal fricatives.[5] Since equalisations of pressure by swallowing is hampered, there is greater susceptibility to middle ear infections. There could be hearing loss.[1,5] Change in dynamics of growth potential affects the growth of maxilla resulting in functional and aesthetic impairment and functions.

PSYCHOSOCIAL ISSUES

A cleft lip and /or palate may impact an individuals self esteem, social skills and behaviour. A larger amount of research is dedicated to the psychological development of individuals with this deformity. Strong parent support networks may help to prevent the development of negative self-concept in these children. They often deal with threats to their quality of life for multiple reasons including unsuccessful social and interpersonal relationships, deviance in social appearance and multiple surgeries.[4]

CLASSIFICATION

Davis and Ritchie in 1922[6] and Veau in 1931 gave simple classifications. But, the internationally accepted classification advocated by Fogh-Anderson in 1942 and confirmed by Kernahan and Stark (1958)[7] is as follows:

A) Group I-Cleft of the anterior (primary) palate

1. Lip: Unilateral Rt/Lt - Total or partial: Bilateral
2. Alveolus: Unilateral Rt/Lt - Total or partial: Bilateral

B) Group II - Cleft of anterior and posterior palate

1. Lip: Unilateral Rt/Lt – Total or partial: Bilateral
2. Alveolus: Unilateral Rt/Lt - Total or partial: Bilateral
3. Hard palate: Unilateral Rt/Lt - Total or partial: Bilateral

C) Group III - Clefts of posterior palate

1. Hard palate: Rt/Lt
2. Soft palate

(Fig. 5-10)

D) Group IV - Rare facial clefts

MANAGEMENT OF CLEFT DEFECTS[8]

Aims and Objectives

1. To correct the defect surgically and make it aesthetically acceptable.
2. To improve speech articulations.
3. To correct malocclusion.
4. To increase the self-esteem of the affected child.

MULTIDISCIPLINARY APPROACH[9]

Management of cleft lip remains an enigma and a challenge. Spatial relationship and function of muscular elements causing this deformity has to be understood for a functional correction of cleft lip and palate. A team of trained doctors including plastic surgeons, Oral and Maxillofacial surgeons, ENT surgeons, speech therapists, child psychiatrists, well-trained nursing staff, Orthodontist, Prosthodontists, Paediatrician are too involved for rehabilitation of a cleft case.

GENERAL GUIDELINES[1,9,12]

1. Immediately after birth: Paediatric consultation, geneticist evaluation to diagnose if any associated syndrome is present. Parent counselling and feeding instructions to be given.
2. After a few weeks: Hearing evaluation and weight evaluations.
3. Depending upon the weight of the baby, according to the rule of 10, i.e. weight minimum of 10lbs, haemoglobin 10mg and 10 weeks, lip repair can be undertaken.
4. Between 1 year and 18 months: Surgical repair of palate after surgical evaluation.
5. Post-palatal repair: Speech and language assessment and speech therapy to be ensued.
6. Psychological evaluation, treatment of middle ear infections.
7. Alveolar bone grafting depending upon the defecting size.
8. Orthodontic treatment after age of 11years.
9. Upto 18 years: End of orthodontic corrections and miscellaneous dental treatments.
10. Evaluation of maxillary growth. If required surgical advancement of maxilla post puberty.
11. Rhinoplasty if required.
12. Psychological support throughout these phases and later.

PRESURGICAL DEVICES

Nasoalveolar moulding prior to surgery can improve long term nasal symmetry with complete unilateral cleft lip - cleft palate patients compared to correction surgically alone; according to a retrospective cohort study.[10,11]

SURGICAL REPAIR

A ideally repaired cleft lip should provide a symmetrical cupid’s bow and philtrum with a minimal scar.[12] The most popular method for unilateral repair is rotation advancement technique introduced by Millard in 1958.[8] Millard repair aimed at lengthening of the skin of the lip reconstructing orbicularis oris muscle and

Table 1: Depending upon the severity of the Cleft, Distraction Osteogenesis, Orthognathic Surgery and Secondary Corrections can be done

age	Treatment	Rationale
At birth	Feeding plate	Sucking milk
2-3 months	Lip repair	Feeding and aesthetics
9-11 months	Palate repair	Sealing the oronasal communication, Enhance speech and feeding
3 years	Pharyngoplasty if needed	Correction of nasal twang
3 years	Premaxillary setback in case of bilateral cases	Creation of labial vestibule,better speech and aesthetics.
3 years	Cleft alveolar bone closure using BMP	Closing alveolar cleft
7years	Cleft alveolar bone closure with bone grafting	Closing alveolar cleft
13 years	Cleft orthodontics	Aligning the teeth correctly
14-16 years	Rhinoplasty if needed	Aesthetics

reconstructing the full height of the labial sulcus. Simultaneous reconstruction of the nasal deformity should be considered in the form of columellar lenghthing and repositioning of the misplaced alar cartilage.[8] To date, several surgeons have modified this basic technique according to their convenience and variations of the cleft. Describing all the techniques is not possible under purview of this article.

Repair of Palate[11]

Primarily it aims at the repositioning of the palatine muscles and surgical repair of palate functionally. Veau (1931) modified the Langenback's operation of pushback. Wardill (1933-37) evolved four flap method of repair. Skoog used a mucoperiosteal flap to cover the raw surface which helps growth of tooth in this segment. Adenwalla and Narayanan (2009) modified the repair of uvula for better velopharyngeal competence. Alveolar bone graft is ideally performed prior to eruption of lateral incisors or canines, to be able to prevent loss of a tooth due to eruption into an ungrafted cleft alveolus. This age is considered to be around 5 to 6 years of age. Bone grafting allows these teeth to erupt in solid bone and provides enhanced stabilization of the maxilla.[13] Transverse maxillary growth is also nearly complete by this age, so that there is no inhibition of maxillary growth in this dimension. Though several different techniques described delayed alveolar bone, an elegant straightforward approach was described by Craven *et al*. The defect is viewed three dimensionally. Both cortical and cancellous bone is utilized. SM Balaji has demonstrated use of rh Bone morphogenetic protein-2 as an ideal bone reconstructive material for alveolar clefts.[14] Comprehensive treatment protocol as laid down by SM Balaji is on Table 1.

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