Effects of Cleft and It's Clinical Implications

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Abstract

The most common malformation of the head and neck region affecting individuals and their surroundings physically, mentally, psychologically and socially is cleft lip and palate. There are of various types with their distinct clinical features and characteristics. Prenatal diagnosis, effective understanding can help formulate a good treatment approach with specialists from various fields working as a team. Parents counseling and follow up forms the mainstay approach of these children.

Keywords: *Lip, Cleft, Harelip, Unilateral, Bilateral.*

Introduction

The most common congenital malformation of the craniofacial region is cleft lip and palate1,2. Clefting in birds is physiologic as it helps them to fly at greater altitudes and equalize pressure in the middle ear.

Epidemiology: In India, the incidence of cleft is 1 IN 800. In white Americans, the incidence is 1 IN 800 too. In comparison, black Americans show an incidence of 1 IN 2000. Isolated cleft palate presents an incidence of 1IN 2500. Sex differences (Male to Female ratio) presents cleft lip as 2:1, CLEFT PALATE as 2:3, cleft lip & palate as 2:1. In Racial differences, Japanese being greater in comparison to Caucasians and Afro-Caribbean. The incidence of occurrence is more on left side as compared to right side. Monozygotic twins show a 40% concordance in comparison to di-zygotic twins (5% concordance).

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Concept of Cleft:

Neutral Zone Concept: The cheek, lip and pharyngeal musclesbalance against the developing upper and lower arches. The force they exert neutralize the tongue forces maintaining dynamic equilibrium[3]. With the change in equilibrium, the form and relationship of the arch with the teeth changes (Figure 1).

Aberrant muscle forces in clefts: The first nine weeks of fetal life is of significance for the development of cleft lip and palate. The re is failure of fusion of the lip and the palatal segments: both right and left. There is break in continuity of the buccinators mechanism. This results in aberrant muscular forces that displace tissue masses (Figure.2).

In unilateral complete clefts of the lip and palate cases, the premaxillary portion of the noncleft segment is pulled antero-laterally. Premaxilla is carried forward in the facial skeleton in the larger segment[4,5]. There is excessive growth in the premaxillary-vomerine suture in bilateral complete cleft lip and palate (CBCLP) patients.

Palatal arch form: The facial versus tongue muscle forces dictate the shape of the palate. Palatal growth rates correlate with the amount of scarring during closure of the cleft space.

Classification: The various types of cleft are explained underneath (Figure. 3):

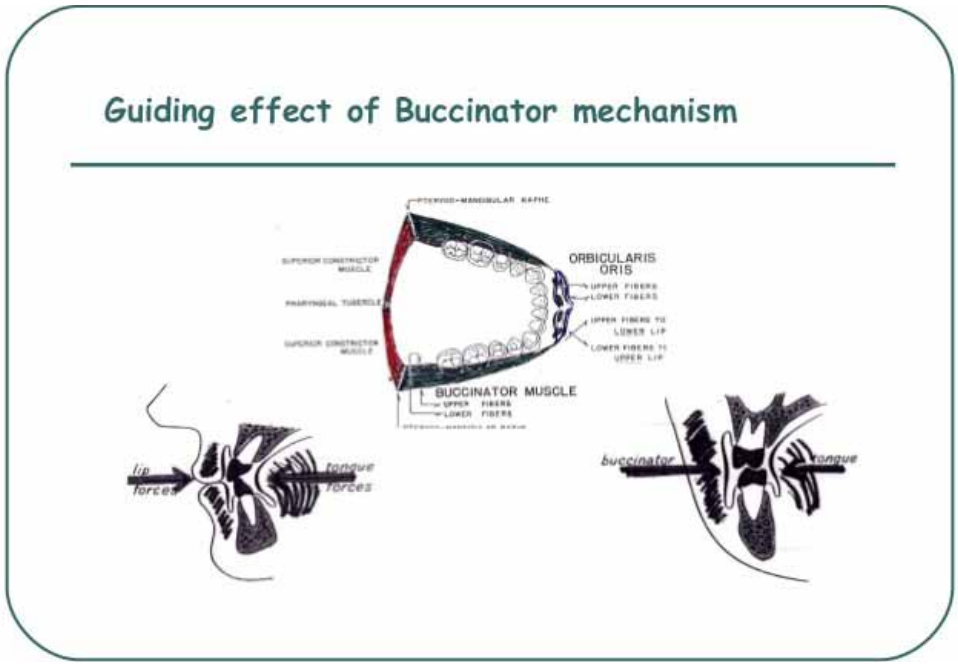

Figure 1. Neutral Zone Concept

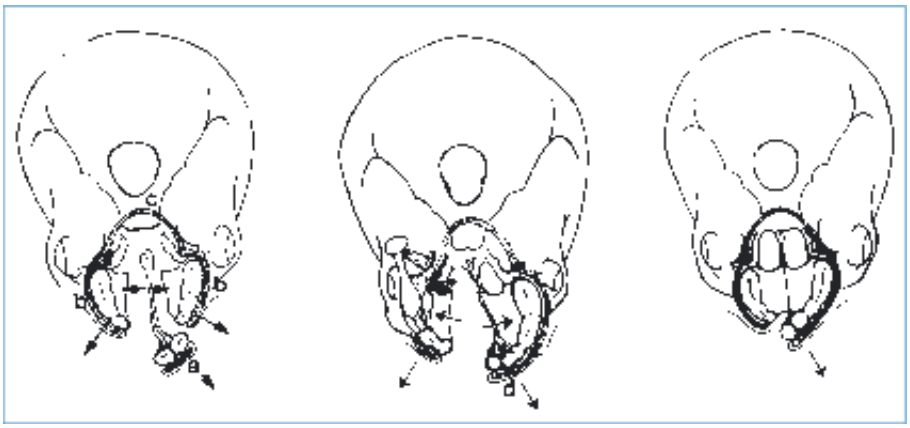

Figure 2. Aberrant Muscle Forces

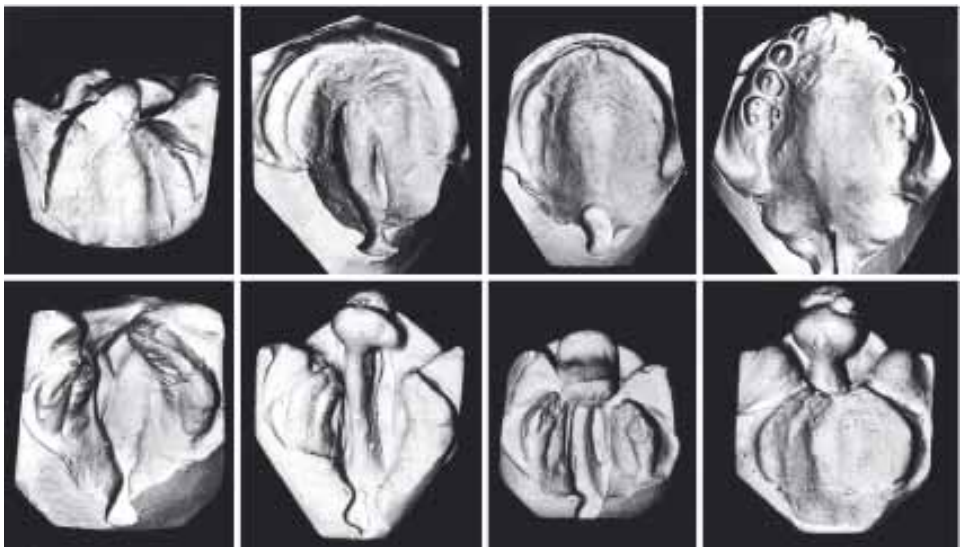

Figure 3. Categories of cleft

1. Clefts affecting lip and palate: simultaneous or separate.

   Cleft affecting the primary and/or secondary palate: complete/incomplete, unilateral/bilateral.

2. Submucous clefts affecting the secondary palate

Clefts of the Lip: Clefts may be unilateral or bilateral (symmetrical or asymmetrical).[6,7] Unilateral cleft lip presents asymmetric and less premaxillary protrusion (Figure 4). Bilateral cleft lip shows marked protrusion of premaxilla.[8]

Figure 4. Unilateral Cleft Lip

Cleft of lip and alveolus: Cleft involves the lip as well as the alveolus.[9,10]

Complete unilateral cleft of the lip and palate: Themedial portion of orbicularis muscle is attached to columella & the lateral portion to the nasal alar cartilage.[11] There is thinness in the medial vermillion border. There is distortion in the perpendicular plates of the sphenoid with flattening of the ipsilateral lower cartilage.[12] Cephalometric findings indicate Increased width of maxilla, increased nasal width withless significant changes in the pharyngeal airway and mandibular length.

Incomplete cleft lip: It presents with a defect in the bulk of orbicularis oris muscle.

Complete bilateral cleft lip: Medial aspect shows complete absence of orbicularis oris muscle.Premaxilla is protruded to varying degree. The various nasal defects include short columella, bilaterally flattened dome, and bilaterally horizontal nostrils(Figure.5). There is protruding premaxilla at birth, marked palatal hypoplasia, retruded maxillary shelves and buccal teeth in class I/class II occlusion. Cephalometric findings indicate increase in width of maxilla, increase in width of nasal cavity, increase in mandibular length and retrognathia.

Figure 5. Bilateral Cleft Lip

Isolated Cleft Palate: It involves hard palate, soft palate and structures of the secondary palate. There is variation of the geometric form. Cephalometric findings show decrease in total length of maxilla, decrease in mandibular length, bimaxillaryretruded jaws with reduction in size of pharyngeal airway.

Submucous Cleft Palate: It shows the presence of bifid uvula (Figure6),zonapellucida.

Figure 6. Bifid Uvula

Syndromes associated with cleft lip & palate: Vonderwoudesyndrome, Downs syndrome, Oral-facial digital syndrome, Treachercollin syndrome, Pierre robin syndrome, Apert syndrome, Trisomy of chromosome 18, Turner's syndrome

Treatment: The treatment is taken care of by a team comprising of Surgeons, Speech and language pathologists, Geneticists, Dysmorphologists and Paramedical specialists.

A. Unilateral cleft of the lip and palate

The treatment protocolfollows age of the patient. At 3 months age, lip adhesion is carried out[13]. At 10 months, lip surgery is carried out. At 18-24 months, palate closure (hard and soft) is done followed by Orthodontic expansion at 5-7 years. Bone grafting is done at 7-9 years. 8 years or later Protraction facial mask is given followed by Maxillary surgical advancement and Lip/ nose revisions technique.[14]

B. Bilateral cleft lip and palate

In such cases, after birth: CBCLP – external elastics is recommended. At 3–4 weeks, lip adhesion surgery is advised. Millard forked flap is done at 6 months of age. At 18-30 months, Von langenbeck repair is done. Buccalcrossbiteis corrected with the help of a quad helix at age of 4-5 years[15].

Fixed palatal retention is attempted at 5-7 years. At 7-8 years, anterior teeth alignment is done followed by bone grafting (secondary).At the age of 9-13 years, full banded treatment is advised. It may be accompaniedby maxillary protraction therapy along with nasaltiprevisionsurgery.

Full Orthodontics is done along with and nasal lip revision at 13-17 years of age. Postsurgical Orthodontics followed by prosthetics andnasal-lip revisions are done at 17-18 years of age.

Treatment sequence of clefts of the uvulae alone and soft palate

Soft tissue clefts are sutured around 3 months. Orthopedic protraction is carried out duringmixed and permanent dentition.

Clefting process and contiguous structures:

A. Skeletal Findings:

(a) Width: Head width is greater than length with orbits further apart than normal. There is a wider maxilla with no crossbites even in adults.

(b) Height: Maxilla on cleft side is smaller than normal, but overall, there is no decrease in vertical facial growth

(c) Depth: Maxilla is placed normally with relation to cranial base. Pre-maxilla is rotated forward & laterally and the protrusion of pre-maxilla increases as age increases.

B. Dental Findings: There is presence of hypodontia, Supernumerary teeth, Microdontia/macrodontia, hypoplasia, natal/neonatal teeth and crossbite in 19% of cases. The teeth adjacent to cleft shows poor palatal support & are susceptible to premature loss. There is delayed eruption of teeth on cleft side.

Effects:

1. Feeding difficulties at birth: Feeding problems arises due to large opening between mouth & nose in case of cleft palate. In case of cleft lip alone problems will arise in efficient sucking. The re are three major concerns:
   - Risk of aspiration because of communication
   - Airway obstruction (Pierre robin syndrome)
   - Difficulty in feeding a child because of nasal regurgitation
2. Hearing difficulties:Hearing defects may be due to sensorineural anomalies and recurrent otitis media.
3. Speech difficulties: Speech pathology may be due to incorrect lip posture, tongue position, velopharyngeal incompetency, abnormal neuromuscular function, abnormal dentition secondary to hearing loss due to sensorineural deficit.
4. Psychosocial issues There is social and emotional adjustment and features of social isolation, obsessive compulsive behavior, lack of cognitive development and school achievement.

Adverse long-term health effects: There are elevated risks for death along with musculoskeletal disorders, autism spectrum disorders and other learning disabilities.

Implications: There is midface retrusion, distortion of dentoalveolar structures, difference in posture and shape of the mandible, crowding of the dentition, missing teeth, concave profile, crossbites.

There are issues like:

1. **Anteroposterior Issues:** The retrusive maxilla along with a normal mandible superimposed on the transverse problem results in a class III tendency.

2. **Vertical Issues:** The position of the mandible varies in rest position and in occlusion. This may generate excessively long or short lower face heights. The functional shift of the mandible results in pseudo prognathism
3. **Transverse Issues:** Crossbites is seen.

Concerns:

In order to minimize the effects of scarring, the various changes that needs to be done include:

a. Biocompatible membranes to inhibit scar tissue formation

b. Epithelial sheets

c. In vivo surgical procedures

Future Perspectives: Information is the quintessential defining factor of this century. It is important to share information among the countries of the world to extend medical benefits equally among developed and developing countries. Japanese Cleft Palate Foundation (JCPF) & the International Cleft Foundation have been established to cater health care needs.Smile train is World's leading cleft charity program, being established in 2000 to help millions of cleft affected families and bring awareness among doctors and medical professionals.

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