An Insight of the Cleft Lip and Palate in Pediatric Dentistry - A Review

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Abstract
Cleft lip and palate is one of the common congenital developmental abnormalities which have a negative impact on the quality of life of the children. It has multifactorial etiology, primarily caused by the interaction between environmental and genetic factors. Management of cleft lip and palate requires a multidisciplinary team involving oral and maxillofacial surgeon, speech therapist, dentist, orthodontists, and so forth. They decide and prepare the best treatment plan depending on the age of the child and the site of defect. This study involves the comprehensive review of the various scientific literatures on cleft lip and palate, discussing the embryology, classification, possible etiology, associated syndromes, clinical features and their management. Parental counseling is essential to create awareness on its preventive measures. Genetic studies should be carried out priorly to identify the potential risk factors which might predispose to this congenital anomaly.

Keywords: Congenital abnormalities; Cleft lip; Cleft palate; Children

Introduction
Cleft lip and cleft palate is the most common congenital developmental deformity that occurs in the primary palate and secondary palate (soft and hard palate). It can be a unilateral, bilateral, complete or incomplete cleft seen at the time of birth in humans [1]. It occurs due to some unknown disturbances during embryogenesis. It is inherited as an autosomal recessive trait i.e., both the parent should have copies of abnormal genes [2]. Some of the genes which are responsible are TBX22, PVRL1, IRF6, P63 and MSX1 [3]. The incidence of cleft lip and palate accounts for about 1:1000 live births [4,5]. The unilateral cleft accounts for about 80% of all the clefts and bilateral cleft accounts for about 20% of all clefts. The incidence of left sided cleft is more compared to right side cleft (R:L - 1:2). Males are most commonly affected by cleft lip compared to females (M:F - 2:1) [6]. The incidence of cleft lip and palate in INDIA is 1.7% among other population in the world [7]. It is increased in children with low birth weight; it is most commonly seen in first born child and in increased age of the mother during conception [8]. The etiology, which is responsible for the occurrence of cleft lip and palate are genetic abnormalities, environmental factors like any maternal infections, intake of teratogenic drugs (trimethadione, methotrexate, and thalidomide), radiation therapy during pregnancy for treatment of malignancies, poisonous diet, maternal alcohol consumption leads to maternal alcohol syndrome [9]. Cleft lip and palate can occur individually or along with congenital deformities such as Down’s syndrome, Pierre Robin syndrome, Velo cardiac facio nasal syndrome, Treacher Collins syndrome, Van der Woudes syndrome, Wardenburg syndrome, Apert syndrome, Electrodactyly- Ectodermal dysplasia- Cleft Syndrome (EEC) etc [10].

Complications Associated with Cleft Lip and Palate
Cosmetic abnormality, feeding difficulty, ear infections and hearing loss due to dysfunction of the eustachian tube connecting the middle ear and throat, recurrent infections can also contribute speech and language delay, dental problems [11].

Embryology
Normal development of cleft lip and palate requires the understanding of neural crest development theory which states that neural crest cells: Specialized embryonic cells gives rise to various connective tissue and neural tissue of the head and face [12]. These cells migrate to different places at variable rates, if there is failure of migration of the neural crest cells, facial abnormalities or cleft may result [12]. The upper lip and premaxilla are formed from three processes i.e., frontonasal process, right and left nasomedial processes of the maxilla. Lip closure occurs during 5th or 6th week.
of embryonic development. Palate is formed by union of two palatal processes that develop from the maxillary processes and closes during 8th or 9th week of gestations [13]. Maxillary processes appear at 5 weeks. Maxillary processes form most of the face, mouth, cheeks, and sides of the upper lip. These processes evolve into most of the hard palate, alveolar ridge, and soft palate. Frontonasal process develops into the nose, central part of upper face and primary palate. Lateral nasal process will form the ala of the nose. Medial nasal process will contribute to the formation of several structures: Mid portion of the nose, upper lip, maxilla, primary palate. The Medial nasal process fuses with maxillary processes by week 7 and forms upper lip and primary palate [14]. Failure of the medial nasal process to fuse with maxillary process leads to development of cleft lip. Prior to 6th week of development, the primary palate forms this consists of prolabium, premaxilla and four maxillary incisor teeth. During 6th week to 10th week of embryological development, the foundation for the hard palate and the floor of the nasal cavities, the palatine shelves, begins their migration toward the midline of the face. This movement toward midline coincides with the lowering of the tongue. Between 5.5 to 8 weeks, the primary palate and the secondary palate (that portion formed by the joining of the palatine shelves behind the incisive foramen) fuse and form hard palate. Failure of the two palatine shelves or processes to unite with the primary palate or with each other will result in a cleft of the hard palate [15].

**Classification**

Several classifications of cleft lip and palate were given by various authors. Basically, it is divided into two categories 1) Morphological classification and 2) Embryological classification.

**Embryological classification of cleft lip**

Unilateral cleft lip results from failure of maxillary process to merge with medial nasal process on the affected side, bilateral cleft lip results from failure of maxillary process to merge with the medial nasal process on both sides, median cleft lip results from failure of the medial nasal process to merge and form the intermaxillary segments, oblique cleft lip results from failure of the maxillary process to fuse with the lateral nasal process.

**Embryological classification of cleft palate**

Anterior cleft palate results from failure of fusion of lateral palatine processes fail to fuse with primary palate, posterior cleft palate results from failure of fusion of the lateral palatine processes with each other with nasal septum, complete cleft palate (Anterior & Posterior) results from failure of fusion of the lateral palatine processes with each other, with nasal septum and primary palate [16].

**Classification of cleft lip and cleft palate**

Davis and Ritchie [17] classified the clefts into three major groups; Group 1: Pre-alveolar cleft; Group 2: Post-alveolar clefts; Group 3: Alveolar cleft. It is in turn sub classified as unilateral cleft, median cleft and bilateral cleft (Figure 1).

Fogh-Anderson classified clefts into three major groups; Group 1: Cleft of lip; Group 2: Cleft of lip and palate; Group 3: Cleft of palate till the incisive foramen (Hard palate and soft palate). It is inturn subclassified as unilateral cleft and bilateral cleft.

Veau [18] classified clefts of lip into four major groups; Group 1: Unilateral notching of vermillion, Group 2: Clefts involving vermillion and lip, Group 3: Clefts involving vermillion, lip and nasal floor, Group 4: Bilateral clefing of lip complete or complete (Figure 2).

Veau classified clefts of palate into four major groups; Group A: Cleft of soft palate, Group B: Cleft of soft and hard palate, Group C: Cleft of hard, soft palate, alveolus and lip (unilateral), Group D: Cleft of hard, soft palate, alveolus and lip (bilateral) (Figure 3).

International Confederation of Plastic and Reconstructive Surgery classified the cleft lip and palate into three major groups based on the structure of the defect, location of defect and extent of defect;

- Group 1: Cleft of lip, cleft of palate and cleft of alveolus.
- Group 2: Unilateral and bilateral cleft.
- Group 3: Complete and incomplete cleft.

Elnassry [19] classified cleft lip and palate patients in to seven classes; Class I: Unilateral cleft lip, Class II: Unilateral cleft lip and alveolus, Class III: Bilateral cleft lip and alveolus, Class IV: Unilateral complete cleft lip and palate, Class V: Bilateral complete cleft lip and palate, Class VI: Cleft hard palate, Class VII: Bifed uvula.

Kernahan and Stark [20] Classified cleft lip and palate and it give the shape of stripped Y letter and include:

- Block 1 and 4 represent right and left cleft lip.
• Block 2 and 5 represent right and left cleft of alveolus.
• Block 3 and 6 represent right and left cleft of hard palate anterior to the incisive foramen.
• Block 7 and 8 represent right and left cleft involving the hard palate posterior to incisive foramen.
• Block 9 represent cleft involving the soft palate.

Millard [21] gave a modification to Kernahan and Stark’s classification. Additional to it, he added two triangles the top of stripped Y such as inverted triangle representing the cleft involving nasal arch or nose the upright triangle representing the cleft involving the nasal floor (Figure 5). Kreins O proposed LAHSAL system for classification of cleft lip and palate patients which was modified on the recommendation of Royal College of Surgeons Britain in 2005 by omitting one “H” from the acronym “LAHSHAL”. LAHSAL system is a diagrammatic classification of cleft lip and palate. According to this classification, mouth is divided into six parts: right lip, right alveolus, hard palate, soft palate (LAHSAL), left alveolus, and left lip. The first character is for patient’s right lip and last character for patient’s left lip. LAHSAL code indicates complete cleft with capital letter and an incomplete cleft with small letter. No cleft is represented with a dot (Figure 6) [22].

Clinical features of patients with cleft lip and palate

Clinical findings seen in cleft lip and palate are natal and neonatal teeth observed in unilateral or bilateral cleft palate, supernumerary teeth, congenitally missing teeth, rotation of permanent central incisor, in bilateral cleft lip and palate, the maxilla is protuberant and mobile, posterior cross bite, Mandibular prognathism, Ectopic eruption of primary lateral incisor, developmental anomalies of tooth morphology, deficiency of alveolar bone along the root surface of the tooth, microdontia, macrodontia, peg laterals, fused teeth, enamel hypoplasia, thick curved hypoplastic incisors, delayed eruption of permanent teeth [22].

Management of Cleft Lip and Palate

The management of cleft lip and palate requires multidisciplinary team approach of dental specialists, medical specialists and allied health science specialists. Dental team, which consists of pediatric dentist, prosthodontist, oral and maxillofacial surgeon and Orthodontist. Medical team, which consists of plastic surgeon, pediatrician, Psychiatrist, medical geneticist and ENT surgeon or otolaryngologist. Allied health science specialists, which consists of speech therapist, audiologist, nursing staff and social worker [23].

The Cleft Lip & Palate Association provides a contact point for parents and patients thereby allowing a free exchange of views regarding progress and possible difficulties. The three major goals in the treatment of cleft lip and palate is to maintain and provide adequate nutrition for infants, performing presurgical orthopedics, parent and patient counseling [24].

Multidisciplinary sequencing of treatment in clefts four stages

- **Stage I**: Maxillary Orthopedic Stage (Birth to 18 months)
- **Stage II**: Primary Dentition Stage (18 months to 5 Years of age)
- **Stage III**: Late Primary or Mixed Dentition Stage (6 to 10 or 11 yrs of age)
- **Stage IV**: Permanent Dentition Stage (12 to 18 years of age) [24,25].

Stage I: maxillary orthopedic stage (Birth to 18 months):
Immediately after birth, patient will have insufficient suction to pull the milk from nipple, there will be excessive air intake during feeding, will lead to regurgitation of fluids, nasal discharge and choking [26]. Spoon feeding can be done. Standard nipple with a cross cut or enlarged cut provides improved ejection of milk and large nipple design which direct milk into pharynx. The child should be in semi-upright position or 30° to 45° to prevent choking [27]. In 1950, prosthetic appliance or maxillary obturator was introduced. It is immediately inserted into the patient’s mouth to facilitate feeding and reduces choking. It aids in providing false palate. It maintains the maxillary cross arch stability [28]. Surgical repair of the lip is done between 10 weeks to three months of age. Prior to that, prepare the child for surgical repair of the lip. Rule of ten is used to ascertain the fitness of the child before the surgery. Child should be 10 weeks old, should have 10 gm of hemoglobin 10 pound weight. Proper feeding during first 3 months of age helps to gain weight for surgical repair of the lip [29].

Premaxillary orthopedics (Birth to 4 or 5 months)

A) Pre-surgical orthopedics

Growth potential of the nasal septum is powerful. In unilateral cleft, there is muscle pull to one side. These two forces combine and pull the major segments to the non-cleft side, if an opposing orthopedic force to the muscle pull can be applied, distortion can be minimized and it prevents lip dehiscence after lip surgery [30]. Active orthopedic force is given by three piece of tape joined together and reduces distortion. It is most effective because it produces scar along the philtrum and is more flexible than other geometric closure techniques [35].

B) Surgical management of cleft lip

Surgical repair of cleft lip is done by three techniques: Millard’s repair, Tennison-Randall’s repair, veau repair [32].

In Millard’s repair, rotation flap and columella flap are planned on the medial side of the cleft and advancement flap on lateral side of cleft. Full thickness of the lip is cut along the marking a rotation flap is rotated towards the cleft defect; rotation gap is produced on the mesial side, which is filled with an advancement flap [33,34].

In Tennison-Randall’s repair, a triangular flap is created on the lateral side of the cleft to fit into the triangular defect produced on the mesial side of the cleft. This procedure can be planned exactly after initial measurement. The result cannot be modified once the lip is cut. The scar is more prominent than in other procedures (Figure 8) [35].

In veau’s repair, simplest one-stage straight line closure is done and produces satisfactory result in a bilateral cleft lip. In this method, vermillion flap from either lateral side of the cleft is brought down over the prolabium to augment the vermillion in the center of the upper lip (Figure 9) [36].

After lip repair surgery, Logan’s bow is given to reduce the tension in the lip wound. It remains in place for 5 days. This repair will improve the infant’s appearance and relieve the parental apprehensions and increase the confidence (Figure 10) [30]. Among the three cleft lip repair techniques, Millard’s technique is most commonly used and it is most effective because it produces scar along the philtrum and it is more than other geometric closure techniques [35].

C) Maxillary orthopedics (3 to 9 months)

After cheiloplasty, maxillary arch collapse can occur unilaterally or bilaterally. Due to increased tension placed on the segments of the repaired lip. To prevent this, maxillary obturator is placed after surgery. It provides cross arch stability and support. Once the segment is aligned, bone grafting of alveolar cleft defect is done. Bone grafting is done at various time periods. Primary bone grafting is done in children less than 2 years of age; early secondary bone grafting is performed between 2 to 4 years of age. Secondary bone grafting is done between 4 to 15 years of age. Late secondary bone grafting is done for reconstruction of residual alveolar cleft defect in adults [37].

D) Surgical management of cleft palate

Surgical closure of cleft palate can be done by one stage or two stage repair. In one stage repair, hard palate repair is done by using mucoperiosteal flap technique at 12 to 18 months. In two stage repair, soft palate repair is done first before 18 months followed by obturation of the hard palate till the second stage repair. In the second stage repair, hard palate repair is done at 4 to 5 years. Palatoplasty is carried out between 12 to 18 months of age. The main objective of it is...
to facilitate normal speech pattern. In 1861, Bernard von Langenbeck described a method of palatoplasty for hard palate repair. It is carried out by elevating flaps from lateral and posterior regions of the cleft. Care should be taken to avoid injury to the greater palatine vessels. The denuded areas of palate are left to epithelialize. Tongue flap from dorsum of the tongue can also be used to close the cleft palate [38,39]. Originally only the cleft edges were incised, a lateral incision was made, the flap was elevated from the hard palate, the palatine musculature was divided and finally the sutures were applied (Figure 11) [39].

E) Complications of primary palate repair

Velopharyngeal insufficiency can occur after primary palate closure. It results in unsatisfactory speech, regurgitation of fluids and facial grimacing. It can be corrected by pharyngoplasty using palatopharyngeal flap [40].

STAGE II: Primary dentition stage (18 months to 5 years of age)

This stage is primarily focused on establishing oral health. Proper oral hygiene measures are practiced to prevent dental caries. During this stage, ectopic eruption of primary maxillary anteriors is common around the cleft defect, so recall and check up is done at 3 to 4 month interval [41].

Stage III: Late primary or mixed dentition stage (6 to 10 or 11 years of age)

During mixed dentition stage, major role is played by the orthodontist. Arch expansion is done by using NiTi arch Expander or Quad Helix. Maxillary Protraction is done using reverse pull Head Gear. Secondary bone graft is placed before the eruption of canine, as the canine erupts through the graft maxillary segment stability is reached [42].

Stage IV: Permanent dentition stage (12to 18 years of age)

Final correction is made at this stage. Orthodontic correction of the malaligned teeth is done if the canine is not erupted; it is exposed as the canine erupts through the graft maxillary segment stability is reached [42].

References


