Cleft Lip and Palate: Clinical Update

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ABSTRACT: Cleft lip and palate are the most common facial deformity. It may involve lip only, lip and palate and palate only. The main reasons of clefting in infants may be either environmental (such as smoking, alcohol, poor nutrition) or genetic factors (such as familial factors and chromosomes). A number of specialists involves in treatment of clefting and decides the best treatment plan depending on the site of defect and age of the infant.

KEYWORDS: Cleft Lip &cleft palate, etiology, treatment

I. Introduction

Clefts of the lip, alveolus and palate are the most common congenital malformations of the head and neck and are the second most common congenital malformation of the entire body, trailing only clubfoot in incidence.[1,2] Great advances have been made in the last 30 years in treatment, but multiple surgical procedures and a lifetime of clinic visits are still required, leading to emotional and physical stress for the patient and their families[1,2].

The ideal treatment of the cleft lip, alveolus and palate involves a multidisciplinary team approach.[3] Members include a pediatrician who oversees the development of the child; the palatal surgeon (usually a general plastic surgeon, sometimes an otolaryngologist); the oral maxillofacial surgeon for the development of the dentition and the palate; the otolaryngologist for diseases of the ear; the psychologist for both patient family; the speech and language therapist; and the clinical coordinating nurse. [3,4]

1. Epidemiology and Genetics

Clefts of the palate, alveolus and lip (CLAP) are syndromic or non-syndromic. Syndromic types are by definition associated with other malformations, and include the Pierre-Robin Sequence, Treacher-Collins Malformation, Trisomies 13 and 18, Apert’s Syndrome, Stickler’s Syndrome and Waardenburg’s Syndrome. At last count, more than 300 syndromes were associated with CLAP.[1,2] Syndromic etiologies include single gene transmission such as trisomies; teratogenic causes such as fetal alcohol syndrome; or environmental causes such as amniotic band syndrome or maternal diabetes mellitus.[4,5,6]

Non-syndromic CLAP is a diagnosis of exclusion, and is considered to be of multifactorial inheritance with known predicted rates of recurrence.[6]

An isolated cleft palate (CP) is genetically distinct from an isolated cleft lip (CL) or a combined CLAP. Cleft lip with or without cleft palate has an incidence of 1:1000 births in the United States and approximately 1:600 births in the United Kingdom, but is different among ethnic groups [3,4]. American Indians have the highest incidence, at 3.6:1000 births, followed by Asians, whites and blacks (0.3:1000). In contradistinction, isolated cleft palate is the same among all racial groups, with an incidence of approximately 1:2000 births. The male:female ratio for CL or CLAP is 2:1, whereas for isolated CP it is 1:2 [1,3].

2. Embryology

The palate is divided by the incisive foramen into the primary and secondary palates, with the primary palate being anterior to the foramen and the secondary palate posterior to the foramen.[4,5] The primary palate develops between 4 and 5 weeks gestation followed by the secondary palate between 8 and 9 weeks gestation. The primary palate, lip and alveolus develop as a mesodermal and ectodermal proliferation of the frontonasal and maxillary processes.[6] A primary cleft is a failure of proliferation, not a failure to meet in the midline. At no time in the development of the normal primary palate is there a separation.[7] Although there may be an isolated cleft lip without a cleft alveolus, a cleft alveolus is always associated with a cleft lip.[2,5]
The secondary palate develops as a medial ingrowth of the lateral maxillae with fusion in the midline. In normal development, therefore, a natural midline cleft exists, which is in contrast to the normal development of the primary palate. Deformities which prevent midline fusion, such as micrognathia and/or macroglossia, may result in a failure of midline fusion and therefore a secondary cleft palate. [1,2,3]

Clefts of the palate and lip are also classified as complete or incomplete. Complete clefts of the lip and alveolus involve extension into the anterior nose. Complete clefts of the secondary palate involve both the hard and soft palates with extension into the nose and exposure of the vomer. As will be discussed, attachment of the vomer may play a crucial role in the success of palatal repair.[1,2] An incomplete cleft has a midline attachment, ranging from a mucosal covering to one with musculature attachment to the midline raphe. The submucous cleft of the soft palate is an extension of the cleft of the secondary palate where mucosa meets in the midline of the soft palate but the velar musculature (levator veli palatini, tensor veli palatini) does not. The classic findings of a submucous cleft include a bifid uvula, absence of musculature attachment to the midline raphe of the soft palate, and a hard palate notch.[1-7]

3. Normal Anatomy

The upper lip is longer than the lower lip, and is shaped liked a flattened ‘M’. The lower lip is shaped like a flattened ‘W’. Cupid’s bow defines the central portion of the upper lip and the apices of the bow join the philtrum. The nadir bisects the apices. [8] The lip extends laterally to approximately the lateral limbus of the eye. Surrounding the lips is the orbicularis oris, the sphincter of the mouth. The maxilla has several distinct anatomical areas. The nasal spine is the anterior projection of the maxilla and alveolus. [7] The alveolar process of the maxilla surrounds the palate and houses the teeth. The incisive canal is located posterior to the incisors, and transmits the lesser palatine artery, one of the distal branches of the internal maxillary artery.[9] Posteriorly and laterally along the palate is the greater palate ton foramina, which transmit the greater palatine artery, a branch of the internal maxillary artery. The palate itself is formed from the maxilla, the horizontal process of the palatine bone and the pterygoid plates. The soft palate attaches to the posterior portion of the hard palate and interdigitates with the lateral pharyngeal wall via several muscular attachments. [5] From the nasopharyngeal to the oral cavity surface, the muscles of the soft palate consist of the palatopharyngeus, the salpingopharyngeus, the levator and tensor palatini, the muscular uvula, the palatoglossus and the superior constrictor muscle. [10]

The superior constrictor muscle is the primary sphincter of the pharyngeal phase of swallowing and is responsible for preventing regurgitation into the nasopharynx (velopharyngeal insufficiency, VPI). [11,12] The tensor veli palatini connects from the eustachian tube to the scaphoid fossa of the sphenoid bone and then to the lateral soft palate. It tenses the palate, but is not believed to play a major role in palatal elevation. Tubal dilatation from the tensor palatini is probably minimal [13,14]. The levator veli palatini originates from the bony cartilaginous junction of the eustachian tube and wraps around the hamulus before connecting to the soft palate. The levator is responsible for palatal elevation and perhaps tubal dilatation. The salpingopharyngeus is a consistently small muscle with probable minimal effects upon palatal and tubal function .[13]

4. Cleft Anatomy

The Unilateral Cleft Lip. The CLAP can be divided into defects of the lip, alveolus and palate. The cleft lip is a failure of mesodermal proliferation resulting in complete or incomplete defects. The complete unilateral cleft lip includes the orbicularis oris muscle, where the medial portion of the muscle attaches to the columella and the lateral portion to the nasal ala cartilage. The medial vermilion border is usually thin (called the white roll). An incomplete cleft lip ranges from a mucosal covering to a slight defect in the bulk of the orbicularis muscle which is barely detectable. [1-5]

The nasal defect of a unilateral cleft lip is fairly constant. The ipsilateral lower lateral cartilage is usually flattened and rotated laterally and inferiorly, resulting in horizontal, short appearance. The columella is short and often bends and dislocates the septum. The overall appearance is a flattened dome with a wide, horizontal ipsilateral nostril.[1-3]

The Bilateral Cleft Lip. The bilateral cleft lip is similar to the unilateral defect with the exception of a complete absence of orbicularis muscle on the medial aspect (the premaxilla or prolabium). [2] The prolabium is usually extruded to a varying degree. The nasal deformity is essentially a duplication of the unilateral defect, with a bilaterally flattened dome, short columella and bilateral horizontal nostrils.[14]

Clefts of the Primary Palate and Alveolus. The primary palate is that portion anterior to the incisive foramen. [15] A cleft of the primary palate results in a gap from the incisive foramen through the alveolus. Clefts of this type are always associated with clefts of the lip. [16]

Clefts of the Secondary Palate. Clefts of the secondary palate are a failure of medial growth of the palatal shelves. Fusion begins at the incisive foramen and progresses posteriorly. [2-4] The vomer is midline,
with various attachments to the remnant palate. The defect of the soft palate is failure of midline fusion. The palatal musculature attaches to the posterior hard palate. [3]

There are a wide clinical range of clefts of the secondary palate, from the submucous cleft to a complete cleft of the hard and soft palate. The submucous cleft palate is as a midline diasthasis of the velar musculature, a bifid uvula and a notch in the posterior hard palate. [1-5]

5. Classification

There is no universally accepted classification of clefts, although the most commonly used is the Veau classification, which was described in 1931. Veau Class I is an isolated soft palate cleft; Class II is a hard/soft cleft palate; Class III is unilateral cleft lip and palate; Class IV is a bilateral cleft of the lip and palate. Most surgeons describe the defect rather than using the Veau system. For example, a Veau Class III would be described as a unilateral complete cleft of the lip, alveolus, primary and secondary palates.[1,2]

6. Complications

Patients with cleft lip and palate experience several functional and morphological problems, depending on the type and extent of the cleft.

● Feeding and swallowing in a baby with CLP can be challenging due to oronasal communication, reduced sucking efficiency and additional structural, airway and neuromotor problems. [17]
● Mastication problems due to the presence of malocclusion which can affect the patient’s quality of life.;[18]
● Hearing problems due to middle ear infections and hearing loss . [17]
● Speech difficulties can be due to residual clefts/fistulae, velopharyngeal (VP) insufficiency, nasal obstruction and abnormal neuromotor and psychosocial development. [19]
● Craniofacial growth may be impaired and can result in midface retrusion
● The main cause of maxillary hypoplasia is scarring from primary palate surgery which restrict facial growth. 17]
● Nasal deformity is also a common problem in CLP patients, affecting quality of life. [19]
● Aesthetic issues due to clefting and protruded premaxillae can further contribute to sociopsychological problems experienced by CLP patients.[19]

7. Dental Abnormalities

Tooth formation can be disturbed in the area surrounding the cleft site and result in malformed crowns/roots, enamel hypoplasia, absent or ectopic teeth and supernumerary formation (Tannure et al., 2012). Maxillary lateral incisors are the teeth most affected (Ranta, 1986). Constricted maxillary dental arch, cross-bite and malocclusion are significantly increased in cleft patients (McCance et al., 1990). These patients also have a high incidence of dental caries and gingivitis (Dahllof et al., 1989).

8. Management of Patients with CLP

The Clinical Standards Advisory Group (CSAG) 1998 report (di Biase and Markus, 1998) showed that the outcome of cleft care within UK was poorer than in other European countries. [9] Therefore, to improve cleft care and outcomes, centralised regional cleft services were developed with multidisciplinary (MDT) teams including paediatricians, plastic surgeons, oral and maxillofacial surgeons, dentists, orthodontists, ear, nose and throat (ENT) physicians, psychiatrists and psychologists, restorative dentistry specialists, speech and language therapists, audiologists and co-ordinators. [14,16]

8.1 Infancy and Primary Dentition Stage

Management of CLP during infancy includes the following aspects

● Prenatal diagnosis (and referral to the regional CLP team).[6]
● At birth, support is provided by specialist nurses to ensure normal bonding and feeding. Further infant feeding interventions may include fabrication of a feeding obturator and premaxilla repositioning appliances to retract and rotate the malposed premaxillae segment in bilateral cleft palate cases [20]
● Lip repair surgery is carried out at 3–6 months after birth to shift the premaxillae back and close the cleft lip.
● Palate repair surgery is performed at 12–18 months of age.
● Formal speech assessment is carried out at 18 months and is monitored throughout childhood.[21]
● Closure of residual fistula and correction of velopharyngeal insufficiency at 4–5 years of age may be necessary to aid speech development.
●● Preventive dental care, including oral hygiene education and dietary advice, is an important part of dental management at this stage[20,21].

8.2 Mixed Dentition

Bony defect of the maxillary alveolus can result in a variety of problems at mixed dentition stage, including displacement, rotation and tipping of the adjacent teeth and their failure to erupt (often maxillary canine). [6]Bony defect can also result in collapse of the maxillary dental arch with loss of alveolar contour. Bony support around the base of nose can be compromised and in bilateral cleft cases, there can be instability and mobility of the premaxilla. Residual oronasal fistulae can also cause functional problems. [4]Therefore, alveolar bone graft (ABG) surgery is carried out at the age of 8–10 years (prior to the eruption of maxillary canines) to address the above problems and restore the continuity and stability of the maxillary arch. Orthodontic differential expansion of the collapsed maxillary arch and some alignment of maxillary incisors can be considered prior to ABG surgery. Cancellous bone harvested from the iliac crest has been shown to provide the best outcome.[22]

8.3 Permanent Dentition

Treatment during permanent dentition may include orthodontic treatment, orthognathic surgery, distraction osteogenesis and finally restorative management.[6]

8.3.1 Orthodontic Treatment

The extent and nature of orthodontic treatment depend on the severity of the malocclusion and the skeletal class III pattern. Orthodontic treatment alone, including dental camouflaging, alignment and retention, may be considered if the skeletal discrepancy is mild. [4]Presurgical orthodontic treatment by simple maxillary dental alignment to improve the dental appearance in patients who require surgical correction of the skeletal problem is often indicated. Postsurgery orthodontic treatment may be required for final alignment of teeth and also can be indicated following any relapse. [6]

8.3.2. Orthognathic Surgery

Le Fort I maxillary advancement osteotomy with or without mandibular set-back surgery is indicated for the correction of skeletal class III pattern due to maxillary hypoplasia in CLP patients following completion of growth.[4,6]

Complications of orthognathic surgery in CLP patients include the following.

●● Increased risk of postoperative infection and ischaemic complications.
●● Maxillary advancement can compromise VP function, producing hyponasal speech.
●● Retromaxillary scarring can occur following surgery.[4]
●● Relapse rate (both horizontal and vertical relapse) is much greater than in the non-cleft population and ranges from 30% to 65%.[7]

8.3.3. Distraction Osteogenesis (DO)

Distraction osteogenesis is used in patients with severe maxillary retrusion which is beyond correction with conventional orthognathic surgery. Following presurgical orthodontic treatment, Le Fort I osteotomy cuts are placed and the maxillae is mobilised to enable placement of the distraction device, which can be either a rigid external (RED) or internal device. The latency period is normally 4–5 days to allow callus formation. Bone lengthening is achieved by gradual mechanical distraction at a rate of 1 mm/day with a consolidation period of 3 months.[23]

The advantages of DO are as follows;

●● Larger corrective movements than orthognathic surgery.
●● Decreased postsurgical relapse.
●● Fewer postoperative problems with speech and language.
●● It can be used in growing patients but overcorrection is required.

The disadvantages of DO include the following.

●● Precise movements and vector control are difficult to achieve, especially with internal DO.[24]
●● Multiple review appointments are required during the distraction phase.[23]
●● It can induce anxiety and stress.
●● Scarring associated with the RED.
●● Risk of infection.
●● Encumbrance of the device can affect the quality of life.[23]
●● Second general anaesthesia is required to remove the device.[25]

9.3.4. Restorative Treatment
Restorative dentistry specialists provide the final active treatment in CLP patients and provide restorative advice to the cleft team.[6] The aim of restorative treatment is to prevent relapse following orthognathic surgery and orthodontic treatment, restore masticatory function and improve speech and aesthetics.[4,6] Restorative challenges in CLP patients include the following.[26] 
- Discrepancy in the maxillo-mandibular relationship.
- Malposition and partial eruption of teeth.
- Malformation and tipping of teeth.
- Caries.
- Flat palate resulting from severe scarring.
- Presence of frenula, absence of keratinised mucosa, gingival recession, shallow vestibule and difficult hygiene.
- Poor condition of supporting periodontal tissues.

Prevention, including fluoride, dietary advice and oral hygiene education, is an integral part of any restorative treatment. Several factors contribute to the difficulty in maintaining oral hygiene of teeth close to the cleft area, including presence of gingival recessions, frenula, tooth malpositioning, dental anomalies and extended use of fixed orthodontic appliances.[27] Non-surgical and surgical periodontal treatments may be indicated. Surgical therapies include gingivoplasty, gingivectomy, gingival grafts, removal of frenula and restoration of biological dimensions.[28]

Different types of prosthodontic treatment can be indicated for replacing the missing teeth and restoring the compromised dentition in CLP patients.[29] 
- Tooth reshaping and composite additions
- Veneers
- Crowns
- Resin-bonded bridges
- Conventional bridges
- Removable partial dentures can be a temporary solution for replacing the missing teeth or can be a definitive treatment option when fixed restorative options are not possible.[30]
- Osseointegrated implants can be successfully used in CLP patients but often require further bone grafting.[4]
- Overdentures and overlay dentures can be useful in extreme cases with severe maxillary hypoplasia and retrusion causing a large skeletal discrepancy that is not amenable to surgical and orthodontic treatment.[31]

Dentures can help to replace missing teeth, cover malpositioned teeth, recover the maxillary arch, close the anterior open bite and provide lip support.[9] 
- Complete dentures are indicated in edentulous patients. Challenges of complete denture treatment in CLP patients include compromised denture stability and retention due to the alveolar ridge anatomy, physical factors (impaired adhesion, surface tension and cohesion) and presence of residual fistula or unrepaired cleft, which would allow the penetration of air and compromise the seal.[31]
- Obturators are indicated for rehabilitation of untreated cleft patients and in patients with residual oronasal communication in large defects following surgery. Obturators can also eliminate hypernasality and improve speech. Obturators are discussed in detail.[30]
- Palatal lift prosthesis is indicated when there is velopharyngeal incompetency following soft palate repair.[32] This is a removable appliance that extends to the soft palate and helps to lift the soft palate to achieve closure to improve speech.[31]
- Speech aids and speech bulbs can be useful when the soft palate has inadequate length and is unable to close, leading to velopharyngeal incompetency. The appliance covers the hard palate and the bulb extends posteriorly to the soft palate, providing velopharyngeal closure and therefore improving speech.[32]

II. Summary

Cleft lip and palate are the most common facial deformity and clefting may involve lip only, lip and palate and palate only. Environmental (such as smoking, alcohol, poor nutrition) and genetic factors (such as familial factors and chromosomes) are the main reasons of clefting in infants. Treatment of clefting involves a number of specialists who decide the best treatment plan depending on the site of defect and age of the infant.
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