CHILDREN'S HOSPITAL
PEDIATRIC REVIEW
FOUNDING MEMBER OF LCMC HEALTH
CHILDREN'S HOSPITAL • NEW ORLEANS
NOVEMBER 2015 • VOL. XXVIII, ISSUE 6

Comprehensive Management of Cleft Lip and Palate

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Objectives

At the end of this activity, the participant should be able to:

1. Discuss the important anatomical elements involved in cleft lip and palate
2. Review the importance of a multidisciplinary team in management of patients with cleft lip and palate
3. Describe the treatment protocol for patients with cleft lip and palate

Introduction

Orofacial clefting is the most common craniofacial anomaly that will be encountered throughout a pediatrician's clinical practice. Cleft lip, with or without cleft palate (CLP), needs to be differentiated from isolated cleft palate (CP) and atypical facial cleft as a clinical entity. Two thirds of all cases of orofacial clefting involve the lip with or without cleft palate while one third has deformity isolated to the palate. LCP is more common in males while isolated CP is seen more often in females. When looking at the laterality of CL, the left side is more commonly affected. The incidence of CLCP in the Caucasian population is 1:1000 while it is double in Asians and half among African-Americans. Of note is that there are no variations throughout ethnicities for isolated CP with a frequency of 0.5 per 1000 live births. Approximately 5% of patients with CLCP and CP will have an associated syndrome, more commonly with isolated CP. Atypical facial cleft are rare conditions and are well defined by Tessier's Classification, but are beyond the scope of this article.

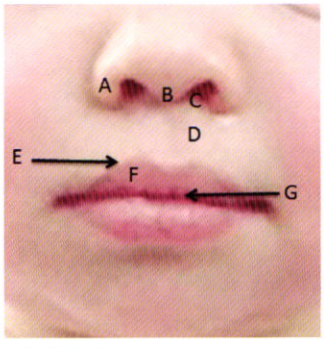


FIGURE 1
Important anatomical landmark of the lip. A-nasal alae; B-columella; C-nasal sill; D-philtral column; E-white roll; F dry mucosal lip; G wet mucosal lip.

Pertinent anatomy

The upper lip has important anatomical structures and subunits that need to be reconstructed in order to provide optimal results. There is a mucosal and a skin component to the lip. The mucosal portion can be further divided into dry and moist components. The white roll of the lip defines the extent of the vermillion border. The orbicularis oris muscle allows the lips to function as a sphincter. In patients with cleft, the later will be discontinuous and inserts at the lateral aspect of the pyriform aperture (bony nasal passage). The orbicularis oris also projects fibers within the cutaneous portion of the lip which creates the philtral columns extending from the medial nasal sill to the height of cupid's bow.

Defining the extent of the cleft lip is important in communication between healthcare providers. Typically, the laterality is identified first followed by the complete or incomplete nature of the cleft. A complete cleft lip extends and includes the nasal sill while an incomplete cleft is anything short of that. Microform or "forme fruste" cleft lip typically referred to a notch in the lip with or without muscle involvement. Finally, asymmetric bilateral cleft lip is seen where the degree of clefting is different on both sides.

It should be noted that despite the clinical presentation being referred as a cleft lip, significant deformities are associated with the nose. In unilateral cleft lip, the ipsilateral alae will be displaced laterally inferiorly as well as being depressed. This is seen both in complete and incomplete situations as well as in bilateral cleft lip.

The palate can be divided into primary and secondary. The incisive foramen is the structure that divides the later. The primary palate extends from the vestibule and includes the dental alveolus while the secondary palate extends to the tip of the uvula. The secondary palate includes the hard and soft palate. The soft palate includes two important muscles, the levator veli palatini and the tensor veli palatini muscles. The former is most important for palatal function and require careful reconstruction in palatal repair (intravelar veloplasty). Palatal cleft presents an abnormal muscle orientation where the levatot veli palatini runs antero-posteriorly. In order to insure adequate velopharyngeal function, the fibers needs to be detached from the posterior palatal shelves and reconstructed in the midline following dissection from the oral and nasal mucosa.

Many classification of palatal cleft exist. We favor a simple anatomical description of the defect. The laterality of the defect is identified first. It is relatively easy to identify in the primary palate while it may be a little more difficult to visualize on the secondary palate. The vomer and nasal septum will define the laterality of the secondary palatal cleft. In bilateral situation, the nasal septum/vomer will not be attached to the palatal shelves while in unilateral situations, it will be attached to the contralateral palatal shelves.

Submucous cleft palate refers to a situation where there is no frank oronasal communication but the levator veli palatini muscle is oriented antero-posteriorly. As such, it creates a central area of thin white mucosa centrally (zona pellucida) and will need to be corrected surgically. Finally, bifid uvula is considered part of the spectrum of cleft palate. It may be associated with submucous cleft but as an isolated finding will not require surgical repair.

FIGURE 2
Left complete cleft lip and palate. The nose shows typical deformity with a depressed alae that is inferiorly and laterally displaced.

Diagnosis

Clearly, a thorough physical exam in the baby nursery will identify if the newborn has a CLCP. Even if difficult, it is important to visualize the entire palate including the tip of the uvula. With improvements in ultrasound technology and easier access, pre-natal diagnosis is now relatively common. While pre-natal diagnosis can be simple for a well-seasoned radiologist, evaluation of the intra-oral structures is often difficult. Pre-natal diagnosis of CL or CLCP should prompt a visit to a craniofacial team, to ensure the parents are well prepared to care for their newborn.

The Importance of the Cleft and Craniofacial Team

The complex and specialized nature of the treatment required by children with CLCP has long been recognized. In order to address these needs appropriately, multidisciplinary teams have been developed to ensure comprehensive care of the children with CLCP. Critical members of the team include audiology, genetic, nursing, otolaryngology, oral and maxillofacial surgery, orthodontics, pediatric dentistry, plastic/craniofacial surgery, speech therapy and social work. Regular meetings with the team ensure that delivery of care is well coordinated and offered in a timely fashion.

Comprehensive Management

The initial intervention in the care of a child born with CLCP remains appropriate feeding counseling to the parents. Breastfeeding may be more difficult, but is certainly possible with cleft lip only. Understanding the mechanics of feeding in an infant as well as the pathology in CLCP will ensure adequate caloric intake. More frequent burping and feeding with the head slightly elevated will optimize feeding. There are multiple specialized bottles available on the market. Our group often recommends a time-proven technique: a bottle with a cross cut nipple. The latter allows adequate flow that can be easily controlled.

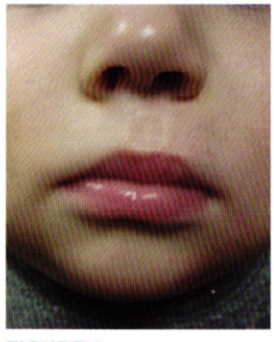


FIGURE 3
Early result of a left cleft lip repair which shows good symmetry of the lip and nose.

Primary Cheilorhinoplasty (Cleft lip repair)

The initial surgical intervention is the repair of the cleft lip. In situations where the cleft lip is complete, a preliminary intervention in the form a lip adhesion and/or alveolar molding (through taping or specialized appliance) will optimize the position of the alveolar segments at the time of the formal cleft lip repair. The latter is typically performed at 3 months of age. Multiple techniques have been designed but all have a common goal, which is to restore the esthetic and function of the lip

while minimizing the appearance of the scar. Furthermore, great attention is given to the nasal deformity. Again, restoration of form and function is paramount in the initial management of the cleft nose. Typically, non-absorbable suture will be used and need to be removed the following week under anesthesia. A nasal conformer, which helps shape the lower lateral nasal cartilage will be worn for two months.

Cleft palate repair (Palatoplasty)

The timing for repair of cleft palate is from 10 to 12 months of age. The surgical goals are to repair the oronasal fistula and restore function of the soft palate to assure adequate velopharyngeal function. Meticulous repair of the levator veli palatini muscle will promote the latter. In order to avoid trauma to the repair while feeding, patients will typically feed using a sippy-cup with a short tip. In conjunction with the cleft palate repair, myringotomy tubes are typically placed in the same setting. The distorted anatomy of the tensor levi palatini predispose to abnormal middle ear drainage and recurrent ear infection.

Velopharyngeal competence

The next important phase in the development of patient with CLCP or CP is the development of appropriate speech. A significant number of patients will benefit from speech therapy in order to prevent the development of a speech disorder in patients where velopharyngeal function is marginal. Speech therapy overseen by an experienced therapist in the management of cleft patients will provide maximum benefit. In refractory velopharyngeal insufficiency, surgical correction may be required. In order to appropriately diagnose the pathology, nasopharyngoscopy is performed. Inadequate nasopharyngeal closure can be directly observed and adequate surgical correction through a superiorly based pharyngeal flap or a sphincter pharyngoplasty can be planned. In the setting of patients with velocardiofacial syndrome, pre-operative imaging should be performed prior to pharyngeal surgery as medialization of the internal carotid is sometimes seen.

Dental management

Appropriate dental management of patients with cleft lips and palate is paramount. The first visit to the pediatric dentist should be by 1 year of age or within six months of the first tooth eruption. This will ensure good dental health but also familiarize the child to the dental office and prepare him or her for the first stage of orthodontic treatment. Patients with CLCP may develop transverse discrepancies in maxillary/mandibular relationship. Initial correction with orthodontic alignment and maxillary expansion is performed at around 7 years of age (once the permanent first molars have erupted). This will set the stage for the alveolar cleft reconstruction.

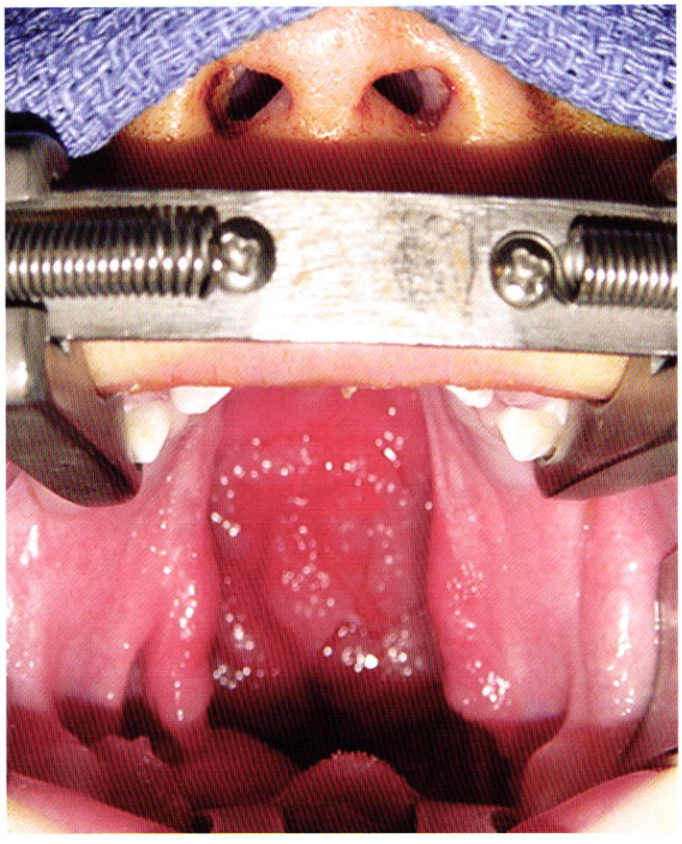


FIGURE 4
Complete secondary cleft of the secondary palate, U-shaped in a patient with Pierre-Robin sequence.

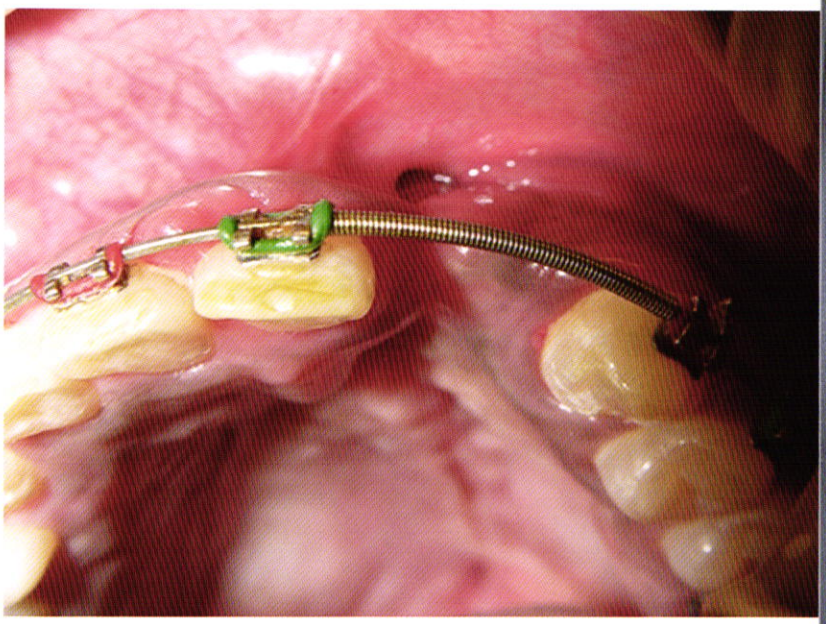
Skeletal reconstruction

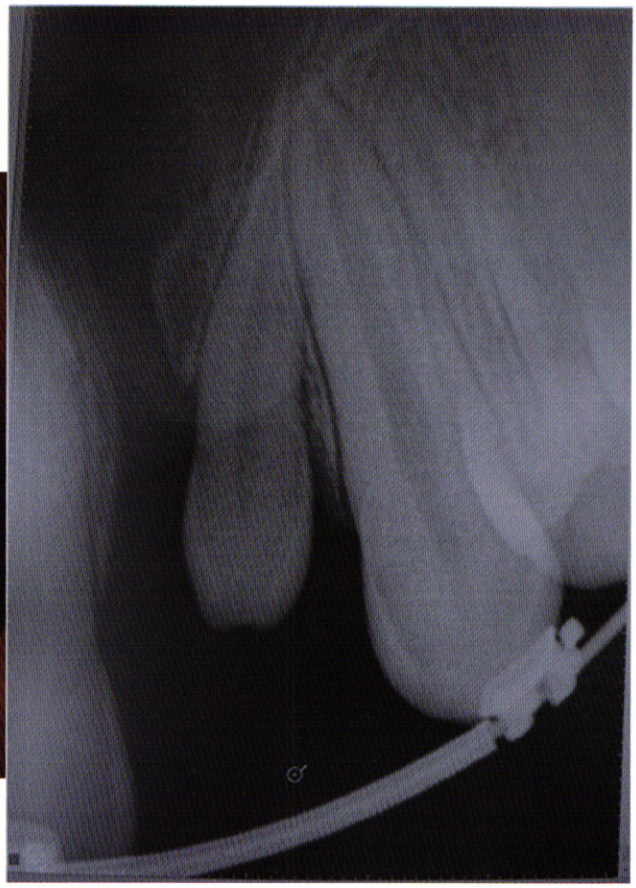
Alveolar cleft reconstruction is typically done at 9-12 years of age and is based on the dental maturity of the patient. It is timed according to the eruption of the ipsilateral permanent canine. Radiograph will allow evaluation of the root formation. Alveolar cleft bone graft, along with closure of persistent oronasal fistula will be performed with bone typically harvested from the iliac crest. There are certainly alternatives available but the later remains the gold standard. Adequate alveolar reconstruction will not only restore the continuity of the maxilla but will also augment the deficient pyriform aperture thereby improving alar support. Furthermore, when the permanent lateral incisor is missing, it should provide enough bone stock for the placement of an osseointegrated dental implant.

The following years will be followed by significant maxillofacial growth. This will need to be monitored to ensure that if disharmony between the maxillary and mandibular arch develops, it can be addressed orthodontically. In situations where the discrepancies are significant, corrective jaw surgery will be needed. The later will not be performed prior to maxillofacial skeletal maturity (16 years old for female and 18 years for male). Once the harmony of the dental arches is achieved by orthognathic surgery, the final stage of surgical treatment will

FIGURE 5

Left alveolar cleft showing a nasolabial fistula. The radiograph shows an unerupted lateral incisor. This patient missed the opportunity of a timely bone graft as the canine is already erupted.





be the completion rhinoplasty. The later will not only address the external nose, improving its appearance but also commonly involves turbinectomy and septoplasty to improve nasal air flow.

Common conditions associated with CP

Van der Woude syndrome (VdWs)

VdWs is a genetic disorder that is characterized by CLCP and lower lip pits. It is inherited in an autosomal dominant fashion and, as such, affected parents have a 50 percent chance of passing their mutation to their offspring.

Velocardiofacial syndrome (VCFs)

VCFs is a genetic condition (22q11.2 deletion) characterized by abnormal development of the pharyngeal arch resulting in defective development of the parathyroid, thymus and heart. Furthermore, an overt or submucous cleft palate may be present. As noted previously, medialization of the internal carotid can be seen.

Pierre Robin sequence (PRS)

PRS is a condition characterized by micrognathia and glossoptosis leading to airway obstruction. The presence of a large U-shaped cleft palate is common but not necessary for diagnosis. In order to alleviate the airway obstruction, prone positioning, tongue lip adhesion, mandibular distraction osteogenesis or tracheostomy may be needed.

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2. The usual timing for the surgical repair of the cleft palate is
   a. 10-12 months
   b. 2 months
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3. The distorted anatomy of the tensor levi palatini muscle predisposed the child with a cleft palate to
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