An Overview of Timeline of Interventions in the Continuum of Cleft Lip and Palate Care

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BACKGROUND

Cleft lip and/or palate (CL/P) is the most common congenital craniofacial anomaly, with a prevalence of 1 in 700 live births.\textsuperscript{1–3} According to the US Centers for Disease Control and Prevention, each year 2650 babies are born with a cleft palate, and 4440 babies are born with a cleft lip with or without a cleft palate in the United States.\textsuperscript{2,3} Clefts can be unilateral or bilateral, complete or incomplete and involve the alveolus, lip, and/or palate in various combinations. The highest rates of CL/P are reported in Asian populations (0.8–3.7 cases per 1000 individuals), while the lowest rates are reported in Africans (0.2–1.7 cases per 1000 individuals).\textsuperscript{4,5} Both genetic and environmental factors have been associated with the development of CL/P. Some of the environmental factors implicated include maternal smoking and alcohol consumption, poor nutrition, and viral infections.\textsuperscript{6} Over 350 genes and 300 syndromes have been associated with CL/P.\textsuperscript{7} Genes associated with nonsyndromic CL/P include IRF6, 8q24, WNT3, 10q25, and RFC1.\textsuperscript{8–11} In addition to traditional polymorphisms, certain methylation patterns have also been associated with an increase risk in CL/\textsuperscript{12,13}. A child born with CL/P is typically followed at a cleft/craniofacial center where many specialists

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are involved in the continuum of care. The objective of this article is to provide an overview of major dental and surgical interventions that are performed in patients with CL/P.

If not treated appropriately in a timely manner, those with CL/P can experience catastrophic events such as premature death and life-long difficulties in feeding, speaking, hearing, self-esteem, and psychosocial relationships. The earliest intervention in those with CL/P starts during the first few weeks of life (infant orthopedic treatment performed by a pediatric dentist or orthodontist in preparation for repair of the lip), and the final phase of treatment is comprehensive orthodontic treatment (with/without orthognathic surgery) that is usually performed in the late teen years. Dentists play a crucial role in the continuum of cleft lip and palate care (Table 1); therefore it becomes critical that dentists are knowledgeable of the treatment protocols and timing. An overview of the timeline of interventions for the CL/P patient is presented in Table 1.

<table>
<thead>
<tr>
<th>Chronologic Age</th>
<th>Dental Development</th>
<th>Interventions</th>
<th>Providers</th>
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<tr>
<td>By 6 mo</td>
<td>Predentition</td>
<td>Infant orthopedic treatment</td>
<td>Orthodontist and/or pediatric dentist</td>
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<td></td>
<td></td>
<td>Lip repair</td>
<td>Cleft and craniofacial surgeon</td>
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<tr>
<td>10–24 mo</td>
<td>Primary dentition</td>
<td>Palate repair</td>
<td>Cleft and craniofacial surgeon</td>
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<tr>
<td>1–2 y</td>
<td>Primary dentition</td>
<td>Establishment of dental home (and follow every 6 mo)</td>
<td>Pediatric dentist</td>
</tr>
<tr>
<td>2.5–3 y</td>
<td>Primary dentition</td>
<td>Speech assessment and velopharyngeal surgery (if indicated)</td>
<td>Cleft/craniofacial surgeon</td>
</tr>
<tr>
<td>5–10 y</td>
<td>Primary dentition and mixed dentition</td>
<td>Assess timing of maxillary (alveolar) bone grafting</td>
<td>Orthodontist/pediatric dentist/cleft and craniofacial surgeon</td>
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<tr>
<td></td>
<td></td>
<td>Maxillary expansion to establish arch forms and correct posterior cross-bites</td>
<td>Orthodontist</td>
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<tr>
<td></td>
<td></td>
<td>Maxillary (alveolar) bone grafting</td>
<td>Cleft and craniofacial surgeon</td>
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<td>9–12 y</td>
<td>Early to late mixed dentition</td>
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<td>Orthodontist</td>
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<td></td>
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<td>Orthopedic treatment using face mask/reverse pull head gear</td>
<td>Orthodontist</td>
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<td>12–14 y</td>
<td>Permanent dentition</td>
<td>Bone plate-supported class 3 elastics to correct maxillary/mandibular antero-posterior discrepancies</td>
<td>Orthodontist and cleft/craniofacial surgeon</td>
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<td></td>
<td></td>
<td>Maxillary distraction osteogenesis (if there is large maxillary/mandibular antero-posterior discrepancy)</td>
<td>Orthodontist and cleft/craniofacial surgeon</td>
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<td></td>
<td></td>
<td>Comprehensive phase of orthodontic treatment (if determined that there will not be a need for orthognathic surgery)</td>
<td>Orthodontist</td>
</tr>
<tr>
<td>&gt;14 y</td>
<td>Permanent dentition</td>
<td>Comprehensive orthodontic treatment (with or without orthognathic surgery)</td>
<td>Orthodontist</td>
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<tr>
<td></td>
<td></td>
<td>Orthognathic surgery (following completion of growth)</td>
<td>Cleft/craniofacial surgeon</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Final restorative treatment</td>
<td>Periodontist/prosthodontist/primary care dentist</td>
</tr>
</tbody>
</table>
PRESURGICAL INFANT ORTHOPEDIC TREATMENT

Presurgical infant orthopedic treatment (PSIOT) is often the first major clinical intervention that is performed on patients with CL/P. PSIOT is initiated within the first few weeks of life, before surgical repair of the lip. PSIOT is purported to restore the skeletal, cartilaginous, and soft tissue anatomic relationship prior to lip repair and consequently enhance the surgical outcomes. Facial tapes, Latham appliances, and Naso alveolar molding (NAM) technique have been widely used for PSIOT (Figs. 1). Nasoalveolar Molding for Unilateral and Bilateral Cleft Lip Repair by Kapadia and colleagues in this issue provide an overview of the NAM approach and Dentofacial Orthopedics for the Cleft Patient: The Latham Approach by Allareddy and colleagues in this issue provide an overview of Latham approach for PSIOT. Certain craniofacial centers elect to perform PSIOT only if there is a large defect, while several others do not perform any type of PSIOT. There has been considerable controversy regarding the long-term efficacy of PSIOT and its adverse impact on maxillary growth. Studies originating from Europe have shown that PSIOT is not an effective intervention and recommend against it. However, several craniofacial centers in the United States elect to perform PSIOT with varying degrees of success. A recent survey suggested that half of craniofacial teams reported offering PSIOT, with the NAM technique being the most popular. Grayson and colleagues have demonstrated that use of NAM is associated with improvements in nasal angle and increases of nostril width, columellar height, and columellar width.

LIP REPAIR

Primary cleft lip repair is the first surgical procedure that is undertaken by the surgical team (Fig. 2). The repair is generally performed between the ages of 3 and 6 months with the purpose of establishing lip competence by the unification of the underlying orbicularis oris muscle. Lip competence is essential for feeding, speech, and control of oral secretions. There are multiple different techniques for closure of the unilateral cleft lip defect, with the most popular including the Millard technique, the Fisher unilateral cleft lip technique, and Mohler technique. All techniques share in common the need to increase lip height on the affected side by regional geometric flaps; however, each technique approaches this problem differently. The surgical technique for a bilateral cleft lip repair is generally approached in a more standard fashion across all centers. The need for primary rhinoplasty at the time of lip surgery has been fiercely debated throughout the years. Most surgeons have incorporated at least a minimal nasal dissection at the time of the primary lip surgery, convinced that it leads to better nasal outcomes and does not significantly increase the risk of nasal stenosis. It is essential that prior to taking the child to the operating
room for a cleft lip repair that the patient has undergone an evaluation to assess hearing. Ninety percent of cleft patients will have conductive hearing loss secondary to incompetent drainage of the eustachian tube into the nasopharynx. Tympanostomy tubes are able to relieve this obstruction and may be placed simultaneously with the lip repair, thereby sparing the child an additional anesthesia event.

**PALATE REPAIR**

The palate is essential for velopharyngeal competence, which leads to proper speech development and feeding. However, every surgical intervention leads to scarring, and scarring can lead to growth restriction of the nasomaxillary complex. This is why the primary palatal repair is recommended to be performed between the ages 10 to 18 months of age. In this way, one allows unrestricted growth of the palate for as long as possible until speech development demands a repair. There are many different approaches to palatal repair, including: Bardach 2-flap technique, Veau-Wardill-Kilner Pushback technique, von Lagenback bipedicle flap technique, and the Furlow double-opposing Z-Plasty. The surgical technique employed should be tailored to each patient, with the goals being to ensure appropriate palatal length, including intravelar veloplasty (repair the levator veli palatini [essential for proper palatal function]), and eliminating anterior palatal fistulae.

**VELOPHARYNGEAL INSUFFICIENCY/INCOMPETENCE**

Velopharyngeal insufficiency/incompetence (VPI) can occur in patients with repaired and unrepaired CL/P. VPI is defined as the ability to completely close the velopharyngeal sphincter that separates the oro- and nasopharynx, which is required for the normal production of all but the nasal consonants. The absence of this ability, termed velopharyngeal insufficiency (VPI), defined as an anatomic or structural defect results from inadequate closure of the velopharyngeal valve. It is seen in a wide range of patients following primary cleft palate repair, with 5% to 40% of cleft palate patients presenting abnormal speech resonance because of residual anatomic structural abnormalities. The primary effects of velopharyngeal insufficiency are nasal air escape and hypernasality. Speech articulation errors (ie, distortions, substitutions, and omissions) are secondary effects of velopharyngeal insufficiency.

Children with VPI may have impaired speech intelligibility because of hypernasality, articulation, and low-speech volume. Secondary effects of VPI include nasal regurgitation of liquids, compensatory misarticulations, and facial grimacing. VPI may impact the child’s confidence, social development, and overall quality of life.

Velopharyngeal insufficiency can be diagnosed by both subjective and objective means. Along with a thorough medical history, a speech assessment and careful physical examination are required in the children with VPI. The most common diagnostic evaluations of the velopharyngeal function and closure pattern are nasoendoscopy and multiview videofluoroscopy. MRI can be used for examining the anatomy of the velopharyngeal mechanism. Perceptual speech assessment is of central importance in diagnosing VPI and predicting postsurgical outcomes.

Treatment of the VPI falls under the umbrella of 3 main modalities: speech therapy, prosthetic devices, and surgical management. The palatal lifts and palatopharyngeal obturators/pharyngeal bulbs are the most commonly used prosthetic devices. These devices are anchored to the dentition and allow closure of the velopharyngeal port, either by altering the position of the velum (palatal
lift prosthesis) or by occupying the pharyngeal gap (pharyngeal bulb).\(^\text{33}\) The primary drawbacks to prosthetics are dental caries with concomitant poor dental hygiene, noncompliance, and emotional distress associated with wearing the prosthesis.\(^\text{39}\) Noncompliance may be exacerbated by the need for frequent device adjustments, and the number of follow-up visits necessary for proper fitting may be responsible for the further compliance burn.\(^\text{33}\) To overcome these drawbacks, a surgical treatment option is available for selected cases. However, the success of the surgical correction of VPI depends as much or more on the selection of the appropriate procedure than on technical expertise.\(^\text{33}\) The most commonly used techniques for the surgical management of VPI are furlow palatoplasty, or double-opposing Z-plasty for palatal lengthening; pharyngeal flap; buccinator flaps for palatal lengthening; and dynamic sphincter pharyngoplasty (DSP).

Thorough knowledge of the velopharyngeal anatomy and physiology is critical for understanding the VPI and selecting a specific treatment option to address this condition. Although many different alternatives are available, there is no universal consensus to guide procedure choice, and recent advances in imaging and VPI treatment modalities continue to evolve.

**PREPARATION FOR CLEFT MAXILLARY BONE GRAFTING**

Patients with CL/P have collapsed maxillary arches that manifest clinically as posterior and anterior cross-bites (Fig. 3). The maxillary arch form is frequently asymmetric, and the minor segment is displaced medially with a collapsed arch adjacent to the cleft. The term alveolar bone grafting is a misnomer, as the entire maxilla is dysmorphic and therefore requires augmentation. A key intervention that an orthodontist or pediatric dentist performs in preparation for the maxillary bone graft procedure is maxillary expansion. Another article provides more details on the techniques of maxillary expansion and associated outcomes. In brief, the indications for maxillary expansion are correction of cross-bites, making maxillary arch compatible with the mandibular arch, and enabling access to the maxillary bone graft site, especially in significantly collapsed arches. Depending on the type and amount of expansion required to achieve the previously mentioned objectives, a wide range of orthodontic appliances (eg, fan-shaped expanders, Haas/Hyrax expanders, and quad helix appliance) may be used.

The timing of maxillary expansion depends to a large extent on the timing of maxillary bone grafting. Thus it is crucial that there is excellent communication among cleft/craniofacial team members, especially between the orthodontist/pediatric dentist and cleft and craniofacial surgeon. It is recommended that maxillary expansion be initiated approximately 6 months before the scheduled maxillary bone graft procedure. The maxillary bone graft procedures must be performed before the eruption of the maxillary permanent canines. The timing of maxillary bone grafts is provided in detail in another article. Typically, the authors evaluate the size and location of the maxillary defect using a combination of periapical, occlusal, and panoramic radiographs. Occasionally, limited field cone beam computed tomography (CBCT) images are also used. If the maxillary defect is present close to the developing permanent central incisor root or permanent lateral incisor root (if this tooth

![Fig. 3. Presentation of a left unilateral complete cleft lip/palate.](image-url)
is present and viable), then the authors recommend grafting before eruption of the permanent lateral incisor. If there is no viable permanent lateral incisor tooth or if the defect is away from root of a developing permanent central incisor, then the grafting can be done once the root of the permanent canine is approximately 50% to 75% developed and starts to erupt.

MAXILLARY BONE GRAFTING

Maxillary bone grafting (MBG) is a critical and time-sensitive component of competent cleft care. A successful bone graft provides unity to the maxilla, closes the remaining oral nasal fistula, establishes the nasal skeletal base, and allows the eruption and maintenance of dentition in the area of the cleft. Timing is critical. Bone is unable to be successfully grafted into a defect if any portion of a tooth is in the cleft site. Therefore, it is critical to bone graft prior to tooth eruption into the cleft site. Bone graft timing is dictated by tooth eruption patterns and location of the cleft. If the cleft is more centrally located and the lateral or central incisor could possibly erupt into the cleft, the bone graft should be performed earlier, between 5 and 7 years of age. If the cleft site is more laterally located in the area of the canine, then it may be safe to wait until the patient is between 7 and 11 years of age. Starting at 5 years of age, a cone beam CT is recommended yearly as part of the overall cleft team examination. This radiological examination provides extensive information regarding the size and orientation of the defect, and more importantly, the development of the adjacent teeth.

Expansion prior to grafting needs to be evaluated on a case-by-case basis. In some patients, expansion increases the size of the defect, making the bone graft unlikely to succeed. The other extreme is demonstrated by severe collapse of the maxillary segments, making it impossible to graft because of the inability to access the cleft site. In the first case, it may be wise to graft first and then expand if necessary, whereas in the second example expansion is an absolute requirement prior to grafting. It is critical for the orthodontist, pediatric dentist, and cleft surgeon to all be competent and collaborative in evaluating the dentition and timing of the maxillary bone graft.

Supernumeraries and teeth in the cleft site present a challenge to successful grafting. Patients with cleft lip and palate are prone to supernumerary teeth in and around the cleft site. These are best removed at least 6 weeks prior to the bone graft procedure. By removing these teeth, bone fill is allowed in the extraction sites prior to grafting, making the size of the defect smaller when the actual graft is performed. It also allows time for the mucosa to heal; this will allow the surgeon the maximum amount of tissue possible to work with and manipulate during the actual maxillary bone graft surgery. Malformed or absent laterals are also an issue in the cleft population. Fifty percent of cleft patients do not have lateral incisors on the affected side. Of those patients who do have laterals, 50% are malformed in the cleft population and may need to be extracted prior to the MBG surgery.

Autogenous bone (generally harvested from the iliac crest) is the gold standard for reconstruction of the maxillary cleft site. This has recently been challenged by the use of bone-morphogenetic protein (BMP) mixed with allograft.

PREMAXILLARY REPOSITIONING

In the bilateral cleft lip and palate patient it is sometimes necessary to perform premaxillary repositioning surgery at the time of the cleft maxillary bone graft (Fig. 4). It is not uncommon for the premaxilla to be so abnormally positioned that a bone graft is actually impossible without its repositioning. Although repositioning of the premaxilla at the time of primary lip surgery is almost always contraindicated and indeed associated with significant maxillary hypoplasia and potential loss, it is sometimes necessary at the bone-grafting stage and can be performed safely and effectively. When performed competently, it will restore the integrity of the maxillary arch, allowing appropriate bone grafting and restoration of the dental arch, aiding in speech and elimination of associated oral nasal fistulae. In the bilateral cleft lip and deformity, it is not uncommon for the premaxilla to be so abnormally positioned that a bone graft is actually impossible without its repositioning.

The blood supply to the premaxilla depends on the nasal septum and the buccal mucosa. The deformity results from the collapse of the lateral alveolar segments and the extension of the premaxilla on the nasal septum. The blood supply to the premaxilla depends on the nasal septum and the buccal mucosa. During the premaxillary setback, a wedge of the nasal septum is removed, and the premaxilla is repositioned and splinted in place. Because of the tenuous blood supply, it is recommended to perform only 1 side of the
bone graft at the time of the setback and to return to the operating room 8 weeks later to perform a bone graft on the contralateral side. Coordination with the orthodontist is critical to determine the appropriate amount of expansion prior to setback. Virtual surgical planning is helpful in determining the amount of possible and necessary movement. This is ideally performed in the late mixed dentition phase of treatment, and creativity needs to be used in stabilizing and splinting the premaxillary segment after repositioning. Wiring the splint to the dentition with the use of orthodontic wires is preferred but not always possible when securing the splint to the teeth; the use of skeletal anchors to attach the splint is sometimes required.\(^42\)

**LIMITED ORTHODONTIC TREATMENT FOLLOWING MAXILLARY BONE GRAFTING**

Frequently, those with CL/P may require a limited phase of orthodontic treatment (usually only in the maxillary arch) following a maxillary bone grafting procedure. This limited phase of orthodontic treatment is performed to facilitate eruption of impacted teeth, correct anterior cross-bites that lead to traumatic occlusion, to align/level the maxillary arch, and to establish compatible arch forms. Occasionally, if the permanent teeth adjacent to a grafted site erupt ectopically, these can be moved into an ideal position with limited orthodontic treatment, and this is by far the most common indication for limited phase of orthodontic treatment. The movement of the roots of the permanent teeth into the grafted site delivers physiologic stress and thus contributes to the longevity of the grafts.\(^{15–47}\) It is recommended that radiographs (limited field CBCT or periapical/occlusal radiographs) be exposed to assure the health of the grafted site and maxillary arch continuity before initiating the limited phase of orthodontic treatment. During recent years, bone anchored plates and class 3 elastics have also become popular adjuncts to comprehensive orthodontic treatment, and use of these is thought to minimize the need for orthognathic surgery. This treatment may be initiated during the late mixed dentition stage.

**COMPREHENSIVE PHASE OF ORTHODONTIC TREATMENT**

The typical clinical features in patients with CL/P include

- Maxillary hypoplasia (this can either be caused by deficient inherent growth potential or restrictions in maxillary growth resulting from scar tissues following the various surgical interventions that occur along the continuum of cleft care)
- Class III dental occlusion
- Anterior crossbite (negative overjet)
Posterior cross-bite (relative or absolute maxillary/mandibular transverse discrepancy) Reduced anterior facial height (caused by overclosure)\textsuperscript{48–51}

Patients with CL/P almost always require a comprehensive phase of orthodontic treatment that is initiated following eruption of all permanent teeth in the teen years. Depending on the amount of skeletal and occlusal discrepancy, the comprehensive phase of orthodontic treatment is done with or without orthognathic surgery. For those requiring a comprehensive phase of orthodontic treatment in conjunction with orthognathic surgery, it is best to initiate treatment either following cessation of growth or close to completion of growth so as to avoid retreatments. It has been reported that 22% to 40% of patients with CL/P require orthognathic surgery.\textsuperscript{48–53} Major drawbacks with large maxillary advancements using classical maxillary osteotomies are high relapse potential and velopharyngeal incompetence and associated speech difficulties.\textsuperscript{50,54,55} An alternative to classical maxillary osteotomy for large maxillary advancement is the distraction osteogenesis procedure. The key to long-term success of distraction osteogenesis is retention within the first 6 months following the procedure. Suzuki and colleagues\textsuperscript{56} examined a cohort of unilateral cleft lip and palate patients and followed them for 12 months after maxillary distraction osteogenesis and reported dentoskeletal relapse rates of 53.7% in the vertical dimension and 22.3% in the horizontal dimension within the first 6 months. There was no significant relapse during the 6 month to 12 months after surgery. Cho and Kyung\textsuperscript{57} followed a cohort of patients with severe cleft maxillary hypoplasia for 6 years after maxillary distraction osteogenesis and reported a 13.5% relapse rate in angular measurements within the first 6 months and only 0.3% relapse from 1 to 6 years.

**RESTORATIVE PHASE**

Patients with CL/P frequently having congenitally missing teeth and enamel defects of permanent teeth.\textsuperscript{58,59} Maxillary permanent lateral incisors adjacent to the cleft side are frequently congenitally missing or diminutive in size, which often necessitates implants and implant-supported crowns and/or extensive restorative work. The implant phase of treatment is initiated following cessation of skeletal growth. The restorative dentists work closely with the orthodontist to determine the ideal space requirements for placement of implants and implant-supported crowns, and the orthodontist completes the comprehensive phase of treatment, keeping the space requirements in perspective.

**SUMMARY**

Patients with CL/P require a multitude of interventions from a myriad of specialists. The earliest intervention is the infant orthopedic treatment of the maxillary alveolus prior to surgical repair of the lip, initiated in the first few months of life. The comprehensive phase of treatment is completed during the late teen years. When a proper team approach to care is taken, excellent outcomes are often realized.

**DISCLOSURE**

The authors have nothing to disclose.

**REFERENCES**


