Lec 13 Treatment of Cleft lip and palate
Part 2

At Birth

Giving birth to a child affected by a cleft can be a distressing experience for the parents, particularly if this condition has not been diagnosed in utero. A multitude of emotions can occur, including shock, anger, guilt, grief and even rejection. It is important that adequate support is given to the parents and that a bond is quickly established between the parents and child.

• The clinical nurse specialist from the regional cleft team provides initial support, help and advice as soon as possible after diagnosis; and

• Patient support groups such as the Cleft Lip and Palate Association (CLAPA) also play an important role in providing continued help and advice.

A baby born with CLP may experience difficulty in feeding at birth. CP produces an open communication between the oral and nasal cavities. Suckling can be slow because the baby will have difficulty generating adequate intraoral pressure and milk can be lost through the nose before it is swallowed. It is important to establish an effective feeding regime as soon as possible:

• Feeding is generally successful using an assisted feeding bottle (Fig. 1) with a standard orthodontic teat, which can be squeezed to generate the necessary pressure; and

• Breastfeeding is occasionally possible, but may need supplementary feeding from a bottle.

Figure 1: Special feeding bottles for CLP.

Presurgical orthopaedics

A period of active presurgical orthopaedic alignment of the cleft alveolar segments is occasionally carried out in the neonate to reduce the size of the cleft defect and facilitate surgical repair. Specialized facial strapping or orthodontic plates (Fig. 2) are used, which can be passive or active and help mould or reposition the divided facial and maxillary segments. In particular, these plates have been used for:

• Reducing protrusion of the premaxillary segment in bilateral CLP cases;

• Reducing the size of an alveolar cleft and approximating the lip margins in unilateral CLP; and

• Reducing the width of an isolated palatal cleft.

Presurgical orthopaedic treatment is usually carried out at the discretion of the operating surgeon. There is currently little substantive evidence to suggest that any of these techniques provide long-term benefit for the dental arch relationship or facial appearance and their use remains controversial.

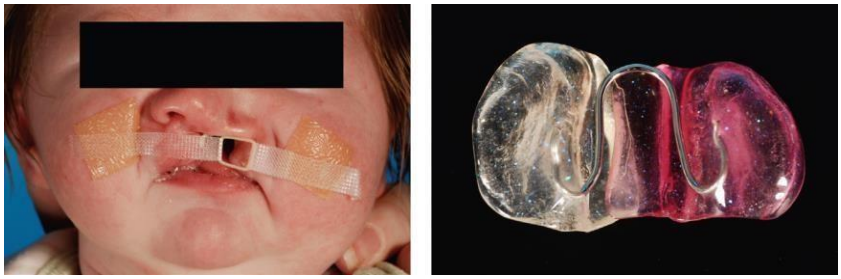


Figure 2: Orthopaedic cleft appliance to approximate the lip segments prior to repair (left panel) and intraoral orthopaedic appliance to approximate the palatal shelves (right panel).

Surgical repair of cleft lip and palate

A number of individual surgical techniques for repairing the embryonic deficits associated with both the lip and palate have been described. However, evaluating which technique, sequence or timing will provide optimum results is difficult and currently no true consensus for any of these criteria exists. Early surgery does allow the child to establish good orofacial function as soon as possible and this is particularly important for the development of normal speech. However, surgical repair can be associated with scarring

in the maxillary region, which can produce growth deficiencies in all three planes of space:

• Midline scar tissue can prevent transverse growth and produce crossbites; and

• Scar tissue within the tuberosity region can tether the maxilla to the sphenoid bone, preventing downward and forwards growth and producing a class III skeletal pattern.

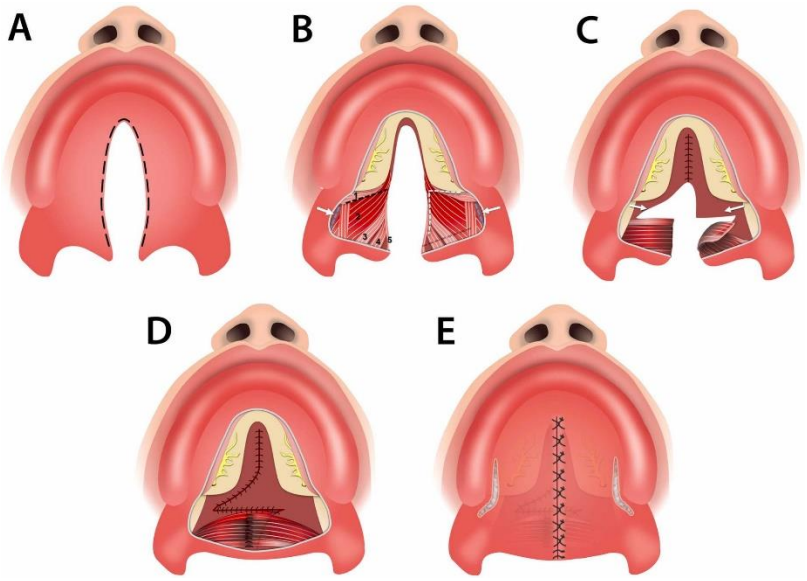
It is clear from comparative studies that facial growth is compromised in operated cleft subjects when compared with those from unoperated samples, particularly those that have undergone palatal repair. The goal of surgical correction is to minimize any potential growth discrepancy, whilst maximizing the aesthetic and functional outcome.

Lip repair

Surgical repair of cleft lip is usually carried out between 3 and 6 months of age as a single procedure, the exact age being dictated by surgeon preference. Classically, the rule of ‘tens’ has been used, with surgery only taking place once the child is at least 10 weeks old, 10 pounds in weight, and having a haemoglobin level of 10%. However, waiting until these criteria are achieved can delay surgery and it has been argued that this can cause problems with both parent–infant bonding and early growth and development. Indeed, advances in neonatal care and paediatric anaesthesia have made it possible to perform cleft surgery during the neonatal period, although there is currently no clear evidence to suggest that this is particularly advantageous.

Palate repair (palatoplasty)

The timing of palate repair represents a balance between maximizing the positive effects of early palate closure on feeding and speech development, whilst minimizing the potentially negative effects of inhibited maxillary growth and development as a result of surgical scarring. Currently, repair of CP is normally undertaken between 9 and 12 months of age and usually involves a palatoplasty to move tissue towards the midline, with or without some lengthening of the palate to improve the posterior soft palate seal.



Speech and language

Following repair of the palate, a speech and language therapist monitors speech development closely. Velopharyngeal insufficiency is the result of an inadequately functioning soft palate, which may be unable to lift and produce a good seal with the posterior pharyngeal wall. Velopharyngeal insufficiency can produce:

• Nasal escape on pressure consonants (i.e. k, p, t); and

• Hypernasality.

The main problem is a lack of mobility in the soft palate, secondary to scarring from the palate repair. In combination with the dental abnormalities, malocclusion and hearing difficulties, which are all often seen in cleft children, velopharyngeal insufficiency can also produce poor articulation, which results in significant difficulties with speech. Surgery is usually carried out as soon as velopharyngeal insufficiency is diagnosed, around the age of 4 before the child begins school, and generally involves either a re-repair of the soft palate or pharyngoplasty. Pharyngoplasty aims to reduce hypernasality by narrowing the velopharyngeal space.

Middle ear disease

Otitis media is also a common finding in children with CP, disruption to the muscles of the soft palate affecting function of the Eustachian tube. This can reduce the acuity of their hearing, causing further potential adverse effects on the development of speech and language. It is important that an audiologist monitors these children and, if necessary,

tympanostomy tubes (or grommets) are placed by an ENT surgeon.

Dental care in the primary dentition

A program of preventive dentistry should be established during early dental development, particularly as many children affected by clefting are vulnerable to caries. Dietary advice should be provided, good oral hygiene established and fluoride supplementation instituted, if necessary. It is vitally important that the dentition of a cleft child is not compromised by dental disease and the cleft team will liaise closely with the general dental practitioner to ensure that this is the case.

There is occasionally delay in the eruption of primary teeth adjacent to the cleft and the lateral incisor can be absent, hypoplastic or even duplicated. Crossbites can occur in the buccal segments; however, active orthodontic treatment is rarely indicated in the primary dentition.

Dental care during the mixed dentition

As the permanent teeth begin to erupt, crossbites affecting both the incisor and molar dentitions can occur and their severity often reflects the degree of disruption that has occurred to maxillary growth and development as a result of previous surgery. The maxillary incisors can also be crowded, rotated and tilted, particularly those adjacent to the cleft, and significant centreline discrepancies are common. In addition, anomalies of dental development affecting teeth around the site of a cleft alveolus can also be seen. These include the presence of supernumerary teeth or agenesis of the permanent lateral incisor; anomalies of shape and size, or enamel defects.

Dental preventive measures should continue during the mixed dentition; in particular, the first molars should be fissure sealed and monitored to ensure these teeth do not become carious.

Alveolar bone grafting

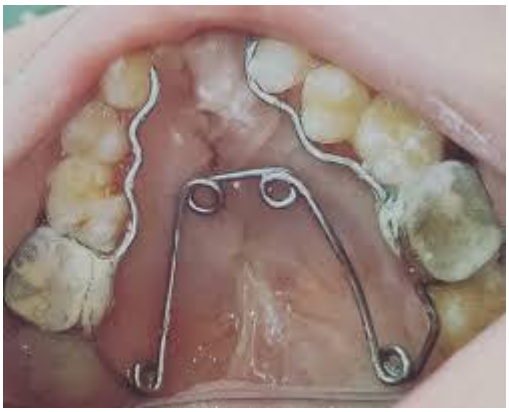
The presence of a residual bony defect in the maxillary alveolus of children affected by complete clefts is a deformity associated with a number of functional and aesthetic problems, which can affect both the occlusion and local orofacial region:

- Adjacent teeth are often displaced, rotated or tipped;
- Teeth in the region of the defect are unable to erupt (particularly the maxillary canine and if present, the lateral incisor);

• The bony defect can lead to collapse of the maxillary dental arch with a loss of alveolar contour;
• Bony support around the base of the nose can also be compromised, with flattening on the cleft side;
• In bilateral cases, there can be instability and mobility of the premaxillary segment; and
• Larger defects can be associated with oronasal fistulae (communications between the oral and nasal cavities in the anterior palate).

Alveolar or secondary bone grafting involves placing cancellous bone, usually harvested from the iliac crest, directly into the maxillary alveolar defect. This procedure is normally carried out at around 8–10 years of age, prior to eruption of the permanent canine, when root formation of this tooth is around two-thirds complete. A period of orthodontic treatment is usually required prior to graft placement to expand the collapsed maxillary arch and create surgical access, maximizing the amount of bone that can be placed. This expansion is often achieved with a quadhelix appliance, followed by a period of retention with a palatal arch. During this phase of orthodontic treatment, some alignment of the maxillary incisors can be achieved by either extending the arms of the helix or using a simple fixed appliance, but care needs to be taken not to move any teeth into the cleft site where there is no bone; if this is a concern, these teeth should be aligned after the bone graft.

Alveolar bone grafting has made a significant contribution to the oral rehabilitation of children with cleft palate. Grafting cancellous bone into the cleft allows teeth to erupt into this region and facilitates tooth movement, which means that orthodontic tooth alignment and space closure can be achieved. Moreover, the timing of alveolar bone grafting means that it does not interfere with growth in the width and length of the anterior maxilla, because this is essentially complete by 8 years of age. Vertical development of the maxilla would appear to continue normally after the insertion of a bone graft.



Dental care during the permanent dentition

Once the permanent dentition is established a decision is made regarding the need for orthodontic treatment alone to correct any malocclusion, or a combination of orthodontics and orthognathic surgery. A key factor is the degree of maxillary and midfacial retrusion, but it should be remembered that these patients also exhibit a full range of mandibular growth patterns and mandibular prognathia can also be seen. A period of time monitoring further facial growth may be required before a final decision is made, but if surgery is indicated, presurgical orthodontic treatment will usually begin once facial growth is complete. Occasionally, in cases with severe maxillary retrusion, osteogenic distraction is employed to move the maxilla forwards in a younger, growing patient. This will provide some early improvement in the profile and reduce the size of the jaw movements that will be required for definitive orthognathic surgery in the late teenage years, once facial growth is complete. Similarly, severe maxillary crowding can be treated prior to definitive surgical set up in younger patients, to provide some improvement in dental aesthetics without compromising later combined orthodontic surgical treatment.

In those cases that can be treated with orthodontics alone, there are often a number of specific problems that exist:

• Crowding associated with a narrow and retrusive maxillary arch;
• Crossbites affecting teeth in the anterior and posterior maxilla; and
• Congenital absence or anomalies associated with teeth in the cleft region.

Orthodontic treatment with fixed appliances is usually indicated in these cases, often in conjunction with some maxillary expansion, but following the same general principles of treatment planning for any malocclusion. Correction of the severely rotated teeth and posterior crossbites often seen in these cases usually requires long-term retention.

Cleft lip nose

In young adults affected by a cleft lip, the nasal aesthetics can be poor; in particular, the nose can be asymmetrical at the tip and the alar base can be collapsed on the side of the cleft repair, both of which may require surgical revision. Primary cleft rhinoplasty can be effective, particularly in unilateral cases and nasal tip correction can also be achieved.

These procedures are commonly undertaken in the later teens. Revision of any surgical scarring associated with the primary lip repair may also be required. These procedures are also usually carried out following the completion of definitive orthodontic or combined treatment in the late teenage years.