Cleft Lip and Palate: A Review for Dentists

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Abstract

The goals of primary closure of cleft lip and palate include not only re-establishing normal insertions for all of the nasolabial muscles but also restoring the normal position of all the other soft tissues, including the mucocutaneous elements. Conventional surgical wisdom, which recommends waiting until growth is complete before undertaking surgical correction of the postoperative sequelae of primary cheiloplasty, carries with it many disadvantages. If, after primary surgery of the lip, orolabial dysfunctions remain, they will exert their nefarious influences during growth and will themselves lead to long term dentofacial imbalances. These imbalances can significantly influence facial harmony. Unless accurate, symmetric and functional reconstruction of the nasolabial muscles is achieved during the primary surgery, not only will the existing dentoskeletal imbalances be exaggerated, but other deformities will be caused during subsequent growth, among which the most important are nasal obstruction and mouth breathing, reduced translation of the maxilla, dysymmetry of the nose and inability of the patient to symmetrically project the upper lip.

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hen assessing children with cleft lip and palate, the general dentist should be on the lookout for oronasal fistulae, inability of the child to project the upper lip symmetrically and deviation of the nasal septum to the noncleft side. All of these problems indicate failure to achieve the goals of primary surgical closure of the cleft, that is, good function of the labial muscles and normal nasal breathing. Early referral for correction of these problems is necessary to ensure the best possible facial growth during childhood. The purpose of this paper is to explain the functional and anatomic bases of inadequate surgery for primary cleft lip and palate, to better prepare dentists to recognize these problems and refer their patients for treatment as early as possible.

Anatomic Features of Cleft Lip and Palate

Congenital labiomaxillary clefts result from the absence of fusion or incomplete fusion of the maxillary and medial nasal processes. It is generally accepted that a congenital labiomaxillary cleft forms about the 36th or 37th day of gestation and that the earliest points of ossification in the maxilla and premaxilla appear about the end of the 6th week of gestation, when the elements of the cartilaginous nasal capsule and facial muscle precursors are already present.¹ In a complete labiomaxillary cleft, the muscles of the nasal floor and the upper lip cannot bridge the gap of the cleft, nor can they unite with their muscular counterparts on the noncleft side. The muscular integrity of the region is considerably disrupted, which has a profound effect on the underlying skeleton.

Normally the anterior muscles of the face form 3 rings:

- 1. a superior nasolabial ring formed on each side by the transverse nasal muscle and the levator muscles of the upper lip;
- 2. a middle labial ring formed on each side by the orbicularis oris muscles of both the upper and the lower lips;
- 3. a lower labiomental ring formed by the depressor anguli oris, depressor labii and mentalis muscles.

The transverse nasal muscle is the most important physiological element in the nasolabial ring. It passes from the anterior border of the nasal bone to the incisive crest and then to the nasal septal perichondrium. Not only is this muscle responsible for nostril constriction, but, together with the external fibres of the orbicularis oris muscle, it

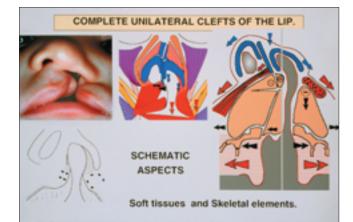


Figure 1: Composite schematic showing the dynamics of distortion in the unoperated cleft lip and palate.

provides support for both its corresponding half of the upper lip and, indirectly, the labial commissure.

In cases of total unilateral labiomaxillary cleft, the muscles on the cleft side remain lateral to the defect and cannot function normally, even if they are correctly formed. Deprived of the nasal septum and the anterior nasal spine as points of anchorage, the structure collapses, much as a circus tent would do if it lost its central pole. Growth of the minor segment of the maxillofacial complex appears reduced, probably as a consequence of the absence of stimulation from the nasolabial muscles.

On the noncleft side (which is frequently but erroneously referred to as the normal side), the nasolabial muscles that are inserted on the nasal septum pull it into the noncleft nostril. Furthermore, the premaxilla on this side is underdeveloped by an amount equal to the degree to which the median interincisive suture is bent to this side (Fig. 1).

The alar cartilage of the cleft side is flattened by divergent muscular traction corresponding to the degree of its missing support. The cartilaginous deformations can be considerable, but except in rare cases, such as holoprosencephaly, there is little or no hypoplasia of the cartilage.¹ Indeed, in many cases there is no cutaneous hypoplasia, and if the skin appears insufficient it is because it has not been distended by function.¹ The sagging nasal capsule induces retrusion of the nasal bone on the cleft side, which gives rise to internal rotation of the anterior maxillary pillar, which in turn causes lateral displacement of the medial canthus. Accordingly, surgery to correct labiomaxillary clefts should reconstruct both form and function of the divided face so that balanced growth of the facial skeleton can take place.

The management of cleft lip and palate is a very broad subject. We examine here the fundamental concepts from which can be determined the best therapeutic approach and the best surgical techniques for cleft lip and clefts of the palate. Surgical correction of cleft lip remains a difficult challenge, principally because the basic surgical problem has not been clearly conceptualized. This failure to accurately define the problem is attributable to the fact that the relevant anatomy is both complex and frequently erroneously described. The goal of primary surgery for cleft lip and palate ought not to be simply reconstruction of the lip but should also encompass reconstruction of all the formative systems of the facial skeleton, to give the best possibility for maxillary development.

Primary Cleft Lip

Several factors influence the optimum timing for closure of a cleft lip. The parents may be persuaded that closure in the first few days after birth is in the baby's best interest. However, any surgeon who believes that physiological surgery is absolutely essential would not be prepared to undertake surgery at this stage. In young infants, it is practically impossible to distinguish the individual muscle bundles, which, because of lack of function, are underdeveloped, friable and unable to adequately support sutures. It is therefore preferable to wait until at least the end of the fourth month, by which time the labial musculature has developed significantly as a result of both growth and function, the function imparted by sucking, crying and other facial activity.¹

The nature of the cleft — unilateral or bilateral — must also be considered. In unilateral complete clefts, early reconstruction of the lip further reduces forward growth of the lesser fragment, which results in retrusion of the entire dentoalveolar segment and underdevelopment of the premaxilla, a problem that is difficult to correct later. It is better, then, to wait until the end of the sixth month, by which time the upper deciduous incisors are better developed or even erupting. This date is physiologically logical.¹⁻³

In bilateral complete clefts, in which the premaxilla is lengthened and often considerably advanced, it is preferable, in our opinion, to close both sides of the cleft lip simultaneously, during the fourth month. Three months later, that is to say, during the seventh month, the dentoalveolar elements of the premaxilla and the lateral segments are usually adequately realigned such that it is possible to perform gingivoperiosteoplasty, a procedure that encourages better transverse development of the maxilla. Careful reconstruction of the nasolabial muscles, establishment of a functional nasal airway on the cleft side and maintenance of the initial surgical result with a nasal retainer worn for at least 6 months provide the best possible functional environment in which good maxillary growth can take place^{2,3} (Figs. 2a, 2b, 2c, 2d, 2e, 2f).

Primary Cleft Palate

Normal development of the inferior part of the maxilla depends on the following factors: the *en bloc* sagittal and vertical movements of the palatal and maxillary bones and

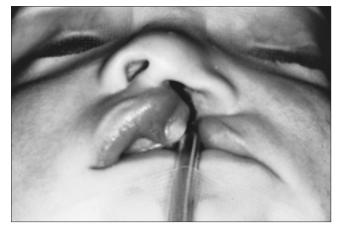


Figure 2a: Unilateral left cleft lip and palate in a 5-month-old baby.



Figure 2c: Appearance 6 days after operation.



Figure 2e: Child at 3 years of age.

the activity of sutures that separate them from the vomer and the cranial base, as well as the transverse and vertical development of the palatal vault.⁴

After closure of a cleft palate by means of the classic techniques, in which vomerine mucosa is used to reconstitute the nasal layer of the secondary palate, the vomer and



Figure 2b: Appearance immediately after functional lip and nose surgery.



Figure 2d: Primary functional soft palate repair carried out concomitantly with lip surgery.



Figure 2f: Lip at 16 years of age.

the palatal shelves are joined by a tight, unyielding, fibrous scar, which seemingly prevents normal sutural activity. This scar reduces the advancement and lowering of the floor of the nasal fossa and the palatal vault and tends to produce troubles of nasal ventilation and a class III deformity.

In palatal clefts, the primordial role of the palatal



Figure 3a: Unrepaired bilateral cleft lip and palate.

maxillary fibromucosa in the transverse and vertical development of the vault is adversely affected by the classic surgical techniques of palatoplasty, in which lateral palatal flaps are medially displaced to the summit of the vault. In this abnormal situation, the maxillary fibromucosa cannot provoke the vertical and lateral development of the palatal vault. It is able only to thicken the roof of the palate and so flattens it.

In summary, the classic techniques of palatoplasty, involving medial displacement of the palatal fibromucosal flaps, are physiologically unacceptable. Instead, we recommend techniques that do not use or do not displace these flaps (or that do so only minimally).

In practice, all of the techniques that use anteromedial displacement of the palatal mucoperiosteal flaps should be rejected. It is preferable to close the palate in 2 operations. The first, performed as early as 6 months, at the same time as the primary lip closure, serves to close the soft palate. The second is performed when the baby is about 1 year of age. At that time, it is possible, after wide subperiosteal dissection, to close the hard palate using only the palatal mucosa covering the cleft margins. In some cases this can be accomplished by medial displacement of only small flaps of the mucoperiosteum of the roof of the palatal vault.

The timing of the first operation described above is suggested because by 6 months of age the muscle is developing well. Thereafter, imparted function will reduce scar development. However, closing the hard palate at the same time would lead to major growth problems.⁴ It is better, therefore, to delay closure of the hard palate until the age of about 12 months, at which time gingivoperiosteoplasty is also performed. The residual cleft of the hard palate is then closed at the age of 12 months, by which time, with rare exception, the cleft has become sufficiently narrow to be closed without or with only minimal displacement of the palatal maxillary fibromucosa⁴ (**Figs. 3a, 3b**).



Figure 3b: Appearance of patient at 7 years of age, after functional palate and lip surgery.

Cleft Alveolus

The use of autogenous bone for grafting of cleft alveolar defects allows improvement in function, speech, psychological well-being and esthetics. The timing of cleft alveolar bone grafting is based on a variety of criteria, including growth, development, eruption patterns, arch form, root development and skeletal maturation. When such grafting is delayed until late in the mixed dentition stage, there are periodontal and restorative challenges because of the significant inadequacies of bone at the cleft alveolar site. Therefore, we advocate early secondary bone grafting of the alveolar cleft during the late primary or early mixed dentition stage, at about 5½ to 6½ years, depending on dental development.

Early secondary bone grafting improves the periodontal health of the erupting permanent dentition. Bone grafting during the late primary dentition stage has periodontal advantages over the current conventional approach of waiting until eruption of the permanent dentition. The incidence of failure and complications associated with this approach is very low.³

Whether the cleft is bilateral or unilateral, when appropriate grafting and soft-tissue procedures accomplish almost normal anatomic reconstruction of the defect, there is usually significant improvement in growth, function and aesthetics^{5,6} (Figs. 4a, 4b, 4c).

As stated above, grafting is usually performed when the maxillary permanent incisor teeth are visible in the mouth but not yet fully erupted. Simultaneous secondary functional cheilorhinoplasty is performed if there is muscular dysfunction of the lip, as determined by deviation of the nasal septum to the noncleft side, presence of vestibular oronasal fistulae or inability of the child to symmetrically project the lips. Total palatal reconstruction is carried out if there are palatal fistulae or distorted palatal anatomy resulting from inaccurate and inadequate primary surgery. Except



Figure 4a: Alveolar defect in a 5¹/₂-year-old child, before alveolar bone graft.



Figure 4c: Anterior view 12 years after functional surgical reconstruction of the cleft alveolus.

in the most severe cases, the bone graft procedure is carried out before orthodontic expansion, which is usually initiated about 8 to 12 weeks after grafting. This delay allows the arch expansion to be accomplished concomitant with a type of distraction osteogenesis, accompanied by appropriate palatal mucosa extension.

The palatal mucosa is reconstructed, from posterior to anterior, with interrupted mattress sutures to ensure optimal results. The reconstructed palatal mucosa is left as a single large palatal flap until nasal reconstruction is completed. From the vestibular side, the nasal mucosa is then reconstructed such that the height of the nasal floor is equal to or slightly superior to that on the noncleft side. This is a very important technical point: unless there is adequate space for insertion of bone between the palatal and nasal layers, the long-term quantity of bone stock will be insufficient. Part of the nasal mucosal suturing is carried out through the access created by delaying final suturing of the palate. Meticulous attention in suturing the nasal mucosa is important so that a smooth and even mucosal layer is apposed to what will be the roof of the bone graft.

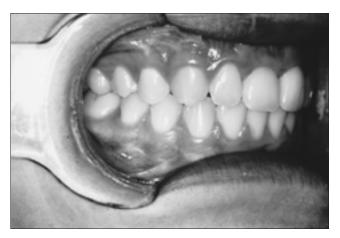


Figure 4b: Right lateral view 12 years after functional surgical reconstruction of the cleft alveolus.

A thin piece of cortical bone is fashioned to fit snugly in the nostril defect, dividing the graft from the nasal mucosa at a height slightly above the nostril floor on the noncleft side. The cortical side of this piece of bone is placed against the nasal mucosa, so that the medullary side faces the graft. Particulate marrow is packed into the defect with pressure. As much bone as possible is placed within the confines of the defect, with particular attention to the alveolar margin of the cleft in the region of the erupting teeth.

At the end of the operation, only palatal mucosa is present on the palate, only nasal mucosa forms the nasal floor, attached gingiva (not alveolar mucosa or any other foreign tissue) covers the teeth and the graft at the alveolar margin, and the maxillary vestibule is lined only by alveolar mucosa. This strict anatomic geography is critical to the long-term success of the operation. No splints are used, but the patient is given a soft diet for about 1 week after surgery.

Conclusion

Cleft lip, with or without cleft palate, occurs more frequently than cleft palate alone and is the most common of the significant orofacial anomalies.⁷ Clefts of the lip and anterior maxilla have been thought to result from a deficiency of mesenchyme in the facial region due to failure of either migration of the neural crest cells or proliferation of facial mesenchyme,⁸⁻¹⁰ but we agree with Mooney and others,¹¹ who have suggested that deficiency of the orbicularis oris muscle may be related more to perinatal functional dysmorphogenesis than to congenital mesenchymal deficiency.

Whether the overall deformity is due to true hypoplasia, hypofunction and attendant hypodevelopment, or a combination of the two, the principal surgical goal is the same: to establish good function through careful muscle reconstruction, which in turn will permit optimum subsequent growth and development of the facial skeleton.¹² This principle is important in both primary and secondary cleft corrections, because good function is a prerequisite for good facial esthetics. *

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