Chapter 83 – Cleft Lip and Palate: Comprehensive Treatment and Technique

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Comprehensive treatment of cleft lip and palate deformities requires thoughtful consideration of the anatomic complexities of the deformity and the delicate balance between intervention and growth. Comprehensive and coordinated care from infancy through adolescence is essential to achieve an ideal outcome. Surgeons with formal training and experience in all phases of care must be actively involved in planning and treatment.[1,2] Specific goals of surgical care for children born with a cleft lip and palate include:

- Normalized aesthetic appearance of the lip and nose
- Intact primary and secondary palates
- Normalized speech, language, and hearing
- Nasal airway patency
- Class I occlusion with normal masticatory function
- Good dental and periodontal health
- Normal psychosocial development

What makes these goals challenging in some children is the wide spectrum of cleft lip, palate, and nasal deformities. The range of manifestations from one affected child to another is highly variable. An asymptomatic bifid uvula is an example of a clinically inconsequential cleft. A relatively simple deformity may include a submucous cleft palate that causes speech abnormality after an uneventful adenoidectomy. An occult cleft lip with an intact philtral ridge and symmetrical Cupid's bow and an otherwise normal-looking child with a small notch in the alveolar ridge are other such examples of minimal deformities. Unfortunately, the other end of the spectrum includes wide bilateral clefts of the lip and palate or facial dysplasia with several associated anomalies (Fig. 83-1).

A cleft surgeon has the tasks of addressing anatomic and functional deficiencies at the initial evaluation and planning for future functional and aesthetic needs. Success in these endeavors is dependent on many factors, including the severity of disruption of bone, cartilage, and soft tissue. Surgical skill and technique matched to the nature and severity of the deformity are the fundamental factors in obtaining superior results. Attention to all aspects of care, such as anatomic variants, the biology of scar formation, parent education, and cooperation, is very important in determining success. A healthy and collaborative team environment is essential in achieving these goals.

This chapter presents a technical overview of the reconstruction of cleft lip and palate deformities. Surgical reconstruction of clefts requires that surgeons undertaking this important work maintain a cognitive understanding of the complex malformation itself, the varied operative techniques used, facial growth considerations, and the psychosocial health of the patient and family. The objectives of this chapter are to present the overall staged reconstructive approach for repair of cleft lip and palate from infancy through the time of skeletal maturity, as well as a focused discussion of the specific surgical procedures involved in primary repair of clefts of the lip and palate.

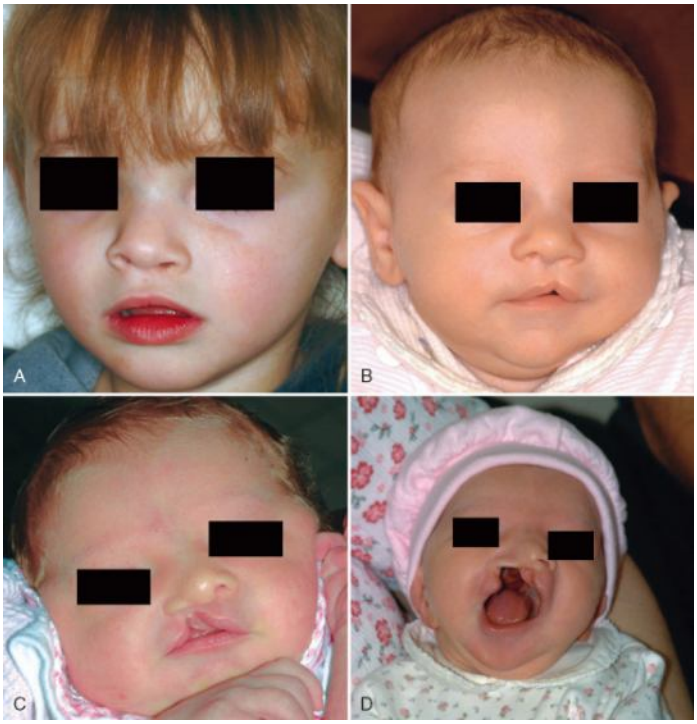

Figure 83-1 Because cleft lips come in a variety of configurations, each repair must be individually customized to establish the most normal morphology. **A,** Microform left unilateral cleft lip only, not requiring repair. **B,** Minor left incomplete unilateral cleft lip only. **C,** Right incomplete unilateral cleft lip and palate with a Simonart band. **D,** Wide right complete unilateral cleft lip and palate.

CLASSIFICATION OF CLEFTS

Classification of clefts is necessary to describe the deformities, to study causative factors, and to compare results of treatment undertaken. Several classification systems have been described.[2–5] Simple classification systems are valuable for everyday use and are easy to understand; they provide broad groups with large numbers of patients in each group for research and analysis. However, simple classification systems fail to detail the variations in severity of cleft manifestations. As a consequence, there has been a tendency to use more complex classification systems, which are more difficult to use clinically but provide better information for research and outcomes analysis.

There is infinite variation in the configuration of clefts, and a compromise between simple and complex classification systems has to be reached. Regardless of complexity, all classification systems use basic nasal (ala, dome, columella, nostril sill, septum, vomer), labial (vermilion, philtrum, Cupid's bow, Lister's tubercle, red and white lines, prolabium), and palatal (incisive foramen, primary and secondary palate, hard and soft palate, alveolar process, attachment to septum) landmarks for reporting.

The LAHSHAL system is a basic and useful classification that allows easy compilation of data. Starting from the patient's right side, the cleft status is recorded as C(omplete), I(ncomplete), or X (absent) for the L(ip), A(lveolus), H(ard palate), S(oft palate), H(ard palate), A(lveolus), and L(ip) (Table 83-1). Craniofacial clefting may be described with Tessier's orbitocentric system of numbering (Fig. 83-2).

Table 83-1 -- CLASSIFICATION OF CLEFTS

Right						Left
L	A	H	S	H	A	L
I	C	C	C	X	X	X

This example is for a unilateral right incomplete cleft lip (I) with a complete cleft alveolus (A) and hard (H) and soft (S) palates. The vomer is attached to the left side of the hard palate. The alveolus and lip on the left side are intact.

C, complete; I, incomplete; X, no cleft.

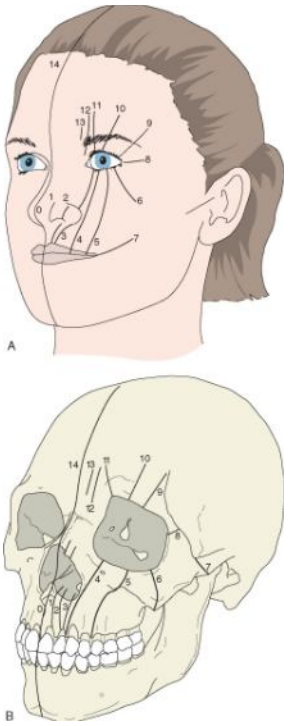

Figure 83-2 **A** and **B,** Complex facial clefts can be classified according to Tessier's orbitocentric system of numbering. Clefts may involve all tissue planes, including the skin, mucosa, bone, teeth, muscle, brain, peripheral nerve, and other specialized structures.

INTERDISCIPLINARY CARE

Successful management of the child born with a cleft lip and palate requires coordinated care provided by individuals from a number of different specialties, including otolaryngology, oral and maxillofacial surgery, plastic surgery, genetics/dysmorphology, speech/language pathology, orthodontics, prosthodontics, and others.[1,2,6] Because care is provided over the entire course of the child's development, long-term follow-up is critical to ensure complete and comprehensive care.

Rehabilitation of a patient with a cleft is best performed as an interdisciplinary effort. Most children with clefts have multiple interrelated issues, and several team models are available to address these issues in an interdisciplinary fashion. Key members of the team are the cleft surgeon or surgeons, orthodontist, and speech-language pathologist. Cleft surgeons may be trained plastic surgeons, oral and maxillofacial surgeons, or otolaryngologists. Other key persons may include the pediatric dentist, geneticist/dysmorphologist, pediatrician, psychologist, and social worker. The team director may or may not be a cleft surgeon but should be a nonbiased participant in care. The formation of interdisciplinary cleft teams serves two key objectives of successful cleft care: (1) coordinated care provided by all the necessary disciplines and (2) continuity of care with close interval follow-up of the patient through-out periods of active growth and ongoing stages of reconstruction.

In a carousel team, patients are seen on the same day by all specialists. Comprehensive evaluation is not performed, at the time of decision making information may be incomplete, and one practitioner's treatment decision may not reflect the complete status of the patient until all the information can be reviewed by everyone involved. In a triage team, one gatekeeper organizes outpatient referrals to specialists through multiple visits. This approach is often costly and requires a team meeting of all the practitioners to provide comprehensive care.

Multidisciplinary teams have many specialists, but they may not necessarily be cooperative or interactive. The best organization of care occurs when multidisciplinary teams are also interdisciplinary. Interdisciplinary teams work together to come up with the best treatment plan for a given patient. In this treatment model, decisions are based on a collaborative process, and each member is cognizant of other members' treatment priorities.

PRENATAL DIAGNOSIS

Team involvement ideally starts in the prenatal period.[1,2,7,8] Prenatal diagnosis by ultrasound presents an opportunity to introduce team members and their roles early in the process and provides reassurance, familiarity, and a structured approach for the various issues, including counseling, treatment, and feeding approaches. Prenatal sonographic detection of clefts is often dependent on cleft type and severity. When a cleft is present, the overall detection rate is approximately 65% (isolated cleft palate, 22%; isolated cleft lip, 67%; cleft lip and palate, 93%).[1]

Additional testing may be warranted to evaluate the possibility of associated deformities, syndromes, and sequences that could affect the birthing process. Exceptionally skilled ultrasonographers can visualize airway development and other abnormalities that may require early intervention with fetal surgery, ex utero intrapartum procedures, extracorporeal membrane oxygenation, or surgical airway management (tracheotomy) at the time of delivery. Evaluations of any associated malformations are completed via three-dimensional ultrasound, magnetic resonance imaging, and other genetic evaluations. These detailed evaluations can be very helpful in understanding the nature of various deformities, the likelihood of an uneventful birth, and the need for additional evaluation and treatment.

SEQUENCING OF PROCEDURES

Because of many different treatment philosophies, the timing of treatment interventions varies from one cleft center to another. Therefore, it is difficult to produce a timing regimen that everyone agrees on.[1,2,9–11] A sample timeline of staged reconstruction of cleft lip and palate deformities is presented in Table 83-2. This is a general timeline and requires an individualized approach based on aesthetic and functional priorities.

Table 83-2 -- TIMELINE FOR STAGED RECONSTRUCTION OF CLEFT LIP AND PALATE DEFORMITIES

Procedure	Timing
Airway evaluation and intervention	After initial assessment when necessary
Presurgical orthopedics, nasoalveolar molding, lip adhesion	Before lip repair in select cases
Cleft lip/nasal repair	After 10 wk
Cleft palate repair	9-18 mo
Myringotomy and tubes	At the time of lip or palate repair, depending on the presence of middle ear effusion and hearing status
Pharyngeal flap or pharyngoplasty	3-5 yr or later, depending on speech development
Maxillary/alveolar reconstruction with bone grafting	6-9 yr, depending on dental development
Cleft orthognathic surgery	14-16 yr in girls, 16-18 yr in boys
Cleft septorhinoplasty	After 5 yr of age but preferably at skeletal maturity after orthognathic surgery when possible
Cleft lip revision	Anytime after initial remodeling and scar maturation, but best after 5 yr of age

The timing of cleft lip and palate repair is controversial. Despite a number of meaningful advancements in the care of patients with cleft lip and palate, there is a lack of consensus regarding the timing and specific techniques used during each stage of cleft reconstruction. Surgeons must continue to carefully balance the functional needs, aesthetic concerns, and the issue of ongoing growth when deciding how and when to intervene. In no other type of deformity is the effect of early surgery on growth more apparent than in the treatment of cleft lip and palate deformities. The decision to surgically manipulate the tissues of a growing child should not be made lightly and should take into account the possible growth restriction that can occur with early surgery. Nevertheless, many patients with congenital deformities will benefit from early surgical intervention for functional or psychosocial reasons. Understanding the growth and development of the craniofacial skeleton is essential to the treatment planning process.[1,2]

MANAGEMENT OF AIRWAY OBSTRUCTION IN PATIENTS WITH CLEFTS

Features of the airway depend on the type of cleft and its severity. An intact palate provides tongue support and prevents glossoptosis. Lacking this support, a child with a wide cleft palate will have obstruction of the upper airway of varying severity. This obstruction is pronounced in children with associated syndromes and the Robin sequence in which mandibular growth is compromised. Prone positioning, feeding adaptation, and time may resolve the mild obstruction. In some cases a nasal airway can provide relief for a few weeks to months. Airway evaluation via flexible and rigid endoscopy is warranted to determine the extent of airway obstruction, as well as additional airway compromise (i.e., choanal atresia, laryngotracheomalacia, subglottic stenosis). In severe cases of obstruction, surgical options include glossopexy, mandibular distraction osteogenesis, and tracheotomy. Difficult intubation should be anticipated, and additional equipment such as a Bullard laryngoscope, laryngeal mask airway, or flexible endoscopic intubation devices should be available.

PRESURGICAL ORTHOPEDICS

Presurgical orthopedic (PSO) devices are used to mechanically manipulate the position of the alveolar segments before definitive lip and nasal repair. A custom acrylic prosthesis is used to mold the lip and maxillary dental arch. As the greater segment is pushed into position, space is cut away within the palatal plate to allow the lesser segment to drift into the proper arch form. When the lip has an incomplete cleft, the alveolar segments are usually molded into a reasonable arch form, and maxillary arch orthopedic devices are not necessary.

Nasoalveolar molding is a technique that combines maxillary arch orthopedics with a PSO device and lip and nasal contour shaping via nasal-vestibular projec- tion (Fig. 83-3).[12–17] The importance of this technique is emphasized by surgeons who advocate primary nasal repair at the time of lip repair for children with wide clefts and severe nasal deformities, especially those with a very short columella. Successful use of PSO devices requires a dedicated pediatric dentist or orthodontist, a good laboratory, and diligent parents. Noncompliance can be a problem, so for this and other reasons, some centers rely on pin-retained prostheses. However, these appliances have been associated with significant midface growth restriction.[17]

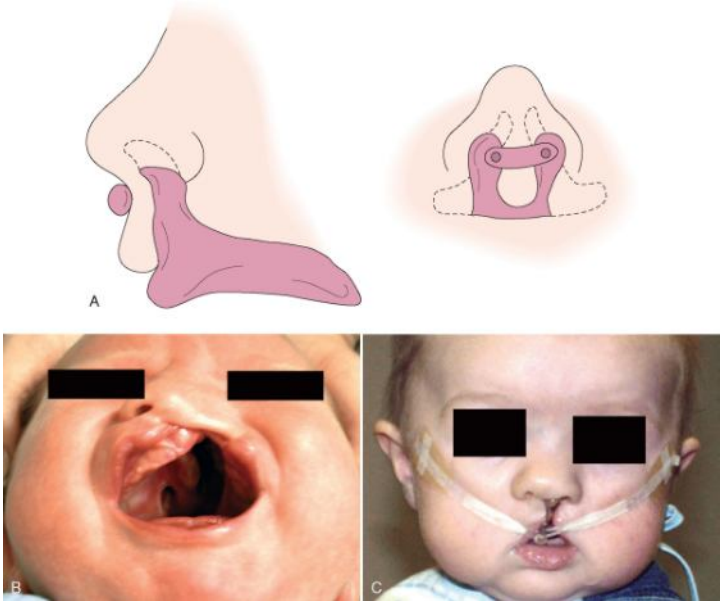

Figure 83-3 **A,** Frontal and lateral views of the Grayson nasoalveolar molding (NAM) appliance showing the nasal projections that help mold the nasal cartilage and maxillary segments into a more appropriate configuration before repair. **B,** Segments before the use of NAM. **C**, Segments after NAM but before closure.

Some surgeons perform a surgical lip adhesion procedure at about 3 months to mold the alveolar segments into better position before definitive repair. Effective PSO devices may obviate the need for a lip adhesion procedure and the additional anesthetic required for it.[18] Although initial results are reportedly good, use of these devices and their efficacy are still controversial because long-term outcome studies showing improved aesthetics, higher retention of dentition, and better dental arch form are still not available.[1,2,14–16,19]

LIP ADHESION

In exceptionally wide unilateral or bilateral clefts or in extremely asymmetrical bilateral clefts, it may be helpful to approximate the segments of the cleft lip before definitive lip repair to achieve a better relationship of both the lip structures and the dental arches. In this technique, which is used by some surgeons, small flaps of tissue are advanced across the cleft site (Fig. 83-4). Care is taken to not disturb natural landmarks, and a through-and-through horizontal tension suture is placed. When used, lip adhesion is usually completed at 3 months of age. The definitive lip repair is then completed 3 to 6 months later by excising the scar and reapproximating the remaining lip structures.

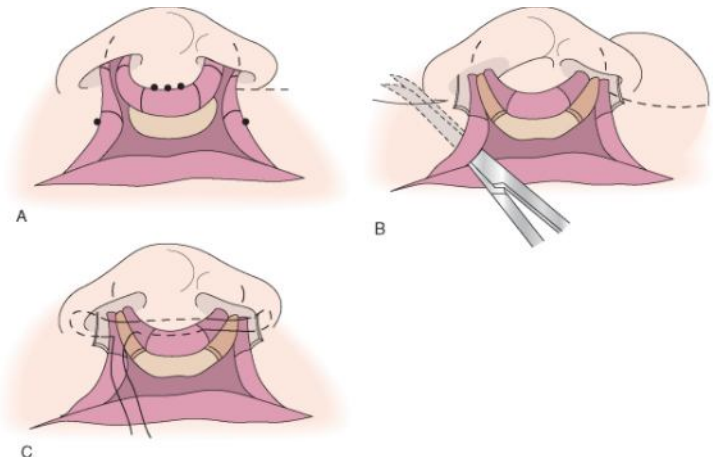

Figure 83-4 Example of the lip adhesion technique used by some surgeons as a first stage in closing wide clefts. **A,** Flaps are designed to bridge the defect. **B,** Undermining allows approximation. **C,** A retention suture may be used before closure for additional support.
(From Senders CW: Presurgical orthopedics. Facial Plast Surg Clinics North Am 4:333-342, 1996.)

PRIMARY UNILATERAL CLEFT LIP REPAIR

Unilateral clefts of the lip and nose have a high degree of variability, and thus each repair design is unique (see Fig. 83-1). One repair technique preferred by the authors for cleft lip and nasal deformities is shown in Figurs 83-5 and 83-6 and is usually performed after 10 weeks of age. The goal of the repair is to create a three-layered closure of skin, muscle, and mucosa in which hypoplastic tissue at the cleft margins is excised and normal tissues are approximated. Critical in the process is reconstruction of the orbicularis oris musculature into a continuous, functional sphincter. The Millard rotation-advancement technique has the advantage of allowing each of the incision lines to fall within the natural contours of the lip and nose.[1,2,20,21] This is an advantage because it is difficult to achieve “mirror image” symmetry in a unilateral cleft lip and nose with the normal side immediately adjacent to the surgical site. A Z-plasty technique such as the Randall-Tennison repair may not achieve this level of symmetry because the Z-shaped scar is directly adjacent to the linear nonclefted philtrum. Achieving symmetry is more difficult when the rotation portion of the cleft is quite short in comparison with the advancement segment. There is a great degree of variability in repair techniques from surgeon to surgeon. Several techniques are

discussed as examples of unilateral and bilateral lip repair.

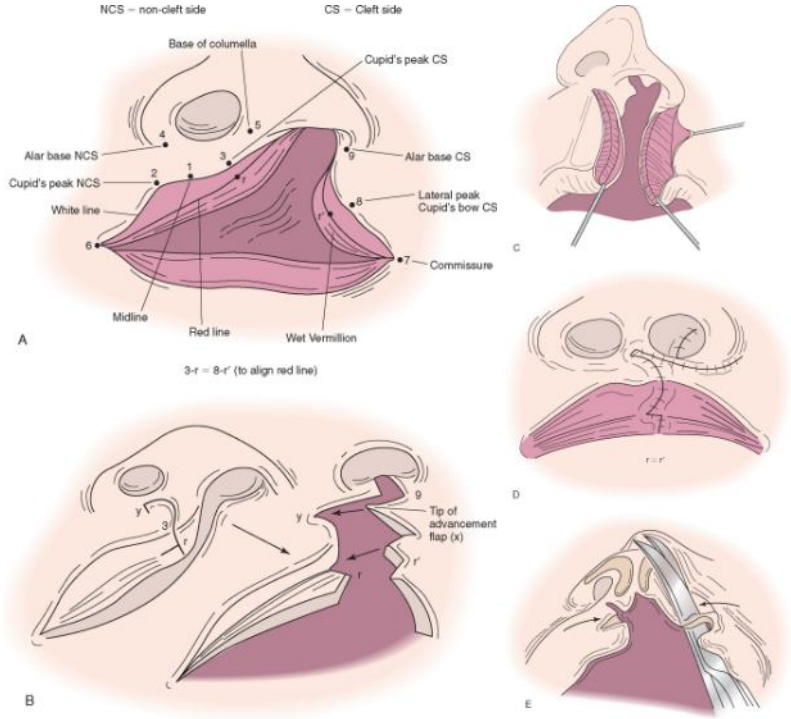

Figure 83-5 **A,** Key landmarks of a unilateral cleft lip repair are marked with a fine marking instrument. **B,** Hypoplastic tissue is excised by following the key landmarks, and a variety of backcuts can be made to address symmetry. **C,** A three-layered closure is achieved by first reconstructing the oral mucosa and nasal floor and then the orbicularis oris musculature. The skin flaps should be passively closed without tension. **D,** The rotation and advancement flaps are inset for optimized symmetry in all three dimensions. **E,** Limited nasal dissection can be used to reposition the lower lateral cartilage, as well as the lateral alar tissue. These tissues must be released from their abnormal insertions and repositioned for better symmetry. A nasal bolster or silicone nasal formers may be placed at the end of the procedure.

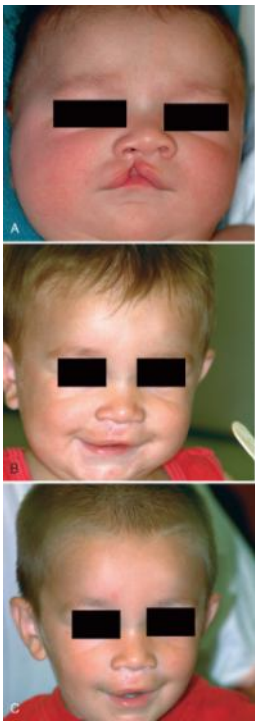

Figure 83-6 **A,** Three-month-old child with a right-sided incomplete unilateral cleft lip. Note the short philtrum near the midline, which must be rotated downward to avoid notching and to improve symmetry. **B,** Nine-month-old boy after rotation-advancement repair of his cleft lip and nasal deformities. **C,** The same child in **B** 2½ years after his cleft lip and nasal repairs.

All landmarks are carefully marked with the use of operating loupes and surgical calipers (Fig. 83-5A). Marking starts with identification of the low point (midline) (1) and the peak (2) of Cupid's bow on the noncleft side (NCS). The distance between these two points is used to determine the position of the peak of Cupid's bow on the cleft side (CS) (3). The alar base on the NCS (4), the columellar base (5), and the commissures (6 and 7) are marked. The lateral peak of Cupid's bow (8) is marked by taking the distance from the CS commissure and the width of the vermilion into account. The combined vertical height of the wet and dry vermilion at point 8 should match the vertical height of the vermilion at point 3. Therefore, 8 can be placed within 1 to 2 mm of the measured distance (matching the distance between 2 and 6) from the commissure to match the vermilion in height.

The tip of the advancement flap (9) is marked so that the distance between points 8 and 9 matches the distance between 2 and 5 (see Fig. 83-5A). The surgeon must avoid discolored, hypoplastic skin with increased vascular patterns when designing the flap. The slightest difference in color here will be highly visible against the contrast provided by the NCS lip and columella skin.

The rotation incision is followed by a very small (<1 mm) releasing cut (y) made high in the lip in a near-perpendicular angle to the rotation incision (see Fig. 83-5B). This should allow the NCS to drop down without tension and create a symmetrical prolabium with minimal transgression of the upper philtral column.

The orbicularis oris is dissected from the skin along the rotation and advancement flaps (see Fig. 83-5C). The dissection is performed to facilitate layered closure. If feasible, the muscle may be interdigitated. The mucosal closure is started in the sulcus and extended toward the wet vermilion with 4-0 chromic or polyglycolic acid suture. The skin is closed with 5-0 or 6-0 nylon or with absorbable suture (see Fig. 83-5D).

Primary nasal reconstruction may be considered at the time of lip repair to reposition the displaced lower lateral cartilage and alar tissue. Several techniques have been advocated, and considerable variation exists with respect to the exact nasal reconstruction performed by each surgeon.[1,2,12,22] Primary nasal repair may be achieved by releasing and reshaping the lower lateral

cartilage and alar base and augmenting the area with allogeneic subdermal grafts or even formal open rhinoplasty. Because lip repair is done at such an early point in growth and development, the authors prefer minimal surgical dissection because of the effects of scarring on subsequent growth of these tissues. McComb described a technique that has become popular in which the lower lateral cartilage is dissected free from the alar base and the surrounding attachments through the incision at the alar crease.[22]

The lower lateral cartilage is dissected free of skin (including that between the medial crura and dome) (see Fig. 83-5E) and suspended to the ipsilateral upper lateral cartilage with a looping suture (see Fig. 83-1D). If necessary, interdomal and suspension sutures (see Fig. 83-1E) may be used to reposition and reorient the nasal tip. This modification requires extended dissection. The dislocated nasal septum is released, transposed, and secured through the membranous septal incision. The nasal ala is then repositioned to address symmetry in all three dimensions. In most complete unilateral clefts, the alar base has to be dissociated from the lateral aspect of the lip and piriform aperture by extending the circumalar incision to allow these elements to move independently. The advancement flap will be advanced more than the ala. The tip of the advancement flap may then be anchored to the membranous septum. The nostril may be stented for several weeks or longer with silicone nostril retainers or bolsters to prevent distortion in nostril shape.

Parents may feed their child immediately by the same methods that were used preoperatively. Arm restraints are routinely used to protect the lip from trauma for approximately 2 weeks. Antibiotic ointment may be applied in the first few days, followed by gentle massage several times per day starting 2 weeks after repair to encourage remodeling of the scar.

PRIMARY BILATERAL LIP REPAIR

Bilateral cleft lip repair can be one of the most challenging technical procedures performed. The lack of quality tissue and the widely displaced segments are major challenges to achieving exceptional results, but superior technique and adequate mobilization of the tissue flaps generally yield excellent aesthetic results (Figs. 83-7 to 83-9). Additionally, the columella may be quite short in length, and the premaxillary segment may be significantly rotated. Adequate mobilization of the segments and attention to the details of using only appropriately developed tissue will yield excellent results, even in individuals with significant asymmetry.

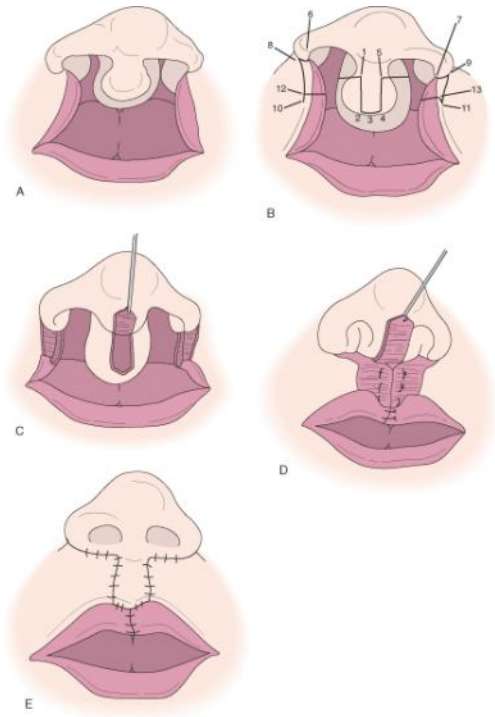

Figure 83-7 **A,** The bilateral cleft of the lip and maxilla shown here is complete and highlights the nature of hypoplastic tissue along the cleft edges. The importance of the nasal deformity is evident in the shorter columella and disrupted nasal complexes. **B,** Markings of the authors' preferred repair are shown with an emphasis on excision of hypoplastic tissue and approximating more normal tissue with the advancement flaps. Measurements should be roughly equal in the following fashion: 1 to 2 = 6 to 10; 5 to 4 = 7 to 11; 1 to 2 = 5 to 4; and 6 to 10 = 7 to 11. **C,** A new philtrum is created by excising the lateral hypoplastic tissue and elevating the philtrum superiorly. Additionally, the lateral advancement flaps are dissected into three distinct layers (skin, muscle, and mucosa). Nasal floor reconstruction also occurs in conjunction with these advancement flaps. **D,** The orbicularis oris musculature is approximated in the midline with multiple interrupted or mattress sutures, or both. This is a critical step in total reconstruction of a functional lip. There is no musculature present in the premaxillary segment, and it must be brought to the midline from each lateral advancement flap. The nasal floor flaps are sutured at this time as well. The new vermilion border is reconstructed in the midline with good white roll tissue advanced from the lateral flaps. **E,** Final approximation of the skin and mucosal tissues is performed in a manner that leaves the healing incision lines in the natural contours of the lip and nose.

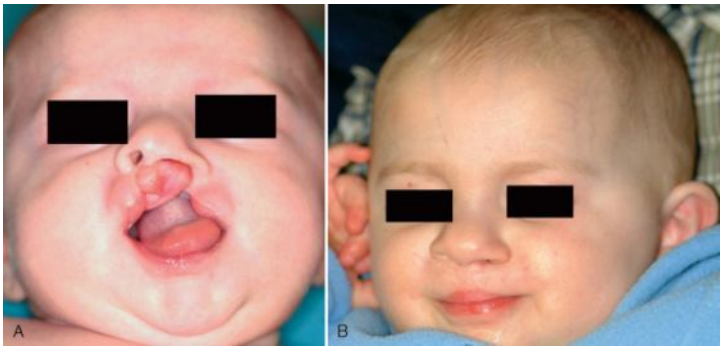

Figure 83-8 **A,** Presurgical appearance of a bilateral cleft lip and palate with impressive asymmetry and rotation of the premaxillary segment. Note the significant nasal asymmetry and bunching of the orbicularis oris laterally. **B,** The same child at 14 months of age. No presurgical orthopedic appliances were used.

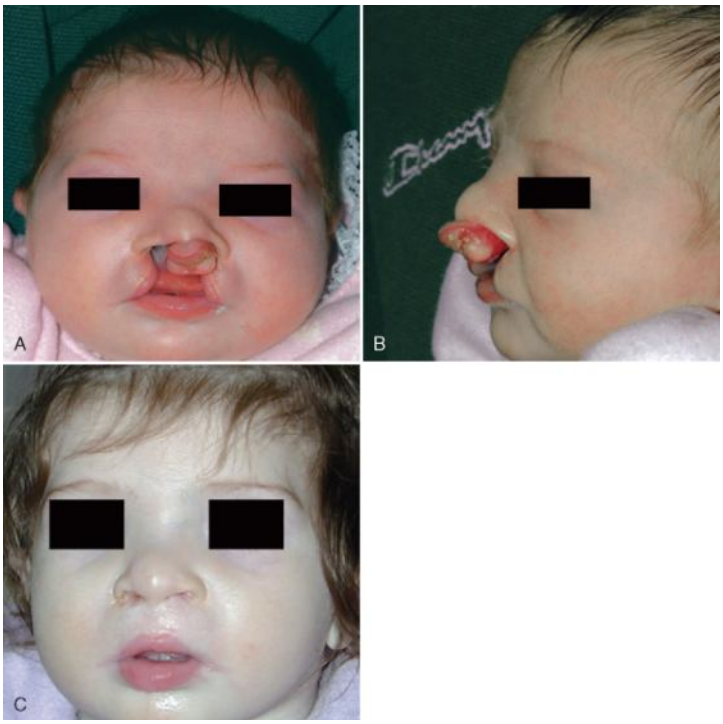

Figure 83-9 **A,** Presurgical frontal view of a wide bilateral cleft lip and palate with significant asymmetry and lack of columella length. **B,** Presurgical left lateral view of a wide bilateral cleft lip and palate with a protrusive premaxillary segment. Note the short length of the columella. **C,** The same child after repair of her bilateral cleft lip and palate. No presurgical taping or orthopedic appliances were used.

Although often more challenging than most unilateral repairs, the technique for bilateral cleft lip and nose reconstruction is similar in concept to that for unilateral repair. Markings are made with a fine marking instrument at key landmarks while being sure to not include any hypoplastic tissue from within the cleft (Fig. 83-7). Points 12 and 13 should be made at the beginning of the good white roll and vermilion, with a small backcut extending laterally to allow advancement under the prolabial tissue. Points 8 to 12 and 7 to 13 should be equal in length, and the measurement at 2 to 4 should be no longer than 3 mm. This tissue will stretch significantly later as remodeling of the lip occurs. Incisions within the nares should be made carefully so that one does not excise too much tissue and make the nares quite small. Backcuts around the alar base should be long enough to allow nasal dissection, release of alar tissue from the piriform rim, and advancement medially to reconstruct the nares.

Some surgeons have used operative techniques involving banked fork flaps to surgically lengthen the columella and preserve hypoplastic tissue. Early and aggressive tissue flaps in the nostril and columella areas do not look natural after significant growth has occurred and result in abnormal tissue contours. Although surgical attempts at lengthening the columella may look good initially, they frequently appear abnormally long and excessively angular later in life. Revision of these iatrogenic deformities is difficult, and some of the contour irregularities will not be able to be revised adequately. Usually, if the hypoplastic tissue is excised and incisions within the medial nasal base and columella are avoided, the long-term aesthetic results are excellent.

The authors prefer a primary nasal reconstruction that can be performed in a fashion similar to the unilateral technique described by McComb.[1,2,22] Other open rhinoplasty techniques involving either a direct incision on the nasal tip or prolabial unwinding techniques have been suggested.[12] As with most early maneuvers, aggressive rhinoplasty at this time may result in early scarring that affects the growth potential of the surrounding tissues and makes revision more difficult and long-term aesthetics less than ideal.

CLEFT LIP REVISION

Despite even the most superb techniques, many patients who undergo cleft lip repair will benefit from at least a minor revision at some point in life. Although revision procedures are often viewed as optional phases of cleft lip reconstruction, surgeons must advise families of this likelihood. The hard and soft tissues of the maxillofacial complex grow and change as a child grows, and the repaired lip is affected. Bilateral clefts will benefit from lip revision more often than unilateral clefts will. The majority of lip growth is complete after the age of 5 years, and this may be the best time to consider revision of the lip because the psychosocial benefits before entering school may be considerable. Alternatively, revision of the lip can be delayed until the teenage years, when most maxillofacial growth is complete. It is preferable to wait until orthognathic surgery is completed (if this becomes necessary) because these procedures will considerably change the contour and shape of the nose and lips.

The surgical objectives of cleft lip revision include excision of residual scar, reapproximation of key anatomic landmarks such as the vermilion-cutaneous junction and vermilion-mucosal junction, and leveling of vertical lip lengths (philtral columns). Repair of the orbicularis oris muscle as a distinct layer is critical to an acceptable outcome. Although small scar revisions may be considered in some patients, many patients will benefit from a revision that completely reconstructs the area by excision of the scar, dissection of the tissues into three layers (skin, muscle, and mucosa), and a reconstruction that achieves improved symmetry and form. As with the primary repair, 6-0 or smaller sutures may be used to minimize stitch marks. Postoperative care includes careful early wound care and avoidance of extended sun exposure, similar to the instructions for primary repairs.

PALATOPLASTY

There are two main goals of cleft palate repair during infancy: (1) closure of the oral-nasal communication involving the embryologic secondary palate and (2) anatomic repair of the musculature within the soft palate, which is important for normal production of speech. The soft palate, or velum, is part of the complex coupling and decoupling of the oral and nasal cavities involved in the production of speech. When a cleft of the soft palate is present, abnormal muscle insertions are located at the posterior edge of the hard palate. Surgery must not simply be aimed at closing the palatal defect, but rather at release of abnormal muscle insertions to create muscle continuity with improved orientation so that the velum may serve as a dynamic structure. Despite successful repair of the palate, a significant number of children who undergo cleft palate repair will still require speech therapy and have difficulty closing the velum for a variety of reasons.

Several techniques of palatoplasty with a substantial number of modifications may be found in the literature. Three popular techniques are discussed, including:

- Two-flap palatoplasty with intravelar veloplasty
- V-Y pushback with intravelar veloplasty
- Furlow palatoplasty

Before each procedure, the type and severity of the cleft, position of the segments, degree of septum deviation, width of the cleft, length and symmetry of the soft palate, and mobility of the soft palate and pharyngeal walls have to be considered. It is helpful to measure and record palatal width, length, and other cleft dimensions with calipers.

The patient is placed supine with the neck slightly extended, and a shoulder roll is used. A cleft mouth retractor is used to maximally open the mouth and at the same time secure the endotracheal tube in a midline position against the tongue. A small throat pack may be fitted in the hypopharynx around the tube. Incisions are marked with a marking pen, gentian violet, or methylene blue. Lidocaine with epinephrine solution is injected into the submucosal tissue and subperiosteal plane and around the palatine foramen. Subperiosteal injections can facilitate undermining of the mucoperiosteal flaps and improve hemostasis. The surgeon should avoid injecting into the neurovascular bundle. If vomer flaps are planned, the vomer is injected.

A variety of blades, including nos. 11, 15, and Beaver 6300 blades, are used to dissect the flaps. A small half-circle cutter is helpful when the cleft space is narrow. The PS 5 is a fine all-purpose ⅜-inch cutter. The PS 4 cutter takes larger bites when required. In careful hands, a cutting needle may be less traumatic than a taper needle. However, if there is any tension on the wound, tapered needles may be used to avoid mucosal damage.

Two-Flap Palatoplasty with Intravelar Veloplasty

In the authors' practice this repair is the most appropriate technique for a complete unilateral cleft palate repair (Fig. 83-10).[1,2,23,24] The nasal flaps may be designed so that a strip of mucoperiosteum is left on the medial edge of the hard palate. The wider the cleft, the wider the strip of mucoperiosteum left on the medial edge of flap. The width and angle of the palatal shelves are also considered when designing the incisions. When these flaps are elevated and mobilized to reconstruct the nasal mucosal layer, they should meet each other without tension. On the soft palate, the incisions should follow the subtle transition line between the nasal and oral mucosa into the tip of the uvula. In a wide cleft (gaps larger than 15 mm), vomer flaps may be designed to aid in closure centrally.

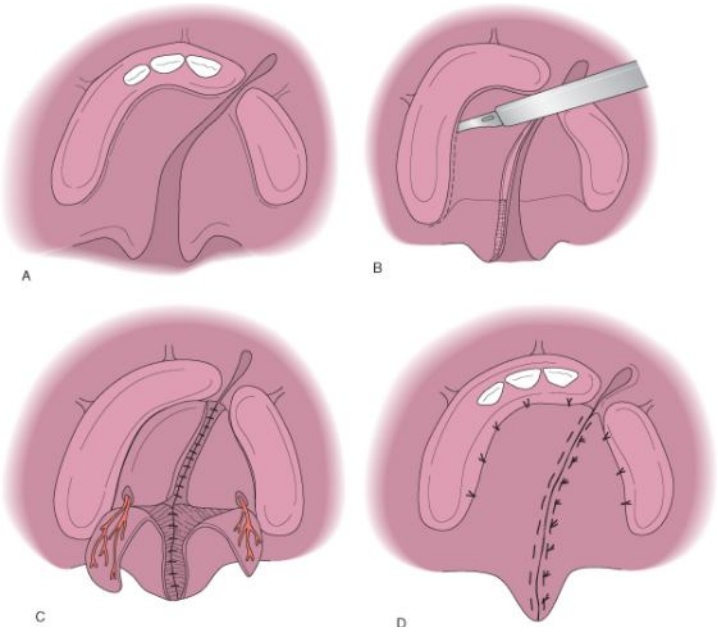

Figure 83-10 **A,** A unilateral cleft of the primary and secondary palates is shown with typical involvement from the anterior vestibule to the uvula. **B,** The Bardach palatoplasty technique requires two large full-thickness mucoperiosteal flaps to be elevated from each palatal shelf. The anterior portion (anterior to the incisive foramen) of the cleft is not reconstructed until the mixed-dentition stage. **C,** A layered closure is performed in the Bardach palatoplasty by reapproximating the nasal mucosa. The muscle bellies of the levator palatini are elevated off their abnormal insertions on the posterior palate. They are then reapproximated in the midline to create a dynamic functional sling for speech purposes. **D,** Once the nasal mucosa and musculature of the soft palate are approximated, the oral mucosa is closed in the midline. The lateral releasing incisions are quite easily closed primarily because of the length gained from the depth of the palate. In rare cases, with very wide clefts, a portion of the lateral incisions may remain open and granulate by secondary intention.

Laterally, the flaps are designed to include oral mucoperiosteum only (see Fig. 83-10B). Gingiva should be avoided. The incision line moves laterally at the junction of the alveolus, and the palatal shelf recovers its posterior direction quickly once the lateral aspect of the alveolus is reached and may continue 0.5 to 1 cm into the soft palate.

The oral mucosa is separated from the nasal mucosa toward the tip of the uvula with a superficial incision. Mucoperiosteal incisions on the hard palate have to be firm, perpendicular to the palatal plane, and should reach the palatal shelf with a continuous clean cut. Hemostasis with a fine needle tip electrocautery or bipolar electrocautery at low setting may be performed.

Elevation of mucoperiosteum is started anterolaterally with a Woodson or other small elevator. Gentle subperiosteal elevation of the tip of the flap is performed. After elevation of the tip, a Freer elevator is used to continue subperiosteal elevation posteriorly to visualize and elevate tissue around the neurovascular bundle. All muscular and tendinous attachments to the hard palate shelf are detached with a combination of sharp and blunt dissection while leaving the flap attached to the hard palate by the neurovascular bundle and lateral pedicle.

The soft palate is transposed medially by entering the space of Ernst with a blunt instrument (Metzenbaum scissors). To obtain more medial mobilization, infracture of the hamulus may be performed but is rarely necessary. Veau's muscle inserts into the periosteum on the posterior edge of the hard palate in concert with the tensor aponeurosis after it rounds the hamulus. (Veau's muscle is a clefted muscle consisting of the levator and the palatopharyngeus.) Some surgeons divide the tendon to mobilize muscles toward the medial aspect.

Gentle traction is used to pull the neurovascular bundle out of the greater palatine foramen (see Fig. 83-10C); the palatine foramen is surrounded by a periosteal cone that can be incised superficially on either side and posteriorly to allow maximal mobilization of the flap. Additional dissection is necessary to provide maximal mobility of the flap.

Complete dissection of Veau's muscle from the hard palate and nasal mucoperiosteum follows. Attachments are stripped from the nasal mucoperiosteum with sharp scissors and scalpel. Precise dissection allows lengthening of the palate and posterior/medial transfer of muscles to create a functional muscle sling. This reconstruction is termed an intravelar velarplasty. Dissection of muscle from the nasal and oral mucosa is more extensive than described in traditional two-flap palatoplasty techniques. Complete dissection with repositioning and tightening of the palatopharyngeus and levator in a posterior position is, for some, theoretically more physiologic.

Once the flaps are mobilized, closure of the nasal layer begins anteriorly and should be tension free (see Fig. 83-10C). Sutures are tied in knots on the nasal surface. The difference in length between the two flaps is considered and spacing of the sutures adjusted at this stage. There should be no tension at the hard palate–soft palate junction. The soft palate muscles are retropositioned (overlapped if necessary) and sutured together with two or three mattress sutures (3-0 to 4-0 resorbable suture). The oral layer is then closed with single resorbable 4-0 or 5-0 interrupted or mattress sutures and knots tied in the oral surface (see Fig. 83-10D).

The combined flaps are anchored (tagged) to the nasal mucoperiosteal closure with a 4-0 resorbable suture at about the midpoint, and the tip is secured to the alveolus with two to four sutures. These steps prevent dead space between the oral and nasal layers and stabilize the flaps. The same objective can be achieved by using three to four vertical mattress sutures through both the oral and nasal layers. Exposed bone laterally may be filled with microfibrillar collagen. A tongue suture (3-0 silk/nylon) is applied to aid in tongue advancement acutely in the rare instance of postoperative obstruction. Surgeons should minimize the time that the mouth gag retractor is activated during the procedure to avoid more severe swelling.

V-Y Pushback with Intravelar Veloplasty

For many surgeons, the V-Y pushback operation is the most appropriate technique for repair of a posterior (soft palate) cleft with limited extension into the hard palate (e.g., Robin-type cleft with unilateral or bilateral incomplete hard palate involvement). Design principles are similar to those for two-flap palatoplasty but include a V to Y incision design on the hard palate for theoretical retropositioning of the soft palate. Mucoperiosteum over the primary palate in front of the incisive foramen is left intact, and flaps are designed to leave this area in a V form. For closure, the flaps slide posteriorly to close the gap, and transposition of this junction into a Y form occurs. (This posterior displacement is limited to the tip of the mucoperiosteal flaps, is minimal, and should not be confused with what is frequently called a push-back palatoplasty in which posterior displacement aims to reposition the velar muscles.) Intravelar veloplasty and the remaining repair are identical to those used for the two-flap technique. Generally, a vomer flap is not necessary. The flaps are secured to the anterior mucoperiosteum with several sutures. This technique is not a good choice for a wide bilateral cleft that involves the incisive foramen area because a residual fistula will remain that is often difficult to close and may affect speech.

Furlow Palatoplasty

The Furlow technique closes the palate by mirror image Z-plasties of the oral and nasal sides of the soft palate (Fig. 83-11A to D).[1,2,25] Palatal muscles are carried in the posteriorly based flaps of each Z-plasty to construct an overlapping retropositioned palatal muscle sling. In some studies, this technique is associated with a lower frequency of velopharyngeal insufficiency (VPI) than noted with other palatoplasty techniques and at times is recommended for the treatment of VPI after another type of repair as a secondary procedure.[1,2,25,26]

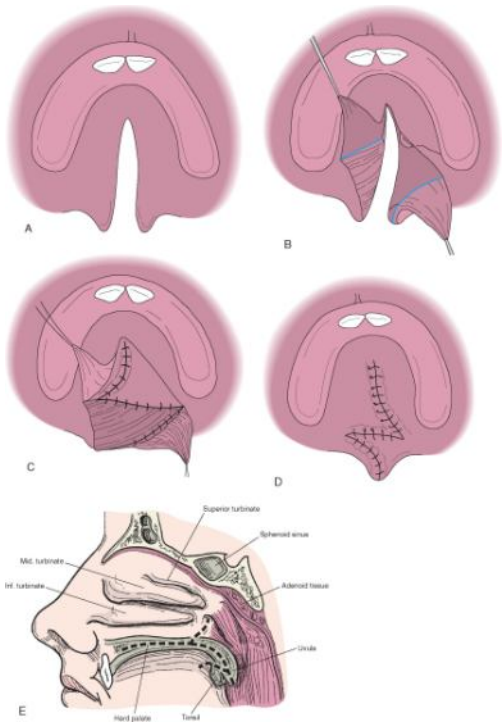

Figure 83-11 **A,** A complete cleft of the secondary palate (both hard and soft) is shown from the incisive foramen to the uvula. **B,** The Furlow double-opposing Z-plasty technique requires that separate Z-plasty flaps be developed on the oral and then the nasal side. Note the cutbacks creating the nasal side flaps, highlighted in blue. **C,** The flaps are then transposed to theoretically lengthen the soft palate. A nasal side closure is completed in standard fashion anterior to the junction of the hard and soft palate. Generally, this junction is the highest area of tension and can be difficult to close. This contributes to the higher fistula rate in this type of repair at this location. **D,** The oral side flaps are then transposed and closed in similar fashion to complete closure of the palate. **E,** Lateral view of nasal and palatal anatomy. The lateral extension of incisions in the Furlow palatoplasty sometimes requires that the incision be extended to the torus tubarius region. Care is taken to reposition the palatal musculature that is abnormally inserted on the posterior palate in conjunction with the double-opposing Z-plasty flaps.

The landmarks that determine the flap angles and flap design include the hamuli, the posterior edge of the hard palate, the base of the uvular halves, and the eustachian tube orifice (torus tubarius) (see Fig. 83-11E). The hamulus is palpable as a small bony eleva tion of the pterygoid plate just medial and posterior to the maxillary tuberosity. Thus, when marking the flaps, the surgeon should remember that the Z-plasties are not two dimensional and that the angles do not need to be at 60 degrees each because flap design is based on the anatomy of the palate, not the geometry of the Z-plasty.

The lateral limbs of the incisions on the oral side and at the level of hamulus are extended to allow advancement of the flaps (see Fig. 83-11B). This incision can be extended around the maxillary tuberosity as a short backcut when necessary. For limited soft palate clefts, only the soft palate incisions and flaps are elevated without the need for hard palate releasing incisions.

The left myomucosal flap (oral) is posteriorly based. The muscle is dissected from the nasal mucosa with scissors, and the levator muscle is carefully preserved. The palatal muscles are separated from the aponeurosis and from the medial side of the superior constrictor to mobilize the flap for rotation. Once the oral flaps are raised, the nasal Z flaps are incised. On the left, the anteriorly based flap (nasal) is elevated as a mucosal flap. On the right, the posteriorly based (nasal) flap is elevated with the muscle. Each lateral limb incision ends just medial to the eustachian tube orifice (torus).

The left anteriorly based (nasal) mucosal flap is incised from the uvula along the dissected edge of the cleft muscle to the left torus. The right posteriorly based (nasal) mucosal-muscle flap is created by an incision from the posterior hard palate to near the torus while taking care to include velar cleft muscle in the flap. However, the mucosal incision on this flap is not extended completely up to the torus to facilitate subsequent closure.

The nasal side is closed by first transposing the left nasal mucosal (anteriorly based) flap transversely to the distal portion of the right nasal incision stepwise from the distal to the proximal aspects to the posterior hard palate nasal mucoperiosteum until the end of the flap reaches near the right torus. When tension is present, avoidance of the natural inclination to sew the distal tip of the flap to its ultimate location first helps protect the fragile tip of the flap. Next, the right myomucosal flap is transposed until the tip of the flap reaches the opposite superior constrictor muscle at the torus (see Fig. 83-11C). This flap is sutured to the nasal mucosal flap anteriorly. The muscle of the posteriorly based oral myomucosal flap on the left side is then secured to the right tonsillar pillar. The right anteriorly based mucosal flap is sutured in stepwise manner across to the oral mucosa on the opposite side.

Retention sutures through the muscle of the posteriorly based right-sided nasal flap may be placed through the base of the oral flap on the opposite side to relieve tension. Mucoperiosteal flaps (when present) are brought into the horizontal plane and sutured. Usually, no bone is exposed and no raw nasal area on the soft palate is left to contract and shorten the palate.

Postoperative care includes a liquid diet and placement of arm restraints for approximately 2 weeks to allow adequate mucosal healing. Careful follow-up and regular visits with a speech pathologist are important.

CLEFT PALATE, MIDDLE EAR, AND SPEECH

Speech characteristics associated with a cleft palate include abnormal nasal resonance, abnormal nasal airflow and altered laryngeal voice quality, nasal or facial grimace, and atypical consonant production (Table 83-3). In patients with clefts, abnormal nasal resonance is typically manifested as hypernasality secondary to VPI. Inadequate nasal resonance (hyponasality) may also occur as result of obstruction. Hypernasality and hyponasality can occur together (mixed nasality). Altered laryngeal voice quality commonly includes hoarseness and reduced volume. Nasal or facial grimace is an unconscious compensation mechanism to inhibit airflow through the nose. As a result of these issues and other factors, there is a risk of articulation disorders and other speech abnormalities.

Table 83-3 -- COMMON SPEECH ABNORMALITIES SEEN IN PATIENTS WITH CLEFT PALATE

Hypernasality: Failure of the palate to separate the oral and nasal cavities during non-nasal consonant production. Oral phonemes substituted by nasal sounds (m, n, ng)
Hyponasality: Reduction of the nasal airflow that occurs with the nasal consonants /m/, /n/, and /ng/. Usually not present with velopharyngeal insufficiency but may be noted when large adenoid or posterior nasal airway obstruction is present with an incompetent velopharyngeal valve
Cul-de-sac resonance: Air enters the nasal cavity but cannot escape because of anterior nasal blockage. Muffled sound quality
Nasal emission: Airflow normal with nasal consonants; abnormal with plosives, fricatives, and affricates. Determined with mirror testing. May be audible or inaudible
Nasal snort,/s/phoneme, and other fricatives: In a patient with an initially closed velopharyngeal valve, as intraoral pressure increases, air escapes from the nose
Stops glottal: Plosive consonant produced by vocal fold valving
Chronic hoarseness: Vocal hyperfunction secondary to compensation at the laryngeal level

Chronic Otitis Media

The cleft palate population has a high incidence of mild to moderate hearing loss. Early onset of otitis media with effusion (OME) is a universal finding in infants with unrepaired cleft palates, and the pathology is causally related to an inherent defect in the opening mechanism of the eustachian tube that results in persistent collapse of the tubal lumen.[27,28] Treatment of this condition requires evacuation of the effusion and insertion of ventilation tubes. In children with a cleft lip and palate, present and persistent OME should be addressed at the time of lip repair, which usually takes place at around 3 months of age. In children with a cleft palate only, persistent OME should be addressed when diagnosed, in a separate procedure, and should not wait for palate repair. In the author's opinion (A.G.), long-term tubes (T tubes) should be used when possible. This option may not be available in a very small ear. In both groups, ears need to be reevaluated shortly before palate repair and regular tubes replaced with long-term ventilation tubes at the time of palate repair. After successful palate repair, most children with a cleft lip and palate will not be scheduled for another surgical procedure for several years. The shorter retaining period and high extrusion rate of regular tubes make them less than ideal in many instances. Even after palate repair, eustachian tube function remains deficient in a large percentage of these children, and ear problems (infections, hearing loss, or both) are most prevalent in the 4- to 6-year-old age group. Ear problems persist at a substantial level until the age of 12 years, and children with a cleft lip and palate have a prolonged recovery and a substantial incidence of late sequelae.[28] T tubes are

retained longer and will therefore obviate the need for repeated tube insertion, which presents additional unnecessary anesthesia risk for the child, as well as increases the cost of care substantially in the first 5 years of the child's life. Evaluation of the ear and hearing should be undertaken every 6 months.

Submucous Cleft Palate

A submucous cleft palate is a microform or incomplete version of a complete cleft of the embryologic secondary palate. It is characterized by the presence of a bifid uvula, a translucent midline known as the zona pellucida (caused by diastasis of the soft palate muscles), and lack of a posterior nasal spine causing palpable notching of the posterior hard palate. A submucous cleft palate may be asymptomatic throughout life or become symptomatic after adenoidectomy, as well as after natural involution of the adenoid tissue. It is important to inspect the uvula carefully and palpate the posterior hard palate before an adenoidectomy. When considering adenoidectomy in any patient, if the posterior palatal spine cannot be palpated and a midline notch is present, partial (anterior) adenoidectomy should be considered. In general, submucous cleft palates are repaired only when they cause symptomatic speech abnormalities.

Velopharyngeal Incompetence

The secondary palate is composed of a hard (bone) palate anteriorly and a soft palate or “velum” posteriorly. Within the soft palate, the levator veli palatini muscle forms a dynamic sling that elevates the velum toward the posterior pharyngeal wall during the production of certain sounds. Other muscle groups within the velum, the tonsillar pillar region, and the pharyngeal walls also affect the quality of resonance during speech formation. The combination of the soft palate and pharyngeal wall musculature jointly forms what is described as the velopharyngeal valve mechanism (Fig. 83-12). This mechanism functions as a sphincter valve for regulating airflow between the oral and nasal cavities to create a combination of orally based and nasally based sounds.

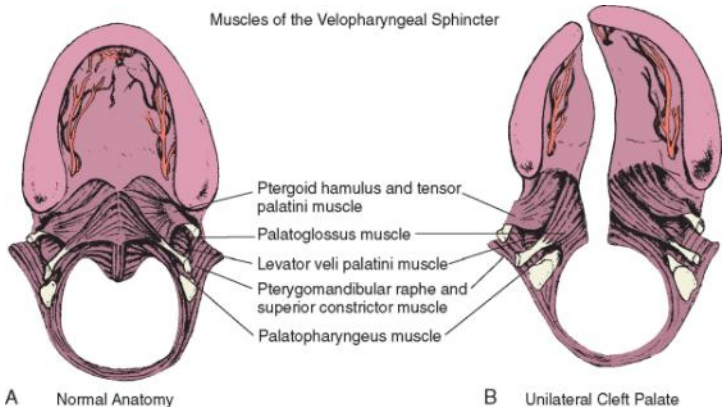

Figure 83-12 Anatomy of the palate and velopharyngeal valve mechanism. **A,** Normal anatomy. **B,** Unilateral cleft of the primary and secondary palate with associated anatomic abnormalities.

Children born with a cleft palate have, by definition, a malformation that has a dramatic impact on the anatomic components of the velopharyngeal valve mechanism. Specifically, clefting of the secondary palate causes division of the musculature of the velum into separate muscle bellies with abnormal insertions along the posterior edge of the hard palate (see Fig. 83-12). VPI is a major functional concern for patients with a cleft. Regardless of the repair type, the incidence of VPI after primary palate repair varies in studies from 5% to higher than 50%.[1,2,29,30]

VPI is defined as inadequate closure of the nasopharyngeal airway port during speech production. The exact cause of VPI after successful cleft palate repair is a complex problem that remains difficult to define completely. Incomplete surgical repair of the musculature is one cause of VPI, but even muscles that have been appropriately realigned and reconstituted may fail to heal normally or function properly. In addition, it must be considered that the repaired cleft palate is only one factor contributing to velopharyngeal valve function and that other abnormalities related to oropharyngeal morphology, nerve innervation, lateral and posterior pharyngeal wall motion, and nasal airway dynamics may all contribute to velopharyngeal valve dysfunction. For example, a short, scarred soft palate that does not elevate well may be compensated by recruitment and hypertrophy of muscular tissue within the posterior pharyngeal wall (Passavant's ridge).

Approximately 20% of children with VPI after palatoplasty will eventually require management involving additional palatal surgery. Left untreated, nasal air escape–related resonance problems will lead to other speech abnormalities, namely, abnormal compensatory articulation. These abnormal, compensatory misarticulations further complicate problems with speech formation and decrease speech intelligibility in patients with cleft palate–related VPI.

After the initial cleft palate repair, periodic evaluations are important to assess the speech and language development of each child. Typically, this involves a standardized screening examination performed by a speech and language pathologist as part of an annual visit to the cleft palate team. Detailed studies that include the use of videofluoroscopy and nasopharyngoscopy may be indicated. Videofluoroscopy is used to radiographically examine the upper airway with the aid of an oral contrast agent. This technique allows dynamic testing of the velopharyngeal valve mechanism with views of the musculature in action. In addition, details of upper airway anatomy, including residual palatal fistulas, can be visualized and their contribution to speech dysfunction evaluated during the study. For a videofluoroscopy study to be of diagnostic value it must include multiple views of the velopharyngeal valve mechanism, and a speech pathologist must be present to administer verbal testing in the radiology suite.

Nasopharyngoscopy allows direct visualization of the upper airway and specifically the velopharyngeal valve mechanism from the nasopharynx. This technique avoids the radiation exposure associated with videofluoroscopy but requires preparation of the nose with a topical anesthetic, skillful maneuvering of the scope, and a compliant patient. Once the endoscope is inserted, observations of palatal function, airway morphology, and pharyngeal wall motion are made while the patient is verbally tested by the speech pathologist. The opportunity for direct visualization of the velopharyngeal valve mechanism in action during speech formation provides information that is critical to clinical decision making related to secondary palatal surgery in cases of confirmed or suspected VPI.

With videofluoroscopy and nasoendoscopy, the closure pattern of the palate is documented and should be differentiated. The closure pattern may help determine the success of various secondary palatal procedures designed to augment anatomic deficiencies.

- The most common pattern of closure (55% of the normal population) is coronal closure. It consists of posterior movement of the soft palate to the posterior pharyngeal wall with little movement from the lateral walls. Approximately 45% of patients with VPI have this pattern of closure.
- The sagittal closure pattern is seen in 10% to 15% of the population. Primary closure is by lateral wall movement without significant anterior-to-posterior closure. This pattern is seen in approximately 10% of children with VPI.
- Circular closure includes lateral wall movement and posterior movement of the soft palate. It occurs in approximately 10% of the population and 20% of children with VPI. Circular closure with Passavant's ridge includes lateral wall and soft palate movement, as well as anterior movement of the posterior pharyngeal wall. This pattern is seen in 20% of the normal population and in 25% of children with VPI.

Secondary palatal surgery in young children is indicated when VPI is causing hypernasal speech on a consistent basis and is related to the anatomic problem. The exact timing of surgery for VPI remains controversial, however, with recommendations ranging from 2.5 to older than 5 years. In such a young age group, variables such as the child's language and articulation development and lack of compliance during speech evaluation compromise the diagnostic accuracy of preoperative assessment. The decision to proceed with additional surgery for VPI is not an isolated surgical judgment. The authors believe that lack of palatal movement in the anterior-posterior direction benefits most often from a pharyngeal flap, and frequently the lack of lateral wall motion is best treated by sphincter pharyngoplasty. There is considerable variation in surgical technique for each of these procedures, and each can be customized to fit the particular closure pattern of each patient.

TONSILLECTOMY AND ADENOIDECTOMY IN CHILDREN WITH CLEFTS

The age group of children evaluated for VPI certainly coincides with the period when enlargement of the tonsils and adenoids is most common. This issue is not well addressed in the literature and, if ignored, has a tendency to disrupt the positive outcome of cleft palate repair.

Severely enlarged tonsils can interfere with elevation and closure of the palate. Careful observation of the tonsil–soft palate relationship during oropharyngeal examination, as well as nasoendoscopy, will disclose the impact of tonsil enlargement. A repeat speech evaluation should be performed 6 to 8 weeks after tonsillectomy and the presence and severity of VPI reassessed.

Enlarged adenoids aid velopharyngeal closure. After adenoidectomy, occult VPI suddenly becomes overt and decompensated speech patterns can be dramatic. VPI after adenoidectomy in the general population occurs in less than 1% of cases. Of these, approximately one third can be observed to have findings of mild VPI before adenoidectomy. The risk for VPI after adenoidectomy is increased in children with developmental delay, generalized hypotonia, mental retardation, submucous cleft palate, family history of VPI, and history of feeding problems in early childhood.

If a subsequent pharyngeal flap is not planned, adenoidectomy in a child with a repaired or submucous cleft palate or in a child with other risk factors as mentioned earlier should be limited to the anterior one half of the adenoid tissue (closest to the choanae and torus) while leaving the height and posterior one half of the adenoid tissue intact for adequate velar closure. Adenoidectomy in this age group is frequently indicated for recurrent or chronic sinonasal infections and for recurrent or chronic otitis media. One of the rare causes of VPI is closure of the palate against asymmetrical adenoid tissue, which can be adequately identified only with nasoendoscopy.

A major cause of obstructive sleep apnea in children is adenotonsillar hypertrophy. Failure to recognize the impact of adenotonsillar hypertrophy in a child with sleep-disordered breathing, snoring, or mild apnea can lead to severe obstructive sleep apnea after creation of a pharyngeal flap to treat VPI. Performing an adenoidectomy after a pharyngeal flap is in place is difficult and may carry a higher risk of bleeding. A moderately enlarged mass of adenoid tissue can become severely obstructive after a few months and interfere with the function of a pharyngeal flap. When adenoidectomy is required before treatment of VPI, appropriate surgical planning may include removal of the obstructive adenoid tissue at least 4 to 6 weeks before creation of a pharyngeal flap.

Enlarged tonsils obstructing the view of the posterior pillars may interfere with the elongation of flaps for pharyngoplasty. When necessary, conservative tonsillectomy (preservation of both the anterior and posterior tonsil pillars) is planned before pharyngoplasty. The lateral pharyngeal ports can become obstructed by enlarged tonsils several months after a successful pharyngeal flap procedure and cause speech disturbance, obstructive sleep apnea, or both. Nasoendoscopy will show this problem and tonsillectomy can alleviate it. It should be noted that adenoidectomy and tonsillectomy in patients with clefting is a controversial area and that treatment plans must be individualized.

OPERATIVE TECHNIQUES FOR VELOPHARYNGEAL INSUFFICIENCY

Contemporary surgical management of VPI generally involves the use of two types of procedures: a pharyngeal flap and sphincter pharyngoplasty.[1,2,29] The use of autogenous or alloplastic implants for augmentation of the posterior pharyngeal wall has been described but is not a commonly used procedure. More recently, some surgeons have advocated performance of a second palatoplasty operation in an attempt at palatal lengthening in a patient with VPI. This may become an alternative in some patients with short gaps posteriorly.

A superiorly based pharyngeal flap remains the standard approach for surgical management of VPI. Using 14F catheters for guidance, the technique advocated by the authors limits the combined size of the lateral ports to less than 20 mm^2, the maximal velopharyngeal port opening during clinically normal speech. Surgical maneuvers are directed at recruiting tissue by developing a superiorly based soft tissue flap from the posterior pharyngeal wall (Fig. 83-13). The soft palate is then divided along the midsagittal plane from the junction of the hard and soft palate to the uvula, and the flap from the posterior pharyngeal wall is inset within the nasal layer of the soft palate. A superiorly based flap may be covered with mucosal flaps on both the dorsal and ventral surfaces, thus increasing the viability of the flap, reducing flap contracture, and expediting mucosal healing. As a result, a large nasopharyngeal opening that cannot be completely closed by the patient's velopharyngeal valve mechanism is converted into two (right and left) lateral pharyngeal ports. Closure of these ports is easier for the patient to accomplish as long as adequate lateral pharyngeal wall motion is present.

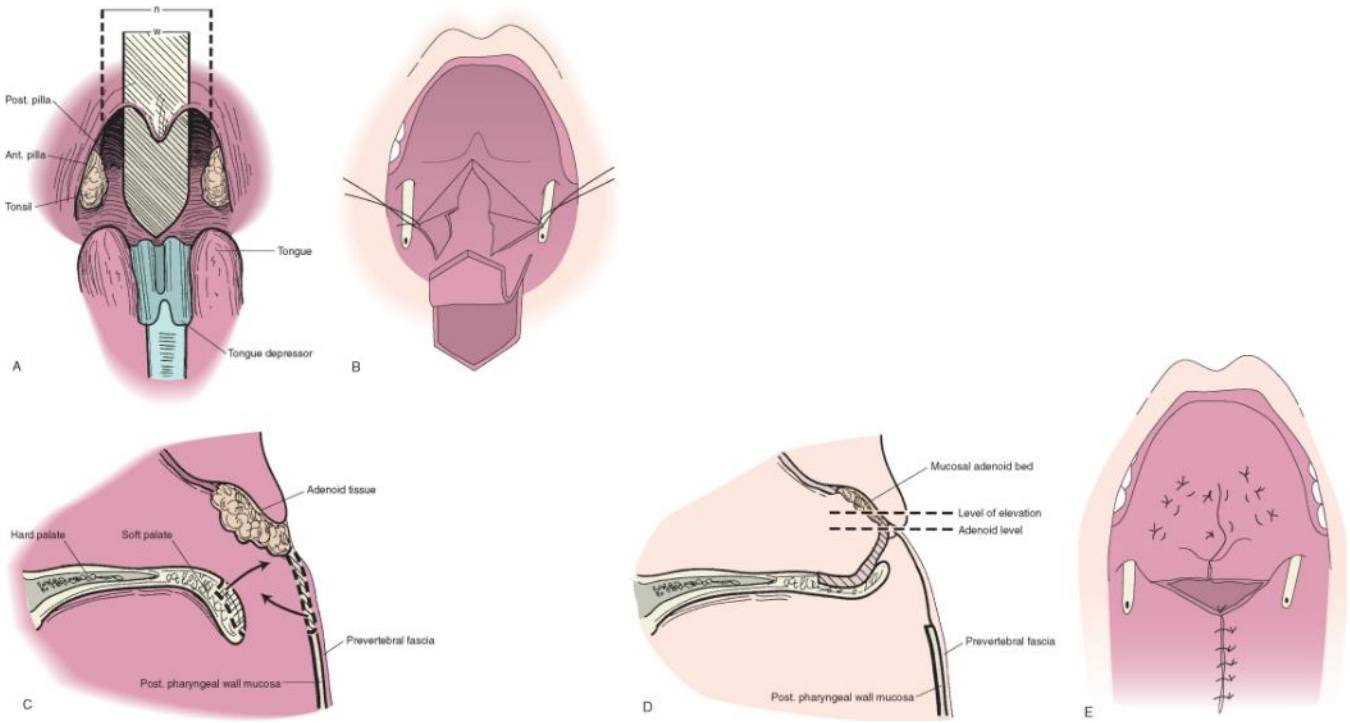

Figure 83-13 Superiorly based pharyngeal flap. **A,** Elevation of a myomucosal flap from the prevertebral fascia and division of the soft palate tissues. **B,** Dissection of the oral, nasal, and muscular layers for insetting the flap. **C,** Sagittal view of soft palatal anatomy in preparation for insetting the pharyngeal flap at the appropriate vertical height. **D,** Sagittal close-up view indicating the relationship between adenoid tissue and the pharyngeal flap. **E,** Insertion of the flap with closure of the oral mucosa over the raw defect to decrease scarring.

The high overall success rate and the flexibility to design the dimensions and position of the flap itself are advantages of the superiorly based pharyngeal flap procedure. Disadvantages of the pharyngeal flap procedure are primarily related to the possibility of nasal obstruction, resulting in trapping of mucus and the potential for exacerbation of obstructive sleep apnea.

Inferiorly based pharyngeal flaps for the management of VPI are rarely used and tend to cause downward pull on the soft palate after healing and contracture of the flap. The result may be a tethered palate with decreased ability to elevate during the formation of speech sounds. Moreover, should postoperative hemorrhage occur, an inferiorly based flap obscures the donor site for access to the bleeding site, and to control the bleeding, the flap may need to be sacrificed in some instances. A superiorly based flap leaves the donor site fully accessible.

Dynamic sphincter pharyngoplasty is another option for the surgical management of VPI. The operative procedure involves the creation of two superiorly based myomucosal flaps that include each posterior tonsil pillar (Fig. 83-14). Each flap is elevated with care taken to include as much of the palatopharyngeal muscle as possible. It may be difficult to raise flaps in patients with velocardiofacial syndrome, because the internal carotid arteries may take a median course in the posterior pharynx. The flaps are then attached and inset within a horizontal incision made high on the posterior pharyngeal wall. These flaps are designed to be anastomosed either end to end (if short) or side to side (overlap) (if long) (see Fig. 83-14).

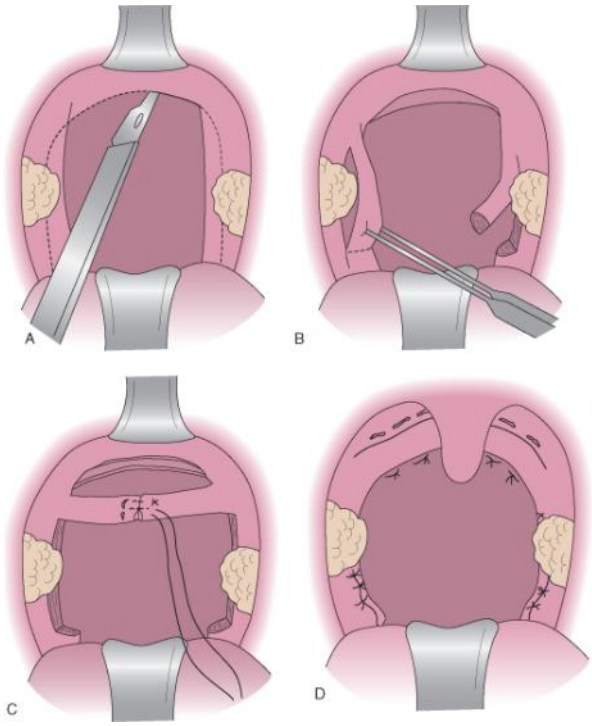

Figure 83-14 Sphincteroplasty. **A,** Incision in the posterior pharyngeal wall and the posterior tonsillar pillars. **B,** Elevation of the tonsillar pillar–myomucosal flaps. **C,** Approximation in the central pharyngeal area to customize the size of the central port. **D,** Sutured flaps placed in the posterior pharyngeal walls and posterior-lateral pharyngeal walls.

The goal of this procedure is creation of a single nasopharyngeal port (instead of the two ports of the pharyngeal flap) that has a contractile ridge posteriorly to improve velopharyngeal valve function. Transposed flaps should be high in the nasopharynx, at the level of the velopharyngeal closure. VPI may persist if positioning is incorrect or narrowing is inadequate. Ideally, at the completion of the procedure, the flaps should not be visible during oropharyngeal examination.

The main advantage of sphincter pharyngoplasty over a superiorly based flap is a lower rate of complications related to nasal airway obstruction, as described earlier. Despite this theoretical advantage, there is no convincing evidence that pharyngoplasty procedures achieve superior outcomes in the resolution of VPI. In addition, the use of a sphincter pharyngoplasty technique may be associated with increased scarring along the tonsillar pillar region.

Some surgeons advocate the use of revision palatoplasty instead of a pharyngeal flap or pharyngoplasty procedure for the management of patients with VPI. Specifically, either aggressive repositioning of the musculature with a two-flap palatoplasty and intravelar velarplasty or a Furlow double-opposing Z-plasty is carried out to facilitate velopharyngeal valve closure. The ideal patient for a revision Furlow procedure is a child younger than 12 years with poor levator reconstruction, a short palate with a gap of less than 10 mm, and 50% to 75% lateral pharyngeal wall movement. An older patient with a 5-mm or smaller gap and good lateral wall movement may also be a good candidate. Long-term speech results of revision Furlow procedures are unavailable at present, but some surgeons strongly advocate this technique. Thus, the exact choice for each patient still remains controversial and is best tailored according to each surgeon's individual experience.

PALATAL FISTULA REPAIR

Fistulas can occur in patients who undergo repair of cleft palates. The recommended timing of closure may vary significantly and remains a controversial topic.[1,2,30–33] Some surgeons or speech pathologists may advocate aggressive management with early closure of any fistula present after the initial palate repair. The authors prefer to take a more long-range view of fistulas and delay surgery for several years whenever possible. Fistulas that have a negative impact on speech may be closed earlier.

In infants, closure of a small, nonfunctional fistula can generally be deferred until later in childhood. In such cases, fistula repair may be incorporated into any future procedures such as pharyngeal surgery for VPI or bone graft reconstruction of the cleft maxilla and alveolus, as long as there is no functional speech or feeding-related concerns. When a large (>5 mm) fistula is present, there is a greater likelihood that functional impairment will be encountered, such as nasal air escape affecting speech, nasal reflux of food and liquids, and hygiene-related difficulties. In clinical situations in which significant functional problems exist, earlier closure of a persistent fistula is indicated. As part of the decision-making process, surgeons must weigh the benefits of repair against the negative effects on subsequent maxillary growth of a second palatal surgery involving stripping of mucoperiosteum.

Another consideration in planning the exact timing of closing a fistula is the type of technique being used for the repair. Closure of a fistula with local flaps or repeat palatoplasty may be undertaken during infancy and early childhood. On the other hand, in cases in which the use of a tongue flap is considered, the child must be old enough to cooperate with the perioperative regimen.

Operations currently used for fistula repair include local palatal flaps, modifications of the von Langenbeck and two-flap palatoplasty techniques, palatoplasty with incorporation of a pharyngeal flap, and the use of a tongue flap. Other regional flaps, including tongue, buccal mucosa, buccinator myomucosal, temporalis muscle, and vascularized tissue transfer, are used less frequently.

When less than 4 mm in diameter, the fistula may be closed with an edge-based turnover flap for nasal lining, followed by a large oral mucoperiosteal rotation flap. In this manner, the nasal and oral suture lines are staggered. In some hard palate fistulas or large fistulas, dissection resembling palate repair with palatal mucoperiosteal flap elevation and separate nasal and oral closure is necessary. Depending on the location, closure of larger fistulas is aided by flaps from the buccal sulcus or pharynx.

When a large fistula is located within the anterior two thirds of the hard palate, another excellent choice for repair is an anteriorly based dorsal tongue flap. First, turnover flaps are developed to close the nasal side of the palatal defect with multiple interrupted sutures. Next, this technique calls for the development of an anteriorly based tongue flap that is approximately 5 cm in length by one third to two thirds the width of the tongue. The tongue flap is elevated along the underlying musculature and then inset with multiple mattress sutures for closure of the oral side. The recipient bed within the tongue is closed primarily. After the initial surgery, the tongue flap is allowed to heal for approximately 2 weeks. At that time the patient is returned to the operating room. The flap is sectioned and the stump at the donor site is freshened and inset into the tongue. The use of laterally and posteriorly based tongue flaps has also been advocated. In our opinion, an anteriorly based flap is better tolerated by most patients and allows the greatest degree of tongue mobility with less risk of tearing the flap from its palatal insertion. Some surgeons use maxillomandibular fixation for 2 weeks while the flap becomes vascularized.

MAXILLARY AND ALVEOLAR CLEFT BONE GRAFTING

Approximately 75% of patients with clefts will have clefts that involve the alveolus, maxilla, nasal floor, and piriform rim. The vast majority of surgeons advocate reconstruction of this portion of the cleft during mixed dentition before eruption of the permanent canine or lateral incisor.[1,2,34] This approach allows continuity of the maxillary arch, support for the erupting teeth, reconstruction of the congenital defect, support of the nasal base, and closure of the residual cleft/fistula. Frequently, orthodontic maxillary expansion is required to match the arch compatibility of the mandible before performing the grafting procedure. Such expansion is performed by a skilled orthodontist over a period of several months before surgery in a coordinated fashion.

The technique used by most surgeons involves layered closure of the defect with a multiple-flap technique that mobilizes attached gingiva into the cleft defect where the tooth/teeth will ideally erupt (Fig. 83-15). Both anterior vestibular and palatal flaps are necessary, because the defect courses from the piriform rim to alveolus and back to the incisive foramen region. Some patients may have a residual hard palatal fistula that can be repaired at the same time.

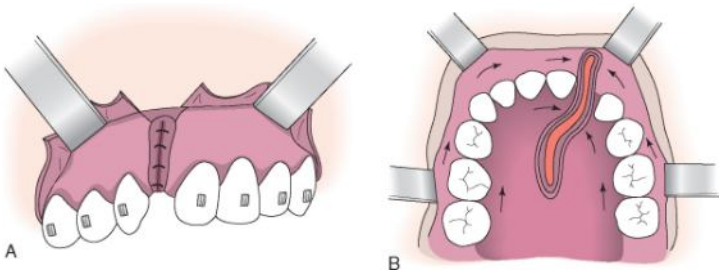

Figure 83-15 **A** and **B,** Four-flap technique of advancing much-needed attached mucosa into the residual cleft of the primary palate with simultaneous closure of the oral-nasal fistula and placement a of cancellous iliac crest bone graft. *(From Fonseca RJ: Davis Reconstructive Preprosthetic OMS. Philadelphia, WB Saunders, 1995, p 996.)*

The iliac crest is the most common donor site for bone graft reconstruction at the cleft site. Other sites have also been recommended, but the best results continue to be achieved with cancellous bone procured from the anterior iliac crest via a minimal 3- to 5-cm incision posterior and lateral to the anterior iliac spine. The cartilaginous cap of the iliac crest is directly incised with a blade, and a small trapdoor is elevated medially. Curettes are used to procure enough bone to fill the defect. Although attempts have been made to use allogeneic bone products or synthetic material, outcomes have been poor in comparison with autogenous iliac crest.[1,2,34]

Watertight closure is desirable for the nasal side of the closure, and resorbable sutures with tapered needles are used throughout. This can be tested by using a bulb syringe with irrigation in the naris. Defects are repaired with horizontal mattress or interrupted suture techniques. The bone graft taken from the iliac crest is then placed in the site while remembering to fill the entire defect up to the piriform rim rather than just at the alveolus. The oral mucosa is then mobilized to close the remaining oral cavity layer in a tension-free manner. Patients with bilateral clefts of the anterior palate benefit from placement of a postoperative acrylic splint made before surgery for stabilization of the premaxillary segment. The splint can be wired to orthodontic mechanics or bonded with a dental bonding agent to the teeth for several weeks during healing of the mucosal tissue.

Patients are placed on a liquid diet for 2 weeks and told to avoid using straws. Once the initial mucosal healing is complete, the diet can be advanced slowly over the next several weeks. The splint is removed when the mucosal tissues are more completely healed. Patients should brush their teeth, but not directly over the repair sites. Gentle mouth rinses of saline or chlorhexidine may be used to keep the dentition and oral cavity clean as well.

CLEFT RHINOPLASTY

Congenital clefts that involve the lip, nose, and underlying skeletal structure will cause a complex three-dimensional deformity of the nasal complex that affects both form and function. In the case of a complete unilateral cleft, the typical nasal deformity is characterized by splaying of the alar base, inferior displacement of the alar rim, deviation of the nasal tip, and irregularity of the caudal nasal septum.[1,2,30] Abnormal fibrous insertions exist between the lateral crus of the lower lateral cartilage and the lateral piriform rim on the cleft side. At the time of the initial lip repair procedure, maneuvers for primary nasal reconstruction include dissection along the lower lateral cartilage to separate the overlying skin from the cartilage and sharp release of the fibrous insertions along the piriform rim so that the nostril can be repositioned appropriately. Despite effective primary cleft lip and nasal repair during infancy, most patients will demonstrate enough residual nasal dysmorphology that secondary nasal surgery for correction of the cleft-associated malformation or improvement in nasal airflow will be beneficial later in life.

The timing of cleft nasal revision surgery also remains controversial. Some surgeons take a more aggressive approach and undertake extensive nasal reconstruction during early childhood. Our philosophy is to delay the definitive cleft rhinoplasty until the nasal complex is close to mature size. When possible, early nasal surgery should be performed after bone graft reconstruction of the maxilla so that a stable bony foundation along the piriform rim and nasal base exists first. If the patient's reconstructive treatment plan also requires maxillary advancement, nasal surgery should be delayed until approximately 6 months after the orthognathic procedure. This allows a more predictable outcome and long-lasting improvement in nasal function and facial aesthetics. Early surgery is reserved for individuals with severe airway or nasal airflow problems or children who endure psychosocial consequences such as teasing at school.

Secondary cleft-nasal reconstruction will often require dorsal reduction, lower lateral cartilage sculpting, cartilage grafting, and nasal osteotomies. Cartilage grafting is a critical component of the final nasal reconstruction and is used for augmentation of the dysmorphic lower lateral cartilage and improvement of nasal tip projection. Several different donor sites may be used, including auricular cartilage, nasal septum, and rib cartilage. Ear cartilage is most useful in situations in which augmentation of hypoplastic cleft-side lower lateral cartilage is required. Septal cartilage is most easily accessible and provides an excellent scaffold for repositioning of the lower lateral cartilage and improvement of nasal tip symmetry and projection. Unfortunately, patients may be scheduled for definitive nasal reconstruction after having previously undergone septal cartilage harvest and not have sufficient quantity for a second septal cartilage graft. In such cases, the use of costochondral cartilage is another excellent option. Rib cartilage provides adequate amounts of graft material but requires a distant donor surgical site. We have found this type of cartilage graft to provide excellent strength for support of the nasal tip and alar complex. These techniques are best carried out through an open approach. A transcolumellar splitting incision is combined with marginal incisions to provide wide access and direct visualization of the nasal dorsum, upper and lower lateral cartilage, and nasal septum.

A similar rationale is applied when considering the timing of secondary nasal reconstruction in a patient with a bilateral cleft lip, but the specific dysmorphology addressed is somewhat different. Generally, nasal asymmetry is less problematic, and the dysmorphology is characterized by deficient columellar length. Many surgeons have focused on secondary lengthening of the columella through the use of banked forked flaps or columellar lengthening with soft tissue flaps from the floor of the nose and alar flaps. Unfortunately, these types of surgical procedures often result in a distorted columellar-labial angle, excessive scars that extend onto the nasal tip, and additional distortion of the broad nasal tip. We find that using septal cartilage strut grafts attached to the caudal nasal septum and lower lateral cartilage yields the most natural-looking results. The objective is correction of the underlying cartilaginous anatomy with stretching of the overlying soft tissue envelope instead of direct surgical manipulation of the columellar skin.

CLEFT ORTHOGNATHIC SURGERY

The technical details and integrated orthodontic-surgical treatment planning for orthognathic correction are beyond the scope of this chapter, but the treatment concepts warrant comment inasmuch as more than 35% of patients who undergo cleft repairs will benefit from orthognathic surgery to treat malocclusion.[35,36] Most commonly, midface hypoplasia results in an underbite that causes functional and aesthetic problems. Additionally, facial asymmetry is common in patients who have previously undergone cleft repairs in infancy. These procedures are performed at the end of growth in the craniomaxillofacial skeleton, usually 14 to 16 years of age in girls and 16 to 18 years in boys. In rare instances, midface advancement may be indicated earlier. Coordinated care with a skilled orthodontist is very important to achieve the optimal result and consists of planning and several phases of orthodontic therapy both before and after orthognathic surgery.

Usually, a Le Fort I osteotomy is indicated for patients with maxillary hypoplasia and malocclusion. In many cases, bilateral sagittal split osteotomies and genioplasty are helpful in treating the malocclusion and improving facial balance (Fig. 83-16). Traditional orthognathic surgery is dependent on precise placement of fixation to ensure the optimal occlusion and facial balance to avoid relapse. Recently, distraction osteogenesis has been used in patients who require exceptionally large advancement of the midface.[37] Although the technique shows little or no improvement over traditional osteotomy techniques in most patients, it may be helpful in some patients who require larger advancement.

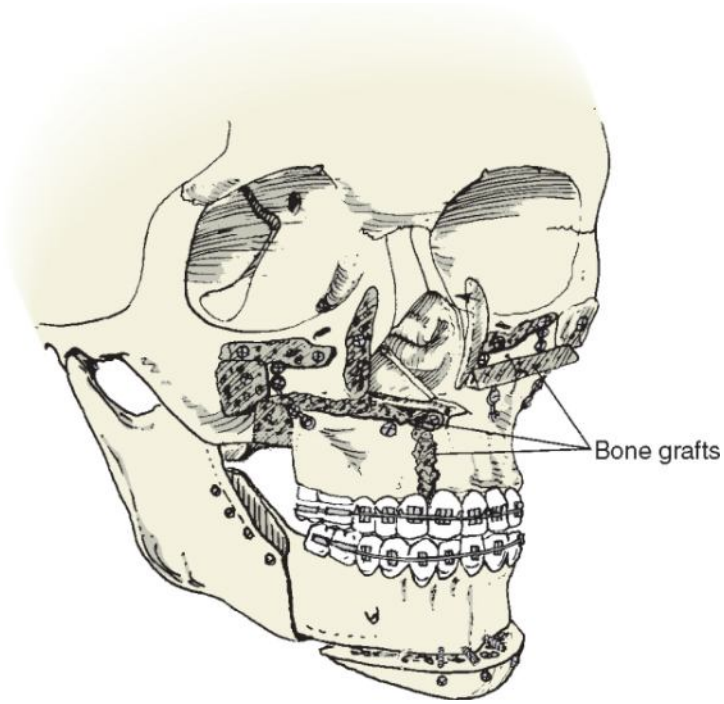

Figure 83-16 A Le Fort I osteotomy, bilateral sagittal splits, and genioplasty osteotomies are shown for a patient with maxillary hypoplasia associated with early surgery required for a unilateral cleft lip and palate. Onlay bone grafts may be helpful for improving facial contour.
(From Ruiz RL, Costello BJ, Turvey T: Orthognathic surgery in the cleft patient. In Oral and Maxillofacial Surgery Clinics of North America: Secondary Cleft Surgery. Philadelphia, WB Saunders, 2002, pp 491-507.)

CONCLUSIONS

Comprehensive care of patients with clefts requires an interdisciplinary approach that demands precise surgical execution of the various procedures necessary to correct cleft deformities, as well as frequent long-term follow-up. Clinicians experienced in the comprehensive interdisciplinary care of patients with clefts are best equipped to deal with these concerns. Treatment of patients with cleft and craniofacial deformities should be free of bias and should include team care that is patient, family, and community oriented. Only in this fashion can the overall treatment be optimally successful. This type of care maximizes patients' ability to grow into adulthood and succeed in life without focusing on their deformity.

PEARLS

- Marking the key landmarks is important to obtain the best result, and the temptation to keep hypoplastic tissue should be discouraged.
- Nasoalveolar molding appliances may be helpful in reapproximating the segments to allow easier surgical repair, and they may improve the aesthetic result of the lip and nose.
- A three-layered closure is the key to both functional and aesthetic repair of the lip.
- Some nasal asymmetry is expected in wide unilateral clefts, but this can be minimized by conservative primary nasal reconstruction.
- Nasal bolsters or silicone nasal stents can be used to help mold the nose in the postoperative period.
- Arm restraints may be used for the first 2 weeks to help avoid injury to the lip repair, but they should be removed occasionally to allow range of motion of the arms.
- Palate repair requires adequate mobilization of tissues to allow the most tension-free closure possible.
- Releasing the nasal and palatal periosteum, as well as the tensor veli palatine muscle or Veau's muscle, is helpful in gaining additional mobilization.

PITFALLS

- Patients with syndromes, dysmorphology, and wide palates are more prone to fistulas.
- A common pitfall is to incompletely reconstruct the musculature of the lip or palate.
- Another common pitfall occurs when the surgeon leaves considerable dead space within the layers of the repair. Placement of at least several sutures through each of the layers is important in preventing dead space.
- Careful wound care is necessary because some bolsters used to help mold the nose may cause pressure necrosis of the columella or nasal base if placed too tightly or kept in place for too long.
- A common pitfall occurs when the surgeon fails to mobilize enough muscle to create an active levator sling that is posteriorly displaced.
- Failure to preserve the vascular pedicle may result in necrosis of the tissues.