Chapter 23

Cleft Lip and Palate

Richard A. Hopper

Court Cutting

Barry Grayson

Cleft lip and palate are the most common congenital craniofacial anomalies treated by plastic surgeons. Successful treatment of these children requires technical skill, in-depth knowledge of the abnormal anatomy, and appreciation of three-dimensional facial aesthetics. Cleft care requires that the plastic surgeon be a member of a collaborative multidisciplinary team. Through close self-scrutiny, disciplined evaluation of the results, and a great deal of imagination, a number of plastic surgeons continue to advance cleft care, seeing as many challenges ahead as they encountered at the beginning of their careers.

Epidemiology and Etiopathogenesis

Among the cleft lip and palate population, the most common diagnosis is cleft lip and palate at 46%, followed by isolated cleft palate at 33%, then isolated cleft lip at 21%. The majority of bilateral cleft lips (86%) and unilateral cleft lips (68%) are associated with a cleft palate. Unilateral clefts are nine times as common as bilateral clefts, and occur twice as frequently on the left side than on the right. Males are predominant in the cleft lip and palate population, whereas isolated cleft palate occurs more commonly in females. In the white population, cleft lip with or without cleft palate occurs in approximately 1 in 1,000 live births. These entities are twice as common in the Asian population, and approximately half as common in African Americans. This racial heterogeneity is not observed for isolated cleft palate, which has an overall incidence of 0.5 per 1,000 live births.

Both environmental teratogens and genetic factors are implicated in the genesis of cleft lip and palate. Intrauterine exposure to the anticonvulsant phenytoin is associated with a 10-fold increase in the incidence of cleft lip. Maternal smoking during pregnancy doubles the incidence of cleft lip. Other teratogens, such as alcohol, anticonvulsants, and retinoic acid, are associated with malformation patterns that include cleft lip and palate, but have not been directly related to isolated clefts.

Genetic abnormalities can result in syndromes that include clefts of the primary or secondary palates among the developmental fields affected. More than 40% of isolated cleft palates are part of malformation syndromes, compared to less than 15% of cleft lip and palate cases. The most common syndrome associated with cleft lip and palate is van der Woude syndrome with or without lower lip pits or blind sinuses. Microdeletions of chromosome 22q resulting in velocardiofacial, DiGeorge, or conotruncal anomaly syndromes are the most common diagnoses associated with isolated cleft palate. Although there is a recognized genetic component to nonsyndromic cleft lip and/or palate, it appears to be multifactorial. Among other recent studies, a meta-analysis of 13 genome scans by Marazita et al. (2004) revealed multiple cleft lip/palate genes on 16 chromosomal regions.

Parents with a child with a nonsyndromic cleft, or a family history of clefting, often ask about their risk of clefts in subsequent pregnancies. The risk depends on whether the proband has a cleft lip alone
Multidisciplinary Cleft Care

Individuals born with cleft lip and or palate require coordinated care from multiple specialties to optimize treatment outcome. The ideal is in a center with a multidisciplinary cleft team, dedicated to treating cleft-related issues from birth to adulthood. Typical members of a cleft team include an audiologist, dentist, geneticist, nurse, nutritionist/dietitian, oral surgeon, orthodontist, otolaryngologist, pediatrician, plastic surgeon, psychologist, social worker, and speech pathologist.

The emphasis is on coordination to minimize the number of surgeries performed while maximizing the benefit to the patient. Although the number of surgical procedures required prior to adulthood has decreased with improved techniques, care of a child with a cleft still requires a complex lengthy surgical treatment plan (Table 23.1). The goal of cleft care is to eliminate as many steps in the treatment plan as possible by optimizing the outcome and benefit of each essential intervention. Recent advances in presurgical orthopedics, such as nasoalveolar molding and gingivoperiosteoplasty, are examples of optimization of early intervention with the goal of minimizing secondary surgeries and eliminating previously essential steps such as secondary alveolar bone grafting and rhinoplasty.

Surgical evaluation and classification

Ideally, the newborn infant with a cleft is evaluated by the cleft team in the first weeks of life. The increasing number of clefts detected by prenatal imaging allows early preparation of the family and introduction to the treatment plan. Patients with cleft lip and/or palate are not a homogenous group. As mentioned above, they can be divided into CL, CP, and CLP; however, the surgical treatment plan requires a more complex classification scheme. The cleft lip deformity is typically divided into unilateral or bilateral, and then subdivided into complete, incomplete, or microform. The width of the cleft deformity and the degree of alveolar arch collapse also play a part in surgical planning, as these directly relate to the degree of associated nasal deformity and the tension and difficulty of the repair. The associated nasal deformity is similarly categorized as mild, moderate, or severe. Mild nasal deformity is characterized by a lateral displacement of the alar base but normal alar contour, minimal columella shortening, and normal dome projection. Moderate nasal deformity has lateral and posterior displacement of the alar base, columella deficiency, and a depressed dome. Severe nasal deformity has an underprojecting alar dome with complete collapse of the lower lateral cartilage and a severe deficiency of columella height. Severe nasal deformities often have a reversed curvature to the alar rim. The nasal deformity is secondary to a three-dimensional distortion of the lower lateral cartilage, described by some as the “tilted tripod.” It is not caused by hypoplasia or deficiency of the cartilage itself.

<table>
<thead>
<tr>
<th>Age</th>
<th>Treatment</th>
<th>Cleft team members</th>
</tr>
</thead>
<tbody>
<tr>
<td>Prenatal</td>
<td>Prenatal imaging, diagnosis, and counseling</td>
<td>Multidisciplinary</td>
</tr>
<tr>
<td>Age Range</td>
<td>Procedure and Treatment</td>
<td>Specialties</td>
</tr>
<tr>
<td>-------------------------------</td>
<td>-----------------------------------------------------------------------------------------</td>
<td>-----------------------------------</td>
</tr>
<tr>
<td>Newborn</td>
<td>Feeding assessment, medical assessment, genetic counseling, treatment information</td>
<td>Multidisciplinary</td>
</tr>
<tr>
<td>0-3 months</td>
<td>Presurgical orthopedics</td>
<td>Orthodontist, plastic surgeon</td>
</tr>
<tr>
<td>3 months (or after presurgical orthopedics)</td>
<td>Primary cleft lip repair and tip rhinoplasty ± gingivoperiosteoplasty</td>
<td>Plastic surgeon</td>
</tr>
<tr>
<td>12 months (delayed if airway or medical concerns)</td>
<td>Primary cleft palate repair with intravelar veloplasty ± bilateral myringotomy and tubes</td>
<td>Plastic surgeon, otolaryngologist</td>
</tr>
<tr>
<td>Diagnosis of velopharyngeal insufficiency (3-4 years)</td>
<td>Secondary palate lengthening or pharyngoplasty, speech obturator</td>
<td>Speech pathologist, plastic surgeon, otolaryngologist, orthodontist</td>
</tr>
<tr>
<td>School-age years</td>
<td>Treatment of secondary lip and nasal deformities</td>
<td>Plastic surgeon</td>
</tr>
<tr>
<td>7-9 years (mixed dentition)</td>
<td>Secondary alveolar bone graft</td>
<td>Orthodontist, plastic surgeon, oral surgeon</td>
</tr>
<tr>
<td>Postalveolar graft</td>
<td>Presurgical orthodontics</td>
<td>Orthodontist</td>
</tr>
<tr>
<td>Puberty</td>
<td>Definitive open rhinoplasty</td>
<td>Plastic surgeon</td>
</tr>
<tr>
<td>Skeletal maturity</td>
<td>LeFort I ± mandible orthognathic surgery</td>
<td>Plastic surgeon, oral surgeon</td>
</tr>
</tbody>
</table>

| Specialties: Orthodontist, plastic surgeon, otolaryngologist, orthodontist |

**a**Essential treatments of cleft lip and palate deformity.

**b**Required if gingivoperiosteoplasty is not done or is unsuccessful.

If a cleft palate is present, it is surgically classified as unilateral, bilateral, or submucous. The width of the cleft is noted as it affects the difficulty of closure.

Although most surgeons use the descriptive classification of cleft deformities during the initial assessment of a patient, other classification systems are often used for outcome research and record keeping. Kernahan and Stark's “stripped Y” diagrammatic classification scheme and its modifications continue to be used in many cleft centers. It recognizes the embryologic division of the primary (lip
and alveolus) and secondary palates at the incisive foramen. Otto Kriens presented a palindromic acronym organization of cleft deformities. The acronym LAHSHAL denotes the bilateral anatomy of lip (L), alveolus (A), hard (H) and soft (S) palates, by convention from right to left. Lowercase letters represent incomplete clefts of the structure; a period denotes no cleft. A bilateral cleft lip with a complete unilateral cleft of the secondary palate, with incomplete clefting of the lip and alveolus on one side would be represented as LAHSal. This system is currently used for the outcomes registry of the American Cleft Palate and Craniofacial Association.

**Microform Cleft Lip**

The microform cleft (Fig. 23.1A) is characterized by a furrow or scar transgressing the vertical length of the lip, a vermilion notch, imperfections in the white roll, and varying degrees of vertical lip shortness. Nasal deformity may be present and is sometimes more extensive than the associated lip problem. Surgery is generally indicated but is approached cautiously to avoid a surgical deformity worse than the congenital defect. If there is isolated disruption of the orbicularis oris sphincter, it can be repaired through an intraoral approach.

**Unilateral Incomplete Cleft Lip**

Unilateral incomplete clefts (Fig. 23.1B) are characterized by varying degrees of vertical separation of the lip, but they all have in common an intact nasal sill, or Simonart band. They require the same surgical technique as a complete cleft lip in order to restore normal nasal and lip anatomy, although the degree of dissection can be tailored to the magnitude of the deformity. As with complete clefts, the best time to address the associated nasal deformity is at the time of the primary lip repair.

Unilateral Complete Cleft Lip

Unilateral complete clefts (Fig. 23.1C) are characterized by disruption of the lip, nostril sill, and alveolus (complete primary palate). Unlike the incomplete cleft lip there is no Simonart band connecting the alar base to the footplates of the lower lateral cartilages of the nose; consequently, the abnormal attachments of the orbicularis oris muscles on either side of the cleft cause a variable degree of collapse of the lower lateral cartilage framework and an associated increased nasal deformity. The critical factors for evaluating unilateral complete clefts are the position of the lesser and greater alveolar segments, the vertical height of the lateral lip element, and the degree of associated nasal deformity. The alveolar (maxillary) segments assume one of four positions: (a) narrow-no collapse; (b) narrow-collapse; (c) wide-no collapse; (d) wide-collapse. “Wide” is determined by an alveolus position lateral to the desired alar base position (i.e., with lip closure the alar base is sitting in the cleft). “Collapse” refers to a palatal displacement of the lateral maxillary segment as predicated by the arch configuration of the medial, noncleft dental ridge.

Clefts characterized as “narrow-no collapse” with minimal nasal deformity are treated with presurgical taping to prevent widening of the cleft with growth and feeding, prior to a primary cleft lip repair with primary tip rhinoplasty. If a gingivoperiosteoplasty is to be performed at the same time, a molding plate can be used to optimize contact of the opposing alveolar ridges. Clefts characterized as “narrow-collapse” or “wide-collapse” benefit from presurgical molding to create the desired arch form, alveolar contact, and nasal anatomy at the time of surgery. Clefts characterized as “wide-collapse” or “wide-no collapse” must be assessed closely by the dental members of the cleft team. If they feel that these cases are deficient in arch mesenchyme, presurgical orthopedics is used to align the arch segments by correcting the collapse, but not to close the alveolar cleft since this will result in a constricted arch. External taping can be used to correct the alar base position over the maintained arch form. The use of presurgical orthopedics or aggressive presurgical taping has eliminated the need for preliminary lip adhesion surgery at most centers. The primary benefit of a balanced noncollapsed arch configuration at the time of primary lip repair is the decreased tension on the lip repair and the secondary benefits to the nasal anatomy by providing a stable skeletal base.

Complete Bilateral Cleft Lip

The most obvious aspect of a complete bilateral cleft is the protruding premaxilla (Fig. 23.1D). Because of the lack of connection of the premaxilla with the lateral palatal shelves, the premaxilla has not been “reined back” into alignment with the lateral arch segments during fetal development. At the time of birth, the premaxilla protrudes on a vomerine stem. Uncontrolled growth at the premaxillary suture results in overprojection of the premaxilla, with or without rotation and angulation of the segment. Just as the premaxilla is not reined back by the lateral palatal shelves, the lateral palatal shelves are not pulled forward by their attachment to the premaxilla. Without the intervening premaxilla to maintain arch width, the lateral palatal shelves collapse toward the midline. The severity of this disruption of arch morphology varies, and will dictate the tension on the repair, the degree of dissection required, and, ultimately, the final aesthetic result unless it is corrected with presurgical orthopedics.

The anterior nasal spine is poorly formed or absent in the bilateral cleft lip deformity, resulting in a retruded area under the base of the septal cartilage and recession of the footplates of the medial crura. The footplates of the lower lateral cartilages are displaced posteriorly and laterally, which in turn pulls the normal junction (genu) of the medial and lateral crura apart resulting in a broad, flat nasal tip. The recession of the medial crural footplates, along with lateralization of the domes, and deficient skin, produces the typical “absent columella.” The most anterior and inferior extent of the
frononasal process, which normally contributes to the skin between the philtral columns of the lip, forms a wide, short disk, called a prolabium, that appears to hang directly from the nasal tip skin. In conventional techniques, linear distance from the inferior tip of the prolabium up to the nasal tip is inadequate to reconstruct both central upper lip and columellar length. This vertically limited tissue is used to create the central lip element at the cost of inadequate columella length and tip projection. A major benefit of nasoalveolar molding (NAM) is the ability to presurgically lengthen both the columella skin and the prolabium, creating enough skin to reconstruct the central lip length without compromising nasal tip projection.

**Incomplete Bilateral Cleft Lip**

Occasionally, bilateral clefts are incomplete with a near-normal nose, a normally positioned premaxilla, Simonart bands across the nasal floors, and clefts involving only the lip (Fig. 23.1E). In such circumstances, a rotation-advancement approach, or a triangular flap approach similar to that used in unilateral repairs, can be used either in a single-stage or a two-stage operation. In two-stage repairs one side is closed first, allowed to heal, and then the other side is repaired a short time later. Symmetry is difficult to achieve with a staged approach, and we prefer a single-stage procedure with a bilateral straight-line technique as described later in the chapter. More patients have complete clefts on one side and incomplete clefts on the other. These cases have both the nasal deformity of a unilateral complete cleft lip and the paucity of lip tissue of a bilateral cleft. If there is a discrepancy in columella height between the two sides, we will consider a rotation-advancement repair on the complete side to increase columella and a straight-line closure on the incomplete side.

**Cleft Lip and Palate**

The primary palate consists of the lip, alveolus, and anterior palate back to the incisive foramen. The secondary palate consists of the hard and soft palates from the incisive foramen back to the uvula. The presence of a cleft palate introduces feeding difficulties, concerns regarding speech development, and the possibility of impaired facial growth. The width of a primary palate cleft and the degree of collapse are typically increased in the presence of a cleft of the secondary palate. The family is counseled about the anticipated increased number of surgical operations that will be required if a cleft palate is present: primary cleft palate repair with intravelar veloplasty; possible secondary surgery on the palatopharyngeal muscle sling, such as a sphincteroplasty or pharyngeal flap; and possible orthognathic surgery at skeletal maturity. The abnormal attachment of the muscles of the soft palate in a cleft palate alters the tension on the pharyngeal drainage of the eustachian canal, increasing the incidence of ear infections. Myringotomy and grommet tube placement is performed in the majority of infants at the time of either the lip repair or the palate repair to prevent the development of hearing abnormalities.

**Isolated Cleft Palate**

The infant with isolated cleft palate is examined carefully to ascertain if there are manifestations of the Pierre Robin sequence (micrognathia, glossoptosis, and airway obstruction). The etiopathogenesis of the cleft palate in the Pierre Robin sequence is thought to be obstruction of the palatal shelves as they swing from a vertical to horizontal orientation during palate fusion. The micrognathia and associated glossoptosis causes this obstruction, resulting in the characteristic wide “horseshoe” cleft palate associated with this sequence. If the Pierre Robin sequence is present, appropriate measures are instituted, the mainstay of which is prone positioning. In severe cases, treatment may include around-the-clock prone positioning, nasopharyngeal airway protection, gavage feedings, and apnea monitoring. Very few of these patients will require temporary endotracheal intubation or tongue–lip adhesion. In extreme cases, tracheostomy is required to manage the airway if the mandible is not lengthened surgically by early distraction osteogenesis. In Pierre Robin patients, palatoplasty may be delayed for several months, compared to other cleft palate closures, to ensure adequacy of the airway.
Submucous Cleft Palate

The submucous cleft palate is traditionally defined by a triad of deformities: a bifid uvula, a notched posterior hard palate, and muscular diastasis of the velum. Submucous clefts vary considerably, however, and muscular diastasis can occur in the absence of a bifid uvula. The majority of patients with submucous cleft palate are asymptomatic, although approximately 15% will develop velopharyngeal insufficiency (VPI). VPI correlates with short palatal length, limited mobility, and easy fatigability of the palate. Because the majority of patients with submucous cleft palate remain asymptomatic, a nonoperative approach is recommended until speech can be adequately evaluated.

Presurgical Orthopedics

The cross-specialty collaboration between plastic surgery and dentistry has produced some of the most exciting advances in cleft care. It has also generated some of the most animated discussions about the perceived advantages and disadvantages of presurgical orthopedics in the literature and at national conferences. Because this field continues to be controversial, it is important to relate advances in presurgical orthopedics from a historical perspective.

Historical Perspective of Combined Presurgical and Surgical Treatments

Surgeons have long recognized the challenge of the bilateral cleft deformity. The main obstacles to the repair are the protruding premaxilla and the deficient columella. During the 16th, 17th, and 18th centuries, the surgical treatment involved excision of the premaxilla followed by a surgical union of the prolabium to the lateral lip segments. At a later age, prosthetic replacement of the anterior dentition was recommended to improve facial appearance. In the 19th century, surgeons finally accepted that excision of the premaxilla removed the upper incisors and deprived the lip of bony support, causing midface deficiency, maxillary constriction, malocclusions, and an apparent mandibular prognathism. The focus became preservation and retraction of the premaxilla to achieve optimal lip repair. Two treatment philosophies evolved: surgical correction alone and surgical correction following presurgical orthopedics.

Surgical Correction

Surgical options for premaxillary retraction included fracture of the vomer, resection of part of the vomer or nasal septum, partial resection of the anterior portion of the premaxilla, and a full-thickness vertical incision of the septum, which allowed the proximal and distal segments to slide over each other. Although these techniques achieved the primary goal of retracting the premaxilla, they were associated with significant complications. Both long-term clinical observations and animal studies demonstrated that resection of the nasal septum produced severe growth arrest of adjacent bones. The technique had other limitations, including lingual inclination of upper incisors caused by a lingually displaced premaxilla, nasal airway obstruction, and flat facies.

Another surgical approach that attempted to achieve premaxillary retraction and lateral segment approximation was lip adhesion, which is still used at some centers. Johanson and Ohlsson (1961) described the use of lip adhesion before primary bone grafting. Millard (4) reported the use of lip adhesion in the upper third of the cleft lip segments in preparation for the rotation-advancement technique of final lip closure. Randall (1965), using short, broad, triangular flaps, claimed that closure of the soft tissues molded the underlying bony structures, reduced tension in the lip, and repositioned the alar base. Randall undermined the lateral lip if necessary when there was a large cleft gap.

The disadvantages of lip adhesion include the risks of an additional surgical procedure, scarring of the involved tissue, loss of the local mucosal flaps used in some techniques for nasal lining repair, and potential dehiscence of the surgical site. In addition, the tension of the surgically adhered lip over the
alveolar segments is an uncontrolled force that does not always align the segments in an ideal position, frequently causing collapse of the dental arches. Lip adhesion should be limited to those cases in which the maxillary segments are expanded without collapse. If the segments are medially collapsed, lip adhesion is of no use and a technique that provides alveolar expansion is preferable.

**Presurgical Infant Orthopedics**

The concept of presurgical treatment originated in the 16th century, when excision of the protruded premaxilla in bilateral clefts was the recommended treatment. Dissatisfied with the long-term results of this treatment modality, surgeons and dentists explored new avenues to achieve more optimal results.

Franco (1561) described a head cap for extraoral therapy. Hoffmann (1686) used a head cap with facial extensions over the cheeks and lips to narrow the cleft by pressing over the premaxilla. Louis (1768), Chaussier (1776), and Desault (1790) used bandages over the prolabium to simulate muscle retraction, compressing the premaxillary region. In 1844, Hullihen, an American dentist, used facial adhesive strapping to “prepare” the cleft before surgery. He believed that closing the alveolar cleft prior to surgery during the first months of life was crucial in order to properly perform lip closure. Other early contributors to this field include Von Bardeleben (1868), who used a compression bandage with a bonnet; Thiesch (1875), who used rubber bands; and Von Eschmarck and Kowalzig (1892), who employed an elastic band attached to a head cap. Brophy (1927) adapted an intraoral approach, passing wires through the alveolar bone proximal to the cleft on both sides. By slow tightening of the wires, he achieved approximation of the segments.

The concept of modern presurgical infant orthopedics (PSIO) started with the work of McNeil, who, disappointed with the maxillary collapse created by the available techniques, used an oral prosthesis similar to an obturator to approximate the cleft alveolar segments. In his technique, a maxillary impression was taken of the newborn and an acrylic appliance was made from a plaster model that was cut and modified with the cleft gap slightly closed. By repeating this step and frequently modifying the appliance, McNeil was able to close not only the alveolar gap, but also the hard palatal cleft by influencing bone growth direction. He believed that alveolar and palatal surgery could be avoided completely, implying that a soft tissue and even a bony continuity could be achieved. He also stated that the technique improved speech function, feeding, and deglutition, and could eliminate the need for orthodontic treatment. McNeil’s exaggerated claims damaged the credibility of the technique, and controversy has surrounded the subject ever since. Moved by the intrigue, W.R. Burston (1958) evaluated McNeil’s work and became one of the most loyal proponents of the technique. At the same time, Schuchardt used the method, especially in preparation for primary bone grafting.

Many variations in presurgical orthopedic techniques have evolved during the last 40 years. The appliances can be described as active or passive, although there is no uniform consensus or universal agreement on their classification. Huebener and Liu classified the appliances as (a) presurgical versus postsurgical, (b) active versus passive, and (c) extraoral versus intraoral.

Generally, active appliances use a hard acrylic plate and controlled forces, sometimes from extraoral traction (bonnet with straps), to move the maxillary alveolar segments into approximation. One of the best-known active appliances is the pin-retained variety used by Latham (1980), which is designed to exert a forward force to the lesser posterior segment of the unilateral cleft maxilla. It consists of a two-piece maxillary splint that overlies the palatal shelves and is retained by short medial pins. An expansion screw connecting the two pieces can be moved to adjust the widths of the lateral palatal segments. An orthodontic elastic chain is used to retract the premaxilla. By adjustment of these independent controls, the premaxilla is brought back into its proper position in the arch before the primary repair. The Latham device requires a surgical procedure to introduce the device and to remove it.

Passive appliances generally consist of an alveolar molding plate made of a hard outer shell and a soft acrylic lining. By gradual alteration of the tissue surface of the acrylic plate, the alveolar segments are gently molded into the desired shape and position by direction of alveolar growth. This method was initially described by Gnoinski and developed by Rosenstein (1963), Rosenstein and Jacobson (1967),
and Monroe (1968). The devices allowed continued growth by a passive molding action without permitting medial movement of the buccal segments. Once the segments were in proper position, early lip repair and bone grafting could be performed. This passive molding approach has evolved into the more recent technique of NAM, which addresses the alveolar, labial, and nasal abnormalities as discussed later in the chapter.

The Controversy

One of the most outspoken opponents of PSIO is Pruzansky. In 1964, he published a dissenting opinion challenging McNeil’s presurgical orthopedic technique. He believed that spontaneous retropositioning of the premaxilla following lip repair obviates the need for intervention with orthopedic devices. It is generally agreed that PSIO does not enhance growth of the maxilla, the orthodontic benefits are limited, and nonsurgical closure of palatal bone and soft tissue is impossible. Proponents of PSIO report that the aesthetic result of the cleft lip and nasal repair is improved and the number of surgeries are minimized. Opponents are concerned about the added cost and the risk of iatrogenic malocclusion and midface retrusion. Despite this ongoing controversy, presurgical orthopedics continues to be widely used, and it has been cited by Brogan and McComb as the superlative example of cooperation within the cleft rehabilitation team. Hotz et al. reported that, in 1984, 22 of the 32 cleft lip and palate rehabilitation centers in Zurich indicated that they used presurgical orthopedics treatment. In 1990, Asher-McDade and Shaw indicated that 40 of 45 British cleft palate teams reported the use of presurgical orthopedics. In a recent unpublished survey of cleft teams in the United States, Huebener and Marsh (1993) showed that appliance use has increased over the past 5 years.

The inability to resolve the controversy surrounding PSIO through clinical trials or reviews of the literature stems from a variety of problems. The variation in the treatment modality and in the timing and use of bone graft, as well as the absence of normative data are some of the difficulties encountered when comparing results from different centers.

The Effect of Presurgical Infant Orthopedics on Maxillary Growth

One of the most controversial issues surrounding PSIO is about its effect on maxillary growth. The dilemma becomes more complicated if the surgeon performs gingivoperiosteoplasty (closure of the alveolar cleft) at the time of the primary lip repair. Ross (1987) showed in a major multicenter study that there is no difference in facial growth between cleft patients treated with or without PSIO. On the other hand, Robertson (1983), in a 10-year follow-up study by a single surgeon, demonstrated that better facial growth was achieved in patients treated with PSIO than in control subjects. In another long-term single-surgeon study, Lee et al. (2004) showed that maxillary growth was not inhibited in patients ages 9 to 13 years who had previously undergone presurgical orthopedics and primary gingivoperiosteoplasty. In contrast, Berkovitch (2004) has been openly critical of the Latham and Millard technique of presurgical orthopedics, gingivoperiosteoplasty (GPP), and presurgical orthopedic, periostoplasty, lip adhesion (POPLA). He reports a higher incidence of anterior and buccal crossbite at 3, 6, 9, and 12 years of age after POPLA when compared to no presurgical orthopedics with no GPP. Millard (1999) reviewed this same clinical database and also reported a higher incidence of anterior crossbite in the POPLA group, but a lower incidence of buccal crossbite. He noted that the two groups had different orthodontic treatment protocols by different orthodontists, and that this could have a confounding effect on the results. These two reports focused on dental relationships as the outcome measure instead of facial skeleton landmarks, which are a more accurate representation of facial growth or impaired growth. It is important to distinguish between dental malocclusion and maxillary hypoplasia. Both can result in anterior crossbite, but dental malocclusion can be treated by orthodontics, whereas marked midface hypoplasia requires orthognathic surgery. This distinction emphasizes the need for a standard outcome measure for PSIO, and the difficulty in interpreting previous studies.

It is logical that restoring the normal anatomy of the maxillary segments presurgically allows lip repair
under less tension. Ross and MacNamera stated that one possible benefit of PSIO is that the lip surgery should be easier, enabling a more precise repair with less tension. If the aesthetic outcome is improved, this is a powerful incentive to adopt presurgical infant orthopedic procedures.

**Presurgical Nasoalveolar Molding**

Presurgical nasal and alveolar molding includes as its objectives the active molding and repositioning of the nasal cartilages and alveolar processes, and lengthening of the deficient columella. A description of the protocol for treatment of the patient with bilateral cleft deformity was introduced by Grayson et al. (1).

This modification of the traditional approach to presurgical molding plate therapy takes advantage of the plasticity of cartilage in the newborn infant during the first 6 weeks after birth. Matsuo, Hirose, and Tonomo postulated that the high degree of plasticity and lack of elasticity in neonatal cartilage is caused by high levels of hyaluronic acid, a component of the proteoglycan intercellular matrix. As the estrogen level increases, the level of hyaluronic acid increases and the elasticity of the cartilage decreases. With the neonatal levels of maternal estrogen highest immediately after birth, the period of plasticity is slowly lost during the first months of postnatal life. It is during this first 2 to 3 months after birth when active soft tissue and cartilage-molding plate therapy is most successful.

After the successful application of molding therapy to deformed auricular cartilage, Nakajima and Yoshimura, and Matsuo and Hirose applied the same method to the unilateral cleft lip nasal structures. A combined technique for nasal and alveolar molding has been refined, critically analyzed, and documented by the Cleft Palate Team of the Institute of Reconstructive Plastic Surgery at New York University Medical Center. This technique has been demonstrated to have a positive influence on the outcome of the primary nasal, labial, and alveolar repair, and has been adopted by an increasing number of cleft teams, including the Craniofacial Center at Children's Hospital in Seattle.

**Correcting the Unilateral Cleft Lip and Nasal Deformity with Nasoalveolar Molding**

In the unilateral cleft, the ipsilateral lower lateral cartilage is depressed and concave, and separated from the contralateral cartilage. This results in depression and displacement of the nasal dome and is associated with overhang of the ipsilateral nostril apex. The columella and nasal septum are inclined over the cleft with the base deviated toward the noncleft side. In addition, the orbicularis oris muscle in the lateral lip element contracts into a bulge with some fibers running along the cleft margin toward the nasal tip.

Shortly after birth, an impression of the intraoral cleft defect is made using an elastomeric material in an acrylic tray. A conventional molding plate is constructed on the maxillary study model from clear orthodontic resin. The molding plate is applied to the palate and alveolar processes, and secured through the use of surgical adhesive tapes applied externally to the cheeks and to an extension from the oral plate that exits the horizontal labial fissure. The molding plate is modified at weekly intervals to gradually approximate the alveolar segments. This is achieved through the selective removal of acrylic from the region into which one desires the alveolar bone to grow ("negative sculpting"). At the same time, soft denture liner is added to line the appliance in the region from which one desires the bone to be moved. The ultimate goal of this sequential addition and selective removal of material from the inner walls of the molding plate is to align the alveolar segments and achieve closure of the alveolar gap. The effectiveness of the molding plate is enhanced by adequately supporting the appliance against the palatal tissues, and by taping the left and right lip segments together between clinical visits.

The goal of presurgical NAM in the patient with unilateral cleft deformity is to align and approximate the alveolar segments, and correct the malposition of the nasal cartilages and the alar base on the affected side, as well as to idealize the position of the philtrum and columella. These changes benefit
the patient and surgeon by decreasing tension on the incision line and subsequent scar, controlling alveolar contact, facilitating creation of a natural curvature to the alar rim, and decreasing the amount of surgical dissection required. Alveolar approximation does occur to a variable degree after lip adhesion or after approximation of the orbicularis muscle with surgical repair of an unmolded cleft lip, but these changes are uncontrolled and often result in collapse of the lesser alveolar segment and variable cleft closure. With NAM, alignment and approximation of the alveolar segments is controlled to create a natural arch form.

NAM also permits the surgeon to offer the patient a limited-dissection GPP at the same time as the cleft lip repair if the alveolar edges are vertically aligned and in close contact (2). The gingivoperiosteal flaps are raised within the alveolar cleft, without any dissection on the anterior face of the alveolus. The two sets of flaps create a periosteum-lined “tunnel” between the exposed bone surfaces of the alveolar cleft. This promotes tissue-guided regeneration as bone forms across the alveolar gap. This limited dissection GPP following NAM differs markedly from the previously reported GPP techniques of Skoog, which required extensive subperiosteal dissection to achieve closure of the unmolded cleft.

The nasal changes of NAM are achieved by the use of a nasal stent rising from the labial vestibular flange of the acrylic intraoral molding plate. The medio-lateral position of the nasal stent is adjusted as it lifts the nasal tip. The shape of the nostrils and alar rims is carefully molded to resemble the normal configuration of these structures through modifications gradually made to the nasal stents.

**Correcting the Bilateral Cleft Lip and Nasal Deformity with Nasoalveolar Molding**

In the bilateral cleft, the alar cartilages have failed to migrate up into the nasal tip and cause growth of the columella. The prolabium lacks muscle tissue and is attached on the end of the short columella. The alar cartilages are positioned along the alar margins and are stretched over the cleft as flaring alae. In the complete bilateral cleft, the premaxilla is suspended from the tip of the nasal septum and the lateral alveolar segments remain behind.

The objective of presurgical nasal and alveolar molding in the patient with bilateral deformity is to lengthen the columella, elongate the prolabium, reposition the nasal cartilages toward the tip, and align the alveolar segments, including the premaxilla. This is accomplished through the use of nasal stents that are based on the border of a conventional oral molding plate, adhesive surgical tape, and elastic bands (Fig. 23.2). The first stage of treatment involves repositioning of the everted premaxilla into the space between the lateral alveolar segments using progressive modifications of an acrylic intraoral plate in conjunction with elastic bands that are adhered to the cheeks. In the second stage, as the alveolar segments gradually approximate one another, the nasal stents are built from the anterior rim of the oral plate and enter the nasal apertures. This provides support and gives shape to the dome and alar cartilages in the immediate neonatal period. The nasal stents advance the lateral alar cartilages into the nasal tip and provide stretch to the columellar skin. In the bilateral cleft deformity, the columella at birth is often from 0 to 2 mm in length; following NAM it is expanded to 4 to 7 mm. The normal columellar, length for children at this age is 3.2 mm. Overcorrection of the columellar length is intended to account for some postsurgical relapse. Attached to the two nasal stents, is a horizontal prolabial band that stabilizes the columellar base at the nasolabial angle, serving as a fulcrum for the distracting forces of the nasal stents upwards on the columella envelope, and of the labial tapes downward on the prolabium. This controlled elongation of the linear distance from the top of the columella to the bottom of the prolabium provides sufficient tissue for the surgeon to create both nasal projection (columella) as well as central lip length (prolabium), a feat that is frustratingly difficult in an unmolded nose.

**Evaluation of Nasoalveolar Molding**

Previous studies on presurgical infant orthopedics, including the Latham device, cannot be extrapolated to the technique of NAM. Previous studies focused on dental relations, such as crossbites, as the outcome measure of facial growth, instead of maxillary dimensions, which are a more
direct measure. NAM also has implications on nasal morphology, soft-tissue relationships, and the ability to perform a limited dissection gingivoperiosteoplasty. These were not considerations in previous studies. To address this lack of objective data on the outcome of NAM patients, the Cleft Team at NYU evaluated their cases with the following findings:

1. The presurgical alignment and correction of deformity in the nasal cartilages minimize the extent of primary nasal surgery required and therefore minimize the extent of scar tissue formation, leading to more consistent postoperative results. An average of 4 years after surgery, superimposition analysis of molded and unmolded unilateral cleft noses demonstrated increased symmetry in the group undergoing NAM as compared with the control group who had alveolar molding without nasal molding.

2. Presurgical closure of the alveolar gap enables the surgeon to perform a GPP at the time of primary lip repair. Santiago et al. have reported that GPP eliminates the need for secondary alveolar bone grafts during the period of the mixed dentition in more than 60% of the cases studied.

3. The quality of the generated bone in the alveolar cleft of NAM + GPP cases is equal to the quality of unmolded clefts that underwent secondary bone grafting. Of the 40% of NAM + GPP clefts that required secondary bone grafting, the outcome of the secondary bone graft was superior to unmolded clefts, presumably because of the intact nasal floor and the bone bridge that had been generated as a result the GPP.

4. The distance from posterior to anterior nasal spine as a measure of maxillary projection an average of 11 years after the primary surgery was not significantly different between molded patients who had a GPP at the time of cleft lip repair and unmolded patients who did not. Although it will be a few more years before this cohort can be examined at skeletal maturity, after this initial study, the alveolar cleft of the control group was closed by secondary bone grafting. Thus both groups will have intact alveolar arches on future analysis.

5. There is a cost benefit to NAM with GPP. The cost savings in not needing to perform secondary alveolar bone grafting in 60% of infants with a GPP outweighs the orthodontic fees for presurgical NAM. This study did not include the possible additional cost benefit of decreased secondary nasal revisions in molded noses.
FIGURE 23.2. Nasoalveolar molding (NAM) of the bilateral cleft deformity. 

A: Bilateral complete cleft lip and palate in a newborn. B: Nasoalveolar molding plate with nasal extensions. The projecting buttons are used to secure the plate to the patient’s cheeks with tape and elastics. The nasal extensions are not added to the molding plate until the alveolar cleft is less than 5 mm wide so as to avoid overstretching the nostril. C: Molding plate and nasal extensions in place. D: Presurgical result of the same patient after a course of NAM. The alveolar segments and premaxilla are aligned, the columella is lengthened, the alar bases are in a more medial position, the alar rims are curved, and the prolabium is of sufficient size to reconstruct the central lip with minimal tension.

Primary Unilateral Cleft Lip Repair

Numerous methods have been described for repair of the cleft lip deformity. Early techniques involved a straight-line closure (Fig. 23.3), and these procedures still find applicability in the repair of microform (forme fruste) clefts. Repairs involving a combined upper and lower lip flap were advocated by Skoog and Trauner and Trauner. Modern repairs have in common the use of a lateral lip flap to fill a medial deficit, a concept that can be accredited to Mirault. The LeMesurier repair involves a lateral,
quadrilateral flap, whereas the Tennison repair employs a lateral triangular flap (Fig. 23.3) (3). Both procedures introduce tissue in the lower part of the lip.

In 1955, Millard described the concept of advancing a lateral flap into the upper portion of the lip combined with downward rotation of the medial segment. The technique preserves both the Cupid's bow and the philtral dimple, and it has the additional advantage of placing the tension of closure under the alar base, thereby reducing flare and promoting better molding of the underlying alveolar processes (4).

At our two institutions we employ a modification of the technique initially described by Mohler, which, in turn, is based on the technique of Millard (5). Compared to the traditional Millard technique, this technique minimizes the alar base skin incisions and places the back-cut used to rotate the medial lip element at the base of the columella instead of the upper lip. With these modifications, the upper lip scar parallels the contralateral philtrum instead of curving across the philtral groove.
There is no agreement on the ideal timing and the technique of repair among established and experienced cleft surgeons. This underscores the fact that more than one treatment plan is acceptable, and that comparable outcomes can be achieved with different philosophies. Successful approaches have in common a surgeon who is knowledgeable about the variation in abnormal anatomy among clefts, is comfortable with the details and limitations of their technique, and is able to combine these two qualities to achieve the optimum surgical result.

The remainder of this section focuses on the modified Mohler technique used by the authors.
Timing and Treatment Planning
Whenever possible, all complete unilateral cleft lips undergo preoperative NAM at our institutions. Presurgical orthodontic treatment is initiated in the first or second week following birth, with the maximum response occurring during the first 6 weeks. The primary lip repair is scheduled when the patient is approximately 12 weeks of age, at which time closure of the anterior nasal floor and a primary tip rhinoplasty are also performed. If the alveolar segments are appropriately aligned and <2 mm apart, the family is offered a GPP at the time of the surgery. Bone grafts are not employed with early closure of the alveolus. If collapse is present or the gap is too wide, the GPP is deferred.

Correction of the nasal deformity in unilateral clefts is coupled with the rotation-advancement repair. Septal repositioning and nasal osteotomies are deferred until late adolescence unless the deformity is severe, in which case they are performed concomitantly with secondary bone grafting of the alveolar cleft (if a GPP was not done) at the time of mixed dentition. We believe that it is important to minimize the number of secondary surgeries to the nose during the growth phase to minimize scarring and to optimize the final result of a formal open rhinoplasty in adolescence.

Anesthesia
General anesthesia is used for all stages of lip repair. A straight, cuffed endotracheal tube is taped to the chin by the surgeon to avoid distortion of the lower lip and alteration of landmarks. The eyes are protected with Tegaderm patches. After markings, 0.5% lidocaine with 1:200,000 epinephrine is injected in the planned dissection planes of the lip, in the supraperiosteal plane of the cleft side maxilla, and between the skin and cartilage of the planned nasal dissection. Accurate injection with a minimal volume of fluid maximizes hemostasis and facilitates dissection.

Unilateral Complete Cleft Lip Operative Technique
The markings for the modified Mohler rotation-advancement repair are applied as shown in Figure 23.4. The depth of the Cupid’s bow on the medial lip segment is marked as point 1, with point 2 being the white roll at the height of the Cupid’s bow on the non-cleft side, and point 3 being equidistant on the cleft side. Ideally, the distance between each point should be approximately 2.5 mm, for a final Cupid’s bow width of 5 mm; however, this can be adjusted based on the patient’s anatomy. Point 4 is selected by a number of considerations, the least important of which is the traditional technique of matching the distance from the commissure to Cupid’s bow on the non-cleft side. Instead, it is selected by matching the vermilion and white roll thickness, or bulk of the lateral lip segment with that of the medial site at the Cupid’s bow peak. This point should coincide as closely as possible with the point on the white roll that intersects the arc of a line drawn from the alar base whose length equals the vertical lip height of the non-cleft side (the height from point 4 to point 5 equals that from point 6 to point 2). The vertical incision of the lateral lip segment that will be approximated to the medial segment to reconstruct the philtral ridge originates from point 4, crossing perpendicular to the white roll, then curves sharply toward point 7 at the nasal sill. The triangle formed by points 4, 5, and 7 is isosceles, with the height from point 4 to point 7 equaling that from point 4 to point 5. It is important that the base of this isosceles triangle does not violate the nasal sill. Consequently, the line from point 5 to point 7 tends to slope slightly inferior. Point 4 can be chosen on most cleft lips using these two guidelines. In some cases, however, the lateral lip element is vertically deficient, resulting in a point 4 that is too laterally displaced (too close to the commissure) to achieve a minimal tension repair. In these cases, the curvature of the lateral vertical incision from point 4 to point 7 can be increased to lengthen the lateral lip segment, or the horizontal incision under the cleft side alar base from point 5 to point 7 needs to borrow skin from the nasal sill. If the dry red lip thickness at point 4 is more than 1 mm less than that under the non-cleft Cupid’s bow, a triangular mucosal flap of dry vermilion from the medial lip segment can be interdigitated into a corresponding cut at the dry-wet lip junction on the lateral lip segment.
For the medial lip segment incisions, point 8 is chosen as the location of the back-cut of the C flap. Unlike the traditional Millard repair, this point is located approximately 1 mm up on the columella and three-fifths along the width of the columella, toward the non-cleft side. This allows the back-cut scar to be hidden at the base of the columella, instead of on the upper lip. It also creates a vertical scar that mirrors the non-cleft philtral ridge and does not violate the philtral groove. The incision from point 3 to point 8 is the vertical philtral incision of the medial lip segment and defines the non-cleft border of the C flap. Unlike the traditional Millard repair, this incision has only a slight medial curvature in order to create a vertical philtrum. The cleft border of the C flap parallels the junction of the medial lip skin and the oral mucosa. It is important not to include any mucosa in the C flap, as it will be rotated into the base of the columella to fill the skin deficiency after downward rotation of the medial lip segment. The cleft border of the C flap terminates at the anterior aspect of the septum, behind the footplates of the lower lateral cartilages.

Points 3 and 4 on the white roll are tattooed with needle and ink to facilitate alignment at the end of the repair. The lip is then infiltrated with lidocaine and epinephrine as described above (see Anesthesia). After the skin incisions are complete, the red lip portions of the medial and lateral segments are everted to equal fullness, and a no. 11 blade is used to transect the red lip from point 3 and 4 respectively, and the superior labial arteries are cauterized. The anterior border of the L flap is marked by the incision from points 4 to 7. This continues as a back-cut of the lateral nasal wall behind the lateral crus of the cleft lower lateral cartilage. The posterior border of the L flap is at the level of the palatal shelf inside the nose, such that when the L flap is elevated, it is a posteriorly based mucosal flap pedicled off the lateral nasal wall, posterior to the lateral crus of the lower lateral cartilage. The base of the L flap is left thick by dissecting in the subperiosteal plane on the piriform aperture. With elevation of the L and M flaps in a submucosal plane, the underlying orbicularis muscle can be judiciously separated from the overlying skin. With dissection of the muscle of the medial lip segment, care must be taken not to violate the midline of the philtrum to avoid distorting the natural groove. The red lip mucosa and white roll are not separated from the underlying muscle in order to permit normal animation of this area. The nasal and perioral components of the orbicularis oris muscle

**FIGURE 23.4.** Markings for unilateral complete primary cleft lip repair. M, medial mucosal flap; L, lateral mucosal flap; C, central cutaneous flap. See text for details.
are separate at the exposed muscle edge of the lateral lip segments. The nasal component will be used to control the position of the alar base, and the perioral component will be rotated inferiorly to join the medial lip muscle in constructing the transverse orbicularis oris muscle sling.

The medial lip segment is lengthened and rotated inferiorly by sequentially releasing the skin with a back-cut at the base of the columella described above, then the muscle with a separation of the nasal and perioral components of the orbicularis oris, and, finally, the mucosa at the frenum. Care is taken not to fully release the frenum if possible to avoid creating a long lip deformity. At the end of the medial lip segment release and rotation, points 1, 2, and 3, the landmarks of the Cupid's bow, should be aligned horizontally with minimal tension.

Angled Converse nasal tip scissors are used to dissect between the footplates of the lower lateral cartilages by accessing them underneath the C flap (Fig. 23.5). A vertical incision is made through the nasal mucosa in the area of the membranous septum between the anterior edge of the cartilaginous septum and the posterior edge of the ascending limb of the lower lateral cartilage within the cleft-side nostril. This releases the cleft-side lower lateral cartilage footplate, allowing differential elevation of this cartilage and associated nasal tip relative to the non-cleft side. Scissor dissection then continues between the ascending limbs of the lower lateral cartilages, over the nasal tip, and along the alar component of the cleft-side lower lateral cartilage. The skin is carefully separated from the lower lateral cartilage over the alar rim to allow the skin envelope to redrape when the cartilage is repositioned. This dissection pocket between the cartilage and overlying skin is extended up to the upper lateral cartilage of the non-cleft side. This continuous dissection plane between the non-cleft upper lateral and cleft lower lateral cartilages will later be used to place subcutaneous Tajima suspension sutures to adjust the alar rim contour.

The final dissection involves releasing the abnormal attachments of the cleft alar base to allow tension-free approximation across the cleft. An upper gingivobuccal sulcus incision is performed on the cleft side and continued as a supraperiosteal dissection over the face of the maxilla. Through this incision, the abnormal fibrous attachments of the cleft side accessory nasal cartilages to the lateral piriform aperture are released. Along with the small back-cut in the nasal lining behind the cleft lower lateral cartilage, this will allow tension-free mobilization of the lateral lip segment and alar base in even the widest unilateral clefts. All areas are checked carefully for hemostasis before the closure begins.

Closure begins with the nasal floor. The L flap is rotated, trimmed, and sutured into the defect created in the lateral nasal lining when the cleft alar base is advanced into the appropriate position (Fig. 23.6). With the alar base advanced, the inferior edge of the L flap and lateral nasal lining is sutured to an opposing septal mucosal flap to close the nasal floor from the nasal sill back to the incisive foramen. At the end of this closure, the posterior displacement of the cleft alar base should be corrected. Closure of the nasal floor to the incisive foramen at the time of primary lip repair will avoid any oronasal or nasolabial communication after the remaining nasal floor reconstruction during the later cleft palate repair. If this detail is omitted from the lip repair, the child will be forced to deal with an anterior nasolabial fistula until closure can be performed at the time of secondary alveolar bone grafting.
FIGURE 23.5. Primary tip rhinoplasty for unilateral cleft deformity. A: Converse nasal tip scissors are used to access the interdomal space by dissecting underneath the C flap between the footplates of the lower lateral cartilages. Note how the back-cut at the base of the columella is opening to allow elevation of the cleft alar rim. The C flap will later be inset into this defect. B: The scissors are then angled to dissect down to the alar rim on the cleft nostril. See text for details.

Lip construction is achieved by everting the red lip on either side of the cleft to even fullness and then advancing and closing the lateral lip segment mucosa to the medial lip segment mucosa. The M flap can be rotated into the defect from the releasing back-cut at the frenum if necessary, or it can be used to augment the labial sulcus. After the lip mucosa is closed, the white roll should be aligned across the cleft, and the red lip should have equal fullness. If the lateral red lip is thin, the lateral mucosal flap had not been adequately advanced toward the midline. The perioral components of the medial and lateral lip segments are approximated across the cleft using buried horizontal mattress sutures of 5-0 Vicryl to create a philtral ridge and construct the oral sphincter. The vertical incision of the lip is closed with sparse, nonstrangulating, 6-0 interrupted nylon sutures, with care taken to ensure that the tattooed marks of the white roll on either side of the cleft are aligned.
FIGURE 23.6. Nasal lining release and inset of L flap. A: The constricted lateral nasal lining is released with an incision behind the lateral crus, and the mucosal L flap is elevated from the lateral lip element. B: The L flap is pedicled off the lateral nasal wall and inset into the lining defect to support the new position of the alar base.

At this point of the repair, the lip anatomy has been corrected, leaving the nasal deformity. Either 4-0 polydioxanone suture (PDS) or clear nylon is used to secure the dermomuscular pennant under the cleft alar base to the muscle and periosteum of the anterior nasal spine. Two or three sutures are used to place the cleft alar base in the desired medial position. A retractor is used to slightly overcorrect the cleft alar rim and underlying lower lateral cartilage in an advanced and superior position. This slides the released cleft lower lateral cartilage footplate toward the nasal dome in relation to the non-cleft side. A series of 4-0 PDS transfixation sutures are used to secure this new relationship of the ascending limbs of the lower lateral cartilages to the anterior septum. This elevation of the cleft side alar rim and lengthening of the columella leaves a defect from the back-cut at the base of the columella. The C flap is trimmed to fit and rotate into this defect. The rotation point of the C flap creates a natural flare to the base of the columella of the cleft nostril.

The final sculpting of the nostril shape is achieved with 4-0 PDS subcutaneous Tajima suspension suture. The needle enters the nasal surface of the cleft lower lateral cartilage at the point of desired elevation, enters into the previously described nasal tip dissection pocket, exits into the non-cleft nostril at the level of the upper lateral cartilage, and then returns on its path, such that tightening of the suture will elevate the cleft alar rim. Lateral alar cinch sutures of 4-0 PDS can also be used to contour the lateral alar rim and nasal lining in the new position, by exiting and entering the same percutaneous hole in the alar groove. The number of suspension and cinch sutures required will depend on the degree of the deformity. We perform a GPP if the alveolar segments are in appropriate alignment following nasoalveolar molding.

With this approach, the lip and nasal deformities can be addressed in a single surgery (Fig. 23.7).

Unilateral Incomplete Cleft Lip Operative Technique

The unilateral incomplete cleft lip deformity is treated with the same surgical technique and dissection that was described for the complete cleft lip, but with a few modifications. Failure to address all the lip and nasal abnormalities in the incomplete cleft lip with the same detail paid to the wide complete cleft will result in a suboptimal result.

Compared to the complete cleft lip repair, the incomplete cleft repair does not involve intranasal incisions. If possible, the nasal sill is not violated by the vertical incision. If the nasal base is wide compared to the non-cleft side, a small wedge can be removed from the nasal sill to create symmetry. If any nasal sill is resected, it is vital that the excision be minimal, because overresection with scarring will result in the recalcitrant micronostril deformity.

The L flap and M flap are not required for the incomplete cleft lip repair because the nasal floor is intact. To correct the alar base malposition, the abnormal attachments of the nasal cartilages to the piriform aperture must be released as in the complete cleft technique, but, also, the nasal floor lining must be dissected free from the piriform rim. The thin nasal floor is firmly attached to the edge of the piriform opening, and can easily be perforated if care is not taken. Failure to release the nasal lining from the underlying bone will make it impossible to mobilize the alar base into the desired advanced and medial position.

The nasal deformity is addressed with the same dissection as the complete cleft; however, the vertical nasal lining incision behind the ascending limb of the cleft lower lateral cartilage is not available for improved access to the nasal tip. Angled nasal tip scissors are used to access the nasal tip between
the footplates of the lower lateral cartilages; if necessary, the nasal tip can be approached laterally from the supraperiosteal maxillary dissection plane.

**Microform Cleft Operative Technique**

The critical factor when evaluating the microform cleft is the vertical height of the lip. If the vertical height of the affected side approximates that of the normal side, imperfections in the vermilion along the skin furrow can be eliminated with an elliptical excision and a straight-line repair. Triangular flaps of the white roll and vermilion can be used to balance the closure.

When the vertical difference exceeds 1 to 2 mm, the modified Mohler rotation-advancement repair described above (see Unilateral Complete Cleft Lip Operative Technique) is used. The additional scar underneath the sill and columella is preferable to a loss of definition in the involved philtral column, which invariably results with straight-line closure when the elliptical excision is extended to provide the desired lengthening.

![Image](image_url)

**FIGURE 23.7.** Mohler unilateral cleft lip repair. A: Preoperative complete cleft lip and nasal deformity. B: Postoperative result 9 months later.

The correction of a very mild nasal deformity is deferred in the microform cleft requiring a straight-line repair, as the repair does not necessitate a perialar incision. If the deformity remains minimal, treatment is postponed until late adolescence, when a definitive rhinoplasty is performed. With a moderate nasal deformity and with mild deformities requiring a rotation-advancement lip repair, correction of the nasal deformity is carried out with the lip repair.

**Lip Adhesion**

Lip adhesion is still used by some centers for wide clefts and those with lateral maxillary collapse that does not respond to presurgical maxillary orthopedics, but lip adhesion is not used by us. Supporters of lip adhesion believe that it improves maxillary arch alignment and enables a more predictable correction of the cleft nasal deformity in select patients. The improved nasal results are thought to be secondary to improved alar base arch support, which reduces the strain and relapse tendency for the mobilized lower lateral cartilage.
The adhesion is classified as a straight line muscle repair and begins with the complete marking of a rotation-advancement cheiloplasty. An L flap is elevated from the lateral segment beginning approximately 3 mm medial to the Cupid’s bow peak. This flap length provides adequate tissue for nasal release. The flap is turned 90 degrees into the nasal release along the lateral floor of the nose, which follows the piriform rim and the lateral portion of the nasal bones. A contiguous, maxillary sulcus incision is made through this nasal mucosal incision, and the lateral lip and cheek muscle mass is elevated in continuity from the maxilla and piriform aperture. The L flap is sutured into the nasal defect, and the lateral lip element is advanced medially for closure.

An M flap is also raised 3 mm from the Cupid’s bow peak to maintain symmetry of repair. The mucosal flap is based on the maxillary alveolus and is turned into the alveolar cleft to augment closure. All dissection is maintained outside the margins for primary lip repair. No medial muscle dissection is done at this stage.

Closure is achieved with sutures placed in the undissected orbicularis layer along the pared margin and is reinforced with a chromic catgut mucosal closure between the M flap and the lateral lip mucosa. Skin is generally closed with interrupted 5-0 chromic catgut, with sutures placed outside the markings for definitive cheiloplasty. The adhesion effectively closes the nasal sill and upper two-thirds of the lip. The forces from the overlying muscle closure have an immediate effect on the position of the alveolar segments.

**Postoperative Care**

The infants are kept in soft arm restraints for 2 weeks after lip repair. An infraorbital nerve block with bupivacaine can be used to minimize the early need for analgesics. Care must be taken not to overuse morphine in these patients.

Some surgeons prefer feedings with a catheter-tip syringe fitted with a small, red, rubber catheter for the first 10 days postoperatively to minimize strain on the muscle and skin sutures and to avoid trauma to the repaired velum. Many others do not impose restrictions and allow return to the preoperative routine immediately. Diet is advanced to full-strength formula or breast milk on the day of surgery to pacify the infant.

Suture line care consists of regular cleansing with half-strength hydrogen peroxide followed with a light coating of polymyxin B-bacitracin ointment. Sutures are removed on the fifth to seventh postoperative day. Postsuture removal taping and silicone scar gel can be used if desired. Parents are told to expect firmness in the lip scar and temporary shortening across the repair that generally becomes maximum 4 to 6 weeks after surgery. Scars typically soften between 3 and 6 months postoperatively.

**Primary Bilateral Cleft Lip Repair**

Bilateral cleft lip repair is recognized by surgeons as more difficult than a unilateral cleft lip repair. As previously described in this chapter (pages 203–204), the anatomy of the bilateral cleft lip deformity creates numerous challenges. Although the lip repair is made more difficult by the deficiency of skin and muscle overlying the premaxilla, it is the associated bilateral nasal deformity that was previously recalcitrant to correction. The treatment of the complete bilateral cleft and associated nasal deformity remains in transition. Only recently, because of NAM, have the results of one-stage primary bilateral cleft lip and nose repair begun to approach those of unilateral cleft lip and nose repair. Previous multistage techniques often produced a lip and nose that were still quite abnormal, with a confluence of scars at the lip-columella junction, a broad nasal tip, an unstable premaxilla, and often large nasolabial fistulas. Results fell short of ideal because the condition was viewed as a purely cutaneous deformity. Over the past decade, techniques have recognized the importance of addressing the contribution of the nasal tip cartilages to the cleft deformity. This shift from a skin-based to a cartilage-based paradigm has produced a number of techniques with improved outcomes.
**Construction of the Central Lip Vermilion**

There are two general methods for constructing the central lip vermilion. One involves using the mucosa visible on the inferior aspect of the prolabial skin to form the central vermilion, such as used in the Manchester repair. The original Manchester repair did not create an orbicularis oris sling across the upper lip, but instead sutured the muscle to the edges of the premaxilla. As there was no muscle under the prolabium or within the buccal mucosa, this approach did not provide sufficient bulk to serve as the central lip vermilion, and resulted in an abnormal appearance with animation of the central upper lip. A number of techniques have been described to address this limitation, including bringing strips of muscle across this area from the lateral lip, and de-epithelializing the buccal mucosa or subcutaneous tissue from the lateral sides of the prolabium and folding them behind the inferior prolabial mucosa. An advantage of using the prolabial or buccal mucosa to create the central vermilion is that very little bulk of the lateral segment of the vermilion is required, thereby decreasing the tension required for closure across the cleft. The disadvantages of this technique are that (a) there are two parallel scars across the vermilion, (b) the central red lip does not have sufficient bulk, resulting in a whistle deformity, and (c) the central buccal mucosa does not possess the same minor salivary gland distribution as the lateral vermilion tissues, often resulting in a dry, chapped, central vermilion segment.

A second approach is to use the vermilion tissue from the lateral lip segments to create the central vermilion as a variation of the technique described by Millard. The muscle of the lateral lip elements rotates down with the full-thickness vermilion flaps and can create a satisfactory central vermilion construction with a single vertical scar under the depth of the Cupid's bow. We prefer to include the white roll with the lateral lip segments that are used to create the central tubercle (Fig. 23.8). By placing two hemicircular incisions at the inferior aspect of the prolabial skin, the combination of the scar on top of the natural white roll and vermilion from the lateral elements produces a satisfactory Cupid's bow.
FIGURE 23.8. Repair of nonmolded bilateral cleft lip deformity. A: Preoperative markings. B: Dissection of nasal cartilages through bilateral alar rim incisions. Elevation of the prolabial flap also allows retrograde central access to the interdomal space. C: Immediate postoperative result. The columella length has been achieved by redistributing the nasal tip skin envelope. D: Nine months after the operation. There is good symmetry with minimal labial scars. The width of the prolabium and interdomal space have slightly increased at the expense of the columella height. Presurgery NAM may have minimized these postoperative changes.

Skin Paradigm of Bilateral Cleft Lip and Nose Repair

Until relatively recently, methods of bilateral cleft lip repair focused on the skin imbalance evident in the primary deformity. The columella is usually short or absent, whereas the prolabium hanging directly from the nasal tip skin is generally too broad and is vertically deficient. A natural approach is to take the excess in prolabial width and use it, usually in two stages, to create both a columella and a central white lip. We refer to this approach as the skin paradigm of bilateral cleft repair.

One early approach was to bring the lateral lip elements together in the midline to close the lip and to use the prolabium to elongate the columella. This approach has been abandoned, because it produces
an unnatural looking lip with a midline scar in place of a Cupid's bow and an upper lip that is tight horizontally. In most of the techniques described in this section,

the wide prolabium is split vertically in some fashion to provide skin for both the area of the white lip between the philtral columns, as well as to form a neocolumella.

The next evolution of techniques involved a staged approach to the lip and the nasal deformity. At the initial operation, the lip is approximated to the wide prolabial tissue. This is followed by a second stage in which the extra width of the prolabial tissue is advanced up into the columella. The lip reconstruction is addressed as either a one-stage or two-stage bilateral triangular or rotation-advancement closure, using the techniques described for unilateral deformities. This approach can result in a lip that is too long; it also produces scars that cross Langer lines and that frequently meet in the midline either under the columella (rotation-advancement approach) or above the Cupid's bow (triangular flap techniques). The muscle is either repaired underneath the prolabium to create an intact orbicularis sling across the cleft, or is attached to the sides of the prolabium. Advocates of not repairing the muscle across the cleft state that if the premaxilla is not retracted before lip repair, muscle repair is often difficult and requires much lateral undermining. Similarly, if the prolabium is hypoplastic, suturing of the muscle to its sides will result in stretching of the prolabium, which facilitates later use for the columella. Other surgeons believe strongly that the muscle should be repaired behind the prolabium. If the muscle is not repaired, the resulting lip is frequently too thin in the anteroposterior dimension. Similarly, the pull on the sides of the prolabium can cause it to get very wide in the postoperative period. In the currently employed one-stage cartilage paradigm repairs, which are described later in the chapter (page 215, Cartilage Paradigm of Bilateral Cleft Lip and Nose Repair) the muscle is always repaired under the prolabium, because widening of the interphiltral distance is undesirable.

After the first-stage procedure, the patient has a prolabium that is too wide and an absent columella. The second-stage procedure, when this extra prolabial width is used to create a columella, can be performed several months after the initial lip repair. The more common approach, however, is to wait until age 4 to 5 years. The second-stage columella reconstruction is usually approached using two different methods. In the first technique, forked flaps from the lip are advanced into the columella (6). In the second method, a midline V-to-Y flap advances the prolabial excess into the columella. The forked flap approach produces a new vertical scar in the midline of the columella. Midline V-to-Y from the prolabium into the columella avoids the new vertical scar on the columella but produces a new vertical scar in the central upper lip. In both instances a confluence of scars at the lip-columella junction is produced, usually under considerable tension. If the lip scars have widened, forked flaps permit revision of the lip scars. The vertical scar in the middle of the columella will be seldom seen in short individuals. In a tall male, a central V-to-Y flap has some advantages. The columella is more visible and will not have a scar, and if the individual grows a moustache, the lip scars will be camouflaged. The prolabium, however, has poor terminal hair growth compared to the lateral lip.

A banked forked flap repair, in which the forked flaps are “banked” under the alar bases at the time of the primary lip repair, previously enjoyed widespread popularity. This “banking” allows a one-stage lip repair that is usually quite satisfactory. The lateral “forks” from either side of the prolabium are inserted into incisions at the alar labial junction. The forks are “unbanked” at a second stage and advanced up to form a columella. This second stage is similar to the procedure described by Cronin (6). Just as in the secondary forked flap approach previously discussed, a new vertical scar in the columella is produced along with a confluence of secondary scars under tension at the lip-columella junction.

### Criticisms of the Skin Paradigm

Although the skin paradigm has been the most common approach to the bilateral repair in the last 100 years, it produces a number of unsatisfactory effects on nasal form. Techniques based on this approach concentrate on redistributing the available skin of the prolabium to correct the deformity, while ignoring the basic underlying anatomic derangement of the nasal tip lower lateral cartilages.
With all of the techniques described above, advancement of the prolabial skin into the columella has the potential of worsening the underlying cartilaginous tip deformity. The neocolumella produced by advancement of the prolabial skin does not contain cartilage to provide support of the nasal tip. The footplates of the medial crura have been advanced into the nasal tip, while the domes of the lower lateral cartilages, which represent the true nasal tip, are thrust laterally. Later growth of these divergent tip cartilages results in a bulbous nasal tip with an abnormal skin envelope, which is difficult to correct in tertiary procedures. The lack of cartilaginous structure in the neocolumella can result in a lip–columella junction that pulls inferiorly, blunting the angle, or conversely may contract up toward the caudal septum, producing a retracted columella. This latter result is compounded by the absence of the anterior nasal spine described earlier.

In addition to the requisite parallel vertical scars at the philtral columns on either side of the prolabium, approaches based on the skin paradigm produce an additional visible vertical scar. The central V-to-Y technique leaves a new vertical scar in the midline of the lip, whereas the forked flap approach produces a new midline scar in the columella. With both of these approaches, a confluence of secondary scars across a tight lip–columella junction is produced.

Dissatisfaction with the long-term results of these earlier skin paradigm techniques prompted surgeons to consider the underlying cartilage displacement to be as important as the skin deformity, creating a shift over the past 10 years to the cartilage paradigm.

**Cartilage Paradigm of Bilateral Cleft Lip and Nose Repair**

A number of techniques have been developed that place primary emphasis on the nasal tip cartilage deformity and assign the skin deformity secondary importance. Common factors to these techniques are that (a) the absent columella skin is recruited from the nasal tip skin instead of the prolabium, (b) the lower lateral cartilage domes are repositioned with the hope that subsequent cartilage growth will create columellar skin and nasal projection, and (c) columella skin can be produced by presurgical columella elongation.

**McComb Method**

McComb described a two-stage approach to correction of the complete bilateral cleft that focuses on the nasal deformity (7). A V-to-Y “gullwing” incision is performed on the skin of the nasal tip at the time of the first procedure. This allows direct visualization of the nasal tip cartilages with elevation of the nasal tip skin. The laterally displaced domes of the lower lateral cartilages are mobilized and sutured together in the midline to a more normal relationship, displacing the intervening fibrofatty tissue into the nasal tip. Closure of this gullwing incision in a V-to-Y manner recruits the width of the nasal tip skin into the production of a columella. A lip adhesion is done at the time of the first nasal procedure. Lip repair is performed at a second stage.

**Mulliken Method**

Mulliken initially described an approach that uses a vertical incision in the midline of the nasal tip along with bilateral incisions at the nostril apices to allow dissection of the nasal tip cartilages. As with the McComb technique, this allows the surgeon to mobilize the domes and suture them together in the midline. The vertical incision in the nasal tip was proposed rather than McComb’s V-to-Y incision to maintain better blood supply to the prolabium, allowing one-stage lip and nose repair. No primary columella elongation was performed at the time of the initial repair save that produced by nostril apex incisions extended out onto the nasal tip.

Mulliken has since modified his initial technique (8). The vertical tip incision is omitted, and all cartilage dissection is performed through bilateral alar rim incisions. Through these limited incisions, the domes of the nasal cartilages can be visualized and approximated at the midline to encourage future anterior growth of the nasal tip. Mulliken also describes skin redundancy in the soft triangles and lateral
columella that becomes apparent after positioning of the nasal cartilages, and should be trimmed.

**Prolabial Unwinding Flap**

Cutting (1993) described the prolabial unwinding flap. In this approach, the wide prolabium is “unwound” to produce columella skin and lip skin without any scar in the nasal tip columella or prolabium. This approach is possible as a result of the circumferential prolabial artery. Nasal tip cartilages are dissected retrograde through transfixion incisions that extend in front of the membranous septum but completely behind the prolabium. This technique is a hybrid of the two paradigms. The problem with this approach was that it is an asymmetric design, is a difficult technique, and still pushes the footplates of the medial crura up into the columella.

**Trott Method**

In this approach, a one-stage open rhinoplasty is performed where the prolabial flap is carried on the end of the columellar skin (9). This allows the best possible exposure to the displaced dome cartilages for mobilization and suturing in the midline. Furthermore, it does not produce any scars in the nasal tip or at the lip-columella junction. The blood supply to the prolabium may be quite precarious. It should be noted that the columella is not elongated in any way by this approach. This method and that of Mulliken share a common feature. Both rely on the growth of the medial crura, which have been placed into a correct anatomic relationship, to produce columellar skin secondarily over time.

**Presurgical Columella Elongation**

Although the techniques focused on correcting the nasal cartilage deformity lead to more favorable nasal tip growth, they are limited in creating sufficient skin for creation of a columella without a nasal tip incision. As described earlier in the chapter (pages 203-204, Complete Bilateral Cleft Lip), the vertical distance from the nasal tip to the inferior prolabium is decreased in the bilateral cleft lip deformity, and this limited skin is needed to create both nasal projection and central lip height. In severe deformities, this leads to a satisfactory lip reconstruction at the cost of a flattened nose.

Presurgical orthopedics is enjoying a revival, largely as a result of the ability to create adequate skin for both an optimum lip and nasal repair. Techniques such as NAM retract the premaxilla, elongate the prolabium, and create the skin for a neocolumella (1). The nasal lining is stretched and the depressed nasal domes are elevated anteriorly and carried toward the midline. This allows one-stage repair of the alveolus on both sides (gingivoperiosteoplasty), lip repair using a narrow central prolabial flap, and nasal repair with retrograde dissection and suturing of the domes in the midline. Initially, it had been hoped that no nasal surgery would be necessary using this method; however, the fibrofatty deposit between the displaced dome cartilages cannot be corrected with presurgical columella elongation alone. After presurgical nasal molding, retrograde dissection of the skin and fibrofat from between the separated nasal domes is necessary along with suturing of the dome cartilages in the midline. No scars from the nasal tip to the Cupid's bow are produced. Presurgical molding requires a dedicated molding and surgical team and parents who are prepared to spend the time to achieve the desired result, but when possible, it is our method of choice.

**Primary Cleft Palate Repair**

Although cleft lip and cleft palate surgeries are linked by a shared patient population, and both require a complete understanding of the abnormal anatomy by the surgeon, they are surprisingly different. A cleft lip repair is an artistic, flexible technique tailored to the unique three-dimensional anatomy of each child, whereas a cleft palate repair is a precise technical exercise whose success is based on performing the dissection reliably and atraumatically. Following a cleft lip repair, the parents appreciate the hours of work of the surgeon because of the visible incisions and facial difference, whereas following a cleft palate repair, the key portions of the operation, namely the nasal closure and the intravelar veloplasty, are hidden in the mouth by the transposed oral flaps. The success of the cleft lip repair can be predicted at the end of the operation; results of the cleft palate repair take
years to assess, and cannot be evaluated definitively until commencement of speech and completion of facial growth. Despite the lack of surgical glamour associated with a palatoplasty, the patient with a cleft palate requires multidisciplinary evaluation and treatment, a precise, technically sound operation, and standardized postoperative care to achieve the desired results while minimizing the potentially severe complications.

**Timing of Surgery**

The optimum timing of cleft palate repair balances the benefit of normal velopharyngeal function to optimize speech development against the potential disadvantage of impaired facial growth secondary to early surgical trauma. Graber’s description in the late 1940s of restricted maxillary growth following early palate closure was accompanied by a recommendation to delay surgery until 4 to 6 years of age. Because of the deleterious implications of this recommendation on speech development, the conventional timing for cleft palate repair was arbitrarily set at 18 to 24 months as a compromise between speech and facial growth. The current consensus, based on an increased understanding of speech development, is that cleft palate repair should be completed before 18 months of age; however, there is no general agreement regarding how early the surgery can be performed. Since Graber’s earlier work, there have been a number of studies supporting the theory that impaired maxillary growth in cleft patients is independent of cleft palate repair, and can result from the lip repair alone or is more a result of intrinsic restriction rather than early surgical trauma.

Results from previous retrospective studies examining the effect of timing of cleft palate repair on speech development are inconsistent and are compromised by small study numbers and potentially confounding variables. Kirschner recently retrospectively reviewed Randall’s and LaRossa’s cases at Children’s Hospital of Philadelphia, using two relatively homogenous cohorts undergoing soft palate repair either before or after 7 months of age, and found no significant benefit of early closure over later repair with respect to speech outcome. These authors emphasized the one thing that the surgical community agrees on: that long-term well-designed prospective studies are required before the optimum timing of cleft palate repair can be decided.

There are currently two common approaches to the timing of cleft palate repair in North America: (a) two-stage repair, with the soft palate repair and veloplasty performed at the time of lip adhesion or primary lip repair, and the hard palate repaired before 18 months, or delayed further with the use of an obturator, and (b) single-stage repair around the age of 11 to 12 months. Our two centers practice the latter approach, delaying the surgery until the time when the child starts to demonstrate the introduction of plosives (b, d, and g) in their speech. It is at this time that they require an intact velopharyngeal sphincter to continue with normal speech mechanics. In children with airway issues, such as that associated with micrognathia of Pierre Robin sequence, the surgery can be delayed until age 14 to 18 months to allow further mandible growth and to decrease the chance of postoperative airway compromise.

**Cleft Palate Repair Operative Technique**

At our two centers we perform a single-stage, two-flap palatoplasty with intravelar veloplasty as a modification of the technique described by Veau, Wardill, and Kilner (the “Oxford” palatoplasty). This technique is described below.

The patient is placed in the supine position, with a shoulder roll to extend the neck. A number of mouth retractors have been designed for the operation, but all retract the lips and tongue, open the jaws, and keep the endotracheal tube out of the operative site. Care must be taken not to hyperextend the neck, not to strangulate the tongue, and not to bruise the lips. The mouth and nasal cavities are cleaned with normal saline and a small throat pack is placed. The hard and soft palates and the nasal septum are infiltrated with lidocaine and epinephrine, avoiding injection directly around the greater palatine vascular pedicle. With pressure, the mucoperiosteum can be hydrodissected from the
hard palate with the injection to facilitate elevation of the flaps.

The bilateral posteriorly based mucoperiosteal flaps are incised laterally at the junction between the hard palate mucosa and the attached gingiva, and then elevated from the hard palate. With a curved elevator, through this lateral incision, the nasal mucosa can be elevated from the lateral nasal wall and posterior nasal spine in continuity with the oral flaps. We find it easier to perform this dissection before separating the oral and nasal flaps along the length of the cleft. The medial aspects of the bilateral mucoperiosteal flaps are then released and the incision continued along the visible line between the oral and nasal mucosa to the tip of the uvula. Care must be taken not to leave the nasal flaps deficient. The oral flaps can always be mobilized to the midline, whereas the mobility of the nasal flaps is limited if they are cut too short. The anterior tips of the mucoperiosteal flaps can then be elevated to expose the greater palatine pedicle, which is carefully preserved and dissected circumferentially.

Two structures tether the mucoperiosteal flaps and limit their mobilization across the cleft at the level of the posterior nasal spine. The first is the greater palatine pedicle, and the second is the abnormal attachment of the levator veli palatini and tensor palatini muscles to the posterior hard palate. A number of techniques have been described for lengthening of the pedicle, including osteotomies of the foramen. Sharp release of the periosteal sheath of the pedicle allows the pedicle to be stretched without compromising perfusion of the flaps. With the pedicle on stretch, a beaver blade is placed behind the pedicle at the edge of the hard palate. The periosteal sheath is lightly stroked until the underlying perivascular fat starts to herniate, at which point the incision follows the length of the pedicle along the long axis of the flap. Scissors are placed behind the pedicle, with one blade resting on the posterior hard palate shelf. Gradual opening of the scissors will create a palpable pop as the remaining periosteal sheath is released by the stretch. Sufficient pedicle length is created to allow tension-free closure of wide clefts. The pedicle dissection must be performed before release of the muscle from the posterior hard palate. If the pedicle is compromised during the dissection, the muscle attachments are required to perfuse the mucoperiosteal flaps.

The nasal lining is then separated from the muscles of the soft palate using sharp fine scissors. There is no reliable dissection plane within the first 2 or 3 mm of the cleft edge, and we prefer to leave this edge of the nasal lining flap thick to help with suturing. Immediately beyond the cleft edge, however, there is a defined, gray, smooth dissection plane under the muscles. Care must be taken not to leave muscle fibers on this undersurface of the nasal mucosa of the soft palate. The dissection continues back to the skull base so that the nasal flaps can be approximated across the cleft with minimal tension.

The final stage of the dissection is the intravelar veloplasty, which is essential for normal speech development. As described by Sommerlad, the normal velum consists of the levator muscle in the middle 40% and the tensor aponeurosis in the anterior 33%. In the cleft palate anomaly, the two muscles are closely related, with the tensor aponeurosis attaching to the posterior border of the hard palate and the levator inserting at the margins of the cleft in the anterior half of the velum. The abnormal attachment of the tensor can be directly visualized at the posterior shelf of the hard palate as obliquely oriented fibers. The fibers are sharply released from the edge of the hard palate, and the tensor tendon is divided medial to the hamulus. This maintains the attachment of the lateral component of the tensor to the hamulus, maintaining muscle tension for normal eustachian function. The medial release of the tensor tendon, however, allows mobilization of the levator muscle, so that it can be retrotransposed across the cleft. The levator is dissected sharply from the palatoglossus on its oral surface to increase its mobility. This leaves a thick, well-perfused oral mucosa flap that can be advanced medially across the cleft, independent of the posterior-medial rotation of the levator sling. The fibrous component of the tensor that was released from the posterior edge of the hard palate travels with the levator to provide more substantial tissue for anterior suture placement when the levator muscle sling is approximated across the cleft. This intravelar veloplasty not only serves to create an intact circumferential levator-pharyngeal sphincter for nasopharyngeal closure and speech, it also serves to lengthen the soft palate.
Sommerlad’s intervelar veloplasty, which was developed independently of the technique of Cutting described above, shares much in common with Cutting’s technique but has a number of features that distinguish it: (a) mucoperiosteal palatal flap elevation is not performed, (b) the velar muscles are exposed by raising oral mucosa in a plane underneath the mucous glands, (c) the nasal closure is completed before muscle dissection and repositioning in order to place the dissection plane on stretch, and (d) the operation is performed under a dissecting microscope with a knife, rather than loupe magnification with scissors. Readers are encouraged to read Sommerlad’s paper for a detailed description of the velar anatomy (10).

The palate is repaired sequentially; the nasal closure from anterior to posterior, followed by the oral closure from posterior to anterior. In a bilateral cleft of the secondary palate, bilateral mucosal flaps are elevated from the caudal edge of the vomer with a midline incision and sutured to the opposing lateral nasal flaps using buried interrupted 5-0 Vicryl sutures. The posterior extent of the vomerine flaps is at the posterior nasal spine. At this point, the nasal closure continues with direct approximation of the nasal lining of the soft palate across the cleft back to the uvula. With a unilateral cleft, only one vomerine flap is required. If there is an associated cleft of the primary palate, the nasal lining is repaired as far anterior as possible. Ideally, the nasal floor should have been repaired by the earlier cleft lip repair back to the incisive foramen, such that oronasal separation can be completed at the time of the palate repair. This saves the child the inconvenience of an anterior oronasal fistula during the years after the palate repair, but also makes a secondary alveolar bone graft easier and potentially more successful as nasal closure has already been achieved.

Various techniques have been described for uvuloplasty, including bilateral Y incisions and truncating the tip of the uvula to create a broad raw surface. None are ideal. With wide cleft repairs under increased tension, the uvula tends to widen at the base and decrease in projection. All techniques have in common accurate eversion of the mucosal lining of the uvula and repair of the muscle bundle at the base of the uvula to decrease postoperative widening and prevent fistula formation.

After nasal closure, the mobilized levator sling is transposed across the cleft. The tension of the muscle repair is based on surgeon experience. A repair that is too tight can lead to a decreased nasopharyngeal aperture and potential postoperative airway compromise. A repair that is too loose will compromise function of the levator sling during speech. We repair the muscle sling with approximately three buried horizontal mattress sutures of 3-0 Vicryl.

Oral closure is achieved using 4-0 chromic vertical mattress sutures. Two 3-0 chromic sutures are used to grasp the underlying nasal lining closure as part of the mattress suture. These are left as delayed ties to close the dead space between the oral and nasal lining. Delayed ties of 3-0 chromic are also used to secure the anterior tips of the mucoperiosteal flaps directly to the alveolus. In a bilateral cleft, the flaps are also secured to the posterior aspect of the premaxilla, where a very limited mucoperiosteal dissection is performed to create an edge to receive a suture. The original description of the two-flap palatoplasty included a “pushback” to lengthen the palate, which left the anterior hard palate exposed. This pushback technique has been discontinued following evidence of impaired facial growth, and is unnecessary for lengthening if a proper levator muscle transposition is performed.

Meticulous hemostasis is essential during the cleft palate repair. If there is any sign of oozing from the flaps or lateral defects, the bleeding is stopped prior to waking the patient. Some surgeons suture absorbable hemostatic material in the lateral defects, but recognize that this does not replace surgical hemostasis. Any blood that has collected in the oropharynx is suctioned. The patient is placed in soft arm restraints, and the endotracheal tube is not removed until spontaneous breathing and purposeful movement is established. We recommend postoperative oxygen saturation monitoring and close observation in the recovery room for 1 to 2 hours prior to discharge to the ward. ICU level care may be indicated in syndromic or other complex patients.

**Other Operative Techniques**
Although the principles of palatoplasty are consistent across techniques, there are variations for mucosal closure. Along with the two-flap modified Veau-Wardill technique described above (see Cleft Palate Repair Operative Technique), variations of the von Langenbeck two-flap technique are also common. With the von Langenbeck closure, the anterior tips of the bilateral mucoperiosteal flaps are left attached to the anterior hard palate and are mobilized as bipedicled flaps that transpose to the midline. The Furlow double opposing Z-palatoplasty can be used for secondary palate lengthening to treat velopharyngeal insufficiency, but is also commonly employed for primary cleft palate closure, especially for isolated clefts of the secondary palate. With this technique the levator muscles are transposed as opposing limbs of a Z-plasty in the soft palate (Fig. 23.9). Lengthening of the palate is achieved by the differential transposition of the Z-plasty. The short limb of the Z is placed on the midline, and the long axis across the cleft. When the flaps are transposed, the lengthening of the palate is achieved at the expense of increased tension in closure of the lateral mucosa.

Complications Following Cleft Palate Repair

Complications of cleft palate repair include bleeding, respiratory obstruction, infection, dehiscence, and oronasal fistula formation. Significant postoperative bleeding is rare, but if it occurs, it requires reintubation and exploration for hemostasis. Respiratory obstruction is also rare in the absence of excessive bleeding, but it may be life-threatening. The airway is monitored carefully in the recovery room and only after adequate assessment should the baby be transferred to the floor. We recommend oxygen saturation monitors to be employed on the floor or the patient can be monitored in an ICU setting if the airway is tenuous. Monitors alone are not a fail-safe prophylaxis. They are only as good as the response of personnel to the alarm. Pain control should be handled by experienced staff, as overmedication with narcotics can easily lead to respiratory arrest in these patients. Infants with Pierre Robin sequence or other congenital anomalies affecting the airway are at highest risk for airway problems.


Palatal fistulas may present as asymptomatic holes or may cause such symptoms as speech problems, nasal regurgitation of fluids, or difficulty with oral hygiene. The most common locations are at the region of the incisive foramen, at the posterior nasal spine, and the uvula. Fistula rate has previously
been reported at 10% to 15%, but in experienced hands is now 5% or less. Meticulous surgical
technique to create intact, well-perfused flaps that are carefully approximated across the cleft with
minimal tension is the best prophylaxis against fistula formation. The use of biomaterials, such as
acellular cadaveric human dermis, has been described as a reinforcing layer on top of the nasal closure
for wide clefts. With a well executed technique this should rarely, if ever, be indicated. Oronasal
fistulas are treated early with local mucosal flaps if they are symptomatic, or are left unrepaired until
the time of another surgery, such as alveolar bone grafting, if they are asymptomatic.

**Orthodontic and Orthognathic Treatment Following Cleft Palate Repair**

Studies on unrepaired cleft palates in developing countries suggest that the surgical intervention of
cleft palate repair impairs future maxillary growth. Some individuals with cleft palate may also have an
intrinsic limited growth potential. Decreased maxillary width and the resulting lingual crossbite are
common and are managed by orthodontic maxillary expansion with a fixed appliance. Once expansion is
completed, the optimum time for bone grafting is chosen according to the stage of canine
development. If the graft is performed too early, it can result in bone resorption as a consequence of
a lack of mechanical stimulation from a tooth. If the graft is performed too late and the erupting
canine root does not have sufficient bone support, the tooth may be lost.

Maxillary retrusion or midface hypoplasia resulting in an Angle class III occlusal relationship with
anterior crossbite can be managed in childhood with a distraction device such as a Delaire mask to aid
horizontal growth, but eventually requires orthognathic advancement. A plan of treatment is
formulated on the basis of clinical examination, photographs, cephalometric studies, and dental
models. If surgery is indicated, presurgical orthodontics is required to align dental arches and to
eliminate crowding and dental compensations. LeFort I maxillary advancement is performed at the time
of epiphyseal closure at skeletal maturity, approximately age 16 years for a girl and 18 years for a boy.
Large advances greater than 1 cm can be difficult as a result of restriction from palatal scarring related
to palate repair or previous pharyngeal flaps, and are prone to relapse. These cases can either be
treated with an accompanying bilateral sagittal split-mandibular osteotomy setback to minimize the
maxillary advancement required, or can be treated with LeFort I maxillary distraction osteogenesis.

Polley and Figueroa have studied and popularized LeFort I maxillary distraction osteogenesis using an
external cranial halo-based device. Once the permanent teeth have descended below the osteotomy
site, the LeFort I osteotomy is performed as with traditional surgery, and the maxillary dentition is
attached to the halo-based distraction device via an occlusal splint or other orthodontic attachment.
The segment can then be advanced at a rate of 1 mm a day into the desired position of
overcorrection. This is followed by a period of wearing the device without advancement during which
ossification occurs within the osteotomy and consolidates the new maxillary position. The original
consolidation period of 8 weeks has been decreased to 2 to 3 weeks by the use of a removable,
estatic-traction, traditional, orthodontic face mask that is attached to the oral splint after removal of
the halo. Polley and Figueroa's studies demonstrated stability of large maxillary advancements with
distraction osteogenesis and that there is minimal detrimental effect on speech.

**Operative Treatment of Velopharyngeal Insufficiency**

**Speech and Language Development**

All children born with a cleft palate require examination by a speech pathologist at regular intervals to
allow timely intervention if a significant delay develops in receptive or expressive language. The
diagnosis and work-up of language difficulties requires the multidisciplinary involvement of the speech
pathologist, audiologist, otolaryngologist, psychologist, and pediatrician, as the delay is not always
secondary to mechanical problems of the velopharynx. Other potential contributing factors include
hearing difficulties, abnormal speech habits, psychosocial delay, and tongue restriction. VPI is the
inability to achieve closure of the velopharyngeal port during sustained speech. The most common
cause of VPI is a cleft of the secondary palate; however, other less-common causes include submucous cleft palate, neuromuscular abnormalities, adenoidectomy, and congenital VPI of unknown etiology. Once other causes of language delay have been ruled out, a formal VPI work-up is performed to diagnose the underlying dynamics of the velopharynx, and to recommend appropriate treatment.

**Velopharyngeal Incompetence**

Intelligible speech production requires reliable and voluntary function of the velopharyngeal valve that controls communication between the oral and nasal cavities. The valve is closed by contraction of the pharyngeal muscles to advance the lateral and posterior pharyngeal walls, as well as the levator sling that pulls the soft palate (velum) posteriorly. If this palatopharyngeal sling is incompetent, abnormal coupling of the nasal and oral cavities occurs, which results in hypernasality, nasal emission, imprecise consonant production, decreased vocal intensity (loudness), and short phrases. These are the typical signs of velopharyngeal incompetence, which may be caused by either a structural defect or a physiologic dysfunction.

Tissue deficiency, pharyngomegaly, and neurogenic paresis of the velopharynx can all cause VPI; however, not all patients who exhibit glottal stops, pharyngeal fricatives, or nasal emission have VPI. Learned articulatory compensations such as glottal stops and pharyngeal fricatives may be confused with velopharyngeal dysfunction. Phoneme-specific nasal emission is often confused with VPI, even though no resonance disorder exists. Other aspects of phonatory, articulatory, and prosodic breakdowns may be unrelated to the competency of the velopharyngeal valve. If opening of the velopharyngeal valve, instead of closing, is the problem, abnormal uncoupling of the nasal and oral cavities results in hyponasality. This can be found in individuals with hypertrophic adenoid tissue and must be recognized before considering surgical intervention. Nonsurgical treatments of VPI include speech therapy, prosthetic management with speech bulb or palatal lift appliances, and posterior pharyngeal injections or implants. This section focuses on the surgical treatment of VPI.

---

**Preoperative Velopharyngeal Insufficiency Evaluation**

The goal of surgical intervention of patients with VPI is to provide a mechanism for functional speech. The design of the surgical procedure is based on the velopharyngeal anatomy and the function, which is determined by a series of clinical and radiographic tests. Clinical examination includes a formal recording of the child’s speech before, during, and after therapeutic intervention. Typical speech samples include isolated phonemes, words, phrases, and nonnasal reading passages with the nares occluded and unoccluded to detect acoustic differences associated with cul-de-sac resonance. Dynamic study of the pharynx by multiview videofluoroscopy and nasopharyngoscopy is usually indicated. This test provides information regarding the posterior and superior movement of the velum as well as the degree of medial excursion of the lateral pharyngeal walls during speech. In patients who have been referred from another center, intraoral examination and nasopharyngoscopy will determine if an intravelar veloplasty was performed at the time of cleft palate repair, and if the levator sling is functioning appropriately. With these tests, the VPI team can determine if the problem is insufficient length and/or excursion of the velum, or poor excursion of the pharynx, which will determine whether correction requires a secondary palate lengthening procedure such as a Furlow palatoplasty, or if pharyngeal surgery is indicated.

After pharyngeal flap surgery, patients are followed closely by both the surgeon and the speech pathologist. Clinical evaluations and tape recordings are obtained at least every 3 months for the first year and then annually for 3 to 5 years. Periodic acoustical analyses with the sound spectrograph are used to monitor speech characteristics postoperatively, and should validate more subjective, perceptual ratings in judging the success of surgery.

**Pharyngeal Surgery for Velopharyngeal Insufficiency**

The nonvelar surgical management of VPI usually consists of pharyngeal flap or sphincter surgery.
pharyngoplasty. At NYU, we prefer a superiorly based pharyngeal flap, whereas at Children’s Hospital, a sphincter pharyngoplasty is employed. Both are useful techniques with advantages and disadvantages.

**Pharyngeal Flaps**

Pharyngeal flaps may be superiorly or inferiorly based. Most studies in the literature have not found a difference on speech outcome between the two dissections. The mucosal flaps are raised from the posterior pharyngeal wall and attached to the soft palate so as to create a midline obstruction of the oral and nasal cavities between two lateral openings (ports). The amount of lateral pharyngeal wall motion will determine how wide the flap needs to be to achieve velopharyngeal competence. If the flap is too narrow, hypernasality will persist from inability of the lateral pharyngeal walls to close the ports on either side of the flap. If the flap is too wide, passive occlusion of the lateral port can occur, and the patient will develop mouth breathing, hyponasality, and possibly obstructive sleep apnea. Hogan (1973) popularized the concept of lateral port control based on his appreciation of the previous work by Warren et al. in the 1960s. These pressure-flow studies demonstrated that oropharyngeal air pressure decreases markedly when the port cross-section exceeds 10 mm$^2$, whereas nasal escape of air is audible above 20 mm$^2$. Sphrintzen et al. introduced the concept of “tailor-made” flaps based on preoperative evaluation of lateral pharyngeal excursion (11).

The technique of pharyngeal flap surgery involves longitudinal incisions through the mucosa and muscle down to the prevertebral fascia on each side of the posterior pharyngeal wall. Dissection is continued along the prevertebral fascia. A superiorly based flap is transversely incised inferiorly and raised to a level above the palatal plane, which usually corresponds to 1 to 2 cm above the tubercle of the atlas. An inferiorly based flap is incised just below the adenoid pad. The flap is usually inset with turn-back flaps on the nasal side of the uvula, with or without opening the midline palate repair. The turn-back flaps from the nasal mucosa are used to line the raw surface of the pharyngeal flap to minimize postoperative contraction. The pharyngeal donor defect of the flap is closed primarily. In patients with velocardiofacial syndrome, the internal carotid arteries can have an anomalous course that approaches the midline. The pharynx is observed and palpated carefully for any abnormal pulsations in the region of the proposed flap. Some authors have advocated preoperative computerized tomography (CT) or magnetic resonance imaging (MRI) angiograms in these select patients.

Complications following pharyngeal flap surgery are considerable compared to those of primary cleft lip and palate repairs. The Hospital for Sick Children in Toronto published retrospective data from a 7-year period in 1992 and reported an 8.2% risk of bleeding, a 9.1% risk of airway obstruction, and a 4.1% risk of sleep apnea. Five percent of their cohort required eventual surgical revision of the flap. With changes instituted by this group based on their review, including closer observation and monitoring, increased education, and decreased number of surgeons performing the procedure, the total rate of complications decreased from 11% to 3.2%. Bleeding decreased to 1.4%, airway obstruction to 3.2%, and hospital stay decreased from 5.8 to 3.8 days. These two valuable reports emphasize the potential complications associated with pharyngeal surgery, and the benefit of constant vigilance and quality improvement at all centers.

As expected, sleep apnea or upper airway obstruction is a potential complication of an operation whose purpose is to decrease the velopharyngeal airway. Although studies report up to a 35% incidence of abnormal polysomnograms following pharyngeal flap surgery, the vast majority of these patients resolve within 5 months. Lesavoy (1996) concluded that “the surgeon may sometimes need to accept some transient upper airway obstruction to achieve correction of velopharyngeal insufficiency.”

**Sphincter Pharyngoplasty**

The sphincter pharyngoplasties performed today are modifications of either the Hynes or the Orticochea techniques. In both techniques, the sphincter is constructed from bilateral superiorly based flaps raised from the posterior tonsillar pillars including mucosa and the palatopharyngeus muscle. In the Hynes pharyngoplasty, the flaps are transposed to the midline and inset into a defect...
created by a transverse incision at the level of the flap base. In the Jackson modification of the Orticochea technique, the flaps are sutured together with a small, superiorly based, posterior pharyngeal flap (12). Subsequent authors and studies have emphasized that the level of the sphincter is the most important predictor of success in both of these techniques. The pharyngeal constriction must be high, at the level of palatopharyngeal closure. The tightness of the pharyngoplasty can be controlled by the degree of overlap of the tonsillar flaps.

The procedure achieves both static and dynamic reduction in the velopharyngeal port with no disruption of the velum. It is ideal when there is poor medial excursion of the lateral pharyngeal walls and a short anteroposterior component of velar competency. It has the advantage of allowing revision if necessary by re-elevating the flaps and adjusting the tightness of the sphincter.

In both sphincteroplasties and pharyngeal flaps, the adenoid pad can limit the superior dissection and placement of the obstruction. For these patients, the otolaryngologist on the cleft team should be consulted to determine if an adenoidectomy is in the best interest of the child.

Pharyngeal Flap Compared to Sphincter Pharyngoplasty

Studies comparing the two pharyngeal surgeries have not documented a significant difference in speech outcome. Both techniques have advantages and disadvantages and potential complications, and require an experienced surgeon for success. An ongoing international prospective trial under the direction of Shaw in Manchester, UK comparing these two surgeries hopefully will improve our understanding of their relative values.

Treatment of the Alveolar Cleft

The preconference symposium of the 2004 American Cleft Palate–Craniofacial Association (ACPA) annual meeting focused on treatment of the alveolar cleft. Three approaches were presented and debated: (a) early alveolar bone grafting in the first year of life with autogenous rib cortical graft as a separate operation; (b) presurgical NAM with primary GPP at the time of primary lip repair; and (c) secondary alveolar bone grafting as a separate operation during mixed dentition with autogenous iliac crest cancellous graft. No conclusions regarding the superiority of one technique over another could be drawn at the end of the symposium. Each approach has been studied by its proponents to provide data justifying its use. Secondary bone grafting at the time of mixed dentition remains by far the most common technique for treatment of the alveolar cleft, and as such remains the standard by which other techniques are compared (13). The ideal treatment for the alveolar cleft would be a minimal surgical intervention performed without an additional anesthetic, with no donor-site morbidity, and no detrimental effect on facial growth or dental eruption. To date, NAM with primary GPP is the closest to this goal, but as described earlier in this chapter (page 206, Presurgical Nasoalveolar Molding), it requires presurgical orthopedics by a trained team, is associated with a 60% chance of avoiding secondary bone grafting, and requires further evaluation to confirm that it has no detrimental effect on maxillary growth. As prospective standardized multicenter data is gathered on the effect of these different surgical approaches on facial growth, bone quality, tooth survival, and patient satisfaction, we will have an increased understanding of their relative advantages and limitations.

Advances in tissue engineering and bone substitutes will generate an increasing number of materials that can be used for “off-the-shelf” replacement of missing alveolar bone. Although exciting, these materials still require a surgical procedure for introduction into the cleft. As with all new techniques, they should undergo the same stringency of evaluation prior to widespread use, including an emphasis on cost-effectiveness and potential side effects.

Secondary Cleft Lip and Nose Surgery

Increased understanding of the primary cleft abnormal anatomy with an associated improvement in the
technique of primary repair has reduced the severity of residual deformities and the need for secondary corrections. Perfection in a single surgery, however, remains elusive. The goals of early cleft lip and nose reconstruction are that the cleft be undetectable by peers at conversational distance by school age to minimize psychosocial stigmata, and that an optimal final surgical result using up-to-date techniques be complete by skeletal maturity. This is rarely achieved by one surgery in wide, complete clefts, resulting in the need for secondary cleft lip and nose surgeries. These secondary interventions should be kept to a minimum while striving to achieve these two goals.

Each surgery is approached with the following guidelines in mind:

- Identify the primary repair that has been performed to appreciate how it will affect the planned revisions.
- Recognize the optimum age to achieve the surgical goals. If the child is too young, small, temporizing procedures are performed to minimize the deformity and scarring until the definitive procedure can be performed.
- Find the normal landmarks and return them to their normal positions.
- Do not remove any tissue until certain that it will not be useful.
- Treat each case individually—there is no routine secondary procedure.
- Use the basic plastic surgery principle of transferring tissue from areas of excess to areas in need.
- Replace lost tissue with similar tissue when prior surgery, growth, or the lack of growth is responsible for the deficiency.

**Indications for Surgery**

The indication for a secondary surgical procedure is a correctable deformity given the age of the patient, which if not repaired, will remain or will result in psychosocial or functional problems. The surgeon must recognize that there are four perspectives of anatomic abnormalities to be considered: those of the surgeon, those of the patient, those of the parent, and those of peers or other members of society. Which perspective is predominant affects both the indication for surgery and the chance of a successful outcome.

When addressing a patient with a secondary deformity, it is first necessary to recognize the cause of the deformity. Steffensen (1953) outlined reasonable requirements for lip repair: (a) accurate skin, muscle, and mucous membrane union; (b) proper rotation of the deflected medial and lateral orbicularis oris muscle into a horizontal position; (c) a symmetric nostril floor and nostril tip; (d) an even vermilion border with reproduction of the Cupid’s bow; (e) slight eversion or pouting of the central upper lip; and (f) a minimal scar. Failure to meet one or more of these requirements may indicate the need for secondary repair.

**Timing of Secondary Repair**

As mentioned above, the goals of secondary repair of cleft lip and nasal deformities are that the cleft be undetectable by a peer at conversational distance by school age, and that an optimal final surgical result is complete by skeletal maturity. To achieve this, we perform presurgical nasoalveolar molding in early infancy, followed by a primary lip repair with repositioning of the nasal cartilages when the patient is approximately 3 months old, and defer any revisions until just prior to school age. At that time, any indicated lip revisions are completed to facilitate the child’s interaction with peers in a school environment. In the case of an obvious residual nasal deformity, such as that following repair of an unmolded wide bilateral cleft, a minor nasal tip rhinoplasty through limited intranasal incisions can be offered. The optimal time to complete the nasal reconstruction, however, is in adolescence, when a formal open rhinoplasty with cartilage grafting,
septoplasty, and/or osteotomies can be done. If orthognathic surgery is anticipated, the rhinoplasty is best deferred until after this is complete, as the appearance of the nose will change following repositioning of the bone that supports the nasal base.

Unilateral Cleft Lip

Deformities of the unilateral lip repair are mainly asymmetries and disproportions. One of the most readily visible deformities is an asymmetry between the vertical heights of the peaks of the Cupid's bow. If the Cupid's bow is not level, the cause should be identified and a surgical solution created. Vertical shortening of the cleft lip scar is not uncommon in the first few months following surgery, but should settle within a year postoperatively.

Deep to the cutaneous cleft lip repair is the orbicularis muscle. Proper reconstruction of the oral sphincter is key to preventing secondary contraction after repair. Discontinuity of the orbicularis oris is seen less often following unilateral repairs than following bilateral repairs. When discontinuity is present, a subcutaneous groove or trough appears and the scar contracture, which is normally seen only in the first few months after a repair, persists. The groove is more readily apparent on lip animation, with bulging of the lateral muscle segments caused by unbalanced contraction.

Secondary deformities of the unilateral cleft lip are diagnosed on physical examination. They include the deficient tubercle, vermilion deficiency and irregularities, the short upper lip, long upper lip, tight upper lip, and unfavorable scars.

Deficient Tubercle

A deficient or poorly pronounced tubercle following unilateral lip repair may be ameliorated by using a V-Y advancement of the labial mucosa. In rare cases, a dermal graft may be used to provide an autogenous lip augmentation. A fine mosquito clamp is used to create a tunnel along the horizontal length of the tubercle and within the substance of the orbicularis oris muscle. This procedure can yield a natural-appearing lip, especially during lip animation, but has the associated risk of asymmetry and fibrosis.

Vermilion Deficiency and Irregularities

The most common irregularity is a notch or whistle deformity. Notching is usually caused by inadequate approximation of the orbicularis marginalis muscle within the red lip. Deficiency of the free edge of the lip can often be treated by reopening of the inferior incision, symmetric eversion of the medial and lateral lip elements, and accurate layered approximation of the orbicularis marginalis and mucosa. If the primary repair had resulted in deficiency of tissue, local rearrangement of tissue is required: V-Y advancement, Z-plasty, or bilateral opposing advancement flaps of the labial mucosa. Z-plasty is especially useful when notching of the vermilion border is combined with scar contracture. The frenulum should always be examined if there is a red lip asymmetry, to ensure that a tight frenulum is not contributing to the problem. If the lip is excessively thick on the cleft side, a transverse ellipse can be excised at the wet-dry junction. Loss of the Cupid's bow after repair of a unilateral cleft can be corrected using the unilateral Gillies operation. This technique involves a triangular skin excision above the mucocutaneous line (to preserve this landmark). The excision is then closed horizontally.

Short Upper Lip

Short lip following unilateral repair refers to a diminished distance from the Cupid's bow to the base of the columella. The most common cause of the short lip is failure to adequately lengthen the lip at the primary repair. A short lip is more common when straight-line and rotation-advancement methods were used for the primary repair if the techniques were not executed properly. It can be a difficult deformity to repair. Careful evaluation is warranted, as recreation of the defect and complete revision is often necessary. As mentioned, shortening of the lip scar is not uncommon in the first few months after surgery, and it may be severe. Asymmetry generally exists during this time and is maximal 6 to 8
weeks after surgery. Softening and relaxation of the scar returns the lip to its immediate postoperative position when a proper muscular repair exists. However, if the lip is short on the operating table, it will always remain short. Techniques available for lip lengthening include (a) rotation-advancement flaps, (b) Z-plasties, (c) V-Y or forked flaps, (d) muscle advancements, and (e) Abbe flaps.

If a straight-line repair was performed primarily, it will not interfere with a subsequent rotation-advancement revision, which will advance the alar base medially and lengthen the columella on the cleft side. The ideal indication for rotation-advancement following a straight-line repair includes (a) the philtral scar on the cleft side is short; (b) the Cupid’s bow is pulled up toward the nostril; (c) the nostril floor is wide; and (d) the ala is displaced laterally and downwards.

A short upper lip following a Millard-type rotation repair usually requires revision with recreation of the defect and repeat rotation-advancement. Simple rerotation and advancement of skin only, without complete takedown of the muscular repair, should be reserved for minimal deficiencies. Additional lengthening may be obtained by adding a Z-plasty placed close to the sill of the nostril so that it is not readily apparent.

### Long Upper Lip

The long lip is more commonly found in bilateral than unilateral clefts. Compared to the rotation-advancement techniques, the quadrangular (LeMesurier) or triangular (Tennison) repair have been blamed for vertically long lips on the side of the cleft. The excess can be easily corrected by excising the exact amount of excess height from the horizontal components of the quadrangular repair.

It is unusual to find a rotation-advancement that has been rotated too much. A long lip deformity following a Millard-type repair may be corrected by total take-down of the repair with partial derotation of the medial segment. It may be necessary to shorten the vertical height of the lateral element to match the new rotation edge by a horizontal excision under the alar base.

### Tight Upper Lip

In a unilateral cleft lip repaired using a triangular method, a common secondary deformity is a tight upper lip. It is a horizontal tightness of the upper lip across the upper alveolus. This tightness is best corrected using a Z-plasty. Many early straight-line designs not only destroy the Cupid’s bow but also result in a horizontal tightness. In time, this tightness might improve as the displaced maxillary segments approximate. The tight upper lip tends to restrain anteroposterior facial growth and gives the relative appearance of a pouting lower lip. Severe tightness requires an Abbe lip switch flap for correction. This is more commonly used for treating bilateral cleft lip secondary deformities and is described in that section (see Bilateral Cleft Lip below).

### Unfavorable Scars

Unfavorable scars alone may require revision. Scars causing concern are generally either hypertrophic or widened, or are in an unacceptable position. Patients with multiple previous procedures may have a network of visible scars on the upper portion of the lip under the columella. If the scars are not hypertrophic and the lip-nose relationship is favorable, there is very little that can be done to improve the appearance. Hypertrophic scars most commonly appear about 1 month after the operation. They are red, raised, and firm. We make an effort to maintain tape on our lip repairs for up to 3 months to prevent these scars. Silicone scar management products can also be used. Wide scars may result from any of the previously discussed techniques, and are often associated with inaccurate approximation of the underlying orbicularis muscle. Revision is the usual solution. A pink scar may persist over time, and may be treated with a laser (see Chapter 56).

Upper lip hair will not grow in cleft lip scars. A patchy moustache formation can either be treated with scar revision, or with individual hair transplants.
Bilateral Cleft Lip

Bilateral cleft lip repair more typically leads to secondary deformity than does unilateral cleft lip repair. The secondary deformities listed above (see Unilateral Cleft Lip) can also occur following bilateral cleft lip repair and some may treated in a similar fashion. In the repair of the bilateral cleft lip, the goal is to achieve a narrow philtrum with a Cupid's bow, an adequate vermilion tubercle, and sufficient horizontal laxity to permit natural animation. Paradoxically, it is often easier to achieve lip symmetry with a bilateral repair than with a unilateral one. Horizontal (distance from philtrum to commissure) asymmetry is usually less obvious than vertical asymmetries. The contribution of the bony components of the cleft to the secondary deformity needs to be considered. It may be necessary to set back a projecting premaxilla before lip or nasal revision can be adequately addressed.

The most frequent secondary deformity of the bilateral cleft lip is paucity of the central lip. The thin central vermilion (whistle deformity) is more commonly seen after a Manchester-type repair, where the central lip has been corrected with abnormal prolabial mucosa that is deficient in bulk and often dry or flaking. The single-stage Millard-type bilateral repair, in which the red lip component of the lateral segments creates the central vermilion, usually leads to better symmetry and a fuller vermilion tubercle.

If a whistle deformity is present following a Manchester-type repair, the best treatment is often to recreate the defect and reconstruct the central lip from the lateral lip elements. If there is insufficient lateral lip bulk, a portion of the old prolabial mucosa may be maintained under the Cupid's bow. Attenuation of the inferior contour of the vermilion free border can also be improved with bilateral mucosal advancements, dermal grafting, or a V-Y advancement.

Once good symmetry is achieved in the bilateral cleft, the result may be compromised by a wide philtrum. We plan our philtrum to be 5 mm wide at the time of primary repair, understanding that this will stretch to a variable degree with time. The narrower the philtrum at the time of primary repair, the more vermilion is available for reconstruction of the central tubercle from the lateral lip elements. If there is excess philtral width in a secondary deformity, this redundant tissue may be used to lengthen the invariably short columella, and should not be excised until the decision has been made not to use it. When narrowing a wide philtrum by direct excision, the redundant dermis can be preserved to provide bulk underneath the new philtral margins.

A tight upper lip that cannot be corrected sufficiently with local flaps requires a donation of tissue from the lower lip via an Abbe flap. The Abbe flap improves the balance between the upper and lower lips by bringing comparatively excessive tissue from the pouting lower lip to the tight upper lip that is deficient of tissue. The scar on the upper lip is excised, creating a central defect. A full-thickness flap is designed centrally on the lower lip to reconstruct the aesthetic subunit of the upper lip philtrum. The donor defect on the lower lip should not violate the mental crease. The flap is rotated on a mucosal bridge containing an intact labial artery and vein that are found at the level of the vermilion border on the lingual (inner) side of the lip. The pedicle is divided after 10 to 14 days, and the flap is inset. The white roll of the flap segment must line up perfectly with that of the lateral lip elements. Up to one-third of the lower lip can be harvested while still achieving primary closure of the donor defect. If the muscle sphincter of the upper lip is in continuity, the Abbe flap can be designed as a skin/mucosal flap to wrap around the native orbicularis muscle.

Deficiency of the central labial sulcus is, unfortunately, a common secondary deformity following a bilateral repair. It results when the labial sulcus height has not been set at the time of the primary repair by suturing the turn-down mucosal flap from the prolabium to the periosteum of the premaxilla. The upper labial sulcus must be of sufficient length to provide a free upper lip for normal animation and speech. If deficiency of the labial sulcus is caused by an adhesion, the adhesion can be split horizontally with the resulting defect closed using Z-plasty flaps from the lateral lip mucosa. If there is insufficient tissue from the lateral lips to reconstruct the central sulcus, a mucosal graft can be used. It is placed inside-out over a stent, which is then inserted into the defect between the premaxilla and lip. This last technique, however, is prone to recurrence of the contracture.
Premaxillary Setback

The complete bilateral deformity is characterized by protrusion of the premaxilla and collapse of the lateral alveolar segments. Following repair of the orbicularis oris at the time of the primary repair, the segments are typically naturally molded by the muscle tension. In some cases, persistent premaxillary protrusion may occur. With the help of the team orthodontist, the decision is made whether a premaxillary setback is required as an orthognathic procedure. Premaxillary setback should only be performed by an experienced surgeon, as the vascular supply of the premaxilla is precarious and loss of the entire premaxilla and central teeth can occur. In some cases, the lip repair has formed a constricting band superior to the premaxilla forcing the premaxilla inferiorly. Not only does the premaxilla continue to project, but its severe inferior malposition may result in the incisor teeth biting into the lower gingivobuccal sulcus. In this circumstance, resection of a short section of vomer stem with repositioning of the premaxilla, mucosal repair, and alveolar bone grafting may be required. Premaxillary setback and repositioning should only be performed with the guidance of an orthodontist to plan for future dental rehabilitation and facial contour aesthetics.

Secondary Cleft Lip Nasal Repair

The literature is replete with numerous approaches to secondary repair of the cleft lip nasal deformity (Fig. 23.10). Many older techniques are still useful in certain circumstances, but should be used within the current paradigm of a systematic anatomic evaluation of the deformity followed by an equally systematic treatment plan. Just as techniques first used in the treatment of cleft patients formed the basis of the aesthetic rhinoplasty, many of the techniques that have recently evolved in aesthetic plastic surgery have been adopted by cleft surgeons. Each component of the deformity must be addressed in an orderly manner: skeletal base, nasal dorsal bone and cartilage, nasal tip cartilage, and, finally, the skin envelope (14).

FIGURE 23.10. Secondary rhinoplasty. A and B: Frontal and worm's-eye view of a nasal deformity secondary to unilateral cleft lip. The nasal tip is wide and underprojected because of divergence of the nasal domes and a shortened left columella. The left alar rim is depressed and flattened secondary to a deformed lower lateral cartilage. The left alar base is laterally and posteriorly displaced. The bony pyramid of the nasal dorsum is asymmetric. The patient also had a deviated septum and collapsed internal nasal valve. C and D: Frontal and worm's-eye view of the same patient after an open Dibbell tip rhinoplasty, submucous resection of the cartilaginous septum, a spreader strut graft, and monobloc nasal osteotomy.
**Skeletal Base**

Like all facial structures, the nose is supported by the underlying skeleton. The cleft deformity is not restricted to the skin and cartilage. In the unilateral deformity, the piriform rim under the ipsilateral alar base is deficient of bone and is retrusive. During the primary cleft lip surgery, the abnormal attachment of the nasal accessory cartilages to the piriform rim is released in order for the alar base to be moved anteriorly, medially, and superiorly into the desired position. Because of the lack of skeletal support, the alar base on the cleft side can, in some cases, become retropositioned with growth, even following an appropriate primary correction. If the patient is undergoing secondary alveolar bone grafting at the time of mixed dentition, this is the best time to augment the deficient piriform rim with autogenous cancellous onlay bone graft. The bone graft will elevate and support the alar base to achieve symmetry, and provide a stable base for the remainder of the nasal reconstruction in the teenage years.

In the bilateral cleft deformity, the anterior nasal spine is absent, and the footplates of the lower lateral cartilages rest on the muscle repair over the premaxilla. Prior to the definitive secondary rhinoplasty, the position of the premaxilla must be assessed. If the patient has not yet undergone orthodontic treatment, the premaxilla can be retrusive or protrusive. Both deformities will affect the appearance of the nose, and should be corrected before a rhinoplasty is undertaken. In the unfortunate event that the premaxilla is absent, either because of inappropriate resection or iatrogenic loss, prosthetic replacement is needed to provide a base support for the nose and lip.

A number of cleft patients will require orthognathic surgery following orthodontics because of midface retrusion. Ideally, the definitive rhinoplasty should be delayed until after the maxillary advancement has been completed. The LeFort segment contains the anterior nasal spine, which will affect the columella-labial angle and nasal tip projection.

**Nasal Dorsal Bone and Cartilage**

The unilateral cleft lip nasal deformity often includes a deviated bony and cartilaginous nasal septum with or without deviation of the nasal bones. All of the techniques described in the cosmetic rhinoplasty literature can be employed. If the nasal bony pyramid is symmetric, it can be mobilized as a "monobloc" and centralized, as described by Vilray Blair (1930). If the pyramid is asymmetric, independent movements of the nasal bones will be required. We used a 3-mm osteotome percutaneously to control the nasal osteotomies.

The deviated nasal septum can be treated with a septoplasty, using sutures and scoring to straighten the nasal passage, or, alternatively, with a submucosal resection if cartilage graft is required for the nasal tip, leaving a 1-cm dorsal and ventral L strut for support. In both cases, the base of the septum is mobilized and centralized using a permanent suture through the periosteum of the nasal spine. If the septal cartilage is too weak to support the new position, onlay strut grafts are used to reinforce the nasal tip projection.

As with any rhinoplasty, the preoperative evaluation includes an intranasal examination. If the patient has collapse of the internal nasal valve with nasal obstruction on inspiration, spreader grafts between the upper lateral cartilage and septum are needed. It is important to inform the patient preoperatively that this will widen the nasal dorsum, which is often opposite to what the patient wants esthetically.

The bilateral cleft lip nasal deformity frequently has a broad nasal dorsum. This can be treated with bilateral nasal osteotomies and infracturing to narrow the nasal pyramid and increase dorsal projection. This maneuver will narrow the nasal passage, and needs to be balanced with the functional goals of the surgery.

**Nasal Tip Cartilages**

The medial and lateral feet of the lower lateral cartilages in the secondary deformity are often displaced posteriorly on the cleft side, with the lateral foot displaced laterally. This causes collapse of
the nasal tripod, alar rim hooding, and lateralization of the genu of the nasal dome. Older techniques transposed subsections of the displaced cartilages, and are rarely indicated. The current consensus is to reposition the entire lower lateral cartilage structure using an open tip rhinoplasty. Unlike a non-cleft rhinoplasty, simple repositioning with nasal tip sutures is typically insufficient to correct the cleft deformity. After the native cartilage framework is reconstructed, autogenous cartilage grafting is required to strengthen the new position and to augment the nasal tip projection.

In the unilateral deformity, a “springboard” graft is often required to maintain alar rim curvature. The graft is harvested from the septum, and is anchored in a subcutaneous pocket at the alar base. The graft is then bent under mild tension over the lower lateral cartilage and secured to the nasal dome. The lateral crus is then secured to the undersurface of the graft. The springboard effect of the graft will create and maintain the desired alar rim curvature when the skin is redraped. Other useful techniques are modifications of the Dibbell (1982) bipedicle chondrocutaneous flap. A bipedicled flap of nasal floor skin is elevated in continuity with a chondrocutaneous flap of medial crus with incisions below the crus and above it in the membranous septum. The depressed medial crus is advanced up into the tip, and the alar base is carried medially by the bipedicle nasal floor flap. This is performed in conjunction with an open approach, using a Tajima (1977) “inverted U” nostril apex incision to address the soft triangle. Tajima suspension sutures anchored to the upper lateral cartilages are useful to elevate the alar rim in both these techniques.

In the bilateral cleft lip nasal deformity the displaced lower lateral cartilages are addressed in a fashion similar to that described above. Compared to the unilateral deformity, however, the nasal tip projection and support is more deficient. Columella cartilage strut grafts or septal extension grafts as described by Byrd (1997) are required to provide support to the nasal tip construct. After medialization of the genu of the lower lateral cartilages to the midline to create a nasal dome, multiple onlay tip or Sheen grafts are usually required to achieve the desired shape and projection.

### Skin Envelope

A principal argument for an aggressive primary rhinoplasty in infancy is the frustration associated with attempts to correct the deformed, deficient, and scarred skin envelope of a secondary or tertiary rhinoplasty. All cleft surgeons have experienced the satisfaction of constructing a formidable cartilage framework, only to see it compromised under compression when the skin is redraped. The delicate anatomy of the natural soft triangle and nasal dome cannot be created with current secondary techniques, but should remain our goal.

In the unilateral deformity, the deformed skin envelope often overhangs the nostril apex. The Tajima “inverted U” nostril apex incision can help to address this problem. The skin flap left attached to the inferior edge of the lower lateral cartilage turns over to form the inner lining of a constructed soft triangle. A similar turn-over flap approach can be used secondarily if the underlying cartilage is already in the correct position.

In the bilateral cleft lip nasal deformity, the skin envelope is deficient vertically, from the nasal tip to the base of the columella. When closing the open rhinoplasty, the relatively lax lateral tip skin is advanced toward the nasal tip when the rim incisions are closed, in order to create sufficient skin for the nasal tip and columella closure. In severe deformities, however, the skin envelope is too tight to drape over the cartilage construct with tension-free closure at the columella incision. As described in previous sections, techniques that borrow skin from the lip, such as a V-Y advance, result in considerable scarring at the lip-columella junction. Conversely, techniques that borrow from the horizontal laxity of the nasal tip skin, such as the McComb and Brauer alar lift incisions, result in additional scars on the nasal tip. Both approaches therefore have limitations, but no ideal alternative currently exists.

### Nostril Stenosis
Nostril stenosis is the most difficult late complication associated with cleft lip repair. It is considerably easier to narrow a nostril than to enlarge it. In general, any circumferential nasal lining incision is associated with a high incidence of nostril stenosis. With a rotation-advancement repair, placement of the L flap in the nasal vestibule helps to provide adequate nasal lining and can reduce the incidence of stenosis. Once formed, nostril stenosis is a challenge to release. Intranasal Z-plasties or composite grafts are frequently used. Long-term postoperative use of nasal stents is required to minimize the chance of recurrence, but, unfortunately, is limited by patient compliance.

Conclusion
Many plastic surgeons were drawn to their surgical specialty after seeing their first cleft lip repair. Cleft care stands out as a rare opportunity in plastic surgery to have a huge impact on an infant’s future psychosocial well-being, and to follow these children over the formative years of their lives. The surgeon is reminded of the success, as well as of the failure, of the surgeon’s primary operations for years to come. Modern cleft surgical techniques, preoperative orthodontics, and specialized multidisciplinary team care enable us to achieve consistently favorable primary surgical results. Repair of secondary nasal deformities, however, remains a challenge, and is still best treated by preventative surgery at the time of the primary repair. Recent “inductive” techniques, such as nasoalveolar molding and distraction osteogenesis, have improved cleft care over the past decade, and as comparable advances in plastic surgery occur in the future, a child born with a cleft can look forward to fewer operations with better aesthetic and functional results.

References


