Cleft Lip: Unilateral Primary Deformities

James D. Burt, M.B.B.S., and H. Steve Byrd, M.D.

Dallas, Texas

Learning Objectives: After studying this article, the participant should be able to: 1. Discuss advantages and disadvantages of the available classification systems for cleft lip deformities. 2. Discuss, in detail, the normal anatomic features of the orbicularis oris muscle and the aberrant features seen in unilateral cleft lip deformities. 3. Define the anatomic features of the unilateral cleft lip and cleft lip nose deformity. 4. Differentiate the advantages and disadvantages of the two commonly used techniques for unilateral cleft lip repair.

The understanding and management of all aspects of unilateral cleft lip deformities continue to evolve. Just as we are entering the era of exciting advances in the understanding of the pathogenesis of craniofacial disorders, expansion of our understanding of the dynamic relationships of the structural and soft-tissue components of cleft deformities has assisted surgeons in achieving progressively improved and consistent outcomes for these patients. The anatomic and physiologic complexity of unilateral cleft lip deformities has been recognized for centuries, and generations of researchers have cumulatively contributed to our current understanding. This article examines the history, classification, anatomy, and controversies in the surgical management of unilateral cleft lip deformities, allowing surgeons to formulate a reasoned, longitudinal management plan for their patients on the basis of the available current data. (Plast. Reconstr. Surg. 105: 1043, 2000.)

Although the term lagocheilos (harelip) is ascribed to Galen, popular use of the term harelip is derived from Johnson’s translation of Ambroise Paré’s writings about bec de lièvre (lip of the hare). 1

The first cleft lip repair reportedly was performed by an unidentified Chinese physician in about 390 A.D. The Flemish surgeon, Yperman (1295–1350), is credited with the original description of a procedure that incised the cleft edges and sutured the margins with needle and twisted waxed thread, reinforcing the closure with harelip needles secured by a figure-of-8 tie. Early techniques of cleft lip repair involved a straight-line closure, such as in the operations proposed by Rose and Thompson. 3

The concept of closure of the cleft lip by local flaps was introduced by Malgaigne in 1843. The next year, Mirault modified Malgaigne’s method by advancing the lateral flap across the cleft. Mirault’s variation utilized the principle of filling the medial deficit with a lateral flap, on which all subsequent methods of lip closure are essentially based. In 1884, Hagedorn applied the Z-plasty technique to cleft lip closure while popularizing the use of a rectangular flap to lengthen the cleft lip in the course of repair.

The first half of the 20th century was devoted to the established straight-line closure. In the 1930s and 1940s, however, the Blair-Brown and Brown-McDowell repairs—modifications of Mirault’s original procedure—dominated the field of cleft lip surgery. Their techniques are based on a triangular flap advanced into the lower portion of the lip.

LeMesurier and Tennison independently modified the technique of lateral flap tissue transferred into the lower portion of the lip. LeMesurier’s innovation consisted of a quadrilateral flap, whereas Tennison’s involved a triangular flap; both introduced tissue into the lower part of the lip and shared the advantage of producing a pouting of the tubercle. Their repairs enjoyed a great deal of popularity in the 1950s and early 1960s.

In 1955, Millard developed the concept of lateral flap advancement into the upper portion of the lip, combined with downward rotation of the medial segment. His technique preserves both the Cupid’s bow and
the philtral dimple and has the additional advantage of placing the tension of the closure under the alar base, reducing flare and promoting better molding of the underlying alveolar process. Subsequently, Wynn\textsuperscript{12} and Davies\textsuperscript{13} described variations of triangular flaps introduced into the upper lip. Nevertheless, Millard’s repair has withstood the test of time and remains the most popular method for closure of the unilateral cleft lip. Skoog\textsuperscript{14} and Trauner and Trauner\textsuperscript{15} independently described procedures that involve a combination of flaps in the upper and lower portions of the lip.

**Classification**

To standardize reporting of cleft lip and palate cases, the Nomenclature Committee of the American Association for Cleft Palate Rehabilitation published in 1962 a comprehensive classification system that was later adopted by the Cleft Palate Association.\textsuperscript{16} The system was complex, however, and was not universally accepted. A few years earlier, Kernahan and Stark\textsuperscript{17} suggested a classification scheme that was simple, if somewhat lacking in detail. In 1971, Kernahan\textsuperscript{18,19} introduced a simplified, symbolic classification scheme—the “striped Y” method—that uses the incisive foramen as its reference point. Since then, minor modifications have been made: the hard palate has been restricted to position 7; the soft palate is identified as 8; and submucous clefts as 9.

Schwartz and colleagues\textsuperscript{20} suggest a modification of Kernahan’s striped-Y system that uses a three-digit number for recording the anatomic sites of clefting. This modification is designed to allow computer data entry while including all appropriate descriptions of each anatomic cleft variant.

The microform (forme fruste) cleft lip deformity has three components, which need not be present altogether for the diagnosis to be made: (1) a small notch within the borders of the vermilion, (2) a band of fibrous tissue running from the edge of the red lip to the nostril floor, and (3) a deformity of the ala on the side of the notch or band. The objectives of surgical repair are to eliminate any notch of the vermilion, correct the drooping or flattened ala, and restore muscle continuity with a minimal amount of scarring.

**Anatomy**

The unilateral cleft lip and cleft lip nose deformities can be summarized as follows.\textsuperscript{21} In osseous deformities:

- The premaxilla is outwardly rotated and projecting.
- The lateral maxillary element is retropositioned and collapsed medially.

In nasal deformities:

- The inferior edge of the septum is dislocated out of the vomer groove and presents with the nasal spine in the floor of the normal nostril.
- There is unilateral shortness in the vertical height of the columella, varying from three-fourths to even one-half that of the normal side.
- The lower lateral cartilage is attenuated, its medial crus lower in the columella and its dome separated from the opposite alar cartilage to rest below it. The lateral segment is flattened and spread across the cleft at an obtuse angle. The alar crease has no alar cartilage bulge to give way to and, consequently, continues obliquely across the tip just lateral to the joint of the columella and through the rim of the ala. This is often responsible for an actual kink in the alar margin itself.
- The alar base is invariably rotated outwardly in a flare.
- The alar rim is invariably distorted by a skin curtain (without cartilage) that droops over the alar rim like a web and further reduces the apparent height of the columella.
- The vestibular lining is deficient on the cleft side.

In lip deformities:

- The orbicularis oris muscle in the lateral lip element ends upward at the margin of the cleft to insert into the alar wing. In cases of incomplete clefting, the muscle does not, as a rule, cross the cleft unless the bridge is at least one-third the height of the lip.\textsuperscript{22}
- The philtrum is short.
- Two-thirds of the Cupid’s bow, one philtral column, and a dimple hollow are preserved.
- The musculature between the philtral midline and the cleft is hypoplastic.
The muscular anatomy of unilateral and bilateral cleft lip has been described,23,24 and differing observations were reported. In contrast to Fara’s23 report, Dado and Kernahan24 found no distinct muscle bundles parallel to the cleft margin and inserted into the alar base and columnella. The muscle bulge in complete and incomplete cleft lips consisted of a haphazard arrangement of muscle fibers running transversely, obliquely, and anteroposteriorly.

The modern trend toward radical muscle mobilization in the surgical repair of unilateral cleft lip25–29 underscores the importance of the muscular anatomy in this deformity. Nicolau26 distinguishes two well-defined parts of the orbicularis oris muscle, deep and superficial. The deep component has a sphincteric function acting in concert with the oropharyngeal muscular apparatus. The superficial orbicularis oris muscle functions in facial expression and provides the very precise movements of the lips needed in speech.

Mulliken et al.30 detail the gross and microscopic anatomy of the skin-mucosal junction (“white roll” of Gillies) at the Cupid’s bow of infants with normal and cleft lips. In the normal upper lip, the anterior projection of the pars marginalis of the orbicularis oris muscle gives rise to the cutaneous-vermilion junction. The normal vermilion is widest at the peaks of Cupid’s bow. On sagittal section beginning anteriorly at the white roll and proceeding orally, the vermilion mucosa exhibits progressively increasing epidermal thickness and size of rete ridges, decreasing melanin, more superficial capillaries, and an abrupt transition from keratinized to nonkeratinized squamous epithelium (“red line” of Noordhoff). In cleft lip specimens, the white roll is absent. There are hypoplasia and disorientation of the underlying pars marginalis component of the orbicularis oris muscle, decreased vermilion width on the medial side of the cleft, and normal to slightly increased width of vermilion laterally (the entire prolabial vermilion component of bilateral cleft lip specimens is hypoplastic). These observations30 endorse Noordhoff’s31 recommendation of a lateral vermilion flap to augment the deficient medial vermilion in cleft lip repair.

Slaughter and coworkers32 detail the blood supply of unilateral and bilateral cleft lip. To various degrees, clefting interrupts the normal anastomoses occurring among the superior labial artery, anterior ethmoidal artery, posterior septal artery, and greater palatine artery.

**SURGICAL MANAGEMENT**

Many controversies still surround the surgical management of unilateral cleft lip and cleft lip nose repair. The recurring themes are the following:

- The results are surgeon-dependent.
- When a given cleft team meticulously pursues a chosen course of management, excellent results may be achieved despite disparate management algorithms.

The major differences between treatment algorithms exist in the following areas:

- Timing of lip repair.
- The use of presurgical orthopedics or lip adhesion to sculpt the maxillary arches and reduce the tension of the definitive lip repair.
- The advisability and efficacy of early gingivoperiosteoplasty or alveolar bone grafting.
- The timing and the approach to primary rhinoplasty for the cleft lip nose deformity.

**Timing of Lip Repair—Intrauterine**

Interest in intrauterine repair of cleft lip has been stimulated by the experimental findings in fetal surgery, wherein wounds created in the ectoderm of the fetus very early in gestation heal clinically and histologically without scar formation. This phenomenon is not clearly understood but is believed to be related to the absence of cellular inflammatory response by the fetus, no collagen deposition, no wound contraction, and healing by what appears to be regeneration.33

Hedrick and associates34 studied delayed in utero repair of surgically created cleft lip and palate in the fetal lamb model. Incisional or excisional unilateral cleft lips were created early in gestation and later repaired. The incisional and excisional clefts were made completely through the lip and alveolus. Two weeks later, the wound edges were freshened and repaired in all but one lamb in each group. In the early third trimester, the fetuses were harvested and studied. The incisional clefts healed spontaneously with replacement of the native reticular collagen pattern and with regeneration of the skin appendages. However, the excisional clefts did not autorepair and when
repaired surgically, they healed without a collagen scar but showed no regeneration of the skin appendages. This study is the first successful large-animal model wherein cleft lips and palates were created, allowed to develop with the cleft, and later repaired in utero. Before repair the edges of the excisional clefts epithelialized. When repaired early in the third trimester, excisional clefts healed without a dermal scar but exhibited incomplete regeneration of the skin appendages. Although the implications of these findings applied to fetal cleft lip repair are obviously attractive, the attendant risk of fetal loss is such that, at the present time, intrauterine fetal surgery should be reserved for life-threatening malformations that cannot be helped significantly by postnatal intervention.

Estes and colleagues discussed the endoscopic repair of fetal cleft lip in lambs. Although human intrauterine surgery for the correction of life-threatening fetal malformations is now a reality, preterm labor is a major complication and one that is directly related to the large hysterotomy required for fetal exposure. Endoscopic surgical manipulation of the fetus through small uterine ports addresses this problem and may eventually lead to prenatal surgical intervention for non-life-threatening malformations in the human fetus, provided that technical precision can be maintained.

**Timing of Lip Repair—Extraterine**

Advocates of neonatal lip repair argue that there is no increase in perioperative morbidity or mortality and that neonatal lip repair provides comparable long-term results to closure later in the first year of life. A secondary consideration has been the potential psychosocial benefits to the family of bringing home a child of “normal” appearance.

Mortality during the first year of life in infants with oral clefts has been studied. Children with cleft palate who did not have additional malformations had a statistically insignificant adjusted risk of 1.2 when compared with children with no malformations. Children with cleft lip with or without cleft palate had a 1.1 adjusted risk. However, 35 percent of children with oral clefts had associated malformations and experienced a much higher rate of mortality.

A retrospective analysis of anesthetic complications of neonatal cleft lip repair included 50 cases of neonatal cleft lip, including 11 prema-

*ture infants of less than 45 weeks postconceptual age. There were no deaths in this group, although there was one case of perioperative hypoxemia, one case of postoperative laryngospasm requiring reintubation, and one case of transient apnea. The authors conclude that neonatal cleft lip repair is safe, provided that the following criteria are met: (1) patients are gestationally mature infants with no intercurrent illness, (2) no opioid analgesia is given during the surgical procedure, (3) experienced nurses care for the infants postoperatively, and (4) there is appropriate monitoring with oximetry and apnea detectors.*

**Presurgical Orthopedics and Lip Adhesion**

Millard and Latham outlined primary surgical and dental techniques for the staged treatment of cleft lips and palate that are based on biologic principles and that theoretically facilitate the continuance of the failed embryonic migration toward a normal end point. Millard was of the opinion that positioning of the alveolar segments that allow closure of the mucoperiosteum across the alveolar and anterior hard palate cleft at the time of repair makes it possible to create a periosteal tunnel across the bony gap and set up a condition conducive to bone formation and eventual tooth eruption in the cleft area. Lip closure by adhesion reduces tension on the primary lip closure and allows gentle molding until the arch solidifies. Thus a complete cleft is essentially converted to an incomplete cleft.

With a balanced, stabilized maxillary platform, the definitive lip and nose correction can be performed. These planned actions bypass a persistent cleft, fistulas, raw areas, malposition of alveolar segments, and, in some instances, the necessity for later bone grafting. The question not totally answered is the effect of this approach on final growth.

The achievement of the symmetrical stable alveolus has become increasingly important as surgeons have sought to correct the cleft lip nose primarily. Ideologic and practical reasons have dictated varying approaches.

Presurgical lip adhesion in the unilateral complete cleft lip, advocated as early as 1954 by Johanson and Ohlsson, was mainly popularized by Randall in the late 1960s. The principle of lip adhesion is to decrease the tension of definitive lip repair by reducing the disparity of the maxillary segments in the interval between adhesion and definitive repair.
Randall\textsuperscript{40} suggested short, broad, triangular flaps interdigitated and sutured to the mucosa, muscularis, and skin. Millard,\textsuperscript{41} on the other hand, proposed a high adhesion that avoids scarring in the area of the repair and permits introduction of lateral lip parings into the lateral vestibule of the nose.

Furnas\textsuperscript{42} discussed the historical evolution of the straight-line repair, which he proposes as a first step before rotation-advancement closure in unilateral clefts. In essence, Furnas performed a preliminary adhesion consisting of straight-line closure, leaving the lip vertically short across the closure. The significance of this adhesion is that it allows lateral lip parings to be introduced into the piriform aperture of the nose, releasing the tethering in this area and providing muscle closure across the alveolar cleft. Because closure is done in the neonatal period, there is rapid molding, which allows rotation-advancement to proceed under little tension so that there is minimal nasal deformity at the time of definitive repair.

Indications for lip adhesion in the unilateral cleft lip deformity are emotionally debated. In general, its main application is in wide unilateral complete clefts and in complete clefts with poorly aligned maxillary segments. Opponents of the lip adhesion principle argue that the scar introduced by the adhesion interferes with the results of subsequent cheiloplasty.

**Orthodontics, Gingivoperiosteoplasty, and Alveolar Bone Grafting**

Kramer and colleagues\textsuperscript{43} studied early (from birth to age 3 months) palatal development in various complete and incomplete forms of cleft lip and palate by means of dental casts. The authors used reproducible reference points to plot palatal shape and dimensions in 128 affected children and 68 normal children, who served as controls. They noted substantial normal palatal growth in the first 3 months of life and concluded that preoperative maxillary orthopedics does not stimulate palatal growth, but rather restricts it.

In a retrospective analysis of 29 patients with early maxillary orthopedics, Gnoinski\textsuperscript{44} found that all patients who had a “balanced” skeletal relationship at age 15 had had lip surgery after 5½ months of age and palatal surgery after age 3. Lip surgery before 5 months of age and palatal surgery before 2 years regularly resulted in an unbalanced skeletal situation at age 15. Although the author stressed the role of timing of surgery in these deleterious effects, it should be recognized that the type of repair is equally important.

Rygh and Tindlund\textsuperscript{45} detailed the appropriate orthodontic treatment plan beginning in the deciduous dentition phase. Conventional correction of the maxilla was mainly by rotation and expansion of the lateral segments, with some lateral movement of the maxillary bones and labial tipping of the lingually positioned upper incisor teeth. These methods failed to yield any appreciable forward movement of the maxillary dentoalveolar arch, and none of the basal parts. The application of external forces through a protraction face-mask technique to encourage an increase in the vertical height as well as the sagittal length of the maxilla has been proposed. Sutural growth in the upper jaw is most active at ages 6 to 7 years and then declines until the pubertal spurt. Maxillary protraction therapy must be instituted before the age of 8 years if significant effect is to be obtained. The basic principle of therapy is short periods of active controlled, efficient treatment and long periods of effective retention. Age 6 seems to be the ideal time to begin therapy, because eruption of the permanent maxillary incisors will take place during the treatment period.

The orthodontic plan for the adolescent cleft patient with a relative class III relationship secondary to a retruded maxilla must be carefully scrutinized. Although the orthodontist may be able to narrow the upper and lower arches through extraction of first molar or bicuspids teeth and improve occlusal relationships, this scheme is a compromise that limits the appropriate orthognathic correction. These children generally benefit from an orthodontic plan that allows the maximum orthognathic advancement of their Le Fort I segment.

The appropriateness of early bone grafts in closure of alveolar clefts remains controversial. A review\textsuperscript{46} of the adolescent facial structure of children who had undergone early bone grafting of their alveolar segment indicates maxillary retrognathia in both unilateral and bilateral cleft categories as well as deficient vertical descent of the maxilla, especially in the anterior part. In about 40 percent of the bilateral and 50 percent of the unilateral cleft patients, midfacial growth attenuation had reached such magnitude that surgical advancement of the maxilla was necessary. Fusion of the suture between the premaxilla and vomer was suggested as the reason for
the typical midfacial structure seen in the patients with the most pronounced growth impairment. The bone grafting sequence consisted of initial lip adhesion and closure of the cleft in the anterior palate by means of vomerine flap. The second surgery consisted of bone grafts to the alveolar process and hard palate clefts with concomitant final lip closure. The grafts were autogenous cancellous bone taken from the tibia. A final closure of the soft palate with the pushback method completed the repair. As a result of this analysis, the authors concluded that early bone grafting of the alveolus should be avoided. Multiple similar studies exist, the common conclusion being that early bone grafting either is not advantageous or is deleterious in its effects.

A dissenting opinion is expressed by Rosenstein et al. 49 and Monroe et al. 50 The surgical sequence that these authors advocate involves premaxillary orthopedics, followed by lip repair at about 6 weeks of age. The appliance is retained following lip repair to mold the greater and lesser maxillary segments into place. (The prosthesis is retained until palate repair is completed at approximately 12 months of age; both the hard and soft palates are closed in one procedure.) At approximately age 4 to 5 months, when the segments abut and good arch alignment has been achieved, an autogenous split-rib bone graft is inserted in the upper buccal sulcus, with margin flaps off the alveolus turned for oral lining. The method avoids the use of any vomerine flaps for closure. A follow-up craniofacial analysis of the grafted and ungrafted comparative groups showed little difference in growth. Accordingly, the authors conclude that there is no real growth disturbance with the procedure and that an improved functional and more stable dental relationship is achieved.

Rosenstein and coworkers 31 subsequently reported the results of early bone grafting and infant maxillary orthopedics in 20 patients who had complete unilateral clefts of the lip, alveolus, and palate and 17 patients who had complete bilateral clefts. Anteroposterior and vertical facial growth was evaluated by cephalometric analysis. Compared with the original series, 49 these patients again showed no adverse growth restraints.

Ten years after primary periosteoplasty, the facial growth of unilateral cleft lip and palate patients was compared with a matched series of primary osteoplasty patients and another group of surgically repaired cleft patients without a bone graft or periosteal flap. This revealed jaw development that was most advantageous after primary periosteoplasty and least satisfactory after bone graft. Facial changes after periosteoplasty consisted of a milder retrusion of the upper jaw, maintenance of the overjet, and a more satisfactory prominence of the upper lip. 52

Similar studies comparing gingivoperiosteoplasty with control groups conclude that “gingivoperiosteoplasty results in a more uniform position of the hard palate.” There was no demonstrable impairment of maxillary growth in the patients treated with gingivoperiosteoplasty. 53

A single procedure performed at 3 months that closes the lip, anterior hard palate, and alveolus has been performed. 54 The secondary palate is repaired by 12 months of age. Early follow-up suggests high rates of osseous union. It is noted that procedures involving mobilization of large regions of periosteum can exaggerate a skeletal deformity, leaving the patient with marked malocclusion and facial disharmony. If, however, the prevomerine suture is not disturbed during surgery, many authors believe that early surgery is no more deleterious to maxillary growth than surgery later in life. 55

In summary, Millard et al.’s 56 latest contribution demonstrates the complexity and trade-offs that arise in managing these patients through to adult life. The use of primary gingivoperiosteoplasty, in addition to lip adhesion, yield the advantages of moving the palate into a normal position, stabilizing the arch, and providing a potential conduit for the eruption of teeth. Gingivoperiosteoplasty provided a symmetrical platform for the early repair of the nose and avoided the difficult anterior fistula. Nonstandard orthodontic care prevented a definitive statement about long-term maxillary growth.

**Unilateral Cleft Lip Repair**

Modifications of the straight-line repair introduced by Thompson 3 continue to enjoy limited use in present-day plastic surgery for the correction of the minimal cleft lip deformity. The technique has widest application in correcting notch deformities at the vermillion.

The Tennison triangular flap repair, with Randall’s 57 suggested revisions, is still popular today. In their long and distinguished experi-
ence, Brauer and Cronin obtained excellent results with the Tennison lip repair, stressing the value of geometric planning as proposed by Randall but noting that the repaired side should be designed 1 mm shorter than the nonleft side to avoid excessive vertical height of the lip.

A review of the various viewpoints and refinements proposed for triangular flap techniques supports the premise that deviations from the correct lip length would be equally divided between too long and too short. No patient from the entire group had a lip that started too short and became equal or that started equal and became too long. It was concluded that a repaired unilateral cleft lip retains the configuration and length determined at the time of the initial repair.

Indications for cleft lip repair by the triangular flap method are impossible to define. Some believe it is never appropriate because the resultant scar crosses the projected line for the philtral column. Others reserve the technique for use primarily in extremely wide clefts, whereas still others use it routinely.

The rotation-advancement repair consists of an upper triangular flap inset into the rotation defect of the medial segment. The technique allows the scar to follow the projected line of the philtral column except in its uppermost portion, where it arcs beneath the columella.

Later refinements of the rotation-advancement principle consist of broader utilization of the mucosal parings to aid in the correction of the associated nasal deformity. This repair, with its more recent modifications, is one of the most popular approaches to the correction of unilateral cleft lip deformity among surgeons today.

Criticisms of the rotation-advancement method relate to its technical difficulty in wide clefts, the necessity for wide soft-tissue undermining, tension across the nostril sill, and a tendency toward a constricted nostril on the side of repair.

Proponents of rotation-advancement emphasize the role of presurgical orthopedics, nasal correction, and, where indicated, lip adhesion as part of the rotation-advancement procedure, as well as the use of an L-flap to increase vestibular lining and the proper use of a C-flap to increase columellar length on the cleft side. Alar base advancement, alar cartilage lift, and medial and lateral alar cartilage freeing must be appreciated to maximize the chances for the anticipated result.

Repairs involving minimal undermining and detachment of the alar base leave fibrous bands between the alar cartilage and the maxilla, and aberrant muscle fibers from the orbicularis and the levator labii superioris alaeque nasi. These bands and muscles inevitably keep the alar cartilage tethered laterally and contribute to subsequent drift of the base of the ala laterally and superiorly.

Unfortunately, comparison of the triangular flap and rotation-advancement techniques for the repair of unilateral cleft lip have yielded a random selection of dissimilar study groups, with a greater proportion of complete clefts for the triangular flap patients than for the rotation-advancement group. Accordingly, observations regarding postoperative nasal deformity and the need for revisions in triangular flap repairs may be related to the fact that the preoperative nasal deformity was more severe. An apparent meaningful difference between the groups may have been due to the presence of hypertrophic scars in the rotation-advancement repair group. No significant difference in vertical lip length was noted, although both groups of patients had lips that were either too short or too long postoperatively. The overall score of results slightly favored the rotation-advancement repair, but there was no statistically significant difference between the two groups. The effect of repair on facial growth, if any, has not been investigated.

Recently, attention has been directed, appropriately, to reconstruction of the vermilion in cleft lip repair. Noordhoff discussed the anatomy of the vermilion and the proper orientation of the white skin roll and the mucosa-vermilion border (red line). The vermilion is widest at the base of the philtral column, and there is always a deficiency of vermilion on the cleft side. The author described a lateral vermilion flap used to augment the deficient vermilion beneath the white skin roll on the cleft side. The author described a lateral vermilion flap used to augment the deficient vermilion beneath the white skin roll on the cleft side of the Cupid’s bow. Although the necessity of the lateral vermilion flap may be argued, as brought out in Millard’s discussion, there is no doubt that attention to the vermilion component of lip repair adds significantly to the result.

The concept of differential reconstruction of the orbicularis oris muscle in unilateral cleft lip repair has evolved and assumed greater emphasis. The most important step in the
treatment of unilateral deformities is the differential rearrangement of the muscle components to correct their insertion and reorient the muscle fibers. The three different components of the orbicularis oris muscle should be repaired independently to permit each to function separately. The repair should consist of insertion of the nasal bundle into the anterior nasal spine or contralateral medial footplate of the alar cartilage, correction of the misdirected nasolabial bundle, and end-to-end union of the deep fibers of the vermillion. Park and Ha emphasized the importance of accurate repair of two different components of the orbicularis oris muscle. The superficial component of the orbicularis muscle serves as a retractor, whereas the deep component serves as a constrictor of the lip. The antagonistic actions of these two orbicularis muscle groups during lip movement affect the balance and dynamics of the repaired lip if not properly and anatomically aligned at the time of surgery.

**Primary Repair of the Unilateral Cleft Lip Nose**

Traditional methods of lip repair did little to address the associated cleft nasal deformity. Although alar base repositioning was common, release, repositioning, and dissection of the nasal complex was avoided because of the general feeling that growth disturbances would ensue. The literature now suggests that many cleft surgeons consider correction of the associated cleft alar deformity to be part of the lip repair procedure.

A comparative study of the skin envelope of the unilateral cleft lip nose subsequent to rotation-advancement and triangular flap lip repairs revealed similar vertical asymmetries of the nasal skin envelope in both groups. The alar dome on the cleft side was depressed, the columnella was short on the cleft side, and there was hooding of the nostril apex. The principal difference between the two lip repairs was observed in the horizontal dimension in the nasal skin envelope. Specifically, the position of the alar base was more normal after the Millard repair, whereas the triangular flap repair left the alar base laterally displaced.

Nonsurgical molding of the cleft alar deformity in the early neonatal period has its advocates. The nose is stented at the time of surgery, which is performed between days 2 and 7 of life. A precedent for molding the embryonic cartilages has been demonstrated in congenital auricular deformities. Advocates of the technique indicate that the alar cartilage exhibits the same elasticity as the auricular cartilage in the early neonate. They believe that there is a good possibility of correcting the cleft lip nasal deformity with a nonsurgical procedure in the early neonatal period. The mold or a retainer was placed in the affected nostril and kept in place for 3 months. Nasal shape and symmetry were considered superior to those conventionally operated on at about 3 months of age. Except for a single nasal infection, there were no complications of the procedure.

Long-term follow-up on the nasal deformity corrected at the time of lip repair suggests that correction of the nasal deformity at the time of lip repair endures, does not disturb growth, and is beneficial to nasal structure. Mobilization of the lower lateral cartilage from the overlying nasal skin, simultaneously releasing it on the cleft side from attachments along the piriform aperture and involving the medial crus in the septal area, is intrinsic to the repair. The method creates a flap of the lining mucosa and cartilage, with the lateral portion of the lower lateral cartilage advanced medially and cephalad and the medial crus advanced toward the tip. Suture-fixation or bolster-suturing techniques are used to hold the cartilage in position. The excellent results depicted should certainly encourage the cleft surgeon to consider correction of the cleft nasal deformity at the time of lip repair.

The Tajima repair, which uses a reverse-U incision along the alar margin with a back-cut along the vestibular lining at the junction of the piriform aperture to release the plica vestibularis, has been variously reviewed.

An objective evaluation of the Tajima secondary cleft lip nose correction is offered by Coghlan and Boorman. The early and late results of surgery were objectively measured by computer on 24 unilateral complete cleft lip and palate patients. Late results ranged from 1 to 4.2 years. Although some groups have had a favorable experience, the concern is that in late follow-up the deformity may recur and the nasal shape may not be satisfactorily distinguished from the preoperative appearance.

Use of a dynamic postoperative nostril splint in surgery of the nasal tip has contributed to efficient maintenance of surgical results by opposing healing contraction. Splinting for 3 to 4 months to retain the corrected contour of the cleft lip nose has been advocated. This in-
cludes recommendations of moldable silicone rubber retainers to add volume to ready-made nostril splints. This method is said to allow precise contouring of the deformity, which, reportedly, can be retained postoperatively. Cartilage memory and scar contraction remain the factors responsible for long-term deterioration of early results. Whether nasal splinting...
(even if continued for 6 months postoperatively) can preserve the immediate operative result permanently will remain contentious. A prospective randomized trial in which objective, reproducible assessment systems for nasal structure are applied is likely to be difficult to achieve.83 Early correction of the nose in unilateral cleft lip patients using an open method has been described.84 The cleft nasal deformity was corrected during the preschool years, and follow-up evaluation was done when the patients were between 15 and 19 years of age. Although early results appeared to be satisfactory, as the patients approached their adolescent growth spurt at approximately age 15, undesirable features became obvious. Patients showed strikingly large noses, with large amounts of subcutaneous fat, thick skin, and a wide nasal tip. These features were considered unique to the patients who had undergone early cleft nasal repair by the open method. Because of these findings, the authors have discontinued open rhinoplasty in preschoolers and now delay secondary correction until patients are 9 to 12 years of age.

The preliminary results of open-tip rhinoplasty85,86 at the time of lip repair in unilateral and bilateral cleft lip have been reported. The method consists of simultaneous lip closure and open-tip rhinoplasty involving nostril and columellar rim incisions. The surgical emphasis is on alar cartilage manipulation, with the skin being adjusted secondarily. The tip of the nose is reconstructed from alar dome repositioning and fixation under direct vision, construction of a subcutaneous soft-tissue pad, and caudal advancement of dorsal nasal skin. Wide dissection of nasal mucosa from the medial wall of the maxilla and piriform margin allows correct positioning of the alar base and prevents intranasal stenosis. Wide dissection and advancement of cheek soft tissues support the alar base and allow for a tension-free closure of the lip. Long-term follow-up is required before definitive assessment of the implications of these extensive procedures for growth.

CONCLUSIONS

It is clear that excellent results in the management of unilateral cleft lip may be achieved by multiple approaches. Our current approach consists of the following:

1. Early assessment in the first days of life and commencement of presurgical orthope-
dics. It is our belief that alignment of the alveolar segments creates the foundation upon which excellent results of lip and primary nasal surgery are dependent.

2. At 3 months of age, we perform a rotation–advancement type lip repair and primary nasal surgery. Where the maxillary segment alignment allows, primary gingivoperiosteoplasty is performed at this time. We do not routinely use nasal splinting following rhinoplasty (Figs. 1 through 3). The accelerating appreciation of normal and abnormal facial anatomy and the implications of this greater understanding as it pertains to cleft lip and cleft lip nose deformities have been examined. The history of cleft lip repair, technical refinements, advantages, disadvantages, and controversies surrounding the prevalent techniques for repair of the unilateral cleft lip and unilateral cleft lip nose, including potential sequelae in terms of facial growth, have been expounded. Although it is obvious that many areas remain incompletely understood, much progress has been made in providing patients with consistent, predictable results from repair of the unilateral cleft lip and cleft lip nose deformities. The key insights are the following:

1. The appreciation of the significance of the component (nasal, superficial, and deep) reconstruction of the orbicularis oris muscle in lip repair.
2. The recognition of the anatomy of the vermilion and the need to consider using a lateral vermilion to augment the deficient medial cleft vermilion.
3. The features of the cleft lip nasal deformity and the recognized benefits of early integrated repair at the time of lip repair.
4. The critical importance of the premaxillary–vomerine suture to facial growth.

Controversy will persist with respect to many areas of management of unilateral cleft lip deformities, but the exciting era of understanding the basic molecular defects responsible for deformity and disease is upon us. As answers to these fundamental questions emerge, enormous innovation in management will ensue. It is likely that fundamental changes in our approach to treating cleft lip and palate are at hand that will bring further benefits to our patients.

REFERENCES


1. THE COMMONLY USED CLASSIFICATION SCHEME DEVISED BY KERNAHAN AND STARK USES WHICH ANATOMIC FEATURE AS ITS FOCAL POINT?
   A) Lip
   B) Nose
   C) Alveolus
   D) Incisive foramen
   E) Velum

2. THE NORMAL VERMILION IS WIDEST AT WHICH POINT?
   A) The tubercle
   B) The peaks of Cupid’s bow
   C) The midportion of the lateral lip segment
   D) The modiolus

3. IN UNILATERAL CLEFT LIP, THE SUPERFICIAL PART OF THE ORBICULARIS ORIS LOCATED IN THE VERMILION
   A) Is distorted along with the deep portion
   B) Inserts into the alar base
   C) Is simply interrupted without distortion
   D) Is hypoplastic and cannot be identified

4. INFORMATION ACCUMULATED SUGGESTS THAT IN UNILATERAL CLEFT LIP DEFORMITIES, THE DEEP SPHINCTER PART OF THE ORBICULARIS ORIS MUSCLE
   A) Does not reach the extremity of the interrupted vermilion
   B) Inserts into the interrupted vermilion
   C) Inserts into the alveolar cleft
   D) Inserts into the alar base
   E) Inserts into the lateral lip dermis

5. ALL OF THE FOLLOWING ARE FEATURES OF THE UNILATERAL CLEFT LIP DEFORMITY EXCEPT
   A) There is a notch in Cupid’s bow on the noncleft side.
   B) The lower lateral cartilage is attenuated.
   C) The alar base is outwardly rotated in a flare.
   D) The alar rim is distorted by a skin curtain.
   E) The inferior edge of the septum is dislocated out of the vomerine groove.

6. A RETROSPECTIVE ANALYSIS OF ANESTHETIC COMPLICATIONS OF NEONATAL CLEFT LIP REPAIR CONCLUDED ALL OF THE FOLLOWING EXCEPT
   A) Patients should be gestationally mature.
   B) No opioid analgesia should be given during the surgical procedure.
   C) Experienced nurses should care for the infants postoperatively.
   D) No oximetry is necessary.

7. ALL OF THE FOLLOWING APPLY TO LIP ADHESION EXCEPT
   A) It was mainly popularized by Mirault in the 19th century.
   B) In principle, it decreases the tension of definitive lip repair.
   C) Randall suggests short, broad, triangular flaps.
   D) Millard proposes a high adhesion that avoids scars on the area of the definitive repair.
   E) Furnas proposes a straight-line adhesion as a first step before rotation advancement repair.
8. CRITICISMS OF THE ROTATION ADVANCEMENT METHOD INCLUDE ALL OF THE FOLLOWING EXCEPT
   A) It is technically difficult in wide clefts.
   B) Wide soft-tissue undermining is necessary.
   C) There is tension across the nostril sill.
   D) Rotation-advancement repair cannot be performed without prior lip adhesion.
   E) There is a tendency toward a constricted nostril on the side of the repair.

9. PRIMARY REPAIR OF THE UNILATERAL CLEFT LIP
   A) Is unimportant
   B) Should not be performed for fear of producing growth disturbance
   C) Has no precedent in the literature
   D) In long-term follow-up by multiple authors, does not disturb growth and is beneficial to nasal structure.

To complete the examination for CME credit, turn to page 1293 for instructions and the response form.