Primary Repair of Bilateral Cleft Lip and Nasal Deformity

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Learning Objectives: After studying this article, the participant should be able to: 1. List five principles that guide synchronous repair of bilateral complete cleft lip and nasal deformity. 2. Explain how different growth rates for the principal nasolabial features are applied during primary repair. 3. Describe two approaches for positioning the alar cartilages to form the columella. 4. Discuss the influences on referral patterns for a newborn with bilateral cleft lip.

Traditional repair of bilateral cleft lip focused on labial closure but accentuated the nasal deformities, which were addressed later. By the end of the past century, single-staged labial closure had replaced the old multistaged procedures and the technical emphasis had begun to shift from secondary to primary nasal correction. Now, presurgical maxillary orthopedics sets the bony foundation for synchronous nasolabial repair and for closure of the alveolar clefts. The study of normal nasolabial growth and the typical stigmata of the conventional methods provides the necessary foreknowledge to guide surgical sculpture in three dimensions and to anticipate the fourth dimension. The convergence of several forces are changing referral lines for children born with bilateral cleft lip. These include affirmation of centers of excellence, surgeons' self-regulation, prenatal diagnosis, economics of health-care delivery, and increasing parental sophistication. These pressures are not necessarily in conflict. Care by a subspecialized plastic surgeon and experienced team is in the best interests of the child and the third-party payer. (Plast. Reconstr. Surg. 108: 181, 2001.)

James Barrett Brown and colleagues introduced their 1947 article with the pithy statement that a bilateral cleft lip is twice as difficult to repair as a unilateral cleft and the results are only half as good.1 Indeed, techniques for correction of bilateral cleft lip have lagged behind those for unilateral cleft lip. The infant with bilateral cleft lip has been subjected to multiple procedures only to endure sundry revisions throughout childhood. Despite the surgeon's best efforts, the child's diagnosis was painfully obvious to all—even at a distance—and these stigmata were not easily erased by revisions. These branded children beckoned surgeons to change their traditional operative strategies.

Over the past decade, two important advances have been made in the repair of bilateral cleft lip and nasal deformity: (1) evolution to single-stage nasolabial closure with positioning of the alar cartilages and sculpting of the soft tissues to shape the columella and nasal lobule, and (2) improved techniques for presurgical maxillary alignment to permit closure of the alveolar clefts and facilitate primary nasolabial repair. Although the principles for single-stage repair are established, craftsmanship continues to evolve. Now, over a half century after Barrett Brown's discomfortable observation, it can be said that the outcome for the infant born with bilateral cleft lip is equal to and can surpass that of its unilateral counterpart.2 Furthermore, given the preoperative advantage of nasolabial symmetry, these children require very few revisions.

Before undertaking primary repair of double cleft lip, there are some lessons to review.

LESSONS FROM SURGICAL HISTORY

Many complex malformations, once only reparable by staged operations, can be corrected, usually more successfully, in a single procedure. This lesson is illustrated in the annals of the bilateral cleft lip and nasal deformity. Emphasis had focused on labial closure and ignored nasal distortion. Surgical text rec-
ommended staged repair, one side of the cleft and then the other. There was a misconception that the diminutive prolabium lacks the potential for growth. Techniques for repairing the double-labial cleft were adapted from those used for the more common unilateral form, and they typically involved the introduction of a rectangular or triangular flap from the lateral labial elements to augment prolabial height. These procedures left geometric (often asymmetric) labial scars and usually resulted in a long-lip deformity and a tight upper lip. Straight-line repair minimized vertical elongation of the central lip but produced an abnormally wide and shield-shaped philtrum. There was longstanding controversy over whether or not to preserve the prolabial vermilion, leave it as a tiny strip, or excise it completely. Apposition of the orbicularis oris muscle was usually not mentioned. Some surgeons thought muscular closure would inhibit premaxillary growth. Nevertheless, with increasing attention to muscular closure in the unilateral deformity, reports began to underscore the importance of orbicular repair in bilateral clefts.

Surgeons conceded to the complexity of the bilateral cleft nasal deformity and deferred correction. The conventional teaching was that the columella is inadequate, and numerous “secondary” procedures were devised to elongate the “short columella.” There are two major strategies. The first, popularized by Cronin, involves rotating bipedicled straps of tissue from the nostril sills. This method gives modest columellar length. A second method, the forked-flap procedure of Millard, involves recruiting labial tissue to the columella. There are two permutations of this method. In infants with a wide prolabium, the tines of the forked flap are banked at the time of labial closure and transposed to the columella in early childhood. More often, Millard prefers three-stage columellar lengthening: (1) bilateral labial adhesions to stretch the prolabium; (2) elevation, rotation, and banking the tines while narrowing the philtrum (at age 18 months); and, finally, (3) retrieval of the prolabial prongs and elevation, along with the medial crura, to augment the columella (at age 2 years).

The forked-flap method, like all secondary procedures, causes peculiar tertiary distortions. Most techniques introduce a nexus of scars across the columellar-labial junction. This produces a midline nasolabial crease that deepens with smiling. The circumferential philtral scar produces a bulge rather than a dimple. The best prolabial scars follow repair in infancy, done under minimal tension. Recruiting tines of a forked flap from the central lip in a child can cause thickened and permanently wide philtral scars. Even in the best of hands, the staged forked-flap procedure results in an unusual appearance: (1) a rectangular columella (with a broad base and without a waist); (2) a sharp columellar-labial angle; (3) abnormally elongated/enlarged nostrils; (4) a tendency to columellar over-elongation with a disproportionate ratio of nostril length-to-nasal tip; and, sometimes, (5) a downward drift of the columellar base. Furthermore, the medial crura become unnaturally positioned in the tip, resulting in a break at the columellar-labular connection. Because of these problems, some surgeons began to wonder whether labial skin belongs in the columella—or if more tissue is needed at all.

Whereas delayed nasal repair was customary for the repair of bilateral cleft lip, the simultaneous correction of the unilateral left lip and nasal deformity became accepted practice. Furthermore, there was no evidence that early manipulation of the alar cartilage impairs the development of the nasal tip.

LESSONS FROM BILATERAL CLEFT STIGMATA

Every child with a repaired bilateral cleft lip has a characteristic appearance whose origins are both intrinsic to the malformation and iatrogenic. The philtrum is bowed, wide, undimpled, overly long, often asymmetric, and lacking a white ridge. If the prolabial vermilion-mucosa is preserved, the free margins of the lateral labial elements hang like swags, flanking a thin median tubercle that is covered by insufficient vermilion and chapped mucosa (“whistling lip deformity”). In profile, the upper lip is flat or convex, whereas the lower lip everts (“cleft lip lower lip deformity”). The child struggles to obtain bilabial closure over a protrusive, retroclined, and vertically elongated premaxilla. The accompanying nasal deformities are primarily deformational but are also postsurgical. The tip is broad, the medial crura are pulled inferior-posteriorly, the nostrils are slumped, the alar domes are buckled and splayed, and the alae nasi are flared, sometimes likened to “cat’s knees.” Often the alar lobules are hypoplastic (a primary deformity).
The caudal margin of the alar cartilage protrudes into the lateral vestibule, producing an oblique ridge or web. Without amends for the vertically long lateral labial elements and normal muscular attachment of the alar bases, an unnatural elevation of the alae nasi will occur and become more pronounced whenever the child smiles. But of all the nasolabial distortions, the short columella is most obvious.

Principles that guide the surgical repair of bilateral cleft lip and nasal deformity have been induced from the stigmata of conventional techniques and by study of the literature: (1) Symmetry. This is foremost. Staged repair portends asymmetry. Even the smallest nasolabial difference on the two sides will magnify with growth. (2) Primary muscular continuity. Orbicularis oris muscular bundles must be completely mobilized from the lateral labial elements and apposed throughout the vertical extent of the upper lip. (3) Proper philtral size and shape. The constructed philtrum widens remarkably (in the upper portion more so than inferiorly) and displays considerable vertical growth. (4) Formation of median tubercle from lateral labial elements. There is no prolabial white roll in the complete deformity, and both central vermilion and mucosa are deficient. (5) Primary positioning of alar cartilages to construct the nasal tip and columella. Techniques based on this principle have dramatically changed the faces of children born with bilateral cleft lip.

The Key: Primary Repair of the Nasal Deformity

Nasal dissection of stillborn infants with bilateral labial clefting reveals that the alar domes and middle crura are splayed, caudally rotated like a “bucket handle,” and subluxed from their normal anatomic position overlying the upper lateral cartilages. Broadbent and Woolf described a case of primary medial advancement of the alar domes combined with excision of skin from the broad tip. However, it was McComb who led the vanguard for primary columellar elongation. He initially tried primary elevation of a forked flap and published his follow-up analysis 10 years later, including measurements of columellar growth. By then he had become disenchanted with this strategy and presented a revised technique for primary nasal repair in 1990—without a forked flap. In the first stage, McComb used an external incision (“flying bird”) to open the nasal tip and allow the apposition and suspension of the splayed alar cartilages. The nasal tip was narrowed by V-Y plasty. Bilateral adhesions were done, followed by definitive labial repair at a second stage. The columellae looked quite normal in McComb’s assessment at 4 years.

Mulliken also focused on early positioning of the alar cartilages. This was initially done at a second stage in conjunction with intranasal transposition of the banked tines of a forked flap. By 1987, banking had been abandoned, and primary columellar lengthening and nasal-tip projection were achieved solely by apposition and elevation of the alar domes and by sculpting the nasal soft tissues. Other surgeons were on the same track to primary nasal correction. In 1991, Trott and Mohan were working in Malaysia, where socioeconomic factors made multistaged repairs impractical. They devised a single-stage nasolabial repair, based on open rhinoplastic exposure of the dislocated alars. Cutting and associates described another variation on the open-tip approach and added presurgical stretching or elongation of the columella.

“The columella is in the nose” became the shibboleth of surgeons who advocated primary nasal repair. The old, non-anatomic techniques that involved secondary recruitment of prolabial skin into the columella were wrong. Instead, the alar cartilages should be placed in the proper position at the time of labial repair, followed by trimming and redraping the soft tissues of the nasal tip. No longer would the columella include labial (often hair-bearing) skin, the nasolabial junction be transgressed by scars, or the philtrum be encircled by scar tissue.

Lessons from the Fourth Dimension

Unlike the sculptor who works in stone, the surgeon must work with a patient who grows and whose nasolabial proportions change. Thus, the surgeon must conceptualize the child’s appearance as a young adult. To do so, the surgeon must have a thorough understanding of three-dimensional form and fourth-dimensional alterations that occur with normal growth. Thankfully, Farkas and colleagues documented the changes in the important nasolabial features, between 1 and 18 years, in 1593 North American Caucasians. Nasal height (n-sn) and width (al-al) develop early, reaching a mean of 76.9 and 87 percent of adult size, respectively, by age 5 years. In contrast, nasal-tip protrusion (sn-prn) and columellar length
(sn-c’) develop slowly, attaining a mean of only two-thirds of adult size by 5 years of age. All the labial landmarks grow rapidly, reaching approximately 90 percent of adult proportions by age 5 years. The cutaneous upper lip attains adult height by 3 years in girls and by 6 years in boys.30

A working fourth-dimensional hypothesis for the repair of bilateral cleft lip is to craft on a small scale those features that are programmed for rapid growth, compared with normal, age-matched infants. The corollary premise is that slow-growing features can be constructed of normal size or slightly larger than normal size.16 This accounting for temporal changes must also include the nasolabial distortions that are particular to children with repaired bilateral cleft lip. Such knowledge can be gained only by observation of older patients, one’s own and those of predecessors and colleagues. The fast-growing features grow even faster in the child with bilateral cleft lip, with the exception of the median tubercle. Conversely, the slow-growing features, specifically nasal-tip projection and columella, seem to grow more slowly in the child with bilateral cleft lip.29

PRESURGICAL PREMAXILLARY–MAXILLARY MANIPULATION

Synchronous nasolabial repair can be accomplished only after proper alignment of the three maxillary segments. Most bilateral cleft lips are complete, but sometimes there is a tiny band on one side that causes rotation of the premaxilla. The protrusive premaxilla must be retracted and centralized, whereas the lateral maxillary segments usually require expansion. There are two basic strategies for presurgical premaxillary orthopedic manipulation, active and passive. The latter method is favored by those who are concerned about the potential deleterious effects of forcing the segments into position. The passive molding plate is retained by undercuts; this maintains the transverse width of the maxillary segments. Because there is no expansion, space is often inadequate for the premaxilla. External force is needed to retract the premaxilla, using either adhesive tape, an elastic band attached to a headcap, or bilateral labial adhesion. Because there is no muscle in the prolabial element of a bilateral complete cleft lip, preliminary labial adhesions are prone to dehisce. Furthermore, external traction techniques tend to focus pressure inferior to the basilar premaxilla, causing it to lingually incline and causing the vomer to bow.

The most commonly used active presurgical device is based on the prototypic design of Georgiade and associates,30,31 refined and popularized by Latham while working in collaboration with Millard (Fig. 1).32,33 The acrylic plates of the custom-made appliance are pinned to the maxillary shelves. A looped wire is passed transversely through the neck of the premaxilla, just behind the premaxillary alveolus and well anterior to the premaxillary-vomerine suture. The maxillary segments are expanded by a ratcheted screw in the midplane of the device, which is turned daily by the child’s parents. Elastic chains on each side are connected to the trans-vomerine wire, looped around a pulley in the posterior section of the appliance, and attached to a cleat on the most anterior point of the maxillary acrylic plates. Tension on the elastic chain retracts the premaxilla; tension may be periodically adjusted. Typically it takes about 6 weeks to align the premaxilla with the expanded palatal segments to effect closure of the alveolar clefts (gingivoperiosteoplasty). Latham’s device is most successful in correcting the premaxillary anteroposterior position; however, the movement is more retroclination than retroposition. The device is somewhat less successful in amending rotation and is least successful in preventing vertical elongation.
Some orthodontists think there are no advantages to premaxillary orthopedics in the management of bilateral cleft lip. However, proponents of active versus passive techniques constitute the two major sides of the ongoing debate. Early longitudinal studies of children managed with a Latham device show no serious deleterious effects on occlusion and growth. However, critics of active premaxillary orthopedics document long-term evidence for minor midfacial retrusion. Whatever the outcome in terms of midfacial position, three advantages of presurgical alignment of the maxillary segments must be underscored: (1) it permits philtral design of proper proportions, (2) it facilitates primary nasal correction, and (3) it allows closure of the alveolar gaps, thus preventing fistulas and (possibly) permitting bony ingrowth and stabilization of the arch. Furthermore, if there is a near-normal maxillary foundation, there could be less postoperative prolabial distortion and interalar widening. If the child exhibits midfacial retrusion, maxillary advancement is a predictably successful procedure when done after completion of growth. In the near future, it is likely that maxillary distraction with an entirely internal device will be available for patients who might benefit in childhood.

Primary Repair of the Primary Palate

Markings

The infant is typically 4 to 5 months old at the time of synchronous repair. The philtral flap is designed with slightly biconcave sides and a dart-shaped tip. The size of the flap depends on the race and age of the infant and on the appearance of the parents. For a Caucasian infant 4 to 6 months of age, suggested dimensions are 6 to 8 mm for philtral flap length: 3 to 4 mm wide between the peaks of Cupid’s bow and 2 mm wide at the columellar-labial junction. A strip of skin is drawn on each side of the philtral flap; these will be deepithelialized to simulate the philtral ridges. The proposed Cupid’s bow peak-points are sited on the lateral labial elements so there will be sufficient central white roll for the handle of the Cupid’s bow and enough vermilion to construct the median tubercle. The alar base flaps are drawn at their junction with the lateral labial elements. The vermilion-mucosal line is tattooed, as are the other important anatomic points to be preserved during repair (Fig. 2, above, left).

Dissection

The philtral flap is incised, the flanking tabs are deepithelialized, and the remaining prolabial skin is discarded. The lateral white line- vermilion-mucosal flaps are incised, and the alar base flaps are elevated. Orbicular muscular bundles are dissected from the lateral labial elements (Fig. 2, above, center). The splayed alar cartilages are exposed through the rim incisions; it is helpful to support the cartilages with a cotton-tipped swab (Fig. 2, above, right).

Labial Closure

Mucosal flaps are elevated from the lateral and medial sides of the cleft defects to construct the nasal floors. Gingivomucoperiosteal flaps are apposed, closing the alveolar clefts. The premaxillary vermilion-mucosa is trimmed to shorten the anterior wall of the gingivolabial sulcus (Fig. 2, second row, left). The remaining premaxillary mucosa is sutured to the periosteum to form the posterior wall of the anterior gingivolabial sulcus. The lateral labial elements are advanced medially as the buccal sulci are closed. The lateral mucosal flaps form the anterior wall of the central sulcus. The orbicular muscles are apposed, inferiorly-to-superiorly, throughout the vertical height of the lip. The uppermost suture suspends pars peripheralis to the periosteum of the anterior nasal spine (Fig. 2, second row, right, left panel).

The redundant tips of the lateral labial flaps are trimmed to form the median tubercle (Fig. 2, second row, right, right panel). The distal end of the philtral flap is inset. The philtral flap is secured to the muscular layer. This helps to depress the philtral flap and to raise the lateral labial flaps in an effort to simulate philtral ridges. Yet, a realistic philtral dimple and flanking columns seem just beyond the surgeon’s craft. The cephalic margin of the cutaneous flaps must be trimmed to correct for lateral labial height; rarely is adjustment necessary at the medial edges. These final steps in cutaneous closure should be done after nasal correction.

Nasal Repair

A midline nasal-tip incision is not necessary, for with experience, it is possible to fully visualize the dislocated alar cartilages through bilateral rim incisions. An interdomal mattress suture is placed to appose the middle crura and genua. One or two mattress sutures sus-
pend each lateral genu (and lateral crus) to the ipsilateral upper lateral cartilage (Fig. 2, third row, left). A cinch suture is placed through each alar base and is tightened until the interalar distance is less than 25 mm (Fig. 2, third row, right). The tips of the alar base flaps are trimmed to form the nasal sills. A suture placed through the dermis of each alar base to the underlying muscle serves to (1) prevent alar elevation with smiling and normal action of the depressor alae nasi muscles, and (2) form the normal cymal shape of the lateral sill (Fig. 2, third row, right panel).

Once the alar cartilages are in proper position, extra skin in the soft triangles becomes obvious and should be excised, including the skin of the lateral columella (Fig. 2, below, left, left panel). This resection narrows the tip, defines the columellar-lobular junction, elongates the nostrils, and narrows the columellar waist. There is also redundancy in the vestibular lining that becomes apparent after position-
ing the alar cartilages. Lenticular excision of this extra mucosa helps to obliterate the lateral vestibular web and supports the lateral crura (Fig. 2, below, left, inset). The completed repair is illustrated in Figure 2, below, right. Clinical examples are shown in Figures 3 and 4.

**TECHNICAL MODIFICATIONS FOR ANATOMIC VARIANTS**

Although most bilateral cleft lips are complete, there are various incomplete forms, symmetric and asymmetric, with a spectrum of clefting of the alveolar ridges and secondary palate.38

**Symmetric Bilateral Incomplete**

Certainly, the symmetric bilateral incomplete variant is the easiest to repair. Usually, the alveolar ridge is intact or there is minor notching. The steps are the same as described for the bilateral complete deformity, with some technical considerations. The most important decision is whether or not to build the median tubercle from the lateral labial elements (aforesaid principle 4). If there is sufficient probabal muscle, white roll, and vermilion, these can be preserved and apposed to the corresponding lateral elements in a bilateral butt joint. However, more often than not, the central lip should be constructed as in the
complete deformity. Careful attention should be given to the width of the philtral flap because it has the same tendency to overgrow transversely, as in the complete deformity.

Positioning the alar cartilages may not be necessary unless the columella measures short and the genua are slumped. Another caveat is to sufficiently narrow the interalar dimension, for it will widen with time.

Asymmetric (Complete/Incomplete)

There is a range of severity in the asymmetric complete/incomplete bilateral cleft lip, depending on the extent of soft-tissue bridging and underlying alveolopalatal disjunction. If a tiny cutaneous mucosal band on the incomplete side pulls over the premaxilla, it is usually best to divide it to allow presurgical centralization of the premaxilla and symmetric repair.

If one side is only partially cleft, the complete side should be addressed first. Unilateral dentofacial orthopedics, followed by a lip-nasal adhesion on the complete side, levels the surgical field before simultaneous bilateral nasolabial (and unilateral alveolar) clo-

![Fig. 4](image-url) (Above, left) Newborn with bilateral cleft lip, right cutaneous band, and intact right alveolus and secondary palate. (Above, right; below) Appearance of the patient at age 5.5 years.
sure. During the second stage, the surgeon should emphasize (overcorrect) repair on the more severely involved side. Furthermore, it is easier to match the “good” side to the “bad” side than vice versa.

**Complete Bilateral with Intact Secondary Palate**

The rare complete bilateral cleft lip with an intact secondary palate is a particular challenge. Neither external elastic traction nor dentofacial orthopedics is possible because the premaxilla is rigidly procumbent. There are two alternatives: (1) try to accomplish bilateral nasolabial repair over the protruding premaxilla; or (2) perform a premaxillary-vomerine ostectomy and positioning and bilateral gingivoperiosteoplasty, in conjunction with nasolabial repair. A word of caution: the risk of premaxillary necrosis is real. The incisions for premaxillary ostectomy/positioning and alveolar closure impair the premaxillary blood supply and limit venous drainage to the septal mucosa and preserved vomerine mucosa. Primary premaxillary positioning in an infant with bilateral complete cleft lip can cause midfacial retrusion. However, this is unlikely in a child with an intact secondary palate.

**Other Technical Variations on the Theme of Single-Stage Repair**

Open rhinoplasty is another way to access and primarily position the subluxed alar cartilages, as first described by Trott and Mohan in 1993. Their dissection plane is anterior to the medial crura. The prolabilial-columellar unit is pedicled on the dorsal nasal skin and based on the paired columellar arteries. Their philtral flap is designed to correspond to the width of the columellar base. Because the lateral philtral incisions extend across the columellar base, this tissue cannot be used to construct the medial sills; the sills are formed almost entirely from the alar flaps. Redundant skin is removed from the lateral labial elements. The interdomal fat is elevated, the middle crura are apposed and secured to the septum, and the soft tissues are gathered to enhance tip projection (Fig. 5). Their postoperative photographs show normally proportioned columellar length and nasal-tip protrusion. The philtra appear to be wide. Perhaps this is acceptable in Asian children, who have a slightly broader Cupid’s bow than Caucasian children. Distal ischemia is a potential problem if the philtral flap were to be designed smaller using Trott’s method.

Furthermore, this technique requires sutural reconstitution of the columellar labial angle, and this could also impede philtral circulation.

Nakajima and coworkers introduced presurgical and postsurgical molding to minimize the bilateral cleft nasal deformity. Nakajima and associates extended this strategy to preoperative stretching of the columella. They fabricated an acrylic, double outrigger and prolabial band, attached it to a passive palatal molding plate, and secured it to the cheeks with tape. The laboral markings are similar to those of Trott and Mohan. However, their open-tip approach differs in that the prolabilial-columellar flap is elevated at a deeper plane (membranous septum). This dissection is less likely to compromise the vascularity of the distal prolabium. Their placement of interdomal sutures is done from the underside of the alar cartilages (Fig. 6). Cutting and associates underscore that preoperative expansion of nasal lining is as important as columellar elongation because it lessens tension on the interdomal apposition and minimizes widening of the nasal tip. They emphasize that the molding prongs must push anteriorly because of the tendency to produce a “turned up” nasal tip. Neither the Trott nor the Cutting methods include cutaneous excision of the domal-columellar rims or resection of vestibular lining.

**Revisions**

Symmetry is the major preoperative advantage of a bilateral cleft lip over a unilateral cleft. In part for this reason, the number of revisions for a double cleft lip should be less than for a single cleft lip. The most common secondary problem is extra mucosa (festooning) in the lateral labial elements. Often this is associated with minor vertical deficiency of the median tubercle. It is best to wait until after the
permanent central incisors have erupted and, if possible, after the premaxilla is in the correct sagittal position and angulation before adjusting the free labial margin to give proper dental show. There is a litany of procedures for the “whistling lip” deformity (e.g., V-Y advancements, double/single Z-plasty, mucosal grafting). Particularly effective is the technique of deepithelialized, medially based submucosal flaps, tunnelled across the midline, to augment the central red lip.49 If there is insufficient lateral submucosa, a thick dermal graft serves to plump the median tubercle. Any excess mucosa can be trimmed to give a normal contour of the free margin in relation to the central incisors.

An uncommon, annoying problem is prolapse of the posterior wall of the anterior gingivolabial sulcus. Resuspension of the sulcal mucosa to the premaxillary periosteum is easily accomplished.

Nasal revisions in childhood are rarely needed. The most common is correction of an abnormally wide nose, requiring readvancement of the alar bases. Sometimes domal divergence must be addressed. The alar cartilages can be readjusted through rim incisions without the need for open rhinoplastic exposure. Final nasal adjustments are done after completion of growth and maxillary advancement (if necessary) (e.g., tip reduction, nasomaxillary narrowing, and septal resection).

Obligation to Periodic Assessment

Not only must surgeons prefigure the rate of growth of nasolabial features, compounded by the distortion of the deformity, but they also are obligated to periodically assess these changes to learn whether or not the predictions are accurate. Photography is the minimal documentation needed. Frontal and lateral views (with the head in a neutral position) are not enough; there must also be a basal view. For the latter, Pigott recommends that the nasal dome be placed well above a line drawn between the medial canthi.50 Lehman suggests
that McComb’s rule be followed in publishing photographs in support of a new method of cleft lip repair, i.e., show 10 consecutive patients with follow-up of 10 or more years. Although admirable, this rule is unrealistic, particularly if applied to the repair of bilateral cleft lip. Less than 10 percent of cleft lips are bilateral, so even a high-volume surgeon might see only a few such new patients each year. Thus, in an average professional career of 25 to 30 years, a surgeon will follow a relatively small number of these children to adulthood.

What is needed is a convenient, objective, and rapid way to evaluate nasolabial symmetry and proportions throughout the growing years. Although clumsy, a panel can be convened to assess the photographs, using a rating scale. Other methodologies are direct anthropometry and computer-aided photogrammetry (indirect anthropometry). There is great potential for laser scanning or a similar advanced technology to assess results. This technology will likely incorporate the soft-tissue landmarks used in medical anthropometry, so until it becomes available, anthropometry can be done the old-fashioned way, by handheld vernier caliper.

Intraoperative anthropometry provides baseline values for subsequent documentation of changes in the nasolabial dimensions. This was done for 45 consecutive infants undergoing single-stage repair of bilateral complete cleft lip and nasal deformity. Fast-growing features, and nasal length and nasal width, were set 88 percent and 96 percent, respectively, shorter than those of age-matched control infants. Slow-growing features, nasal protrusion and columellar length, were constructed longer than normal (130 percent and 167 percent, respectively). Because all labial features grow rapidly, these were downsized, with the exception of central vermilion-mucosal height that was deliberately made full.

Follow-up anthropometry required first measuring a cohort of normal children; this showed no differences from the results of the larger samples determined by Farkas and colleagues. Thirty-two children with repaired bilateral complete cleft lip were assessed from age 1 to 12 years. Nasal-tip protrusion and columellar height were at the mean or longer. Interalar distance was about 2 SD above the mean. Cutaneous upper labial height remained short in children younger than age 5 years but tended to scatter around the mean in the older children. A short cutaneous upper lip is attractive, provided there is sufficient height of the central red lip. Although vermilion-mucosal height was made full, in one-half of the older children this dimension was just below the mean. Nevertheless, the total upper labial height was either normal or slightly more than normal. Admittedly, there are problems with this study. First, the author’s technique evolved through three technical phases in the study period, from two-stage to single-stage repair—but the principles did not change. Second, the study was serial and retrospective, so the measurements in the three phases cannot be compared at the same point in time. Third, only 12 of 32 children had undergone the one-stage repair described herein, and their mean age at evaluation was 2.5 years.

A proviso for any anthropometric study is that normal scalar measurements do not necessarily equate with normal appearance. Nevertheless, it is the author’s strong impression that a child with measurements within 1 SD of normal looks better than a child with abnormal values. Furthermore, inclination of the upper lip influences appearance; i.e., a vertical upper lip looks longer than a protrusive lip. Thus, if the premaxilla is lingually inclined, which many are, this causes the upper lip to appear longer than it may be by mensuration.

It is difficult to compare one surgical method with another without a standard way to assess anatomic outcome. Kohout and colleagues used photogrammetric analysis of two surgeons’ results; one used the Mulliken method (group I) and the other used the Trott method (group II). The authors attempted to distinguish results attributable to methodologic design from those produced by execution of the particular method. Nasal-tip projection was above normal with both methods but more so in group I. Interalar dimension was abnormally wide in both groups. Columellar length, as a proportion of tip projection, approached normal in group I but was short in group II. Philtral width (in proportion to nasal width) was normal in group I but abnormally high in group II. An overly wide Cupid’s bow in group II was attributed to design.

Photogrammetry also permits the determination of nasolabial angle and nasal-tip angle, provided the head is not rotated. Kohout and associates found that the nasolabial angle was obtuse in early childhood but narrowed in late childhood and adolescence, presumably be-
cause of an increase in upper labial and columellar obliquity. Nasal-tip angle was blunted after both the Mulliken and Trott methods; there were no long-term measures of possible change in the slow-growing lobule.57

EPILOGUE

Older children with bilateral cleft lip continue to walk through the doors of cleft centers having undergone many procedures and requiring more. They are unhappy because of their appearance and functional problems, but the patterns of cleft care are changing. Fewer general plastic surgeons take on primary cleft lip repair. Two possible reasons for this trend are (1) established postgraduate fellowship training in pediatric plastic surgery, and (2) increased number of craniofacial teams and opportunities for pediatric plastic surgeons. Remarkably, this increased focus on care by a few is occurring despite diminished reimbursement for cleft work.

Outcome analyses in other pediatric surgical specialities underscore the value of subspecialization and high-volume operators. For example, there are lower inpatient costs and fewer complications if ureteroneocystostomy is done by fellowship-trained pediatric urologists as compared with general urologists.58 For pediatric cardiac surgeons, there is an inverse relationship between the annual number of procedures (or a surgeon’s case load) and inpatient mortality.59,60 In these examples, the determinant of outcome is indisputable; the results of cleft treatment are more difficult to assess.

Surely every infant born with cleft lip/palate, particularly a complete deformity, deserves the care of an experienced team. Scandinavia has the longest tradition of centralized cleft care, beginning in 1933. There is only one center in Denmark, two in Finland, two in Norway, and six in Sweden. The six-center Euro Cleft Study showed that standardization, centralization, and participation of high-volume operators were associated with good outcomes (and fewer revisions).61 In the United Kingdom, the “threshold volume” for primary cleft repair has recently been mandated, the result of an outcome assessment of the country’s cleft centers under the auspices of the National Health Service. The investigating committee concluded that the number of cleft units should be reduced drastically and that children should be sent only to teams composed of two surgeons, each seeing 40 to 50 new patients annually.62

Boorman goes further by raising the question of whether the rarity of the bilateral deformity and its attendant problems might argue for further control of referral.62

Such sweeping regulations would be unlikely in America’s decentralized health-care system, where change occurs slowly and, usually, voluntarily. Parameters for cleft care have been established by the American Cleft Palate-Craniofacial Association (ACPA).63 This organization also surveyed North American teams and determined standards-of-care delivery.64 Of 220 responding teams, the mean number of new cleft lip cases seen per year was 17. However, 37 percent of U.S. and Canadian centers reported performing fewer than 10 primary labial repairs each year. These numbers would be even lower with more than one plastic surgeon on a team. The ACPA evaluation did not address the level of activity necessary to maintain competency or provide optimal care. These issues are on the agenda of the Craniofacial Outcomes Registry, which is supported by a grant from The National Institute of Dental and Craniofacial Research. This program is being conducted with the cooperation of ACPA and its members. Prospective data, collected, and centralized by the Craniofacial Outcomes Registry, will permit inter-team comparisons of outcome, the purpose being to promote higher-quality care in the future.

In the meantime, the traditional referral lines for a newborn with cleft lip/palate are changing at the grassroots level. Rather than a pediatrician, the prenatal ultrasonographer or perinatologist is increasingly the first to guide parents to a plastic surgeon. Savvy parents often seek information (albeit non-peer-reviewed) in medical cyberspace, and parental networks advise these “Internauts” where to search. Having completed a cram course in cleft care, parents are likely to demand to take their infant to a cleft specialist outside their insurance network. Countercurrent to this rising tide of parental self-referral stand the health maintenance organizations that insist that all children stay within the network and accept assignment to their general plastic surgeon.

Rather than take up arms against third-party payers, why not join them? Consider collaborative outcome studies. One hypothesis would be that primary repair of a cleft lip in a restrictive insurance setting is penny-wise and pound-foolish. Perhaps if initial closure were done by
subspecialists, there would be fewer procedures and revisions, and decreased costs to all parties. It is easy to document efficacy in terms of speech, dentition, and facial growth. Standardized evaluation of aesthetic results should soon be possible using advanced technology, such as laser scanning. A child’s happiness and parental acceptance could be assessed by a quality-of-life outcome study.

Who should care for an infant with cleft lip? At a societal level, this question exemplifies the larger issue of what we want our health-care system to be—and for whom. But the same question also could be asked by the surgeon called to see a newborn with cleft lip. Remember the golden rule of pediatric care: Do for the child what you would want done for your own.

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REFERENCES
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Self-Assessment Examination follows on page 195.
Primary Repair of Bilateral Cleft Lip and Nasal Deformity
by John B. Mulliken, M.D.

1. ALL THE FOLLOWING ARE COMMON STIGMATA AFTER TRADITIONAL REPAIR OF BILATERAL CLEFT LIP, EXCEPT:
   A) Slumped genu
   B) Elevated alae nasi
   C) Bowed philtrum
   D) Proclined premaxilla
   E) Thin median tubercle

2. THE FOLLOWING ARE CRITICISMS OF THE FORKED FLAP PROCEDURE FOR COLUMELLAR ELONGATION, EXCEPT:
   A) Scars at columellar-labial angle
   B) Inadequate columellar length
   C) Wide philtral scars
   D) Abnormal columellar shape
   E) Medial crura at nostril-lobular junction

3. PRINCIPLES FOR PRIMARY REPAIR OF BILATERAL COMPLETE CLEFT LIP AND NOSE INCLUDE THE FOLLOWING, EXCEPT:
   A) Maintain symmetry
   B) Secure primary muscular continuity
   C) Design proper prolabial size and configuration
   D) Construct median tubercle from prolabial vermilion-mucosa
   E) Form columella and nasal tip by positioning alar cartilages

4. ALL OF THE FOLLOWING ARE POTENTIAL ADVANTAGES OF PRESURGICAL ORTHOPEDICS, EXCEPT:
   A) Permit gingivoperiosteoplasty
   B) Correct protrusive basal premaxilla
   C) Facilitate primary nasal correction
   D) Stabilize dental arch
   E) Allow proper design of philtrum

5. ALL OF THE FOLLOWING ARE FAST-GROWING NASOLABIAL FEATURES, EXCEPT:
   A) Interalar width
   B) Columellar length
   C) Cutaneous upper lip height
   D) Cupid’s bow width
   E) Median tubercle

6. WHICH OF THE FOLLOWING NASOLABIAL FEATURES IN A CHILD WITH REPAIRED BILATERAL COMPLETE CLEFT LIP IS LEAST LIKELY TO APPROXIMATE THE AGE-MATCHED NORMAL MEASUREMENT?
   A) Interalar width
   B) Nasal protrusion
   C) Columellar length
   D) Width of Cupid’s bow
   E) Total upper lip height
7. WHICH OF THESE NASOLABIAL FEATURES SHOULD BE CONSTRUCTED CONTRARY TO ITS PREDICTED GROWTH RATE?
   A) Nasal-tip protrusion
   B) Columellar length
   C) Interalar width
   D) Cupid's bow
   E) Median tubercle

8. WHICH IS THE EASIEST AND MOST AVAILABLE METHODOLOGY TO ASSESS NASOLABIAL FEATURES AFTER REPAIR OF BILATERAL CLEFT LIP?
   A) Anthropometry
   B) Computer-aided photogrammetry (indirect anthropometry)
   C) Laser surface scanning
   D) Panel evaluation
   E) Digital three-dimensional photography

9. WHICH (OR WHO) IS LEAST LIKELY TO GUIDE PARENTS OF AN INFANT WITH A BILATERAL COMPLETE CLEFT LIP TO A SUBSPECIALIST PLASTIC SURGEON?
   A) American Cleft Palate-Craniofacial Association
   B) Health maintenance organization
   C) General plastic surgeon
   D) Internet
   E) Another parent

To complete the examination for CME credit, turn to page 283 for instructions and the response form.