CME

Cleft Lip, Cleft Palate, and Velopharyngeal Insufficiency

David M. Fisher, F.R.C.S.(C.), F.A.C.S.
Brian C. Sommerlad, F.R.C.S., F.R.C.S.Ed.(Hon.)

Toronto, Ontario, Canada; and London, United Kingdom

Learning Objectives: After reading this article, the participant should be able to: 1. Recognize the clinical features associated with unilateral cleft lip, bilateral cleft lip, the cleft lip nasal deformity, cleft palate, and velopharyngeal insufficiency. 2. Describe the most frequently used techniques for repair of cleft lip and palate. 3. Diagnose and treat velopharyngeal insufficiency.

Summary: This article provides an introduction to the anatomical and clinical features of the primary deformities associated with unilateral cleft lip–cleft palate, bilateral cleft lip–cleft palate, and cleft palate. The diagnosis and management of secondary velopharyngeal insufficiency are discussed. The accompanying videos demonstrate the features of the cleft lip nasal deformities and reliable surgical techniques for unilateral cleft lip repair, bilateral cleft lip repair, and radical intravelar veloplasty. (*Plast. Reconstr. Surg.* 128: 342e, 2011.)

There is a wide spectrum of cleft deformities. The care of the patient with a cleft deformity requires a team effort that is multidisciplinary, long term, and tailored to the individual. This article focuses on the features of the deformities and plastic surgical interventions performed during the early years of childhood.

ANATOMY

Unilateral Cleft Lip

Cupid's bow is preserved in the medial lip element. With experience, the midline and the peaks of Cupid's bow can be easily identified. The lip height, as measured from the cleft side peak of Cupid's bow to the base of the columella, is characteristically deficient and the bow is sloped. The cutaneous roll of the entire length of the bow is well preserved. However, there is vermilion height deficiency below the cleft-side half of Cupid's bow.

The proposed peak of Cupid's bow on the lateral lip element has traditionally been less well defined. However, Noordhoff[1] considers the proposed base of the philtral column an anatomically determined landmark that should be preserved. This point, "Noordhoff's point" (Fig. 1), is identified on the vermilion-cutaneous junction where the cutaneous roll and the vermilion-mucosal junction lines start to converge medially. At this point, there is adequate vermilion height (usually matching the height of the vermilion below the non–cleft-side peak of Cupid's bow) and good quality cutaneous roll. Both of these features, vermilion height and quality of the cutaneous roll, diminish if this point is moved medially.

The lateral lip element height as measured from the subalare to Noordhoff's point is often different from the height of the noncleft side (subalare to Cupid's bow peak); it is often excessive in incomplete clefts and usually shorter in complete clefts.[2,3] The transverse length of the lateral lip element (Noordhoff's point to the commissure) is almost always shorter than that of the noncleft side (Cupid's bow to the commissure). Lateral lip element hypoplasia, defined as combined height and transverse length deficiency, is present in 63 percent of patients with unilateral cleft.[3]

From the Cleft Lip and Palate Program, The Hospital for Sick Children, University of Toronto.
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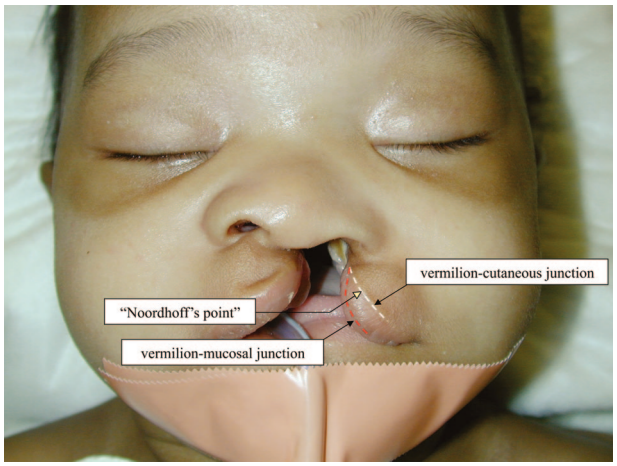


Fig. 1. Noordhoff's point. According to Noordhoff, the proposed peak of Cupid's bow on the lateral lip element is an anatomically determined landmark that should be preserved. This point is chosen on the vermilion-cutaneous junction where the cutaneous roll and the vermilion-mucosal junction lines start to converge medially. At this point, there is adequate vermilion height and good quality cutaneous roll. Both of these features, vermilion height and quality of the cutaneous roll, diminish if this point is moved medially. Choosing a point more lateral will only further shorten the lateral lip in its transverse length.

The orbicularis oris muscle is misdirected by the cleft.[4,5] On the cleft side, the lower bundle of the pars superficialis changes direction as it approaches the cleft to run almost vertically to find insertion to the cleft side nostril and periosteum of the piriform aperture. The upper bundle inserts into the lateral aspect of the ala and the nasolabial fold. Contraction of the lateral lip element orbicularis produces a noticeable bulge in the lateral lip and contributes to the nasal deformation.

Bilateral Cleft Lip

In complete bilateral cleft lip–cleft palate, the prolabium and premaxilla, both derivatives of the frontonasal process, remain entirely separated from the lateral lip and maxillary arch elements derived from the maxillary processes. The prolabium will vary in dimensions between affected individuals, but there are some universal findings. The cutaneous portion is void of the median groove and the philtral ridges which, in the noncleft lip, define the shape and dimensions of the philtrum. The cutaneous roll of the prolabium is of poor quality when compared with that of the lateral lip elements. The height of the vermilion is inadequate. The labial sulcus is shallow if at all present. The prolabium, lacking orbicularis muscle, sits flaccid on the premaxilla, rotated anteriorly into elevation and in extreme cases even adopts a horizontal posture overlying an advanced and anteriorly rotated premaxilla. The lateral lip elements in the bilateral cleft are similar to the lateral lip element of a unilateral cleft.

Cleft Lip Nasal Deformity

Fisher and Mann[6] have described a model for the cleft lip nasal deformity. With progressive degrees of clefting, the model predicts the following nasal deformities observed with clefting of the lip (Fig. 2). (**See Video 1,** which demonstrates the anatomical features of unilateral and bilateral cleft lip nasal deformities, available in the "Related Videos" section of the full-text article on PRS Journal.com or, for Ovid users, at ***http://links.lww.com/PRS/A377***.)

Cleft Palate

In complete unilateral cleft lip–cleft palate, the vomer is attached to the palate on the noncleft side. The cranial portion of the vomer is almost vertical, whereas the caudal portion approaches the horizontal toward its attachment to the palatal shelf. In complete bilateral cleft lip–cleft palate, the vomer is attached anteriorly to the premaxilla and remains free of attachments to the palatal shelves. In cleft palate, the septum is usually midline and is attached to the hard palate as far back as the anterior extent of the hard palate cleft. The submucous cleft palate presents with the following classic triad of bifid uvula, midline notching of the posterior hard palate, and diastasis of the velar musculature. Sommerlad et al. have described a scoring system to further describe the spectrum of submucous cleft palate.[7]

The musculature of the cleft velum is abnormal. The levator veli palatini arises from its normal skull base origin, passes inferomedially above the upper concave margin of the superior constrictor to enter the velum, to be inserted into the cleft margin in the anterior half of the velum. This is in contradistinction to the noncleft palate, where the levator reaches the midline in the middle 40 percent of the velum.[8] The muscle fibers of the palatopharyngeus and palatoglossus are also attached somewhat anteriorly. Their fibers fan out, with their more lateral fibers running anteroposterior and attaching to the posterior hard palate shelf and the more medial fibers running more obliquely and attaching to the cleft margin. The

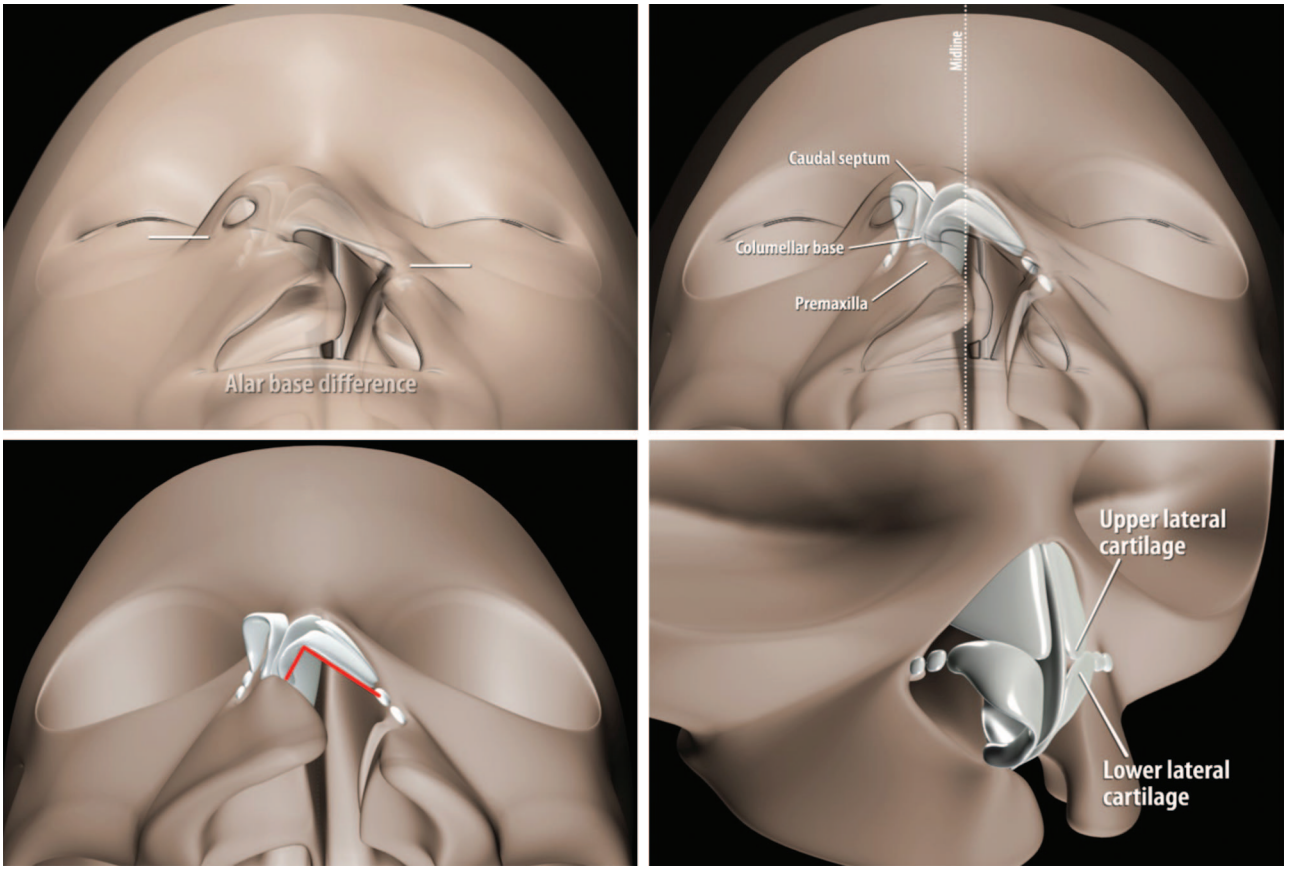
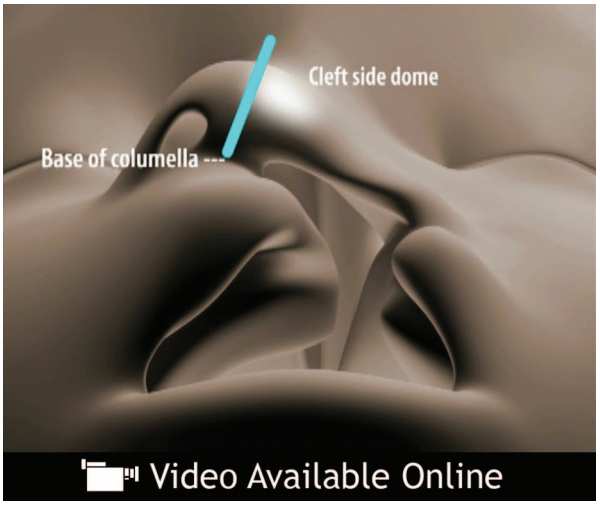


Fig. 2. Cleft lip nasal deformity. Asymmetry of the underlying skeletal base and muscular diastasis produce the typical deformities observed within the spectrum of the cleft lip nasal deformity. For a complete description of the features of the deformities, watch the video from which these still frames have been taken.



Video 1. Video 1, which demonstrates the anatomical features of unilateral and bilateral cleft lip nasal deformities, is available in the "Related Videos" section of the full-text article on PRSJournal.com or, for Ovid users, at ***http://links.lww.com/PRS/A377***.

muscles can be identified in the anterior two-thirds to three-quarters of the cleft velum, in contrast to the normal velum, where these fibers rarely meet the posterior edge of the hard palate. The muscle of the tensor veli palatini does not enter the palate. It arises from the scaphoid fossa of the pterygoid process, the lateral lamina of the cartilage of the auditory tube, and the medial aspect of the spine of the sphenoid. Its tendon passes through the origin of the buccinator and passes around the hamulus to enter the velum. In the noncleft palate, the tensor tendon fans out to form the palatal aponeurosis in the anterior third of the velum. In the cleft velum, the anterior tendinous fibers form the "nasal component" of the tensor, which attaches to the lateral aspect of the posterior hard palate shelf. When the velar musculature and the nasal component of the tensor are identified during cleft palate repair (Fig. 3), the insertions of the nasal component of the tensor tendon and the fibers of the velar musculature are parallel and side-by-side, with the former lying lateral to the latter. Posteriorly, these two structures can be seen

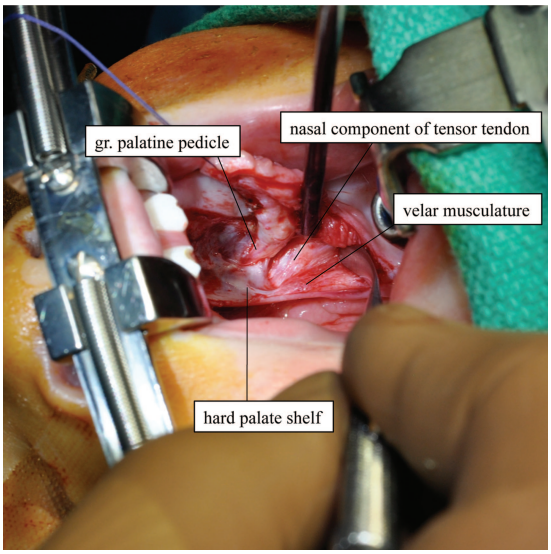
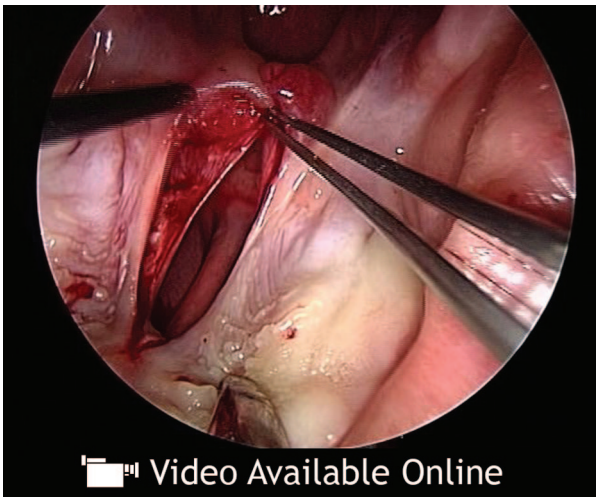


Fig. 3. Nasal component of the tensor tendon and the velar musculature. The white fibers of the nasal component of the tensor tendon pass around the hamulus (deep to the suction cannula tip) and insert into the lateral aspect of the hard palate shelf. The velar musculature (i.e., palatoglossus, palatopharyngeus, and levator) seen deep to the elevator occupies a more medial position in the anterior velum. Anteriorly, the tendinous fibers of the nasal component of the tensor and the muscular fibers of the velar musculature are almost parallel but diverge posteriorly to their respective origins.

to diverge as the tensor tendon passes caudally toward the hamulus and the velar musculature (i.e., palatoglossus, levator, and palatopharyngeus) passes laterally toward its origins. [**See Video 2,** which demonstrates the relationship of the velar musculature and the nasal component of the tensor, velar muscle release, and velar muscle repositioning (D.F.M.), available in the "Related Videos" section of the full-text article on PRSJournal.com or, for Ovid users, at ***http://links.lww.com/PRS/A378***.]

MANAGEMENT OF THE CLEFT ALVEOLUS

The alveolar cleft gap results from both deficiency of bone and displacement of the alveolar segments. Management protocols for the alveolar cleft vary widely between centers, and this remains a subject of controversy. Many surgeons prefer the alveolar cleft to be narrowed before definitive cleft lip repair, and this can be accomplished by lip adhesion or by various methods of presurgical orthodontics. Nasoalveolar molding[9–11] offers the potential for some additional correction of the associated nasal deformity. Presurgical orthodontics requires access to specialized orthodontic care



Video 2. Video 2, which demonstrates the relationship of the velar musculature and the nasal component of the tensor, velar muscle release, and velar muscle repositioning (D.M.F.), is available in the "Related Videos" section of the full-text article on PRSJournal.com or, for Ovid users, at ***http://links.lww.com/PRS/A378***.

and is time consuming, and its long-term benefits have yet to be proven. Lip adhesion, in contrast, requires an additional operation, introduces scar, and delays definitive lip repair. Bony reconstruction of the alveolar cleft can be performed at various time points:

I. Gingivoperiosteoplasty, at the time of cleft lip repair
II. Primary bone grafting, at the time of cleft lip repair
III. Secondary gone grafting
    A. Early, for preservation of the lateral incisor
    B. Late, for preservation of the canine
IV. Segmental Le Fort osteotomy and bone grafting, at skeletal maturity

Primary bone grafting may have a negative effect on maxillary growth,[12,13] and there may remain a need for secondary bone grafting because of insufficient alveolar bulk. Subperiosteal degloving of the maxilla is not a requirement for gingivoperiosteoplasty, provided that presurgical orthodontics has been able to achieve abutment of alveolar segments. The advantage of gingivoperiosteoplasty is the reported elimination of the requirement for secondary bone grafting in 60 percent of cases.[14] The authors' preference for secondary bone grafting over gingivoperiosteoplasty is based on the following: (1) ideal abut-

ment of alveolar segments is not always achievable by presurgical infant orthodontics; (2) both hypoplasia and the separation of parts contribute to the alveolar cleft gap, and in the setting of alveolar hypoplasia, narrowing the cleft to allow for gingivoperiosteoplasty may overly constrict the arch; (3) there is the potential for alveolar surgery at this young age to injure tooth buds and contribute to growth restriction; (4) secondary bone grafting in our experience is a reliable operation and offers an opportunity to augment the alar base and piriform margin deficiencies; and (5) gingivoperiosteoplasty has been shown to result in bone of less quantity and poorer location when compared with secondary bone grafting.[15] The authors' preferred algorithm for management of the alveolar cleft involves presurgical nasoalveolar molding (D.M.F.) or no presurgical orthodontics (B.C.S.), lip repair at 3 to 5 months of age, and secondary alveolar bone grafting before eruption of the adult cleft side canine.

CLEFT LIP REPAIR

Unilateral Cleft Lip Repair

"All cleft lip surgeons have their favorite surgical technique for repairing the unilateral cleft lip. It is usually a hybrid of training experience and imagination."[16] Although this is very true to a great extent, principles exist that should be followed, and several different repair techniques have gained greater popularity over others.

It is worthwhile to review the ideal goals of cleft lip repair. Approximation of medial and lateral lip elements should be achieved without loss of natural landmarks. There should be thoughtful discard of tissue so as to excise the poor quality cleft marginal tissues and achieve a balanced lip. The scar of union should be placed along natural lines, with appreciation of the anatomical subunits of the lip-nose complex. The muscle should be repaired. The nostril margins should be of equal circumference and the alar bases should be symmetric from the anterior view.

The ideal repair should approximate the medial and lateral lip elements appropriately at all levels (i.e., nostril sill, cutaneous roll, vermilion-cutaneous junction, and vermilion-mucosal junction) without interruption or loss of landmarks and achieve balance by providing length where tissue is short and excision where height is excessive. Lalonde[17] has coined the term "nostril sill roll." Although not always present in complete clefts, medial and lateral components of the roll can be identified in most incomplete clefts. Preservation and direct side-to-side approximation of these roll components will reconstruct the roll, producing the most natural-appearing nostril sill. The noncleft side heights (subalare to Cupid's bow peak and subnasale to Cupid's bow peak) provide the measures that must be created by the repair on the cleft side. The cleft side medial height (subnasale to Cupid's bow peak) will need to be lengthened and the lateral lip height (subalare to proposed base of the philtral column) will often need to be altered to match that of the noncleft side. At the level of the cutaneous roll, the entire length of Cupid's bow should be preserved in the medial lip element. In the lateral lip element, Noordhoff's point should be preserved and used to form the base of the philtral column incision. The cutaneous roll of the medial and lateral lip elements should be approximated in side-to-side fashion. Vermilion height deficiency below the cleft-side half of Cupid's bow should be augmented and the red lip elements should be approximated with attention to creating a level vermilion-mucosal junction.

Early lip repairs were limited in their abilities to achieve these goals. Paré, as early as 1564,[18] pared and then approximated the cleft margins with a straight needle wrapped in a thread in a figure-of-eight. This closed the lip but failed to address the height deficiency of the medial lip and left a residual notch. Rose[19] and Thompson[20] each achieved modest length in the repair by approximating curved and angled excisions, respectively. Mirault[21] used a low triangular flap from the lateral lip to provide increased length. Although this repair ignored Cupid's bow, it introduced an important principle that has been adopted by most techniques that followed; any substantial medial lip deficiency needs to be augmented with tissue from the lateral lip. Le Mesurier[22] used a quadrilateral flap from the lateral lip element to create a half-bow from the lateral lip element. The results were admirable; however, the technique discarded the cleft-side half of the true Cupid's bow and created a nonanatomical stairstep scar.

Inferior Triangle Repair of Tennison Modified by Randall

The inferior triangle repair is a Z-plasty where the middle limb of the Z is shared by the incised cleft margins (Fig. 4). Tennison[23] bent a wire and used this as a stencil to produce a zigzag scar, the limbs of which were equal. Randall[24] used calipers and simple mathematics. He created the required length and reduced the size of the inferior triangle. In this repair, the total lip height of the noncleft side is measured from a reference point in the

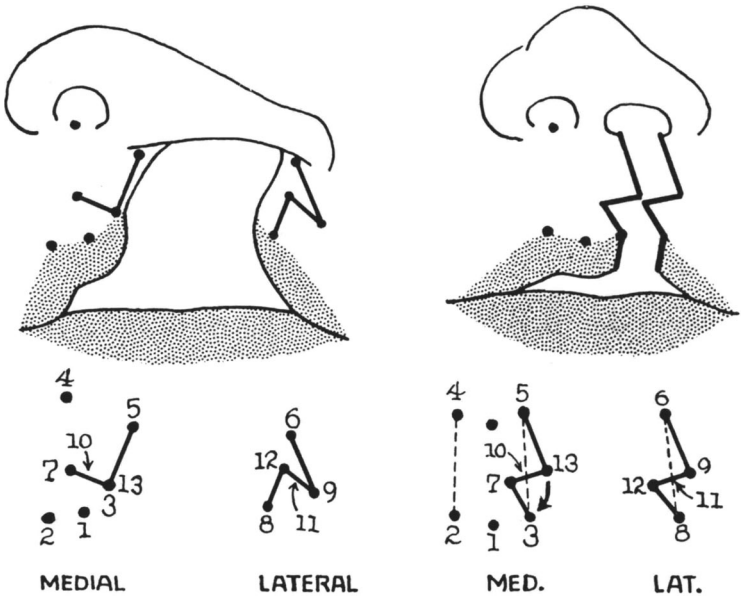


Fig. 4. Inferior triangle repair of Tennison modified by Randall. (Reprinted from Randall P. A triangular flap operation for the primary repair of unilateral clefts of the lip. *Plast Reconstr Surg.* 1959;23:331–347.)

nostril sill to the non–cleft-side peak of Cupid's bow. The greater lip height is measured on the cleft side from the proposed medial point of closure at the height of the lip in the nostril sill (symmetric with the reference point in the non–cleft-side nostril sill) to the cleft side peak of Cupid's bow. The difference between the total lip height and the greater lip height gives the approximate height of the lesser height, the base width of the inferior triangle required to level Cupid's bow. There are several advantages of the inferior triangle repair. Nostril sill closure is accomplished by simple side-to-side approximation of medial and lateral nostril sill elements, and scarring at the base of the nose is kept to a minimum. Lip length can be accomplished even when the medial lip height is very short. When the lateral lip height is short, which is often the case in complete clefts, lateral lip height can be accomplished without compromising on Noordhoff's point. The disadvantage is a nonanatomical zigzag scar. In addition, secondary revisions may be challenging because the Z-plasty is in the middle of the lip.

Rotation-Advancement Repair

In the rotation-advancement repair reported by Millard,[25] a curvilinear (rotation) incision is made in the medial lip element (Figs. 5 and 6). The rotation incision mirrors the non–cleft-side philtral column in its lower half. The incision then skirts along the lip columellar crease and approaches but should not cross the non–cleft-side philtral column. The incision allows for caudal rotation of the cleft side peak of Cupid's bow, and the resultant defect is filled with a large triangular

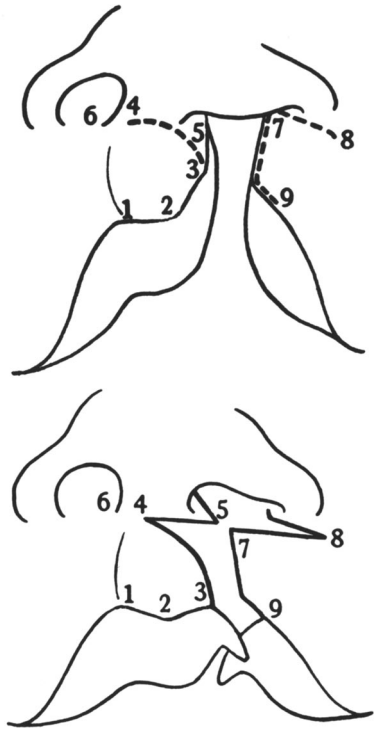


Fig. 5. Rotation advancement repair. (Reproduced from Millard DR. Complete unilateral clefts of the lip. *Plast Reconstr Surg.* 1960; 25:595–605.)

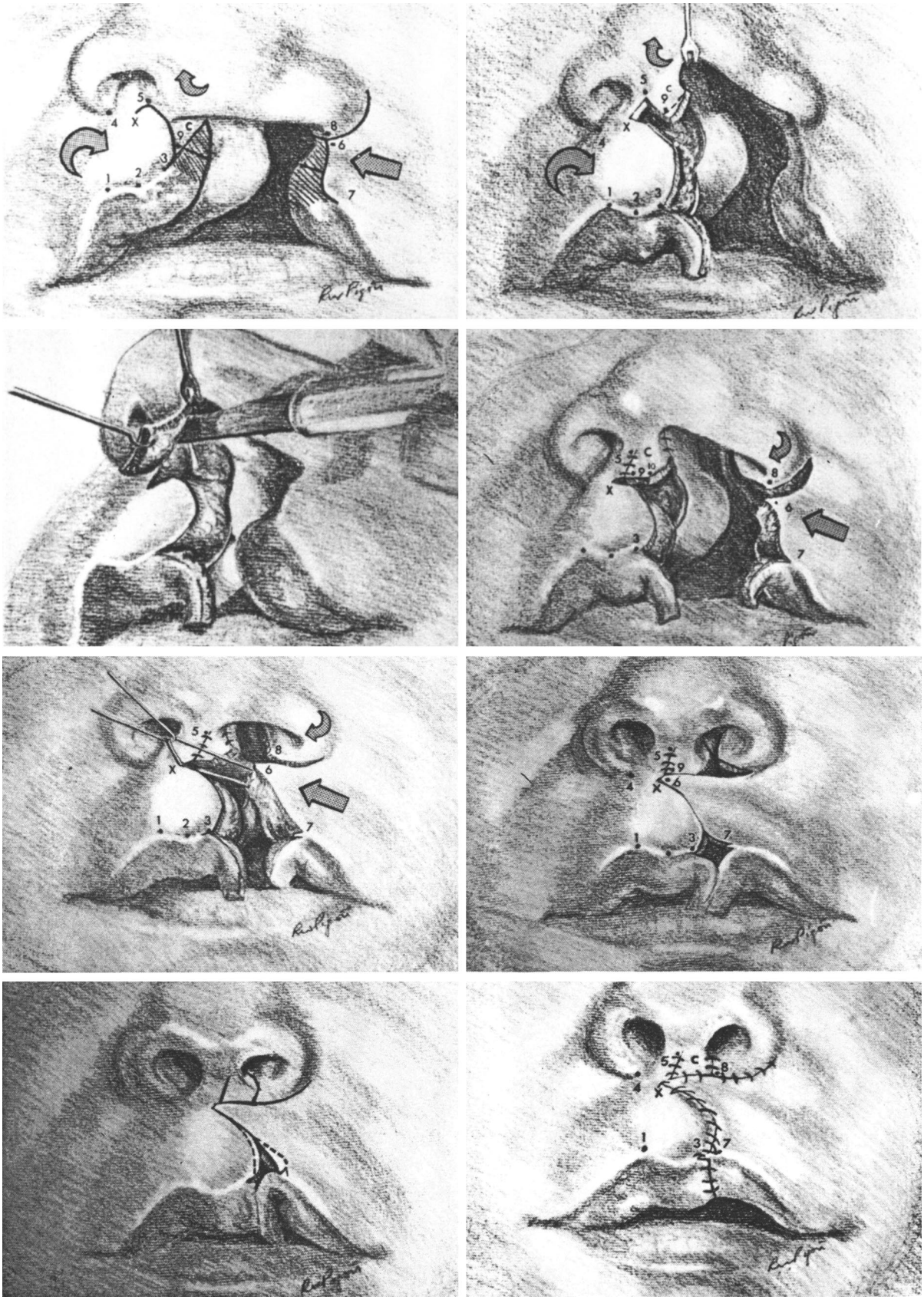


Fig. 6. Rotation advancement repair. (Reproduced from Millard R. Extensions of the rotation-advancement principle for wide unilateral cleft lips. *Plast Reconstr Surg*. 1968;42:535–544.)

(advancement) flap from the lateral lip element. Prolabial skin lateral to the rotation incision forms the "c-flap." It is rotated laterally and contributes to closure of the nostril sill.

Millard observed that the rotation of Cupid's bow was often inadequate and subsequently introduced a "back-cut."[26] This back-cut extended from the most cranial extreme of the rotation incision down the lip just medial to the noncleft side philtral column. The back-cut is extended caudally as much as needed to level Cupid's bow. The back-cut leaves a quadrilateral-shaped defect that is then filled by the c-flap. With this latter maneuver, the c-flap can be advanced to elongate the columella on the cleft side.

The main advantage of this repair is that it preserves the Cupid's bow and philtral dimple. At least in the lower half of the lip, the scar mirrors the non–cleft-side philtral column. In addition, many surgeons like the so-called cut-as-you-go execution of the repair. Despite its advantages, there are several drawbacks. The scar is not the anatomical mirror image of the philtral column in the upper half of the lip. The repair introduces excessive scar at the base of the nose (i.e., columellar base, nostril sill, and alar crease). When the c-flap is used to fill in the back-cut and support the columella, it no longer can be used to the same extent to close the nostril sill. In this setting, nostril stenosis can result. Some would argue that the medial and lateral elements of the nostril sill are present in the unrepaired state and that there is no need to augment nostril sill closure with skin from the lip and that the c-flap only interrupts the nostril sill roll. A long incision around the base of the nose is unnecessary and produces an obvious scar. Many surgeons have now abandoned this element of the repair, deeming the alar crease incision unnecessary. The length of the cleft marginal incision of the lateral lip must be sufficiently long to meet up with the full length of the rotation incision. When the lateral lip element is vertically short, the surgeon will need to extend the incision beyond Noordhoff's point. The transverse length of the lateral lip, usually already short, is thus further compromised to achieve vertical height.

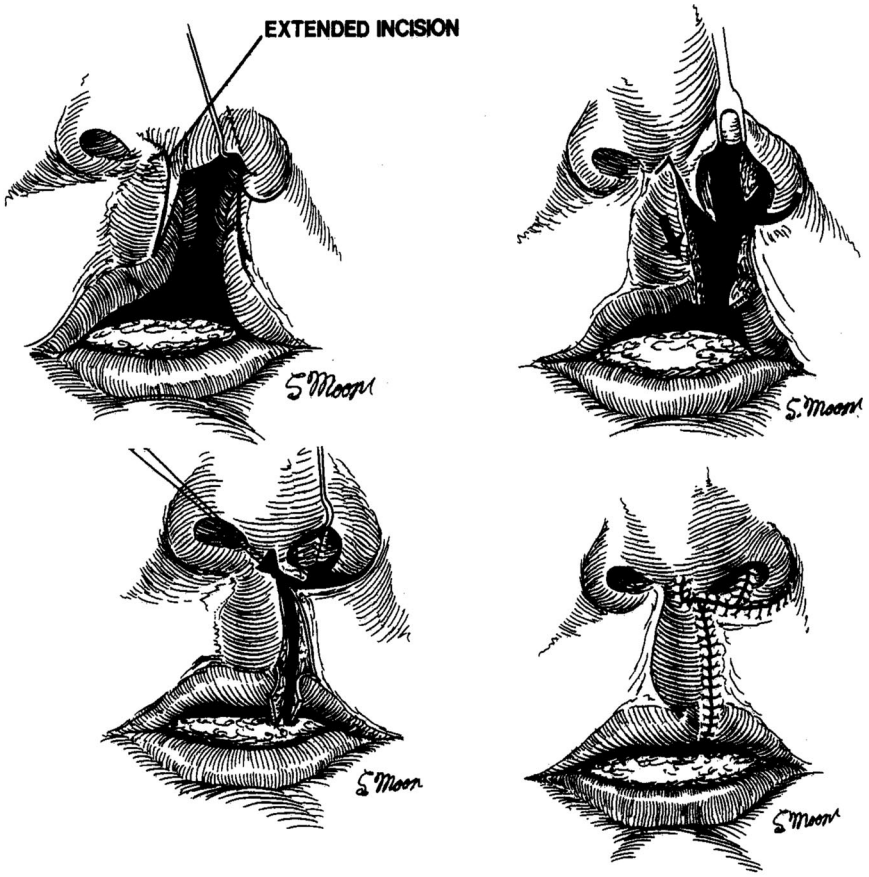


Fig. 7. Modified rotation advancement repair. (Reprinted from Mohler LR. Unilateral cleft lip repair. *Plast Reconstr Surg*. 1987;80:511–517.)

Modified Rotation-Advancement Repair

Mohler[27] altered the markings of the rotation-advancement repair to yield a scar that is more symmetric with the non–cleft-side philtral column (Fig. 7). The change was made by straightening the curve of the rotation incision and extending the incision into the columella. The back-cut then turned 90 degrees and finished at the lip columellar crease. The resultant transverse limb of the repair is elevated to the lip-nose junction, an improvement over the original version. Mohler has stated that the columella seems to be a rather silent donor site; however, Noordhoff[1] warns that when the columella is narrow, the Mohler repair is contraindicated. The advantage of the repair over Millard's rotation-advancement is a more anatomically positioned scar. However, it shares with all rotation-advancement repairs a necessary compromise of transverse lateral lip length when the lateral lip is vertically short.

Modified Rotation-Advancement Repair: Chang Gung Repair

Noordhoff[1] modified Millard's original repair (Fig. 8). In this repair, the alar crease incision is not used. When rotation of Cupid's bow is inadequate, a small opening cut is made above the cutaneous roll above the cleft-side peak of Cupid's bow. Into this defect, a small triangular flap from the lateral lip is introduced. This triangle is inconspicuous and provides tension in the repair in the ideal position, above the roll, which accentuates the pout of the lip. Noordhoff also stressed the importance of achieving balance in the red lip and a level vermilion-mucosal junction ("red line"). Vermilion is almost always deficient in height below the cleft-side half of Cupid's bow. Here, Noordhoff augments the deficiency with a laterally based vermilion flap from the lateral lip.

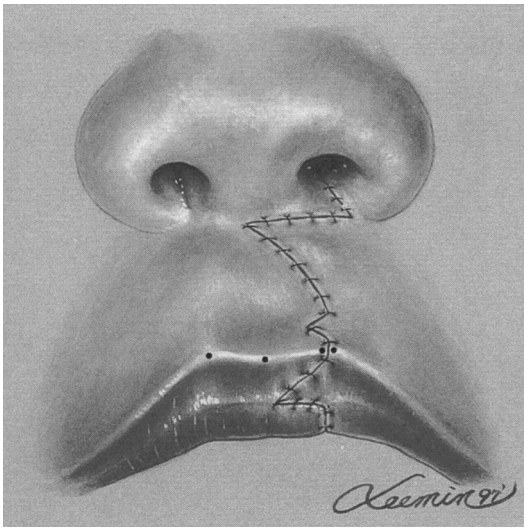


Fig. 8. Modified rotation advancement repair: Chang Gung repair. (Reprinted with permission from Noordhoff MS. *The Surgical Technique for the Unilateral Cleft Lip-Nasal Deformity*. Taipei, Taiwan: Noordhoff Craniofacial Foundation; 1997.)

Anatomical Subunit Approximation Technique

The anatomical subunit repair reported by Fisher[28] aims to produce a cutaneous scar along the "ideal line of repair" (Figs. 9 and 10). [**See Video 3,** which demonstrates the surgical markings used for unilateral cleft lip repair using the anatomical subunit approximation (D.M.F.) technique, available in the "Related Videos" section of the full-text article on PRSJournal.com or, for Ovid users, at ***http://links.lww.com/PRS/A379***.] Above the level of the roll, the repair ascends the lip along a line drawn to mimic the philtral column of the noncleft side. It then curves superolaterally along the lip columellar crease to the point of closure in the nostril sill. Lip length is achieved by two mechanisms. In all cases, a Rose-Thompson lengthening occurs just above the level of the roll as angled lines approximate in the vertical. In the minority of cases, this is sufficient. In most cases, medial lip height is more deficient, and a small triangle is positioned above the roll. Approximately 1 mm of lengthening is accomplished by the Rose-Thompson effect, which must be considered when calculating the required height of the inferior triangle. Noordhoff's vermilion triangle is incorporated in the repair.

The anatomical subunit repair was developed by the first author (D.M.F.), who uses the repair for all unilateral cleft lip repairs. This author's bias toward this repair is based on the following advantages. The scar at the base of the nose is minimized and nostril sill closure is uninterrupted. The scar is positioned long the seams of anatomical subunits with the exception of, when required, a small triangle above the roll. Tension is ideally positioned above the roll. Continuity of the roll is achieved by side-to-side approximation of roll elements. Like the inferior triangle repairs, lateral lip transverse length need not be compromised to achieve vertical height.

Hybrid Repairs

Many surgeons, including the second author (B.C.S.), perform an amalgamation of what they see as the best features of several described techniques. At lip repair, Sommerlad combines the techniques of single-layer closure of the hard palate with vomerine flap,[29] elevation and release of anterior maxillary periosteum and lateral nasal

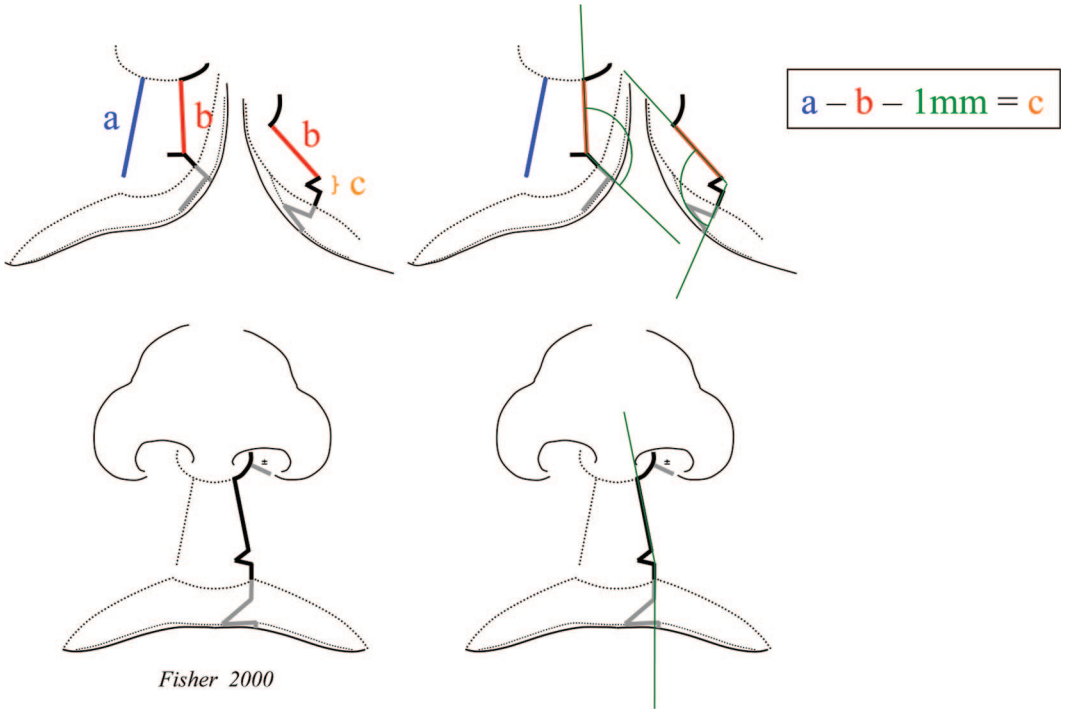


Fig. 9. Anatomical subunit approximation technique. (Reproduced from Fisher DM. Unilateral cleft lip repair: An anatomical subunit approximation technique. *Plast Reconstr Surg*. 2005;116:61–71.)

wall,[30] overlapping of the reorientated orbicularis musculature, a modification of the Millard repair with a small inferior Tennison triangular flap, and closed nasal dissection with the use of long-acting absorbable sutures to attempt to maintain the nasal correction.

Bilateral Cleft Lip Repair

The principles of contemporary bilateral cleft lip repair have been outlined by Mulliken[31,32] and Chen and Noordhoff.[33] The authors' preferred method for bilateral cleft lip repair[34] (Fig. 11) is based on these teachings but with some modification and differences. (**See Video 4,** which demonstrates Fisher's surgical markings for bilateral cleft lip repair, available in the "Related Videos" section of the full-text article on PRSJournal.com or, for Ovid users, at ***http://links.lww.com/PRS/A380***.) Principles common to these techniques include the following: (1) presurgical orthodontics (routinely, D.M.F.; occasionally, B.C.S.), (2) simultaneous bilateral cleft repair, (3) reduction of the prolabium, (4) formation of Cupid's bow and tubercle from lateral lip elements, (5) muscle repair, (6) use of prolabial mucosa to deepen central the gingivolabial sulcus, and (7) primary rhinoplasty.

PALATOPLASTY

The aim of cleft palate repair is to create an anatomically intact and functional palate to improve feeding, achieve normal speech, and minimize maxillary growth restriction. There is considerable support in the literature to advise early palate repair.[35,36] The authors aim to perform palatoplasty at 6 months (B.C.S.) and between 6 and 12 months (D.M.F).

The management of palatal cleft associated with cleft lip and alveolus varies depending on the sequence and type of repairs used to close the lip, alveolus, hard palate, and soft palate. All possible combinations have been described in various protocols. Such protocols include the following:

Closure of the lip followed by closure of the hard and soft palate (used by B.C.S. until 1993 and currently by D.M.F.).
Closure of the lip, alveolus, and hard palate followed by closure of the soft palate (B.C.S.'s present technique).
Delayed hard palate closure; closure of the lip and soft palate followed by closure of the hard palate.[37]
Closure of the soft palate followed by closure of the hard palate followed by closure of the lip.[38]
One-stage closure of the lip, palate, and alveolus.[39]

Fig. 10. Complete left unilateral cleft lip repair by the anatomical subunit approximation technique reported by Fisher. (*Above*) Preoperative appearance at 5 months (following nasoalveolar molding). (*Below*) Postoperative follow-up at 3 years.

Fig. 11. Complete bilateral cleft lip repair [first author's (D.M.F.) preferred technique]. (*Above*) Initial presentation at 1 week. (*Below*) Postoperative follow-up at 4 years.

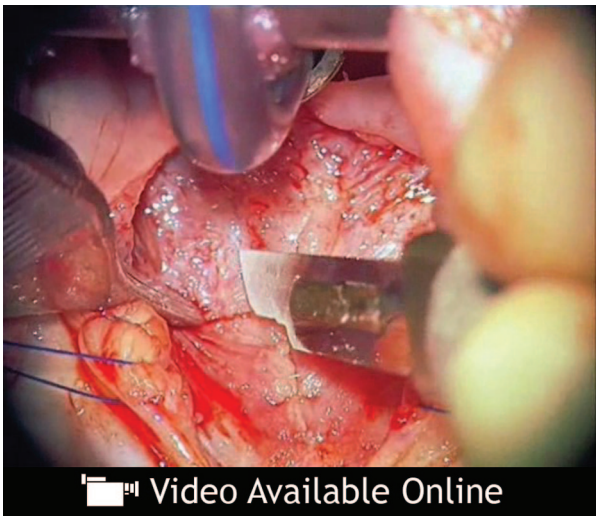
Video 3. Video 3, which demonstrates the surgical markings used for unilateral cleft lip repair using the anatomical subunit approximation (D.M.F.) technique, is available in the "Related Videos" section of the full-text article on PRSJournal.com or, for Ovid users, at ***http://links.lww.com/PRS/A379***.

Video 4. Video 4, which demonstrates Fisher's surgical markings for bilateral cleft lip repair, is available in the "Related Videos" section of the full-text article on PRSJournal.com or, for Ovid users, at ***http://links.lww.com/PRS/A380***.

Hard Palate Repair

Techniques for hard palate closure include von Langenbeck (Fig. 12), Veau-Wardill-Kilner pushback, two-flap (Bardach), hybrid repair (Clarke) (Fig. 13), and no palatal incisions (Sommerlad). [**See Videos 5 through 10,** which demonstrate Sommerlad's method of cleft palate repair, available in the "Related Videos" section of the full-text article on PRSJournal.com or, for Ovid users, available at the following links: part 1, planning the incisions (Video 5), ***http://links.lww.com/PRS/A381***; part 2, raising the oral mucosa (Video 6), ***http://links.lww.com/PRS/A382***; part 3, repairing the nasal mucosa (Video 7), ***http://links.lww.com/PRS/A383***; part 4, dissecting the muscles (Video 8), ***http://links.lww.com/PRS/A384***; part 5, repairing the muscles (Video 9), ***http://links.lww.com/PRS/A385***; and part 6, closing the oral mucosa (Video 10), ***http://links.lww.com/PRS/A386***.]

In addition to cleft marginal palatal incisions, von Langenbeck[40] introduced the concept of lateral relaxing incisions to relieve tension in the midline repair. Incisions are made bilaterally at the junction of the gingival and oral palatal mucoperiosteum down to bone and lateral to the greater palatine canal. Incisions extend into the anterior aspect of the oral-side soft palate mucosa. Bipedicle mucoperiosteal flaps are thus produced, each based both anteriorly and posteriorly and carrying the greater palatine artery as an axial supply. Although relaxing incisions carry a theoretical risk of increased anteroposterior maxillary growth restriction, this has not been proven. The authors of this article have different opinions in this regard. Mahoney and Fisher[41] use relaxing incisions liberally (in all cases except for isolated soft palate clefts) and have a fistula rate of 0.8 percent in a series of 485 consecutive cases of primary palatoplasty. Sommerlad has been able to avoid relaxing incisions in 90 percent of cases. After the introduction of a posteriorly based flap of neomucosa from the epithelialized vomer to augment closure of the nasal layer at palate repair, his fistula rate in complete unilateral clefts has been 3.9 percent. There may be a tradeoff between the possible effect of lateral releasing incisions on maxillary growth and their benefit in reducing fistulae.



Videos 5 through 10. Videos 5 through 10, which demonstrate Sommerlad's method of cleft palate repair, are available in the "Related Videos" section of the full-text article on PRSJournal.com or, for Ovid users, at the following links: part 1, planning the incisions (Video 5), ***http://links.lww.com/PRS/A381***; part 2, raising the oral mucosa (Video 6), ***http://links.lww.com/PRS/A382***; part 3, repairing the nasal mucosa (Video 7), ***http://links.lww.com/PRS/A383***; part 4, dissecting the muscles (Video 8), ***http://links.lww.com/PRS/A384***; part 5, repairing the muscles (Video 9), ***http://links.lww.com/PRS/A385***; and part 6, closing the oral mucosa (Video 10), ***http://links.lww.com/PRS/A386***.

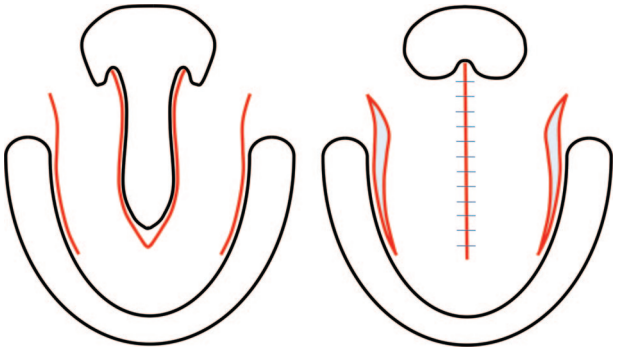


Fig. 12. Von Langenbeck palatoplasty showing elevation of bipedicle oral side flaps.

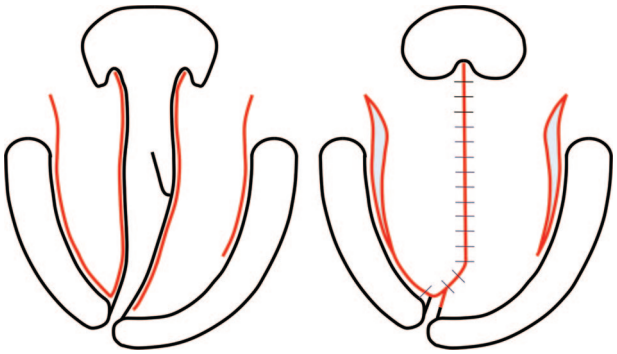


Fig. 13. Hybrid repair showing elevation of a bipedicle flap from the greater segment oral side with elevation of a posteriorly based flap from the lesser segment oral side. (Adapted from Gillett DA, Clarke HM. The hybrid palatoplasty: A preliminary report. *Can J Plast Surg*. 1996;4:157–160.)

The Veau-Wardill-Kilner[42–45] pushback procedures were devised with the belief that V-Y pushback of posteriorly based oral mucoperiosteal flaps of the hard palate would result in effective palatal lengthening and improved speech outcome. There is evidence, however, that this did not improve speech outcome.[46] The pushback leaves extensive raw area over bone anteriorly and has several potential consequences such as greater transverse collapse, increased anteroposterior maxillary growth restriction, and large anterior fistulas.

The two-flap repair[47] is widely used for unilateral complete clefts. In this technique, the bilateral lateral relaxing incisions are extended anteriorly as far as the cleft margins. This produces two flaps based posteriorly on the greater palatine pedicles. Flaps are elevated both to aid in exposure and to provide closure without tension. There is no attempt to push-back the repair, and flaps returned to their original anterior position. A welcome alternative is the hybrid repair described by Gillett and Clarke.[48] In this technique, a posteriorly based flap is raised on the cleft side (lesser segment) and a von Langenbeck bipedicle flap is raised on the noncleft side (greater segment). The advantage of the hybrid repair over the two-flap repair is that there is no incision posterior to the incisors. There is no risk of complete anterior flap dehiscence and theoretically less impairment of maxillary growth and anterior dental occlusion.

In unilateral complete clefts, Sommerlad[49] achieves a one-layer closure of the anterior hard palate with a superiorly based vomerine flap. The subsequent definitive palate repair is then very similar to repair of an isolated secondary palate cleft.

Soft Palate Repair

Muscle repair has become almost universally accepted as a critical component of contemporary palatoplasty. Veau brought our attention to the abnormal insertions of the velar musculature in the cleft state, and Kriens[50] coined the term "intravelar veloplasty," which consists of levator muscle repositioning and levator sling reconstruction. Sommerlad[49,51,52] has introduced the concept of "radical intravelar veloplasty" and the use of an operating microscope.[53] (**See Videos 5 through 10,** ***http://links.lww.com/PRS/A381***, ***http://links.lww.com/PRS/A382***, ***http://links.lww.com/PRS/A383***, ***http://links.lww.com/PRS/A384***, ***http://links.lww.com/PRS/A385***, and ***http://links.lww.com/PRS/A386***, respectively.) The operation can also be performed with high loupe magnification and good lighting. Critical to this radical muscle repair is separation of the velar muscle mass (i.e., levator, palatopharyngeus, and palatoglossus) from the oral and nasal mucosa and from the back of the hard palate. The levator is identified within the velar muscle mass and traced laterally to the levator tunnel where the levator enters the velum by passing above

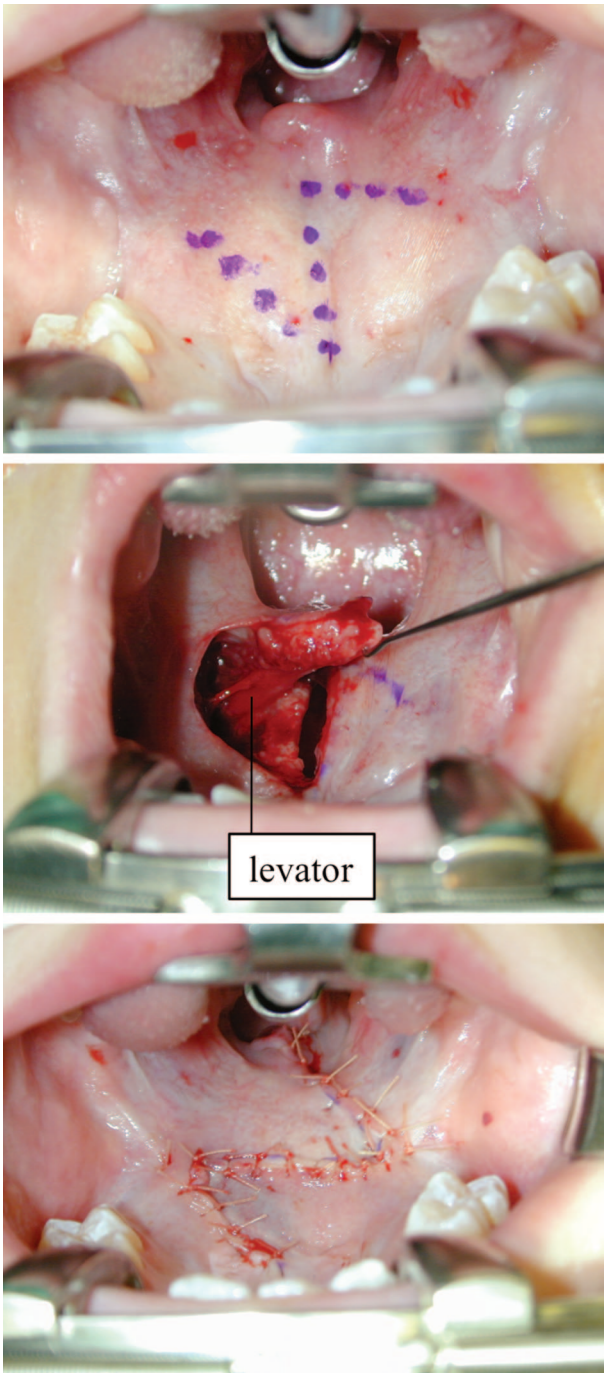


Fig. 14. Double opposing Z-plasty. (Adapted from Furlow LT Jr. Cleft palate repair by double opposing Z-plasty. *Plast Reconstr Surg*. 1986;78:724–738.)

the cranial margin of the superior constrictor. Separation of the velar muscle mass from the nasal component of the tensor then allows for untethered retropositioning of the levator. (**See Video 2, *http://links.lww.com/PRS/A378*.**)

The double opposing Z-plasty (Fig. 14) is popular in many centers. Readers are encouraged to read Furlow's description of the technique.[54,55] The repair has the advantages of lengthening the soft palate and reconstructing the muscle sling. Because the muscle is not exposed on both oral and nasal sides, there may be some preservation of the vascularity to the musculature. In addition, the technique can be used secondarily for treatment of velopharyngeal insufficiency[56] when the muscles have not been adequately repositioned by the primary operation and when the closure defect is small. The main disadvantage of the technique is that length is achieved at the expense of lateral tightening. Although many surgeons consider the technique to be inadequate for wider clefts, the Children's Hospital of Pennsylvania modification[57] in-

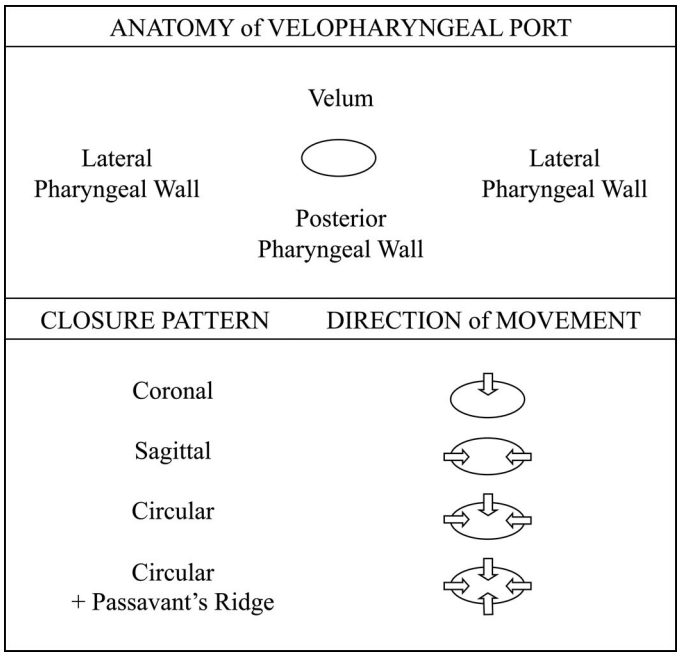


Fig. 15. Patterns of velopharyngeal closure. (Adapted from Skolnick ML, McCall GN, Barnes M. The sphincteric mechanism of velopharyngeal closure. *Cleft Palate J.* 1973;10:286–305.)

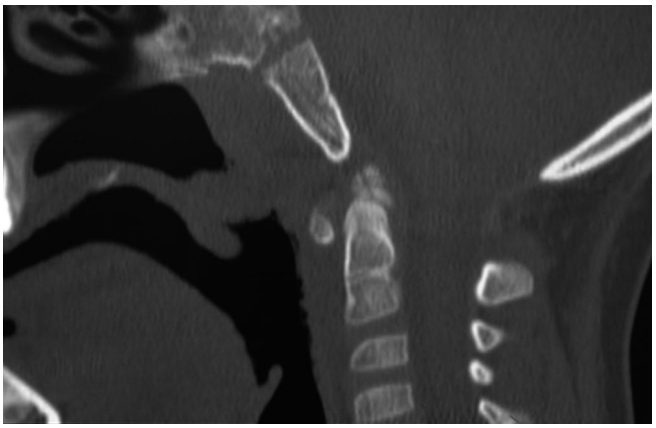


Fig. 16. Postoperative computed tomographic scan (sagittal view) obtained after pharyngeal flap pharyngoplasty. Note that the base of the flap is positioned high on the posterior nasopharyngeal wall at the level of attempted velopharyngeal closure.

corporates lateral relaxing incisions, extending its application for all clefts.

VELOPHARYNGEAL INSUFFICIENCY

The primary goal of cleft palate repair is normal speech. Velopharyngeal competence, the ability to completely close the velopharyngeal sphincter, is required for the normal production of all but the nasal consonants (in English: /m/, /n/, and /ng/). Velopharyngeal insufficiency is defined as the inability to completely close the velopharyngeal sphincter. The primary effects of velopharyngeal insufficiency are nasal air escape and hypernasality. Speech articulation errors (i.e., dis-

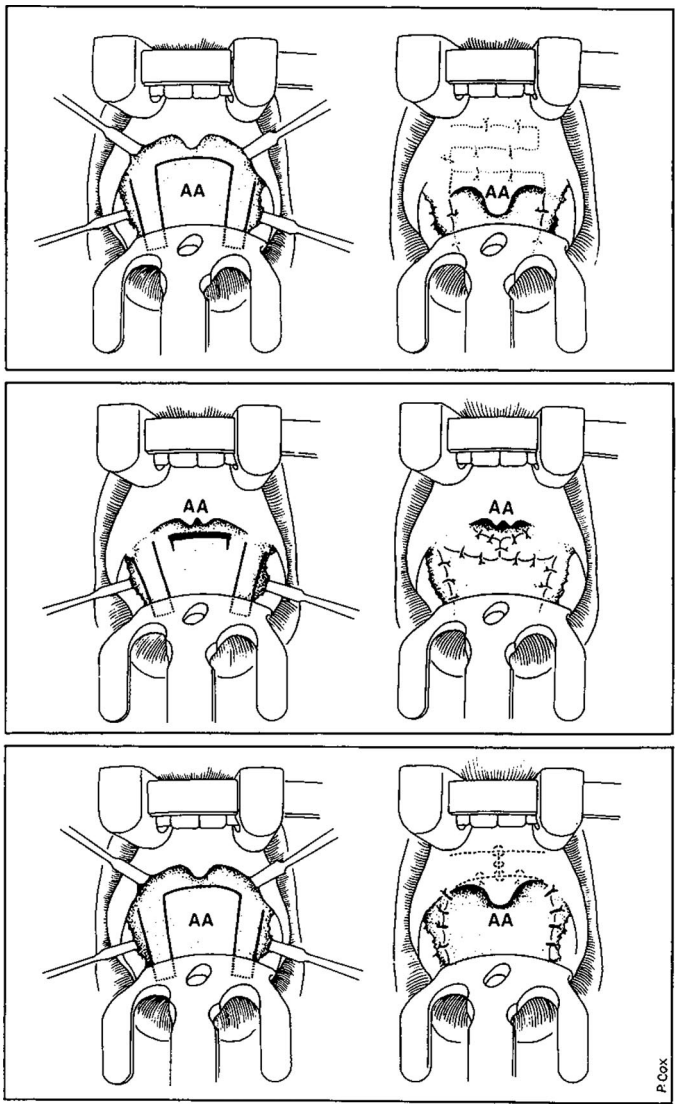


Fig. 17. Artist's rendition of sphincter pharyngoplasties. (*Above*) In the Hynes pharyngoplasty, lateral pharyngeal wall flaps (including the posterior tonsillar pillars with palatopharyngeus in addition to the salpingopharyngeus and the underlying superior constrictor) are sutured high on the posterior pharyngeal wall in a crossover fashion. (*Center*) Orticochea transposed the palatopharyngeus into a low, inferiorly based flap in the oropharynx. (*Below*) The Hynes pharyngoplasty can be modified by suturing the tips of the flaps with their underlying muscle in end-to-end fashion. (Reproduced from Moss ALH, Pigott RW, Albery EH. Hynes pharyngoplasty revisited. *Plast Reconstr Surg*. 1987;79:346–353.)

tortions, substitutions, and omissions) are secondary effects of velopharyngeal insufficiency. The result is decreased intelligibility of speech. The velopharyngeal port is bordered anteriorly by the velum, bilaterally by the lateral pharyngeal walls, and posteriorly by the posterior pharyngeal wall. Each of these structures can contribute to velopharyngeal closure. Skolnick et al. have described four patterns of velopharyngeal closure[58] (Fig. 15).

Velopharyngeal insufficiency can be diagnosed by both subjective and objective means. Perceptual evaluation of speech by an experienced speech language pathologist remains the criterion standard.[59] Multiview videofluoroscopy and nasendoscopy provide visual information (i.e., closure pattern and closure rating) that is valuable for surgical planning. Velopharyngeal closure rating is the fraction of the diameter of the velopharyngeal port that is closed off during attempted sphincter closure, expressed in increments of 0.1 from 0 to 1.0.[60] Nasometry provides an objective measure, *nasalance*, which is a ratio of nasal acoustic energy over nasal plus oral acoustic energies expressed as a percentage. There are published population norms for nasometry scores for picture-cued passages.[61]

Speech therapy to correct articulation errors forms the foundation of management of cleft speech disorders. Surgery for velopharyngeal insufficiency is directed at correcting nasal air escape and hypernasality. Prosthetics, palatal lifts, and pharyngeal obturators offer an alternative to surgery but tend to be poorly tolerated in these young patients. Surgical interventions include posterior pharyngeal wall augmentation, redirection of anteriorly displaced levator muscles, palatal lengthening, sphincter pharyngoplasty, and pharyngeal flap pharyngoplasty (Fig. 16). If an adequate intravelar veloplasty has not been performed and the palatal muscles are observed to

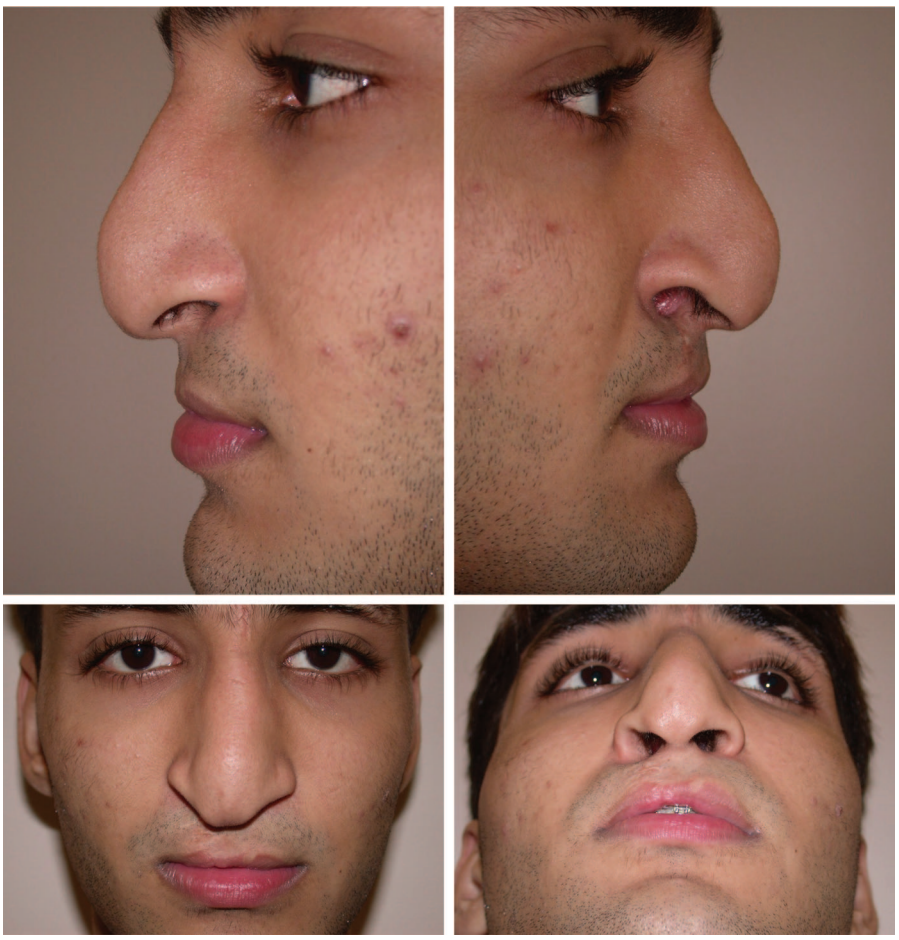


Fig. 18. Preoperatively, this patient displays typical stigmata of a bilateral cleft lip nasal deformity. The tip is broad and underprojected and the columella is short. The nares are oval (longer in the transverse dimension).

remain anteriorly directed, secondary palatoplasty is indicated as a primary intervention.[62,63]

There is evidence to suggest that the preoperative closure pattern can influence the outcome of pharyngeal flap pharyngoplasty.[64] This argues for a selective approach to the surgical correction of velopharyngeal insufficiency. At The Hospital for Sick Children in Toronto, the preoperative closure pattern and closure rating dictate the surgical procedure. For small central defects with a high closure rating, a secondary Furlow palatoplasty is recommended. For coronal closure patterns (poor lateral wall movement and lateral port deficiency), a sphincter pharyngoplasty is performed. Pharyngeal flap pharyngoplasty is indicated for patients with a noncoronal closure pattern and low closure rating. At Great Ormond Street Hospital for Children in London, much emphasis is given to lateral videofluoroscopy. If there is evidence of anteriorly inserted levators, palate re-repair is usually the first surgical option. If not, or if palate re-repair has not adequately corrected the velopharyngeal insufficiency, a posterior pharyngeal wall augmentation (modified Hynes) pharyngoplasty[65,66] (Fig. 17) is usually performed.

RHINOPLASTY

A full discussion of cleft lip rhinoplasty does not fit within the scope of this article. The abnormal anatomy of the nose in the cleft state has been presented. All interventions to correct the associated nasal deformity should be aimed at reversing

Fig. 19. The same patient as shown in Figure 18 is shown postoperatively, after definitive septorhinoplasty [V-Y advancement of lateral crural mucosal flaps (after Potter[67]), columellar strut graft from septum, and tip graft from septum; no reduction of the nasal dorsum was performed]. Note the increase in nasal tip to nasal base length, with the majority of increased tip projection achieved by increase in the projection of the infratip lobule rather than by columellar lengthening.

the deformation of the structures of the nose caused by the asymmetry of the underlying skeleton and muscular action. There are many opportunities to address these nasal deformities: presurgical orthodontia (e.g., arch alignment and nasal molding), primary cleft lip repair (e.g., alar base and columellar base repositioning, caudal septal repositioning, muscle repair), alveolar bone grafting (e.g., alar base and piriform aperture augmentation), and definitive septorhinoplasty (e.g., straightening and or narrowing of the bony pyramid, alignment of the septum, repositioning of the lower lateral cartilages and domes). The primary goal for the surgeon should be to introduce no iatrogenic deformity. Definitive septorhinoplasty will be more satisfying and successful if the structures of the nose have not been scarred or taken away by previous surgery.

For correction of the bilateral cleft lip nasal deformity, older techniques of columellar lengthening that involved recruitment of skin from the prolabium or nostril sills have been abandoned. The authors warn against any visible cutaneous nasal incisions and excision of "excess" nostril rim skin at the time of primary rhinoplasty. There is no excess of skin envelope; rather, there is a deficiency of underlying cartilage support. When the child is older and the cartilages can be repositioned, the surgeon will appreciate a full and unscarred skin envelope (Figs. 18 and 19).

David M. Fisher, M.D.
Cleft Lip and Palate Program
The Hospital for Sick Children
University of Toronto
5424-555 University Avenue
Toronto, Ontario M5G 1X8, Canada
david.fisher@utoronto.ca

PATIENT CONSENT

Parents or guardians or the patient provided written consent for the use of the patients' images.

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