Correction of Secondary Cleft Lip Deformities

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Learning Objectives: After studying this article, the practitioner should be able to (1) describe the common secondary deformities of the cleft lip, (2) determine the appropriate timing for surgical intervention to correct the deformities, and (3) determine the best method of addressing each of the individual secondary deformities of the cleft lip.

Secondary deformities are common in children born with a cleft lip and palate. Patients with cleft lip deformity will undergo multiple surgical procedures early in life, so it is imperative to prioritize treatment of their secondary deformities and minimize the number of interventions needed. Of the many approaches used to correct these problems, surprisingly few work well consistently. As with all plastic surgery, the timing and procedure should be predicated on the severity of the deformity. (Plast. Reconstr. Surg. 109: 1672, 2002.)

Secondary deformities of the cleft lip are the rule rather than the exception. As the patient with a cleft lip deformity will undergo multiple surgical procedures throughout the course of his or her early life, it is imperative to prioritize treatment of the secondary deformities and therefore minimize the number of interventions necessary. A myriad of approaches have been used to correct these problems, but surprisingly few work well consistently. As with all plastic surgery, the timing and surgical treatment should be predicated on the severity of the deformity.

Essentially all patients with primary cleft lip deformities are operated on within the first year of life. After this, there is a long period of dramatic growth. Although this early surgery is mandatory to minimize secondary functional problems, the powerful variable of growth may ultimately distort the immediate surgical result. Consequently, the correction of these secondary deformities is an integral part of the care of these patients. Indeed, these secondary procedures are frequently more complex than the initial surgery. The problems are so widely varied that the choice of an appropriate technique to correct them is challenging. Another extremely important variable is that of appropriate timing of surgery. Determining the age to surgically intervene is an important component of a successful outcome. As will be discussed, this must be determined in large part on the basis of the severity of the deformity inasmuch as it relates to normal growth and development.

EVALUATION

Before embarking on secondary surgery for the cleft deformity, one must accurately diagnose all problems associated with the lip and nose. With respect to the lip, one must carefully examine the lip scar, the status of the orbicularis muscle, the orientation of the vermilion and white roll, the Cupid’s bow, and the mucosa. The nasal deformity must be documented and examined for symmetry of the alar bases and nostril shape, length of the columella, and any deformations or deficiencies of the nasal lining. It is imperative that all of these elements be related to the underlying skeletal deformity on which they are based. Skeletal imbalance from maxillary hypoplasia or malposition contributes significantly to the secondary problems commonly seen after initial lip repair.1

Once the deformity has been accurately diagnosed, one must make every effort to determine the underlying cause. Although hypoplasia and distortion occur as a result of the malformation and set the stage for many of the commonly seen secondary deformities, poor preoperative design of the primary operation
contributes as well. For example, a short lip may be caused by something as simple as a tight contracted scar. On the other hand, under-rotation of the initial lip repair may be the source of the problem, as may be failure to anatomically reunite the underlying orbicularis. The design of the secondary surgery should be focused to correct the specific problem. The solution must take into account the soft tissue, muscle, and the underlying skeleton. Addressing all short lips with the same operation is not sufficient. Numerous classification schemes have been devised to simplify the evaluation of secondary cleft lip and nasal deformities. However, they tend to be very cumbersome and do very little to facilitate the diagnosis and its underlying cause.2,3

Timing

There is no absolute rule for delineating the precise timing of surgery for secondary deformities. Although Millard’s admonition that surgery may result in exaggerated scar formation in patients aged 8 to 18 years must be considered, many patients will absolutely require surgical revision during this time period.4 The severity of the individual patient’s deformity and its effect on the child from a psychosocial or functional standpoint is also a critical factor to be considered. Secondary procedures to improve functional problems, such as speech, breathing, or eating, are frequently indicated and need to be aggressively addressed regardless of age. Perhaps the most common age for revisional soft-tissue surgery in our clinic is in the preschool period from 4 to 5 years of age. Peer interactions begin to develop at this time, and the child is extremely impressionable and vulnerable. Another common period for surgical correction is during early adolescence. The nature of peer interaction is frequently changing at this time, and teenagers often become extremely self-conscious. In addition, teenage patients usually express their opinions more vocally regarding their appearance and should be involved in the decision-making process regarding secondary surgery.

The cessation of facial growth is another critical landmark in the timing of secondary operations. However, it must be remembered that different components of the face cease growing at different ages. Whereas mandibular growth may not be complete until age 16 or older, nasal growth in girls has been conclusively shown to stop at approximately 11 to 12 years of age and in boys at approximately 13 to 14 years of age.5

Cleft Lip

Normal lip anatomy. The anatomy of the upper lip and its relation to the lower lip is remarkably constant between individuals. The central defining structure of the upper lip is the Cupid’s bow. The gentle curve of this structure between the Cupid’s bow peaks, and its relationship to the white roll is important to reconstruct. In the 3-month-old child, the Cupid’s bow is generally less than or equal to 5 to 6 mm. As one ascends the philtrum above this, the philtral columns on either side gradually narrow as the columella is approached, where they are generally 10 to 11 mm apart. These are important measurements in the repair of the cleft lip deformity, because the reconstructed philtrum has a tendency to stretch significantly over time. This is particularly true with the bilateral cleft lip deformity, in which the orbicularis muscle is usually sutured together, or to either side of the prolabium. This causes the new philtrum to stretch. As such, the width of a Cupid’s bow should be designed no greater than 4 to 5 mm at the time of primary surgery to avoid the commonly seen wide philtrum.

Another important component to upper lip anatomy is the length of the upper lip. At rest, the upper lip normally projects 2 to 3 mm anterior to the lower lip. The upper and lower lips rest together in the relaxed state. A competent seal is easily created with light pressure. Failure to achieve adequate upper lip length at the time of primary surgeries results in a fore-shortened tight-lip appearance. In the wide cleft, this is sometimes inevitable, because there is a primary deficiency of tissue. However, inappropriate design of the lip rotation may also limit potential lip length.

Finally, the vermilion border, the distinct convex white roll, and the precise relation between these two structures are absolutely critical in lip repair. The white roll is a structure that cannot be duplicated surgically. In secondary surgery, incisions should be carefully designed, preserving and aligning the white roll precisely, because a malalignment of more than 1 mm is clearly noticeable at a conversational distance. Any intervening scar, whether hypopigmented or hyperpigmented, interrupting the continuous anatomic line is very noticeable. This is also true for the line of demarcation between the wet and dry vermilion. The
The vermilion lip line is normally straight, with an increased fullness in the midline tubercle. Any deviation from this with a notch or convexity is abnormal.

**Prevention of Secondary Cleft Lip Deformities**

The ideal time to address secondary deformities is at the time of the initial operation, when every effort should be made to prevent them. We have recently seen a shift in philosophy from staged, delayed procedures to an early aggressive repair of clefts to minimize deformities early. As such, it is important even before the surgery to use the techniques of surgical orthopedics to align the cleft margins, minimizing the tension on the repair and improving the bony platform for the nasal correction. In the past, many devices have been described that allow either passive or active manipulation of the cleft segment into a more anatomic relationship. The passive plate maintains the transverse width of the maxillary segments using external forces to retract the premaxilla or malpositioned segment using adhesive tape and elastic bands. This technique is most successful when there is adequate space for the premaxilla or maxillary segments. There is also some benefit from the device as a feeding appliance. The Latham device is a custom-made appliance that is pinned to the maxillary segments. A ratcheted screw in the device expands the segments, and elastic traction helps in retracting the protruding premaxilla. Typically, it takes about 6 weeks to align the expanded segments to effect closure of the alveolar clefts. Many orthodontists think that premaxillary orthopedics has no real long-term advantage in the management of the cleft patient and may even have some deleterious effects. Nevertheless, whatever the ultimate outcome of the midfacial position, the significant advantages of presurgical alignment include (1) allowing approximation of alveolar segments, (2) facilitating nasal repair, and (3) decreasing tension on lip repair.

In our experience, the most successful method of presurgical treatment has been the use of the passive appliance as described by Grayson and colleagues. As in standard techniques, this procedure uses an introral prostheses made of acrylic that is shaped to the cleft margins. It is gradually reduced in size to guide the cleft margins to a more anatomic orientation. In addition, a nasal extension on the anterior portion of the device fits the nasal vestibule. This is thought to gradually reshape the collapsed ala by changing the shape of the lower lateral cartilage, which under the effect of maternal estrogen is still relatively malleable. By better aligning the cleft margins, not only is the lip and nasal repair facilitated, but also primary approximation of the alveolus (gingivoperiosteoplasty) may be possible in some cases. Studies evaluating this technique have found bony continuity frequently reestablished across the alveolar segments, eliminating the need for secondary bone graft procedures.

The primary drawback to this technique is its labor-intensive nature. These children must be evaluated every 1 to 2 weeks for reshaping of the appliance. Furthermore, the parents must participate actively in the care by cleaning, replacing, and retaping the appliance in position daily. Many clinics are not equipped to provide this level of care, and there is a learning curve to successfully use this technique.

At the time of lip repair, one of the most critical steps is precisely marking the lip. One must take care to make all markings for the flaps indelible by applying methylene blue ink with either a 25-gauge needle or a Beaver blade to prevent the markings from being washed off during the surgical preparation or distorted secondary to local infiltration. Skin incisions should be beveled slightly to allow for eversion of the margins during closure. In addition, all tissue should be handled using skin hooks to avoid pressure necrosis, which can compromise the final result. Specific attention must be given at the time of closure to the oribcularis muscle. Accurate release and anatomic approximation of both the superficial and deep components is imperative to both the appearance and the function of the lip postoperatively.

This meticulous attention to detail should extend to reconstruction of the floor of the nose. Although this has very little effect on the appearance of the repair, it allows for using tissue for the nasal sill and helps to better close the anterior fistula seen in so many postoperative cleft lip patients. The exposure afforded by the primary lip operation is frequently never achieved again, and a secondary operation for this is consequently much more difficult.

In the bilateral lip, the universal stigma associated with staged repairs was so consistent and unacceptable that a change of thinking advocated by Mulliken, Cutting, McComb, and others led to a new era of prevention of bilateral...
deformities and a dramatic improvement in results (Fig. 1).5–11,22 The new priorities for a successful repair include (1) symmetry, (2) muscular continuity, (3) appropriate philtral size and shape, (4) formation of a median tubercle from lateral labial elements, and (5) primary positioning of alar cartilages. These surgical maxims, coupled with premaxillary orthopedics, show great promise.

Deformities of the Vermilion Border

The vermilion border at the level of the cleft repair may be malaligned or appear peaked because of a short vertical dimension to the lip. Mild deformities of the vermilion are commonly seen early after the rotation advancement repair.23 This is thought to be somewhat attributable to a degree of superficial scar contracture, which usually resolves with maturation of that scar. We think it is beneficial to have the parents participate in active massage of the scar to facilitate resolution of this problem. However, if the problem persists beyond 1 year postoperatively, we consider correction. In cases in which the lip is short by only 1 to 2 mm, we prefer a diamond-shaped excision of the scar and closure, which increases the length of the scar and brings the vermilion border down. It is also possible to design a small Z-plasty along the affected column; however, we do not use this technique because it places additional scars on the lip that do not fall along anatomic landmarks.

In more severe cases (3-mm or greater discrepancy), the problem is most likely because of inadequate rotation of the lip. In these cases, the repair should be taken down and repeat rotation and advancement performed using available adjacent tissue to achieve a greater length. This represents a problem with the design of the initial operation. Any technique short of this will not achieve adequate lengthening.

Malalignment of the vermilion or the white roll by as little as 1 mm is noticeable (Fig. 2). As has been pointed out previously, indelibly tattooing the border at the time of the initial lip repair is mandatory. If the problem requires correction, a simple Z-plasty designed along the border is usually successful in correcting the malalignment (Figs. 3 and 4). This procedure also follows the principle that only vermilion should be used to reconstruct vermilion, and it allows for recreation of the vermilion white-roll transition.

Deficient Vermilion

The treatment of vermilion deficiencies is quite different in unilateral and bilateral deformities. The problem may be somewhat prevented at the time of the initial lip operation by back-cutting the mucosa in the gingivobuccal sulcus along the medial lip element, where the deficiency is most common, and advancing it inferiorly. The resulting defect in the sulcus may be filled using local tissue flaps (half Z-plasties). Placing skin hooks on the inferior border of the vermilion and then pulling down helps in estimating the amount of vermilion advancement that is necessary.

Once the problem becomes established, it
most frequently presents as a “whistle” deformity or notch (Fig. 5). When the problem is very mild, particularly when the lateral lip element appears to have a relative excess of vermilion bulk, we perform a horizontal linear excision on the inner aspect of the adjacent lip to diminish the discrepancy. To augment the deficiency, however, local tissue rearrangement is usually sufficient. This frequently takes the form of a V-Y advancement. Great care must be taken to design the flap wide enough to adequately fill the defect. As with any similar situation, the defect should be created before designing any flap.

Vermilion augmentation with a graft is also an option. In the past, we have used dermal fat grafts with some success. These grafts have been harvested from the groin and placed into pockets created within the area of a deficiency. In approximately 50 percent of our patients, the appearance has been improved, but the firmness and lumpiness of the lip that developed was unacceptable to the patient. As has been demonstrated by Coleman, it may be most beneficial to minimize the amount of fat that is placed along any one area of the recipient bed to maximize vascularity and graft take.\(^{24}\) As such, fat harvested from the periumbilical area may be centrifuged and injected with a needle with multiple passes along the area of deficiency. We have used this technique for mild-to-moderate problems in 25 patients with mixed success (Fig. 6). Clearly, this type of procedure is best for relatively mild deformities. Deep, tight scarring prevents adequate expansion of the lip where it is most needed and provides a poor bed to revascularize the graft. Severe notching of the unilateral lip accompanied by scarring is often not amenable to local tissue rearrangement. Although this is relatively rare, such severe deficiencies need fresh tissue. Even in the unilateral deformity, we think the Abbé flap is the best choice to correct the problem.

Vermilion deficiencies found in bilateral cleft lip are much more common than unilateral deformities and are frequently more severe. This should be expected, because the vermilion from the lateral lip element is routinely used to replace the hypoplastic vermilion of the prolabial segment. Horizontal stretching of the prolabial segment with time and growth may also contribute to the problem.

Mild irregularities may be treated with techniques described above, such as V-Y advancements, double or single Z-plasty, or mucosal grafts. The technique of deepithelialized, medially based submucosal flaps tunneled across the midline to augment the defects is also particularly effective, and we have used this with success.\(^{7}\) A common problem in even a well-planned bilateral lip is a relative excess of mucosa in the adjacent lateral lip elements, which should be treated with conservative excision at the time of any midline augmentation. Definitive lip touch-ups are often best delayed until the permanent central incisors have erupted and after the premaxilla is in the correct anatomic position to more accurately judge appropriate lip position.\(^{7}\)

Severe bilateral cleft lip vermilion deficiencies require Abbé cross-lip flap transfer. In addition to augmenting the upper lip vermilion, this helps restore a balance by removing the relative excess vermilion from the lower lip. The technique of Abbé flap transfer is quite straightforward; however, several points merit discussion. The flap should be designed smaller than the actual defect to facilitate closure of the lower lip and to allow for stretching.

Fig. 3. Diagram illustrating Z-plasty rearrangement of white-roll vermilion border to correct malalignment and scar contracture (limited to white roll).
of the flap once in position. Great care must be taken to tattoo the vermilion borders and white roll of both the upper and lower lips before beginning this procedure, much as in a primary lip repair. The location of the labial artery on the pedicle side can be gauged using the cut side as a guide. Tissue from the discarded philtrum, if of good color and consistency, may be used to augment the floor of the nose and the columella only if absolutely necessary. Postoperatively, a nasal trumpet can be used to improve the airway. The flap may be further protected from disruption by using Ivy loops for fixation, tying the loop loose enough so that patients can insert food, spoons, and straws in their mouth but not wide enough to overstretch the flap. This is particularly useful in the immediate postoperative period during emergence from anesthesia, when the patient is not capable of protecting the flap (Fig. 7).

Cupid’s Bow Deformity

The Cupid’s bow itself is a difficult structure to recreate surgically. The problem usually arises at the level of the Cupid’s bow peak because of either malalignment of the vermilion and white roll or a short lip scar. In these situations, the problem should be addressed by either a Z-plasty or a repeat lip rotation as previously described. Although direct recreation of the bow by skin excision and vermillion advancement has been described, this is generally an unacceptable procedure because of the artificial appearance that is created and the scar that the procedure induces. In our opinion, it is much better to simply focus on resetting the proper height for symmetry of the Cupid’s bow and tolerate the scar. In severe deformities, use of the Abbé flap is the best option.

Mucosal Deficiencies

Sulcus deformities most frequently present for treatment around the time of initiation of orthodontic care in anticipation of a bone graft. They are more commonly seen in the bilateral deformity. The prolabial vermilion used to recreate the sulcus is most often inadequate. Although local flaps as described above may be useful, most frequently opening the area of scarring and grafting the resulting defect is the most direct solution. Although skin may be used, we think buccal mucosa is a better option, given the match to the surrounding tissue and the innocuous nature of the donor site. It is always necessary to stent the graft site to promote take, and we have found dental amalgam useful for this purpose. Although there is some secondary graft contracture that can be expected, prolonged stenting as has been advocated by some authors is impractical in most patients.
Short Lip

A lip is considered short when the philtral column on the cleft side is at least 3 mm shorter than the contralateral noncleft philtrum. In very minor cases, the cause may be active underlying scar contracture that improves over time (6 to 8 months). More severe cases are usually caused by inadequate rotation of the lip at the time of the primary operation and are most often seen with an improperly designed rotation advancement repair. As previously described, a diamond-shaped excision of the scar and closure may augment the lip as much as 2 mm. A Z-plasty transfer along the line of the previous scar, although introducing new scars on the lip, may be beneficial for as much as 3 mm of advancement. In the most severe cases, the entire lip repair must be taken down as before and repeat rotation advancement performed.

When evaluating these patients, consideration must be given to the status of the orbicularis muscle, which may not have been repaired anatomically at the time of the primary lip surgery. This may contribute to shortening of the lip. Preoperatively, this may be diagnosed by asking the patient to purse the lips. With this maneuver, the orbicularis will noticeably bulge on either side when it has not been united. When this is the case, the lip must be taken down and the muscle fully released from its abnormal vertical insertions and reunited with sutures. Dissection of the orbicularis on the medial lip should proceed no further than half the width of the philtrum to preserve the integrity and depth of the philtral dimple.

Long Lip

Very rarely does the rotation advancement lip repair produce a long lip. Long lip was

Fig. 5. Vermilion deficiency or notching in cleft lip deformity treated with local rearrangement of tissue. (Above) Unilateral; (below) bilateral.
more frequently seen with the Tennison repair, which is still a useful technique but not as popular as the rotation advancement repair. In treating the long lip, there is a temptation to simply superficially excise tissue from the alar base and elevate the lip on the cleft side. However, this frequently is insufficient and any reduction in length achieved short lasting. We think it is necessary to excise tissue in all dimensions to correct this deformity. The entire repair should be taken down and the lip reduced both vertically and transversely. We have found permanent suspension sutures to the periosteum helpful in maintaining the elevation.

**Tight Lip**

The tight lip may be seen in unilateral or bilateral cases, although more frequently in the bilateral deformity. In unilateral cases, the cause is usually a wide cleft not treated with presurgical orthopedics. When a significant deficiency in tissue is present in the lip, the only acceptable option is to bring in additional tissue in the form of a cross-lip transfer. Again, although used most frequently for the bilateral deformity, severe deficiencies in the unilateral patient may also be treated with the Abbé flap. However, this problem is being seen with decreasing frequency because of presurgical treatment of wide clefts, which helps correct the underlying skeletal imbalance before surgery is performed.

**Wide Lip**

The wide lip deformity is almost exclusively seen in the bilateral cleft lip patient and is always caused by designing the new philtrum too widely at the time of the initial lip repair. At the age most lips are first repaired (3 to 6 months), no more than 4 to 5 mm of prolabium should be used to create the philtrum. Because the orbicularis is most frequently approximated to the lateral border of the prolabium, there is a great tendency for this to stretch over time (Fig. 8).
To correct this problem, the excess philtrum should be excised along the previous lip scar. Just as in the primary operation, the philtrum should be designed slightly narrower than the ultimate desired width in anticipation of stretching. In the past, consideration was always given to using scar tissue or nostril sill with reasonable color and consistency rather than discarding it (forked-flap technique of Millard). This tissue often became part of the new columella and nostril sill. We strongly believe that labial skin (often hair-bearing and thick) should not be used to reconstruct the columella. The forked flap technique causes its own deformities, introducing severe scars across the central columnellar-labial junction, and the circumferential philtral scar produces a contracted bulge rather than a dimple. 

Conclusions

Secondary deformities are common in children born with a cleft lip and palate. They are both intrinsic to the malformation and iatrogenic. The planning and procedures for a newborn with cleft lip and palate should be done only by an experienced team of professionals using the latest techniques to minimize the problems and number of operations and to enhance normal growth and development. When a secondary deformity is observed, treatment should be based on the degree of the deformity and the severity of the impact on the normal functions and growth of a child.

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References

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Fig. 8. The philtrum should be created from a prolabial segment no larger than 4 to 6 mm at the time of primary lip repair. (Above) Philtral appearance in the early postoperative period. (Below) Postoperative result several years later demonstrates significant stretching of the reconstructed philtrum.


Self-Assessment Examination follows on the next page.
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1. ALL OF THE FOLLOWING ARE TRUE ABOUT PALATAL FISTULAS EXCEPT:
   A) Nasoalveolar fistula most frequently represents a residual cleft not addressed at the time of initial repair.
   B) Posterior fistula is secondary to a breakdown in closure.
   C) There is no such thing as a fistula that is simple to close.
   D) All fistulas should be immediately repaired regardless of symptoms.
   E) Bone grafting should only be considered if a good lining closure is present.

2. WHICH OF THE FOLLOWING SEEM TO CONTRIBUTE TO A POSITIVE OUTCOME IN CLEFT SURGERY?
   A) Premaxillary orthopedics
   B) Nasoalveolar molding
   C) Careful dissection and handling of tissues
   D) Lip scar massage
   E) All of the above

3. ALL OF THE FOLLOWING ARE TRUE ABOUT AN ABBÉ FLAP EXCEPT:
   A) The lower lip has a philtrum, white roll, and Cupid’s bow.
   B) Local tissue rearrangement works in most cases of vermilion deficiencies, with the Abbé flap reserved for severe defects.
   C) An Abbé flap also improves lip balance by removing relative excess vermilion from the lower lip.
   D) An Abbé flap should be larger by 25 percent than the proposed new philtrum to compensate for contracture.
   E) The location of the labial artery can be anatomically gauged by using the cut side as a guide.

4. WHICH OF THE FOLLOWING SECONDARY DEFORMITIES IS MOST LIKELY TO BE CAUSED BY FAILURE TO UNITE THE ORBICULARIS ORIS MUSCLE?
   A) A short lip
   B) Vermilion border malalignment
   C) Vermilion notching
   D) A long lip
   E) Mucosal deficiency

5. A SIGNIFICANTLY SHORT LIP FOLLOWING PRIMARY CLEFT REPAIR USUALLY REQUIRES WHICH OF THE FOLLOWING PROCEDURES?
   A) Diamond-shaped excision and closure
   B) Abbé flap transfer
   C) Z-plasty rearrangement
   D) Takedown of the repair and repeat rotation

6. THE LONG LIP DEFORMITY CAN MOST FREQUENTLY BE CORRECTED BY EXCISING SKIN FROM THE ALAR BASE AND SUTURE SUSPENDING THE LIP CEPHALAD.
   A) True
   B) False