

# Update in Unilateral Cleft Lip Surgery

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**Learning Objectives:** After studying this article, the participant should be able to: 1. Describe the unilateral cleft lip and nasal deformity and associated anatomical variations. 2. Understand the history and evolution of the unilateral cleft lip repair. 3. List different presurgical treatment options. 4. Differentiate between surgical techniques.

**Summary:** This article describes characteristics of the unilateral cleft lip and nasal deformity and its management, including presurgical orthopedics, operative techniques, and postsurgical care. The rotation-advancement and straight-line repairs are discussed in detail, as are the current concepts in primary cleft nose repair. (*Plast. Reconstr. Surg.* 148: 262e, 2021.)

*Semper investigans, nunquam perficiens.* (Always searching, never quite achieving perfection.)

—D. Ralph Millard

Isolated unilateral cleft lip with or without cleft palate is one of the most common birth defects, with a reported incidence of 0.1 to 2.1 per 1000 births,<sup>1,2</sup> varying across ethnicities. The rate of cleft lip and palate ranges from one in 2000 among African Americans, to one in 1000 among Caucasians, to one in 450 among Asians and Native Americans. Left-side clefts are twice as common as those on the right, and unilateral clefts are nine times more common than bilateral. Approximately 30 percent of cleft lip cases are associated with other birth defects.<sup>3-6</sup> Of the 70 percent of nonsyndromic cases, 20 percent are familial and 80 percent are sporadic.<sup>7</sup>

There have been many risk factors linked to development of cleft lip, ranging from genetic to environmental. The strongest is family history. For parents with one child with cleft lip, there is a 4 percent risk for the next child; this risk increases to 9 percent if there are two children affected. A parent with cleft lip has a 4 percent risk of giving birth to an affected child, and this risk increases to 15 percent if the first child has a cleft. Many environmental factors have been linked to cleft lip and palate development, including maternal age, prenatal medications (e.g., steroids, antiepileptic drugs), maternal smoking or alcohol consumption during pregnancy,<sup>8</sup> and prenatal maternal malnutrition.<sup>9,10</sup>

None of these factors demonstrated definitive causal relationships.

Similarly, there have been many genetic studies that identified abnormalities on a chromosomal and genomic level. Mutation in interferon regulatory factor 6, or Van der Woude syndrome with cleft lip and congenital lip pits, is the most common syndrome, occurring in 7.6 percent of cleft lip patients.<sup>11,12</sup> Interferon regulatory factor 6 is thought to be responsible for keratinocyte proliferation and differentiation.<sup>13</sup> *MSX1* on chromosome 4 (one of the muscle segment homeobox genes) has been associated with nonsyndromic cleft lip with or without cleft palate,<sup>14</sup> particularly following exposure to environmental factors such as prenatal maternal alcohol consumption and cigarette smoking.<sup>15</sup> Methylene tetrahydrofolate reductase on chromosome 1 is a major enzyme of folic acid metabolism. Mutations in this gene are found at a higher frequency in the setting of maternal hyperhomocysteinemia, leading to syndromic cleft lip and palate, likely caused by folate insufficiency.<sup>16</sup> There are many other loci identified that have been linked to orofacial clefts by means of many different pathways. Expectedly, development of cleft lip and palate appears to be a complex interplay between genetic and environmental factors, many of which have yet to be fully identified.

**Disclosure:** Dr. Hollier serves as chair of the Global Medical Advisory Board for Smile Train. Drs. Xue and Buchanan have no financial interests to report.

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**CLEFT CARE: PRESURGICAL**

The care of patients with unilateral cleft lip with or without cleft palate is performed by a multidisciplinary team (Table 1). One-third of cleft patients have other congenital defects. In addition, a cleft diagnosis is often distressing for families, which benefit from education and psychological support.<sup>17</sup> Multidisciplinary care begins with pre-natal consultation and education. It is important for the cleft surgeon to realize his or her role as a member of this team. Most cleft lips are repaired at 3 to 6 months of age. It is before this time that the patient and family need the most support from other specialties.

One of the key members of the team is the cleft/craniofacial orthodontist. In addition to the more traditional role of restoring dentofacial balance, the cleft orthodontist manages presurgical infant orthopedics. For patients with very wide clefts, and with significant discrepancy between the alveolar segments, presurgical manipulation can optimize surgical outcome.<sup>18</sup> This can be accomplished passively by means of lip taping and lip adhesions, or actively by means of Latham devices and nasolabial molding. Ultimately, the goal for all forms of presurgical infant orthopedics is the same: to narrow the wide lip and alveolar cleft segments, and improve nasolabial symmetry.

Lip taping is the simplest and least labor-intensive method of presurgical orthopedics, and is best when begun shortly after birth. Typically, a Steri-Strip (3M, St. Paul, Minn.) is placed across the lip under tension, stretching between the two cheeks, each with a hydrocolloid bandage to maintain adhesion while protecting the underlying skin. For wider clefts, two strips can be used connected with an orthodontic elastic to increase and maintain tension. The tape is maintained up to 1 week, and can be changed as needed. The goal is to bring the alveolar segments into closer approximation. At our own institution, this method is often used for children who are not amenable to nasolabial molding because

of medical or social reasons. External taping can also be combined with a palatal plate or nasolabial molding.<sup>1</sup>

The Latham device uses a pinned intraoral appliance with a mechanical screw that is turned to narrow the width of the palatal cleft, thereby bringing together the alveolar segments.<sup>19,20</sup> Nasolabial molding combines an intraoral acrylic appliance<sup>21</sup> with a nasal stent. The device is then progressively reshaped to narrow the cleft and reshape the nose.<sup>22,23</sup> Therapy using the Latham device is typically shorter in duration (4 to 6 weeks), whereas nasolabial molding is usually used for 3 to 4 months until the time of repair. Both require meticulous planning and weekly to biweekly adjustments. Nasolabial molding does have the added benefit of shaping the collapsed cleft-side ala, improving nasal form and symmetry<sup>18,24</sup> and increasing alar dome height<sup>25,26</sup> (Figs. 1 and 2). A 2008 survey<sup>27</sup> of 622 cleft surgeons showed that 71 percent used presurgical orthopedics. Of these, 38 percent of patients were treated with nasolabial molding and 26 percent of patients were treated with the Latham device. Of these surgeons, 61 percent reported having never performed lip adhesions. A 2019 American Cleft Palate–Craniofacial Association survey demonstrated that half of the responding cleft teams offer presurgical infant orthopedics, with nasolabial molding being the most common (88.2 percent) and Latham appliance making up 14.7 percent.<sup>28</sup> Recent multicenter study demonstrated that both devices are effective at improving nasolabial aesthetics, but Latham devices have been associated with maxillary growth restriction when combined with gingivoperiosteoplasty.<sup>18,29</sup> The exact mechanism for this growth restriction remains unclear.

Because of the labor-intensive quality of these presurgical measures, some patients may not be candidates. Patients with wide clefts who for whatever reason are not candidates for presurgical orthopedics or lip taping may require lip adhesion. Tissue conservation is an essential component of this procedure, with all markings kept within the tissue that will eventually be discarded in the definitive repair. Undermining around the cleft side pyriform aperture and gingivobuccal sulcus may be necessary to gain enough laxity to allow for a strong orbicularis oris muscle closure and a tension-free skin reapproximation. Skin-only adhesions will not provide the appropriate strength to narrow the cleft, and may lead to significant landmark distortions and scarring.

**Table 1. Multidisciplinary Cleft Team for Pediatric Plastic and Reconstructive Surgery**

Craniofacial orthodontics
Otolaryngology
Pediatrics
Speech pathology
Pediatric dentistry
Audiology
Pediatric neuropsychology
Genetics
Advanced practice providers
Nurse coordinators

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**Fig. 1.** (Above) Female patient with wide right unilateral cleft lip and palate. (Above, left) Before treatment. (Second from above, left) Pretreatment image showing wide alveolar and palatal cleft and displaced right nasal ala. (Above, right) Two months after treatment with nasoalveolar molding. (Second from above, right) Two months after treatment with nasoalveolar molding showing approximated dentoalveolar arch and improved nasal alar contour. (Below) Female patient with wide right unilateral cleft lip and palate. (Second from below, left) Before treatment. (Second from below, right) Two months after nasoalveolar molding treatment. (Below, left) One month postoperatively. (Below, right) Six months postoperatively.

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**Fig. 2.** Female patient with wide right unilateral cleft lip and palate, demonstrating basal views. (Above) Two months after nasoalveolar molding treatment. (Center) One month postoperatively. (Below) Six months postoperatively.

### UNILATERAL CLEFT LIP DEFORMITY AND REPAIR

The unilateral cleft lip deformity is characterized by a deficiency of lip height, and an off-centered and rotated Cupid’s bow/philtral complex (Fig. 3). The deformity demonstrates variable expression, ranging from microform, to minor, to incomplete, to complete forms. In the complete unilateral cleft lip, the orbicularis oris muscle on the medial lip element attaches abnormally to the anterior nasal spine, the medial crura of the nasal lower lateral cartilage, and the anterior nasal

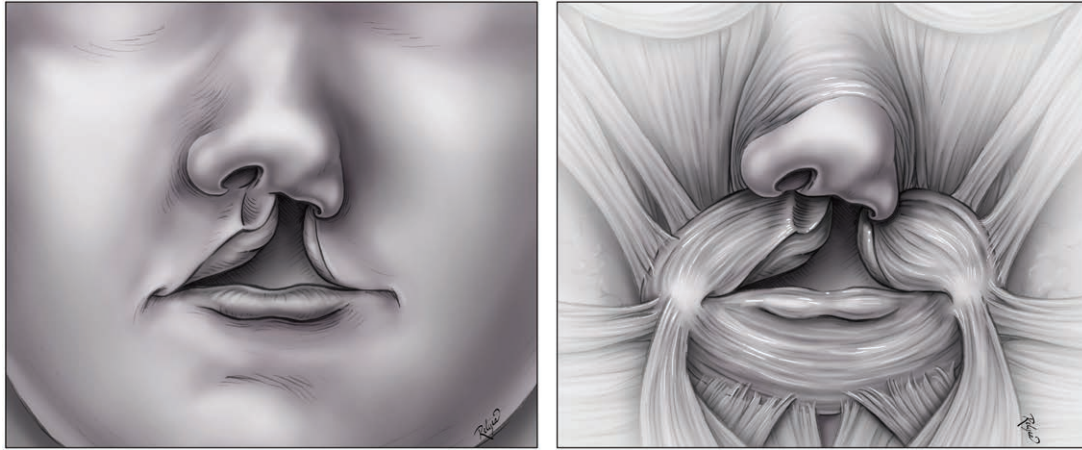
septum, whereas the lateral lip muscle inserts along the nasal sill and periosteum of the piriform aperture. This aberrant muscle orientation contributes to the cleft lip nasal deformity. Inadequate correction of the orbicularis oris muscle results in persistent lip and nose asymmetry. All three layers of the lip—mucosa, muscle, and skin—should be addressed in the surgical repair.

An incomplete cleft lip deformity by definition demonstrates an intact nasal sill, or Simonart band, but with variable degree of separation in the lip. Minor form refers to a vermilion notch that is greater than 3 mm,<sup>30</sup> often associated with a vertical skin depression extending into the nasal sill. A microform, or forme fruste, manifests as a vermilion notch that is less than 3 mm. Anatomically, unlike in the complete cleft lip, the orbicularis oris muscle may be in continuity, but thinner and weaker. Most forms demonstrate some level of nasal asymmetry despite less severe lip deformity.

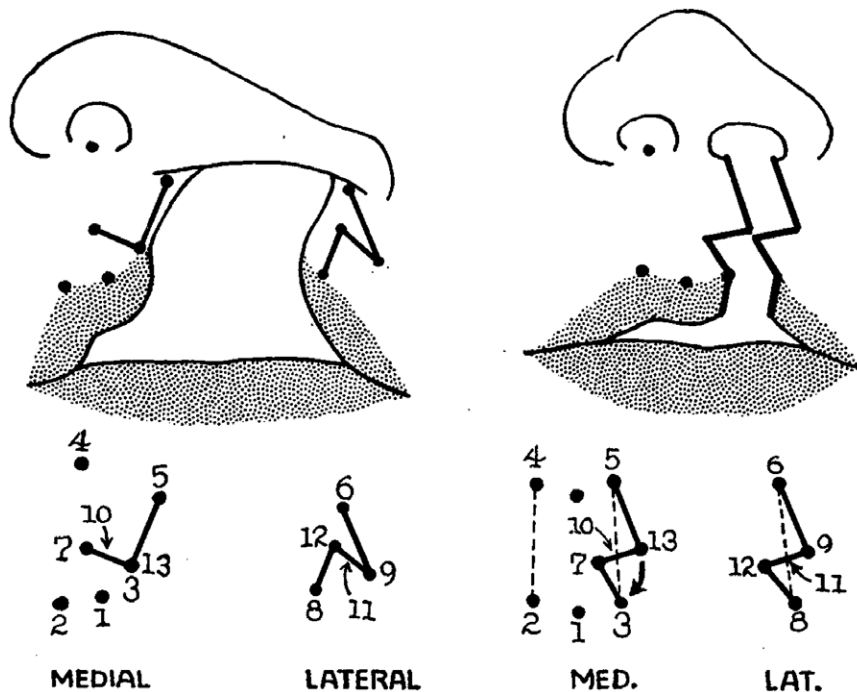
The surgical evolution of the unilateral cleft lip repair has been a continuous quest toward what Millard called the “ideal normal,”<sup>31</sup> referring to a “normal” appearing lip and nose. It reflects knowledge gained over years, ingenuity of new solutions, and an unwavering desire toward perfection. There have been many heroes along the way, each paving the path for the next toward solving a puzzle that requires “imagination ... [to] project beyond surgical stages, growth, heredity and time.”<sup>31</sup>

Early cleft lip repairs date back centuries. The first clearly written account was in 1564,<sup>32</sup> in which Paré recognized that a cleft lip can be repaired if the cleft margins are excised and sewn together. From these direct closures came the realization that although the lip is closed, the height discrepancy of the medial and lateral lip elements creates a notable notch. In the early twentieth century, Rose<sup>33</sup> and Thompson<sup>34</sup> were firsts to address this discrepancy by designing curved and angled tissue excisions, which provided a modest lengthening of the lip. However, this too was inadequate, particularly in severe deformities.

Next came the recognition that the short medial lip element needs to be augmented using local tissue rearrangement. Mirault<sup>35</sup> added an inferior triangular flap from the lateral lip element in addition to angled skin excision. Le Mesurier<sup>36</sup> created a quadrilateral flap from the lateral lip to reconstruct the Cupid’s bow. Tennison<sup>37</sup> and Randall<sup>38</sup> applied the principle of the Z-plasty to cleft lip repair (Fig. 4). Although these techniques were good solutions to the height deficiency problem, they failed to respect natural landmarks, and



**Fig. 3.** (Left) Unilateral cleft lip is characterized by lip height deficiency and off-center/rotated Cupid's bow and philtral complex. (Right) Note the discontinuous orbicularis oris muscle with aberrant muscle insertions.



**Fig. 4.** Tension/Randall lip repair. (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.)

often created noticeable nonanatomical scars. From these earlier techniques arose the modern ones, which can be largely categorized into rotation-advancement and straight-line (Table 2).

**Rotation-Advancement**

Ralph Millard introduced the rotation-advancement repair in 1955 to address the problems created by the geometric techniques. His repair balances the Cupid's bow with minimal interruption to the continuity of the philtral column (Fig. 5). The original Millard I design results

in a more oblique cleft-side philtral scar, accentuating the asymmetry. Millard II modification added a back-cut at the noncleft side philtral column, which allowed for more rotation of the philtral complex and a less oblique scar (Figs. 6 and 7). [See Video 1 (online), which displays surgical marking for the modified Millard repair for unilateral cleft lip. See Video 2 (online), which displays the operative details for the modified Millard repair for unilateral cleft lip.]

Since its advent more than 60 years ago, there have been numerous modifications seeking to

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**Table 2. Comparison of Basic Characteristics of Two Major Repair Techniques**

Characteristic	Straight-Line	Rotational-Advancement
Lip height discrepancy (medial lip)	Back-cut within dimple	Back-cut along contralateral philtral column
Lip height discrepancy (lateral lip)	Adjust angle of lateral lip dart	Adjust by moving the Noordhoff point
Horizontal lip discrepancy	Minimal	Affected by Noordhoff point selection

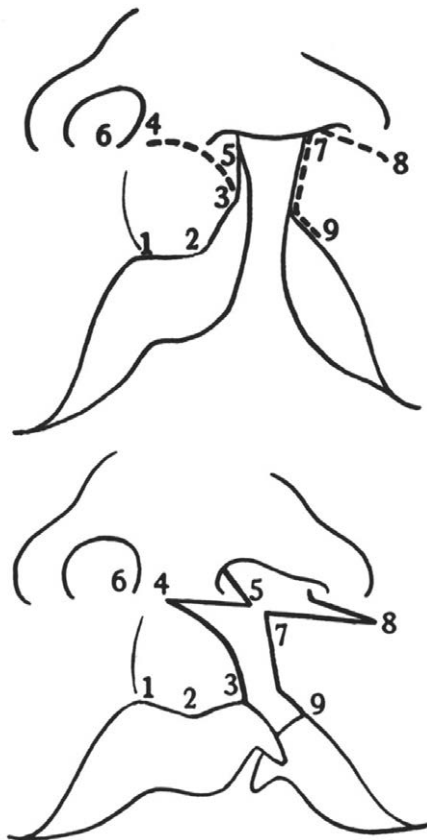
refine the results, including but not limited to Byrd,<sup>39</sup> Cutting,<sup>40</sup> Losee,<sup>1</sup> Mohler,<sup>41</sup> Mulliken,<sup>42</sup> Noordhoff,<sup>43</sup> and Stal,<sup>44</sup> making the advancement-rotation technique the most commonly taught in training programs and the most frequently used. However, it is not without its limitations. Because the technique relies on rotation to lengthen the medial lip element, inadequate rotation and/or scar-shortening can result in a short and notched lip deformity. In addition, to match the length of the medial lip after rotation, more lateral lip tissue may be needed for closure, thereby resulting in a tight lip.<sup>45</sup> Scarring around the nose can be significant, particularly when using the modifications that make additional incisions on the columella.

**Straight-Line**

Unlike rotation-advancement techniques, the straight-line techniques result in a vertical linear scar. Because simple straight-line excision and closure of cleft margins fail to augment the cleft-side height deficiency, techniques evolved to recruit additional tissue from the lateral lip, most often in the form of a small triangular<sup>35,37,46</sup> or quadrangular<sup>35,36</sup> tissue flap inserted into a back-cut in the medial lip. Fisher’s anatomical subunit repair<sup>47,48</sup> is one of the most commonly used straight-line repair techniques. It evolved from the geometric repairs, with improved placement of scars within natural anatomical boundaries. The medial lip is lengthened with a small back-cut and a lateral triangular flap in the nadir of the Cupid’s bow (Fig. 8). Because the lengthening occurs at the concavity above the white roll, there are minimal nonanatomical scars. The lateral lip triangular flap breaks up the straight-line scar, preventing contracture (Fig. 9). [See Video 3 (online), which displays surgical marking for straight-line repair for unilateral cleft lip (part 1). See Video 4 (online), which displays surgical marking for straight-line repair for unilateral cleft lip (part 2). See Video 5 (online), which displays operative details for straight-line repair for unilateral cleft lip (part 1). See Video 6 (online), which displays operative details for straight-line repair for unilateral cleft lip (part 2).]

The central tenets of both techniques are the same: identify and preserve the existing philtral complex (philtral columns and Cupid’s bow); elongate the medial lip; dissect and repair skin, muscle, and mucosa; and hide scars within natural anatomical boundaries. The primary difference lies in the understanding of the skin deformity. The rotation-advancement technique effectively rotates the entire philtral complex downward and fills a superior defect using a lateral lip flap (Figs. 5 and 10). The presumption, therefore, is that the philtral complex comes together at an apex at the base of the columella.

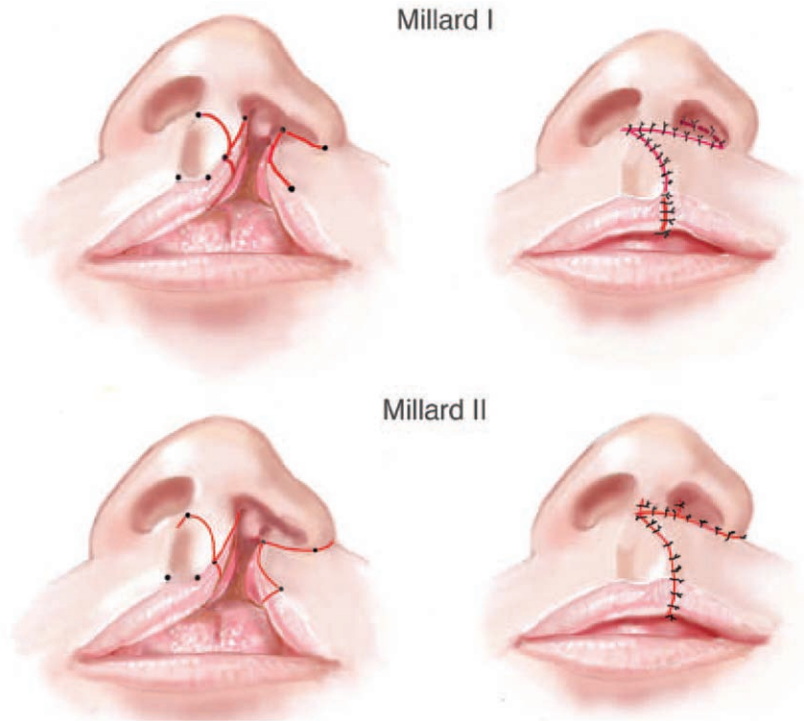
Alternatively, the straight-line technique presumes that the philtral deformity is caused by a length discrepancy between the two philtral columns, instead of a rotational defect. In other words, the Cupid’s bow appears rotated



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

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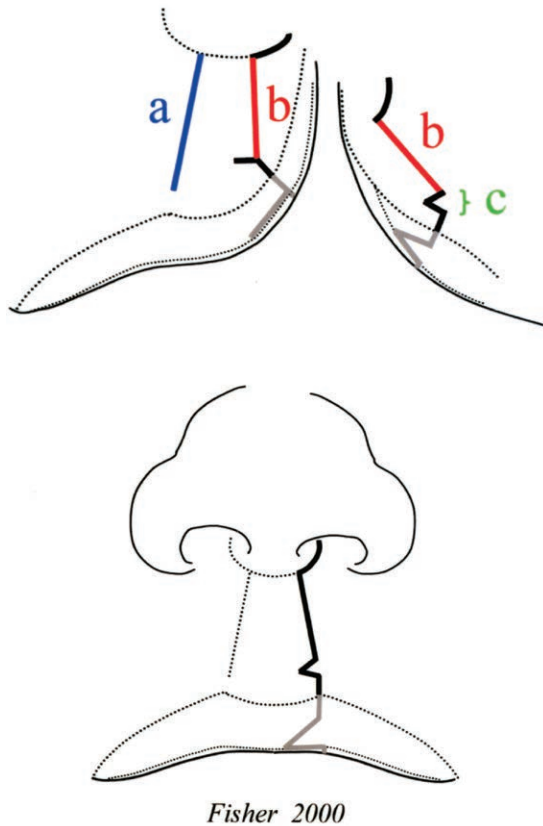


**Fig. 6.** Comparing Millard I and Millard II unilateral cleft lip repair. (Reprinted from Marcus JR, Allori AC, Santiago PE. Principles of cleft lip repair: Conventions, commonalities, and controversies. *Plast Reconstr Surg.* 2017;139:764e–780e.)



**Fig. 7.** Rotation-advancement repair for left cleft lip in a male patient. (Left) Preoperative, (center) 3-month postoperatively, and (right) 1-year postoperative views. (Courtesy of Dr. Laura Monson.)

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**Fig. 8.** Anatomical subunit repair for unilateral cleft lip: Fisher repair. (Reprinted from Fisher DM. Unilateral cleft lip repair: An anatomical subunit approximation technique. *Plast Reconstr Surg.* 2005;116:61–71.)

because one column is shorter than the other. Lengthening the shorter column with a back-cut and a small lateral lip dart effectively repositions the Cupid's bow (Fig. 11). This presumes the philtral columns are parallel to each other and do not join at a peak at the base of the columella.

### THE CLEFT LIP NASAL DEFORMITY

Understanding the orientation of the orbicularis oris muscle under the cleft lip skin is one of the best ways to understand the cleft nose deformity. The foundation of the nose is unbalanced, and the nasal structure is deviated toward the noncleft side because of the asymmetrical pull of the muscle (Fig. 12). The greater the severity of the cleft, the more aberrant the muscle insertion. The nasal structures tend to follow this anatomy. For the complete unilateral cleft lip, the ipsilateral nasal ala straddles the cleft. The cleft side maxilla is displaced inferoposteriorly compared with the noncleft side. Latham<sup>49</sup> postulated in 1973 that the embryonic nasal septum and its ligamentous attachment to the premaxilla (the

septopremaxillary ligament) were responsible for this maxillary asymmetry. The body of the nasal septum is deviated and warped toward the cleft, and during the early embryologic period, this leads to decreased anterior growth of the maxilla and later inadequate inferior growth of the non-cleft-side maxilla because of a tethering effect. The end result is a nose with asymmetric alar base positions, a tip that is depressed and rotated toward the noncleft side, and a depressed cleft-side ala.

Although it is tempting to accept that the nasal deformity is a product of the cleft lip anatomy, it does not fully explain the persistent (albeit milder) nasal deformities seen in incomplete and even microform cleft lips. These minor deformities suggest that, aside from displacement, there may be inherent deficiency in the nasal structures themselves. One theory is that weaker and thinner cleft-side lower lateral cartilage is present, perhaps caused by inadequate mesodermal migration during nasal formation<sup>50</sup> (Fig. 13).

Significant controversy exists regarding the timing of correction of nasal deformity. Furthermore, there remains no consensus regarding open versus closed primary rhinoplasty, or the effect of primary rhinoplasty on nasal growth.<sup>51</sup> Management varies widely, from no primary cleft nose correction to aggressive cartilage reshaping. In addition, the advent and wide use of nasoalveolar molding for presurgical shaping of the cleft-side ala has assisted surgical repair over the past several decades. In general, most primary rhinoplasty techniques involve hidden access incisions,<sup>40,52,53</sup> release of the lower lateral cartilage by undermining either in the supracartilaginous<sup>54</sup> and/or infracartilaginous plane, and suture suspensions either to the contralateral ala or upper lateral cartilage to reshape the ala and nasal tip.<sup>42,55</sup> Most rely on nasal stents to maintain postoperative form. All begin with defining the nasal base and ala position with precise repair of the nasal component of the orbicularis oris muscle.

The goal for primary nasal correction should be to improve the aesthetic appearance of the nose without interrupting its growth potential. All primary nasal repairs focus on the lower third of the nose and are specifically designed to address the collapsed and deficient cleft-side ala and alar base. Unfortunately, the long-term outcome of the unilateral cleft lip nasal repair remains one area of dissatisfaction for many cleft surgeons.<sup>52,54,56,57</sup> Relapse of the cleft-side alar deformity occurs more frequently than desired.





**Fig. 9.** Straight-line repair of unilateral cleft lip in a male patient shown (*above, left*) preoperatively, (*above, right*) intraoperatively, (*below, left*) 1 month postoperatively, and (*below, right*) 3 months postoperatively.

Secondary revisions are needed 35 to 74 percent of the time, most of the time at skeletal maturity. In these cases, a formal septorhinoplasty is performed. One large long-term outcome study<sup>26</sup> demonstrated a 20 percent rate of collapse of nostril height at 5 years after repair, and recommended overcorrection of nostril height and undercorrection of width at time of primary repair to compensate. Recognizing that the inherently weaker and malformed cleft-side cartilage is likely contributing to the relapse despite suture suspension and postoperative stenting, Lu et al.<sup>58</sup> used septal cartilage graft, harvested from the excess caudal septal at the time of septal repositioning, as a rim graft for support. The study demonstrated sustained symmetry at more than 3 years' follow-up, with no disruption to nasal and facial growth at 7 years. The authors admit that more specific studies with controlled groups and longer follow-up are needed.

Alternatively, Tse et al.<sup>56</sup> focus on the foundation of the nose and correction of the structural displacement by septal reposition, adequate muscular reconstruction, sidewall advancement, and nasal sill closure extending into the nasal floor. The nasal tip is left unchanged. The authors report that only two of 102 cases underwent nasal revision at 5-year follow-up.

At our own institution, management varies depending on the surgeon. The consensus is that nasoalveolar molding can improve the deformity in the early setting, and postoperative nasal stenting is essential to maintain the shape of the surgical correction.

### POSTOPERATIVE MANAGEMENT

Postoperatively, families are given the option to stay or go home, with most choosing to stay overnight to ensure adequate oral intake. As

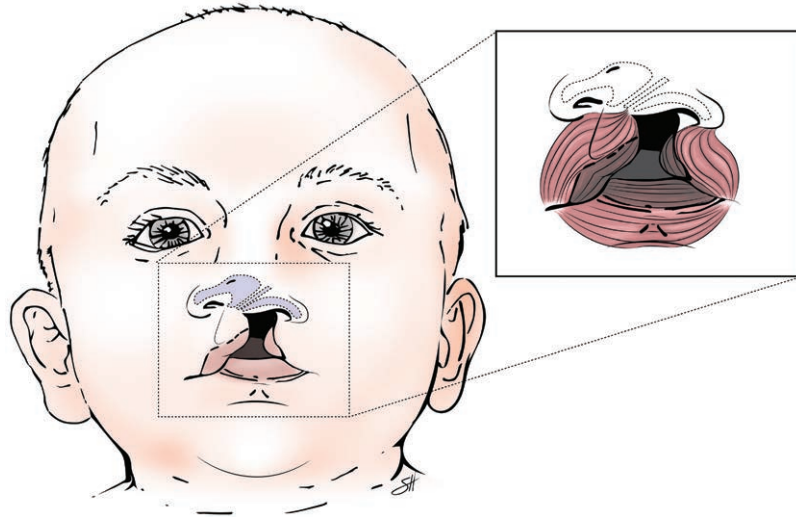


**Fig. 10.** Rotation-advancement repair of left cleft lip in a male patient shown preoperatively (*left*) at 3 months of age and (*right*) at follow-up 4 months postoperatively. (Courtesy of Renata Maricevich, M.D.)

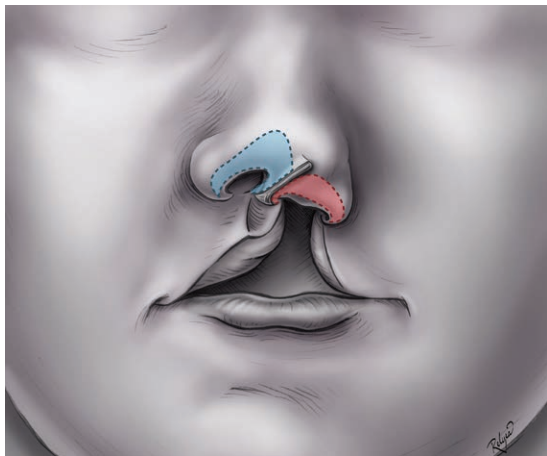


**Fig. 11.** Straight-line repair of right cleft lip of a male patient shown preoperatively (*left*) and 2 years postoperatively (*right*).

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**Fig. 12.** Cleft nose deformity, demonstrating abnormal insertion of the orbicularis oris muscle affecting the severity of deformity.



**Fig. 13.** Inherent discrepancy of the lower lateral cartilage contributes to the cleft nose deformity.

become hypertrophic in the early postoperative period are candidates for topical steroid taping with aggressive scar therapy. If there is no improvement, intralesional steroid injection is offered. Scar management continues until scars are fully matured.

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**PATIENT CONSENT**

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

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