Advances of Plastic & Reconstructive Surgery

Chapter 1

Cleft lip nasal deformity: Analysis and treatment

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Abstract

Cleft lip and palate (CLP) are the most frequent congenital craniofacial defects and are usually associated with craniofacial defects and nose deformities that alter the facial aesthetic configuration.. The aetiology of CLP is thought to be multifactorial (genetic, teratogenic and/or environmental factors), 3 although there are no studies which have determined the exact causes that produce it. This chapter describes the nasal deformities associated with congenital clefting and outlines the timing and techniques used to correct these deformities.

1. Introduction and objectives

The clinical spectrum of this disorder ranges from the lesser degree which constitutes CLP to the maximum extent represented by a bilateral CLP, and which correlates with the severity of nasal alterations. CLP is usually accompanied by a characteristic nasal dysmorphia and hemifacial growth disorders. The causes of this nasal dysmorphia can be primary or secondary; the primary are intrinsic to those of CLP, whilst the secondary are associated with varying degrees of partial recurrence of the original deformity, scarring and/or sequelae from previous surgical procedures.

Patients with nasal dysmorphia secondary to CLP present aesthetic involvement and functional impairment leading to a psychological conflict which in some cases causes isolation and/or social exclusion. This, in turn, can cause a need for early intervention due to the functional and aesthetic alterations suffered by these patients.

The decision to repair a cleft lip or palate deformity is based on a variety of factors: speech development, facial growth, psychological impact on the child and family, and safety to undergo anesthesia. The cleft lip nasal deformity is a complex, three-dimensional problem that challenges any rhinoplasty surgeon. The extent of the nasal deformity is related to the severity of the original cleft malformation and ranges from mild to severe [1]. To perform nasal surgery is determined by the amount of functional breathing issues and aesthetic concerns of the patient.

After initial surgical treatment, further surgery (both nasal and maxillary) is often required to reduce the physical impact in these patients. Many techniques for the correction of this nasal dysmorphia have been described, but the only correct procedure is likely to be the use of cartilage grafts; preferably through an open rhinoplasty, which should not be performed until the development of the facial skeleton is complete (16–18 years).

This chapter describes the nasal deformities associated with congenital clefting and outlines the timing and techniques used to correct these deformities.

2. Anatomy of the cleft nasal deformity

The nasal deformities associated with congenital unilateral cleft lips have been well described and are consistent [2,3,4]. The deformity begins with the deficiency of tissue in the nasal base related to the maxillary hypoplasia and continues with findings related to the external pressures applied after surgical repair and during development. The extent of the typical nasal deformity is related to the degree of deficiency of alar base support on the cleft side.

2.1 Unilateral cleft nasal deformity

The typical characteristics of the unilateral cleft lip nose are described in Table 1. The hallmark of the unilateral cleft lip nasal deformity is a three-dimensional asymmetry of the nasal tip and alar base (Figure 1). The nasal tip refers to the subunit composed by the alar bases, the columella, and the lower lateral cartilages. The nasal tip is also asymmetric, with the cleft side lower lateral cartilage (LLC) having a shorter medial crus and longer lateral crus than the LLC on the noncleft side. The columellar complex, which is created by the medial crura and feet of the LLC, the caudal septum, and soft tissue, is typically deviated toward the noncleft side, secondary to an asymmetric, unopposed pull of the orbicularis oris muscle. The cleft alar base is asymmetric and the cleft ala is displaced laterally, inferiorly, and posteriorly to its noncleft counterpart [5]. The weakened and malpositioned cleft side LLC produces a nostril that is wide and horizontally oriented. This changes the three-dimensional configuration of the entire tip.

The nasal septum is deflected caudally into the noncleft nasal airway due to the unop-

posed pull of the orbicularis oris muscle and the septopremaxillary ligament. Further posteriorly, the lack of these attachments to the middle and posterior cartilaginous septum leads to bowing of the septum into the cleft side airway [3]. In the unilateral cleft condition the nasal airway is compromised on both the cleft and noncleft sides.

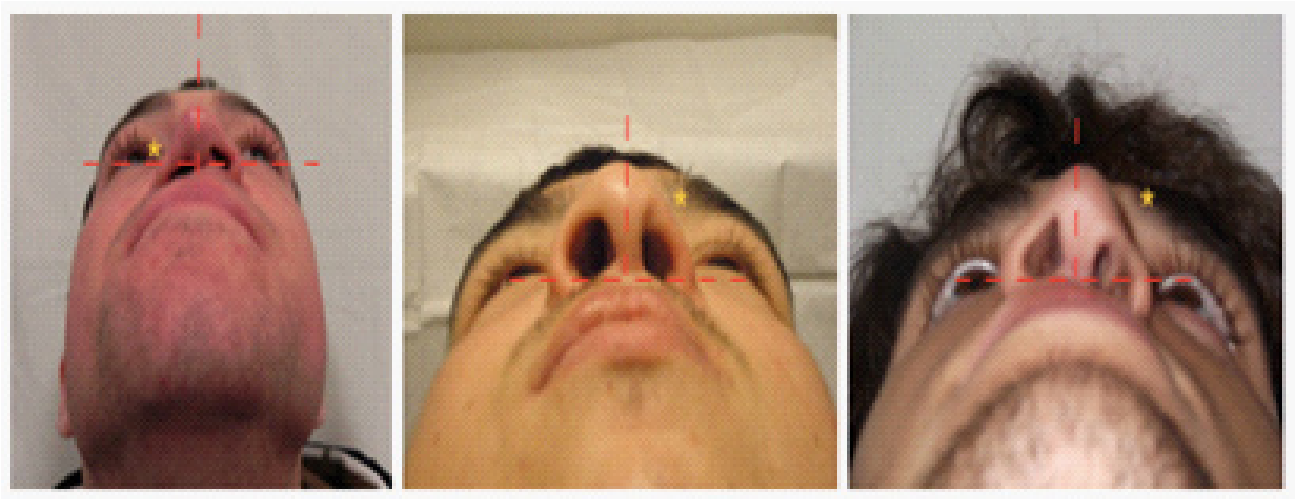
In the unilateral cleft deformity the external nasal valve is compromised by two related factors: introversion of the nasal ala and webbing of the nasal vestibule. Introversion of the cleft nasal ala is the result of posterior inferior rotation of the lower lateral cartilage due to the distortional pressures on the cartilage from the position of the columella and alar base [13]. The introversion leads to hooding and thickening of the ala; it also contributes, along with surgical scarring, to webbing of the nasal vestibule. An oblique fold is formed by posterolateral displacement of the piriform margin and introversion of the lower lateral cartilage. This bulk influences airflow and alters the relationship of the upper and lower lat- eral cartilages.

The middle one third of the nasal deformity can be characterized by interrelated changes to the upper lateral cartilages and to the internal nasal valve. The internal nasal valve is formed by the relationship of the upper lateral cartilage, the nasal septum, and the inferior turbinate. This weakness results from inadequate skeletal support and is often manifest by concave of the upper lateral cartilages. This weakness typically affects the internal nasal valve on the cleft side. On the cleft side, there is limited attachment of the upper and lower cartilage and a side to side relationship rather than the more typical overlap seen on the noncleft side [12]. Both of these factors lead to decreased support of the upper lateral cartilage and collapse of the upper lateral cartilage with deep inspiration. In the cleft lip nasal deformity, the septum is bowed into the cleft side at the internal nasal value, and the upper lateral cartilage support is weak, causing the cartilage to bow or collapse with respiration. Therefore, the internal nasal valve can significantly limit the nasal airway on the cleft side.

The upper one third, there is no classic deformity to this portion of the nose in the cleft lip nasal deformity, the osseous pyramid is typically reduced in width at the time of definitive rhino- plasty to enhance the overall appearance of the nose.

Table 1: Characteristics of unilateral cleft lip nasal deformity

Lower lateral cartilage and Nasal tip (Lower Lateral Cartilages The lower lateral cartilages have four components: the medial crus, middle crus, lateral crus, and dome)	Medial crus of lower lateral cartilage shorter on cleft side Lateral crus of lower lateral cartilage longer on cleft side (total length of lower lateral cartilage is same) Lateral crus of lower lateral cartilage may be caudallydisplaced and may produce hooding of alar rim Alar dome on cleft side is flat and displaced laterally Columella deviates to the noncleft side- Short on cleft side Base directed to noncleft side (secondary to contraction of orbicularis oris muscle) . Nasal floor and sill are often absent on the cleft side. Bony deficiency on the cleft side of the Skeletal base
Nostril	Horizontal orientation on cleft side
Alar base	Displaced laterally, posteriorly, and inferiorly
External nasal value	Compromised by introversion of the lower lateral cartilage and webbing of the nasal vestibule
Septum	Caudal septum deflects toward the noncleft side. Cartilaginous and bony septum deviates toward the cleft side.
Upper lateral cartilages	Weakened support leads to bowing or collapse with deep inspiration on the cleft side. Abnormal relationship between the cleft upper and lower lateral cartilage.
Internal nasal valve	Frequently compromised by weakened support of the upper lateral cartilage and the deviation of the septum.



2.2. Bilateral cleft nasal deformity

The bilateral cleft lip nasal deformity is also caused by a lack of skeletal support. The bilateral cleft lip nose is usually not grossly asymmetric. Of course, if a marked difference exists on the two sides of the lip, there can be gross asymmetry of the cleft nasal tip and alar base in the bilateral cleft lip patient (Table 2).

The nasal tip is typically in the midline in the bilateral complete deformity. If one side of the lip is more involved than the other, the short columella is typically deviated toward the less involved side, pulling the tip in that direction (Fig.). The lower lateral cartilages demonstrate short medial crura and long lateral crura. The domes of the lower lateral crura are splayed, con-

tributing to a poorly defined, frequently bifid tip (Fig.). The angle at the dome is obtuse. The alar bases are posterior, lateral, and inferior, giving rise to flaring of the base and widening of the nostril. The tension on the lower lateral cartilage leads to introversion and webbing of the vestibular floor. The septum is in the midline in the complete bilateral deformity and deviated caudally toward the less involved side if an asymmetry exists.

The nostrils in bilateral cleft lip patients are more horizontal than those in noncleft patients. The nasal septum is usually midline, being deviated caudally to the less involved side if asymmetry exists. The middle nasal third exhibits poor cartilaginous support, compromising the internal nasal valve and affecting functional nasal breathing.

The middle one third is analogous to the unilateral deformity, with poor support to the upper lateral cartilage leading to bowing and possible collapse of the upper lateral cartilage with deep inspiration. However, because the septum is typically in the midline, the compromise of the internal nasal valve is often not as significant.

The upper one third is typically not involved in the bilateral nasal deformity.

Table 2: Characteristics of the Bilateral Cleft Lip Nasal Deformity

Lower lateral cartilage and Nasal tip (Lower Lateral Cartilages Thelower lateral cartilageshavefourcomponents: the medial crus, middle crus, lateral crus, and dome)w	Deviates toward less involved side if discrepancy exists. Columella is short and deviates toward less involved side if discrepancy exists. Lateral steel of lower lateral cartilage on the cleft side produces a long lateral crus and a short medial crus. This also causes blunting of the dome with a more obtuse angle. Medial crura are splayed, producing a poorly defined, bifid tip. Alar bases are displaced posteriorly, laterally, and inferiorly Nasal floor and sill are often absent on the cleft side. Bony deficiency on the both side of the Skeletal base
Nostril	Nostrils are wide and horizontally oriented.
Alar base	Displaced laterally, posteriorly, and inferiorly
External nasal value	Compromised by introversion of the lower lateral cartilage and webbing of the nasal vestibule
Septum	Deviated to less involved side if a discrepancy exists.
Upper lateral cartilages	Weakened support leads to bowing or collapse with deep inspiration. Abnormal relationship with the lower lateral cartilage.
Internal nasal valve	Compromised by weakened support of the upper lateral cartilage.

3. Timing of cleft nasal repair

Cleft nasal reconstruction can be divided into primary and secondary repairs [1]. Primary rhinoplasty refers to nasal surgery performed at the time of the initial cleft lip repair. Secondary rhinoplasty refers to any cleft nasal surgery performed after the initial cleft lip re-

pair. Open septorhinoplasty carried out at the age of 16–18 years is an appropriate procedure to correct the cleft lip nasal and palate deformity.

The decision to perform surgery on patients with cleft deformities is the product of many factors. These include the fact that the child with a nasal malformation is exposed to ridicule during childhood. This philosophy is counterpointed by the surgeon's knowledge that nasal and midfacial growth is not completed until the mid to late teen years. Early surgical intervention is thought to interfere with subsequent nasal growth. Although traditional philosophy avoids aggressive early nasal surgery fearing nasal growth inhibition, there is a growing trend toward primary cleft nasal repair [5-13].

In many centers, rhinoplasty at the time of initial cleft repair is the accepted surgical treatment. This philosophy recognizes that maximizing nasal tip projection and nasal and alar base symmetry during lip repair allows the nose to grow in a symmetric fashion. For this reason, most con- temporary cleft surgeons perform primary nasal repair on the tip and alar base.

3.1 Primary cleft rhinoplasty

The goals of primary rhinoplasty are:

- Maximize symmetry of the nasal tip and alar base. Surgical challenge:

- The base of the nose in the unilateral deformity involves gross asymmetries of both the skeleton and soft tissues, the soft tissue attachments of the alar base must be completely freed from the skeletal base (pyriform aperture) [5]. This allows repositioning of the nasal and alar base in a more symmetric fashion [13-18].

- Dissection of the cleft alar base from the pyriform aperture involves an internal alotomy. The cleft alar base is entirely separated from its skeletal attachments, the floor of the nose is approximated and the alar base is medialized, thereby creating improved three-dimensional symmetry.

- Nasal tip plasty. The cutaneous attachments of the lateral cruras are separated. This is accomplished by creating medial and lateral tunnels using the incisions of the standard rotation–advancements lip repair [19].

- After the lip is sutured closed and the alar base is repositioned, the LLC is molded into its new shape. This can be accomplished with interdomal sutures or with the application of nasal bolsters. Increase nasal tip projection and improve tip symmetry [20].

3.2. Secondary cleft nasal reconstruction

Several special considerations must be made for secondary rhinoplasty. The external approach is an excellent method to gain exposure of the cartilaginous structures as dense scar often impedes dissection. In such cases the direct visualization provided by the external approach may be needed in addition to the tactile feedback upon which endonasal dissection depends. In cases with severe scar formation, even with direct visualization, it may be difficult to differentiate scar from cartilage. As in primary cases, three-pointretraction while applying downward pressure with the tips of a pair of Converse scissors will aid in finding and maintaining the correct plane. The goals of secondaryrhinoplasty are:

- Complete and aggressive restructuring of the internal and external nasal anatomy can be performed. The purpose of the surgery is to create symmetry and definition of the nasal base and tip, to relieve nasal obstruction, and to manage nasal scarring and webbing.

- The difficulty of the repair lies in the complex interaction of the original three-dimensional pathophysiologic process and the previous surgeries and lip repair same the time of definitive rhinoplasty.

- Correction of short columellar skin.

- Augmentation rhinoplasty, especially in bilateral cleft lip-nose.

4. Techniques for the repair of cleft lip nasal deformity

4.1. Surgical keys:

- One of the most serious problems of the nose is a deviated and depressed nasal tip.

- Correction procedures: nasal tip correction is achieved by repositioning of the lower lateral cartilage through open rhinoplasty and a caudal septal extension graft, and these procedures are the same as those previously reported.

- The first step in the repair of the nasal deformity, whether unilateral or bilateral, is to establish a solid nasal base on which to build the remainder of the reconstruction.

- Vestibular expansion graft also ensures prevention of postoperative collapse of the support of the lower lateral cartilage.

- Additionally, the improvement of the maxillary platform by bone graft is important to correct the dislocated nasal ala.

Figure: The incisions used to open the nosevary, but a combination of bilateral marginal incisions with an inverted-V mid-columellar incision is typically used

4.2. Incisions

The approach used to perform cleft nasal reconstruction varies with the deformity. The incisions used to open the nose vary, but a combination of bilateral marginal incisions with an inverted-V mid-columellar incision is typically used [22].

However, if additional skin is needed in the columella, modification of the typical external columellar incision can be used. In the unilateral cleft nasal deformity, an asymmetric V-to-Y on the cleft side can be designed to increase columellar soft tissue (and length). If significant columellar deficiency is present in the bilateral cleft nose, a midline V-to-Y upper lip incision can be used, recruiting skin from the upper lip into the columella. An alternative incision choice is to use upper lip forked flaps [23]. These flaps have the advantage of narrowing the central segment while lengthening the columella.

4.3. Nasal base

The first step in the repair of the nasal deformity, whether unilateral or bilateral, is to establish a solid nasal base on which to build the remainder of the reconstruction. The first step in the creation of the base occurs with primary rhinoplasty. Closure of the nasal floor with the lip represents the beginning of the base. The next step is to correct the bone deficiency of the maxilla and premaxilla and restore the abnormal posteroanterior position of the alar base.3 Furthermore, augmentation of the nasal sill and base may be required at the time of the definitive rhinoplasty and can usually be accomplished with local tissue flaps [25].

4.4. Septal reconstruction

The cleft lip nose is usually associated with significant septal deformity. In the unilateral cleft nose, the pyriform aperture is very asymmetric and usually has a bony deficiency on the cleft side. This bony asymmetry, coupled with an unopposed pull of the orbicularis oris muscle results in a caudal nasal septum significantly deviated to the noncleft side. In the bilateral deformity, bony deficiency on both sides of the pyriform aperture causes a wide and poorly supported caudal nasal septum. This results in a wide and weakly supported caudal septal deformity.

Nasal reconstruction begins with septal reconstruction and repositioning. If the caudal septum is dislocated, it must be freed from its bony and soft tissue attachments, and repositioned into the midline 6. This often requires resection of a small amount of inferior cartilage from the maxilla and suture repositioning of the caudal septum. The caudal septum is usually sutured to the nasal spine. If the septum requires further stabilization or straightening, a caudal septal extension graft can be used. This provides support to the base of the nose, and can be a pillar upon which to suture and stabilize the nasal tip.

The posterior septal deformity should also be corrected to maximize the nasal airway and to obtain cartilage grafts. In the unilateral cleft nose, the posterior septum is usually deflected significantly to the cleft side. The septum is usually approached through an open septoplasty, by separating the LLCs. This allows correction of both the caudal and posterior deformities and gives access to obtain cartilage for graft material.

5. Treatment of the asymmetric alar base

The alar base on the cleft side is usually malpositioned. In the original congenital malformation, the position of the affected alar base is posterior, lateral, and inferior to the unaffected side. Once the lip is repaired, however, the alar base is repositioned. This new position may be symmetric with the noncleft side, but usually is in a new asymmetric position. The alar base width on the cleft side is often narrower than the noncleft side and malpositioned in either

a superior or an inferior position versus the unaffected alar base.

An additional deformity that often presents with the secondary cleft defect is a lack of normal configuration of the affected nasal sill. During initial lip closure, the floor of the nose is reconstructed. A very small deformity (malposition) of the nasal sill (0.5 to 1 mm) at the time of lip closure usually creates a very noticeable deformity in the adult nose. At the time of definitive nasal reconstruction, the nasal sill and anterior floor should be closed in as symmetric a position to the noncleft side as possible. This often requires incision of the nasal sill and layered repair of the nasal floor. The cleft alar base is then repositioned in a symmetric fashion.

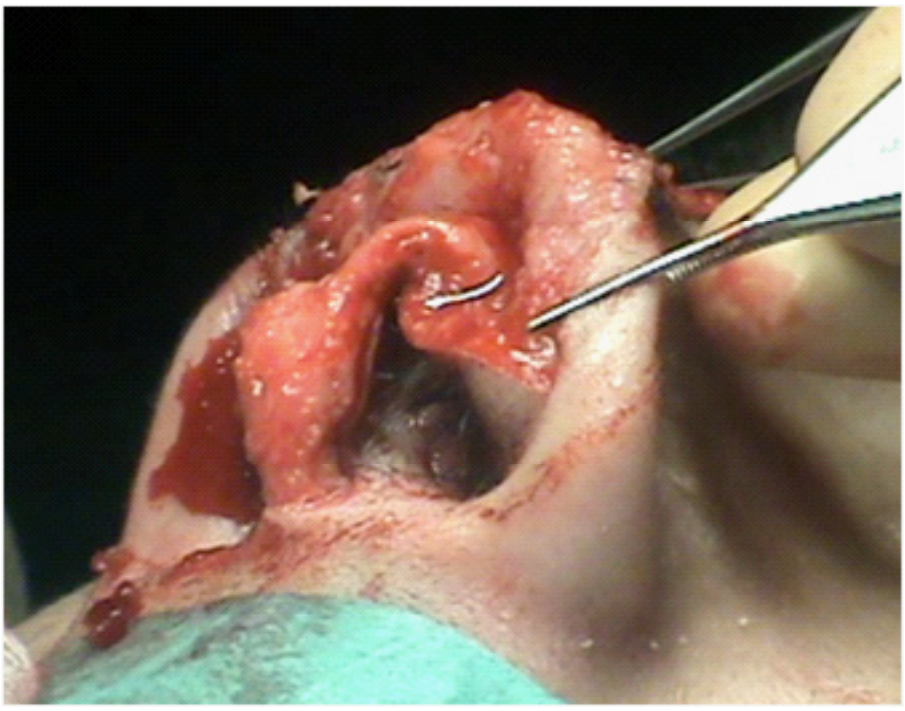


Figure: The cephalic view demonstrating malposition of the lower lateral cartilages.

5.1. Lower nasal third

After stabilizing the caudal nasal septum and providing three-dimensional alar base symmetry, the external rhinoplasty is performed. The lower third of the nose requires support and symmetry.

Support and symmetry can be established by stabilizing the nasal tip complex to the nasal septum. This can be accomplished by (1) a extended spreader grafts, (2) a septal extension graft, or (3) a columellar strut to createthree-dimensional symmetry.

One difficulty cleft lip rhinoplasty is the lack of septal cartilage available for grafting material. In these cases it is often necessary to harvest cartilage from one or both ears. A vertical skin incision is made approximately 1 cm in front of the post auricular sulcuson the posterior conchal bowl. The skin and perichondrium is then elevated from the posterior concha with Converse scissors. Retraction with a small skin hook and blunt dissection with a cotton-tipped

applicator aids in this process. Care should be taken to leave the peripheral vertical component of the concha intact so that no change inshape of the ear occurs. The harvested segment may extend toward the canal meatus, but the emenentiacor responding to the root of the helixshould not be excised. The resulting pieceis usually kidney shaped and will vary from 2−4 cm in largest dimension. The skin flaps should be judiciously cauterized to prevent thermal injury. Closure with a few subcutaneous 4.0PDS sutures should be placed. A bolster in the anterior conchal bowl may be fashioned from a dental roll and sutured through the ear with a 3.0 nylon suture.

In cases when ear cartilage is also in sufficient or exceptionally strong grafting material is needed, costal cartilage may be harvested. Typically, the cartilage is taken from rib VII, VIII,or IX. A 3−5 cm incisionis placed over the medial aspect of the rib. The muscle is separated in the direction of its fibers to access the rib surface. Subperichondrial dissection around the rib is performed with an elevator. It is important to retain a subperichondrial dissection on the deep surface of the rib in order to avoid injury to the pleura.

Under direct visualization, the graft is freed from the surrounding perichondrium and the desired segment sharply excised. Am all eable retractor may be placed deep to rib to protect the pleura. A needle may be inserted into areas of the rib in which it is unclear whether bone or cartilage is present. Closure should be performed in a layered fashion after hemostasisis achieved.

After stabilizing the medial crura to the nasal septum, vertical division of the LLC can be performed lateral to the nasal dome. This allows an increase in nasal tip projection. Camouflage of nasal tip asymmetries is then accomplished with a shield-style nasal tip graft [24]. In cleft nasal deformities, tip grafts provide support, an increase in nasal projection, and camouflage for tip asymmetries.

Vertical dome division both narrows the nasal tip and provides elevation of the cleft side LLC. If there is residual alar hooding of the cleft side LLC after completing division of the LLC, the LLC can be sutured superiorly to elevate the cleft side alar rim. The LLC may be sutured to the ipsilateral ULC for superior repositioning.

Table 3: Major and minor tip support mechanisms

Major tip support mechanisms
- Attachment of medial crura to the septum - Resilience of the alar cartilage - Attachment of cephalic alar cartilage to caudal upper lateral cartilage
Minor tip support mechanisms
- lnterdomal ligaments - Cartilaginous and membranous septum - Anterior nasal spine - Skin and soft tissue envelope - Lateraralcrural attachment to the pyriform aperture

5.2. Middle third of the cleft nose

The middle third of the nose in cleft patients often is weak, resulting in both aesthetic and functional issues. Concavity of the cleft side upper lateral cartilage (ULC) often results in internal nasal valve dysfunction [25]. This weakness can be treated with onlay grafts, spreader grafts, autogenous spreader flaps, or flaring sutures. Unilateral, or asymmetric spreader grafts, placed between the ULC and the nasal septum, often improve nasal function and help to straighten the slumped nose.

6. Treatment of the alar rim (lateral crus of the LLC)

The cleft side LLC is poorly supported with lack of normal skeletal support medially and laterally. This lack of support usually causes malposition of the cleft alar rim. The cleft side lateral crus of the LLC is concave and results in an introverted alar contour [26].

Various techniques exists to treat the LLC concavity. These include suture techniques, underlay alar strut grafts, on lay grafts, and autocartilage flaps [27]. Additionally, the lateral crus can be dissected from the underlying vestibular skin, removed, flipped, and resutured in a convex fashion. Alar strut grafts are placed between the existing concave-outward LLC and the underlying vestibular skin. This graft strengthens the lateral crus of the LLC and flattens the concave LLC.

Suture techniques such as horizontal mattress sutures also strengthen and flatten the lateral crus of the LLC. Last, the cephalic margin of the LLC can be made into an advancement flap. This cartilage flap can be advanced to support the remaining LLC. Each of these techniques supports the lateral crus of the LLC and strengthens the external nasal valve.

Figure A: patient with bilateral cleft lip related nose deformity before and 3years after. The illustrations demonstrate the soft tissue and frame alterations to achieve the intended goals.Patient satisfaction and good nasal breathing are the best result

7. Conclusion

The cleft lip nasal deformity will likely continue to frustrate and challenge reconstructive surgeons for years to come. The concepts seem clear: restore symmetry and definition to the nasal tip and base, realign and open the nasal airway, and prevent scarring and webbing from compromising the results. In practice, however, the three-dimensional nature of the deformity and the involvement of all nasal layers make a de- pendable result elusive. Preoperative analysis for this condition should include an aesthetic and functional nasal study and an anthropometric and psychological study. Open septorhinoplasty carried out at the age of 16–18 years is an appropriate procedure to correct the cleft lip nasal and palate deformity. Structural grafting is used to maximize function, structure, support and symmetry.

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