Management of Secondary Deformities in Cleft Patients

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Contents

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S. O. Keyhan et al. (eds.), *Handbook of Oral and Maxillofacial Surgery and Implantology*, https://doi.org/10.1007/978-3-031-36962-9_216-1

Abstract

Secondary or residual deformities following primary cleft repair are encountered in almost every patient with a cleft lip and palate. Treatment of cleft always includes multiple staged procedures and the final assessment of the results of this surgery must wait till the child grows up fully. The primary surgical procedures have an influence on the growth pattern of these children resulting in various secondary deformities like lip distortion, deformity of the nose, cleft in the alveolus, fistulae, malocclusion, and dysfunction of the palate. Management of such deformities is difficult and usually requires a multidisciplinary approach with careful systematic evaluation, integrated treatment plan, and high-quality surgical technique. This chapter aims to discuss the various secondary cleft deformities and its management in brief.

Introduction

It must be the principal goal of cleft surgeon to restore the deformed and displaced anatomy as close to normal as possible. The pathogenesis of these deformities is related to the presence of scar tissues into the basal bone area of the cleft, which inhibits alveolar growth; palatal soft tissue scarring, which inhibits growth of palate and also causes palatal orientation of dentoalveolar elements; and the excess lip tension, which may inhibit growth of maxilla along the dentoalveolar structures (Berkowitz 2013; Chow et al. 2003; Marcks et al. 1958).

The first step in management includes a systematic evaluation of the problem at hand. A format for evaluating has been presented in the following table:

As per Wilson L.F., correction of residual deformities of the lip and nose in repaired clefts of the primary palate (lip and alveolus). Clin. Plast. Surg. 12:719, 1985 (Marcks et al. 1958).

I. The foundation of the lip and nose
   A. Evaluate the alveolar defect.
   B. Determine the position of the maxilla.
   C. Identify missing or malpositioned dental units.
II. Lip
   A. Assess the total amount of lip tissue.
   B. Evaluate the freedom of buccal aspect of the lip from the anterior aspect of the alveolar process. Does the upper lip pout?

C. Evaluate the vermilion mucosa for equal bulk along the entire margin.
D. Cupid's bow.
   1. Determine if all components are present.
   2. Determine the position of components.
E. Philtral Structures.
   1. Assess the length of philtral edges.
   2. Evaluate the direction of ridges as well as the location and direction of lip scars.
   3. Assess the width of the philtrum column.
F. Check the alignment of the circumoral muscles.

III. Nose
A. From the front
   1. Locate the lower lateral cartilages.
   2. Locate the alar bases: to each other, to the eyes, and to the structures of the Cupid's bow.
   3. Determine the size and shape of alae.
   4. Locate the upper lateral cartilages.
   5. Determine the width of the bony pyramid.
B. From both profiles
   1. Determine the nasolabial angle.
   2. Assess the length and direction of the septum.
   3. Evaluate the visibility of the columella.
   4. Assess the length of columella and the relative position of nasal tip.
C. From below
   1. Determine the size, shape, and direction of nostrils.
   2. Assess the length of both sides of the columella and the direction of nostrils.
   3. Evaluate deviation with respect to the most anterior portion of the septum.
   4. Evaluate the vestibules.
   5. Evaluate the alae for size, shape, and position.
D. From above
   1. Assess the straightness of the septum.
   2. Determine dimensions of lateral cartilages.
   3. Similarly, assess the alae.

The management of various secondary cleft deformities will be discussed in this chapter with emphasis on pre-treatment considerations and patient's preparations, surgical techniques, post-treatment considerations, common complications, and case scenarios.

Cleft Lip Deformities

Residual or secondary deformities post cleft lip repair can either be esthetic or functional and include scars, skin shortage or excess (vertical and transverse), orbicularis oris muscle malposition or diastasis, lip landmark abnormalities, vermilion deficiencies, and buccal sulcus obliteration (Lip 2013; Marcks et al. 1958; Miloro et al. 2004; Monson et al. 2014; Sittah et al. 2017, 2018).

Pre-Treatment Considerations: Timing, Surgical Planning and Patient Preparations

Timing for a secondary lip repair surgery is not rigid and may vary from patient to patient. It can be done as a part of long-term follow up or earlier, on request, if possible, depending on the fitness of the patient. These procedures may be done either under local anesthesia or general anesthesia. Pre-surgical planning and preparations are done as per that. Comprehending the normal anatomy and the abnormal anatomy guides the patient into planning the required secondary surgical procedure.

Unilateral Cleft Lip Deformities

- Lip Scars: Z /W-plasty or wave-line incisions are the usual revision techniques for such scars. Sometimes during revision, scar can be de-epithelized and buried to get philtral prominence in vest-over-pants fashion.
- Long Lip: it is not a very commonly encountered defect. It is mostly attributed to primary cleft repair and occurs as result of excessive rotation. In such cases, scar should be excised, derotated, and re-advanced with repair of incision that went to normal side. Full thickness horizontal excision from superior portion of lip below nostril sil can be used as a corrective procedure. Transverse scar of philtral area correction has been documented using excision in triangular horizontal fashion for lip shortening.
- Short Lip: it is the most common deformity which occurs due to the vertical scar contracture along the suture line subsequent to primary lip repair, and components of this defect are usually seen in each layer—skin, muscle, and mucosa. Thorough clinical assessment with total secondary cleft repair is indicated. Straight line or slightly curved repair causes shortening. Millard's rotation-advancement produces temporary shortness in wide clefts which usually settles into normal position with resolution of the scar. Permanent shortening of lip happens due to technical errors like—insufficient rotation, incision not extending across base of columella, not using backcut, insufficient dissection, repositioning, and lengthening of muscle layer. Surgical management will require excision of scar and rotation of flap with a back cut. For major discrepancies, revision rotation-advancement is needed. Rose-Thompson scar excision also helps in gaining additional length in case of lower vertical scar.

- Tight Lip: loss of soft tissue in excess during initial lip repair can result in tight lip. To correct these deformities with significant horizontal deficiencies an Abbe Flap may be required.
- Vermilion Deficiency or Deformity: corrective procedures include Z plasties, V-Y advancements, transposition flaps, free grafts, and cross lip flaps. Whistle deformities can be corrected using V-Y advancement flaps. The techniques used in unilateral cases can be applied to bilateral as well.
- Deficient Buccal Sulcus: corrective procedure include widely undermined laterally based mucosal advancement flaps.
- Orbicularis oris abnormalities revision includes redirecting and introducing muscles keeping in mind the anatomy of cleft and normal lips. Techniques for realigning have been described by many including Randall, 1959; Fara, 1968; Millard 1968; Climo 1969; Delaire, 1975; and Kernahan, 1978.
- Reconstruction of philtrum and cupid's bow mainly requires a centrally placed Abbe's Flap.

Bilateral Cleft Lip Deformities

Most commonly encountered deformities include v-shaped whistle deformity, short or wide prolabium, asymmetric prolabium, inadequate orbicularis repair, deficient gingivobuccal sulcus, and tight or long upper lip. Secondary repair involves completely opening the scar to narrow prolabium, reorientation, and positioning of muscular bands correctly, using resulting lateral fork flaps for columellar lengthening. Whistle deformity can be corrected using triangular island flaps based on orbicularis muscle and advanced from lip segments in a v-y fashion. Nasal deformities are corrected either simultaneously with columellar lengthening or at a later stage.

- Lip Scars: minor scar revisions or white roll mismatch requires same correction techniques as unilateral deformities. Adequate philtrum width and acceptability of moustache growth is an indicator of successful bilateral cleft lip repair in males. Such cases can be corrected using Abbe flaps and temporal island flaps bring hair-bearing areas from lower lip and scalp.
- Long Lips: multiple techniques have been described over the years. Ragnell's design has been shown to be the most satisfactory which involves excision of scar and advancement of alar bases medially after adequate undermining. Shortening of lip with increase in transverse width and reconstruction of a mucomuscular lip can be done with a switch flap. Transverse wedge excision at nasolabial junction also known to achieve shortening of excess length.
- Short Lip: minor corrections can be done using scar excision and z-plasty techniques. If enough tissues are available, major corrections can be done with total reoperation. In case of deficiency in tissues, lengthening requires flaps like abbe flap, Muir and Bodenham's technique using full thickness ear lobe graft, fork flaps, and cheek advancement flaps beneath alae as described by Barron.

- Tight Lip: excessive tissue excision and usage of prolabium for lengthening during primary correction results in tight lip having excess tension. Abbe flap and its various modifications are recommended for the correction of the same. Webster's perialar crescentic flaps may also be used. Good muscle function can be obtained with advancing orbicularis muscle to midline and covering with a sandwich Abbe flap.
- Orbicularis Oris Deformity: muscle needs to be separated from its vertical attachment and distinct horizontal muscle flaps need to be developed as far as nasolabial folds if required for correction of such defects.
- Philtral Abnormalities: if not corrected at the primary repair stage, these can be corrected at the secondary stage using abbe flap (in case of extreme defect) or other less extensive methods in case of absence of need for tissue augmentation. Other methods include transposed inferiorly based flaps from midline to lateral horizontal position to create the hollow, bilateral vermilion advancement in V-Y fashion for lip eversion and construction of central tubercle of cupid's bow, and midline flap (Onizuka and associates, 1978).
- Vermilion Deficiency: most common deformity includes whistle deformity which can be corrected using a bilateral V-Y advancement (Robinson, Ketchum, and Masters. 1970) or bilateral vermilion island flaps-based orbicularis oris muscles.
- Buccal Sulcus Abnormalities: this deformity requires separation of prolabium from premaxilla by lining posterior aspect of prolabium with lateral flaps and leaving premaxilla to epithelize secondarily.

Case Scenarios

See (Figs. 1 and 2)

Alveolar Cleft Deformities

A clinically apparent gap is noticed in the alveolus, solely on either the labial aspect or involving both cortices. Tissue hypoplasia and subsequent aberration of anatomy on the cleft side is an expected finding in patients presenting with unilateral clefts (Bonanthaya et al. 2021).

Abnormalities of the Alveolus and Dentition

- Crossbites result from collapse of the arch medially.
- Premaxilla is either normal or rotated away from the cleft side.
- Rotation of central incisor toward the cleft side.
- Congenital absence of lateral incisor is a common feature. Sometimes a hypoplastic tooth is seen or a tooth resembling a supernumerary tooth.

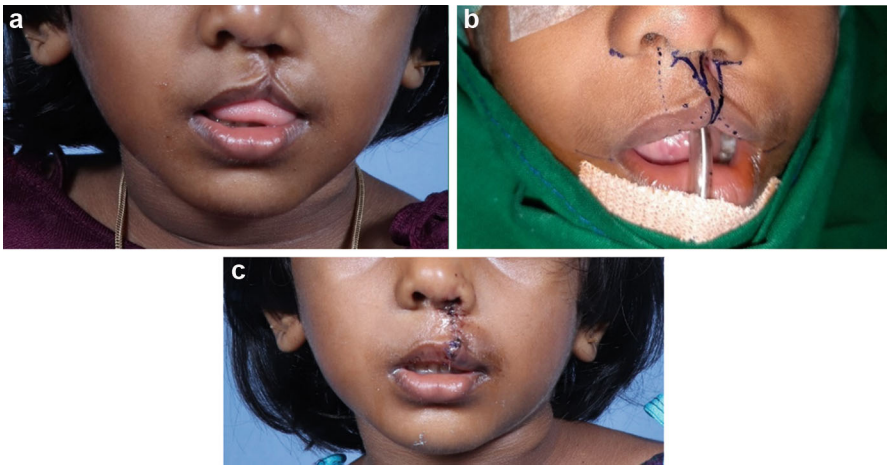


Fig. 1 (**a**) An 8-year-old with scar over the upper lip post primary cleft lip repair surgery. Problems noted include lip scar, vermilion deformity, notching, and nasal floor insufficiency. (**b**) Modified Millard's markings made, scar excised, and reorientation of muscles done. (**c**) Immediate post-op picture

- Bone loss secondary to sub-optimal attachment of investing tissue on the cleft side, with a higher risk of predisposition attributed to central incisors and cuspids (Fig. 3) (Abyholm et al. 1981; Bertz 1981; Bonanthaya et al. 2021).

Alveolar Bone Grafting

Alveolar bone grafting helps canine and lateral incisor tooth to erupt into position and stabilizes the alveolar arch from collapsing. This provides a bony framework for the alar base, helps separate the oral cavity from the nasal, while also enabling a strong base for maxillary advancement to correct retrusion of midface.

Timing of Alveolar Bone Grafting (Boyne and Sands 1972)

Primary	In infancy, less than 2 years of age after lip repair, but before repair of cleft palate
Early secondary	Between 2 years and 5 years of age, before incisors eruption
Secondary	Between 8 years and 11 years of age, prior to the predicted eruption of upper cuspids
Late secondary	After completion of 12 years

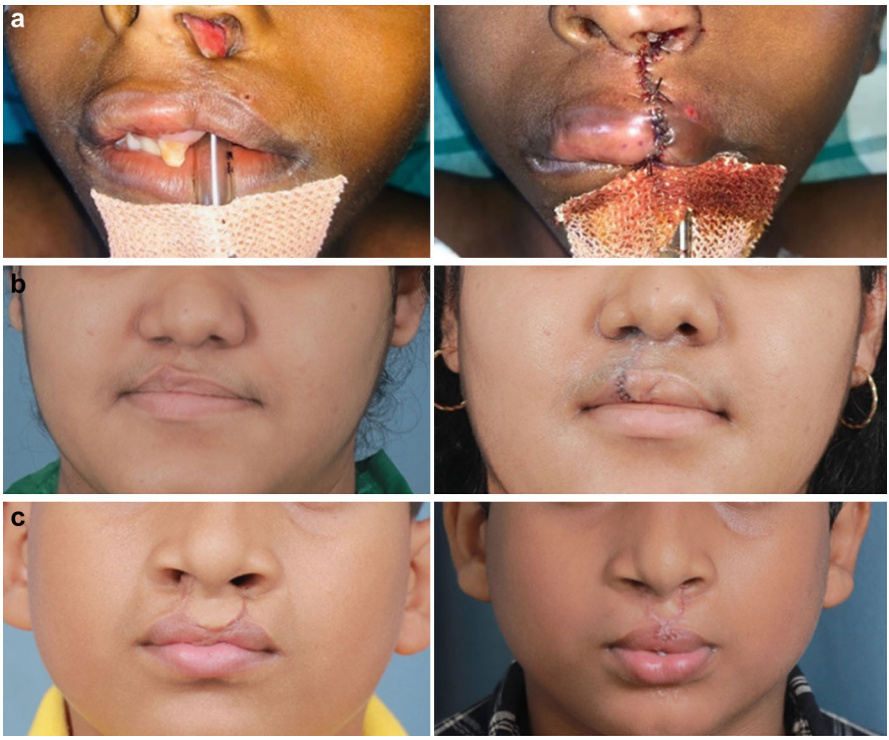


Fig. 2 Series of secondary lip correction cases. (**a**) Unilateral lip with secondary vermilion notching and nasal floor gaping correction, (**b**) unilateral lip with notching correction, and (**c**) bilateral cleft lip scarring correction

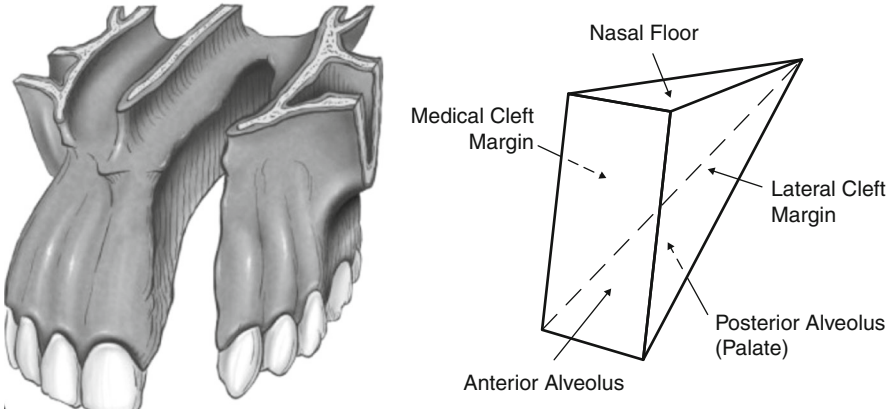


Fig. 3 Surgical correction should require reconstruction of all the surfaces of the cleft, which includes the floor of the nose, medial and lateral cleft margins, and anterior and the posterior alveolus (palate)

Rationale of Alveolar Bone Grafting

- To provide stability to maxillary arch.
  It is easier to advance maxilla in one piece by Lefort 1 osteotomy than three separate osseous segments.
- For closure of oronasal fistula and anterior palatal cleft.
  When bone grafts are used to construct a pyriform rim which is symmetric to normal side, it provides a better platform for alar base improving nasal symmetry and prevents inferior turbinate prolapsing in to cleft.
- For ensuring better periodontal support for teeth bordering the cleft
  Bone grafting before eruption of permanent dentition will create a bony matrix for teeth to erupt improving orthodontic result and reduces the need for any permanent fixed prosthodontic appliances (Bergland et al. 1986).

Sources of Bone Graft

Alloplastic	Allogenic	Autologous
rhBMP-2 TEOM Glasses—Available commercially (SiO_2, Na_2O, CaO, P_4O_{10})	Undecalcifed freeze-dried bone Allograft cells-and-Tissuebank-Austria Osteograft Maxgraft	Iliac crest—Anterior or posterior Mandible Calvarium Rib Proximal tibia

Surgical Technique

There are three main designs for soft tissue flaps which include buccal finger flap, lateral sliding flap, and oblique sliding flap. As per Posnick et al., two factors which rule the design of flap (Fig. 4)

1. Depth of vestibule needs to be preserved.
2. Flap design should ensure maximum attached mucosa in the region of the alveolar cleft which in turn will aid in development of an adequate periodontal sulcus and attachment of erupting canine.

- Surgical correction of alveolar cleft is performed under general anesthesia. A two-team approach may be adopted where iliac crest is considered as the donor site.
- The graft has to be inserted into a so-called “pocket,” the boundaries or walls of which are essentially made up of the labial, palatal, or nasal flaps, the dissection of which has to be done meticulously.

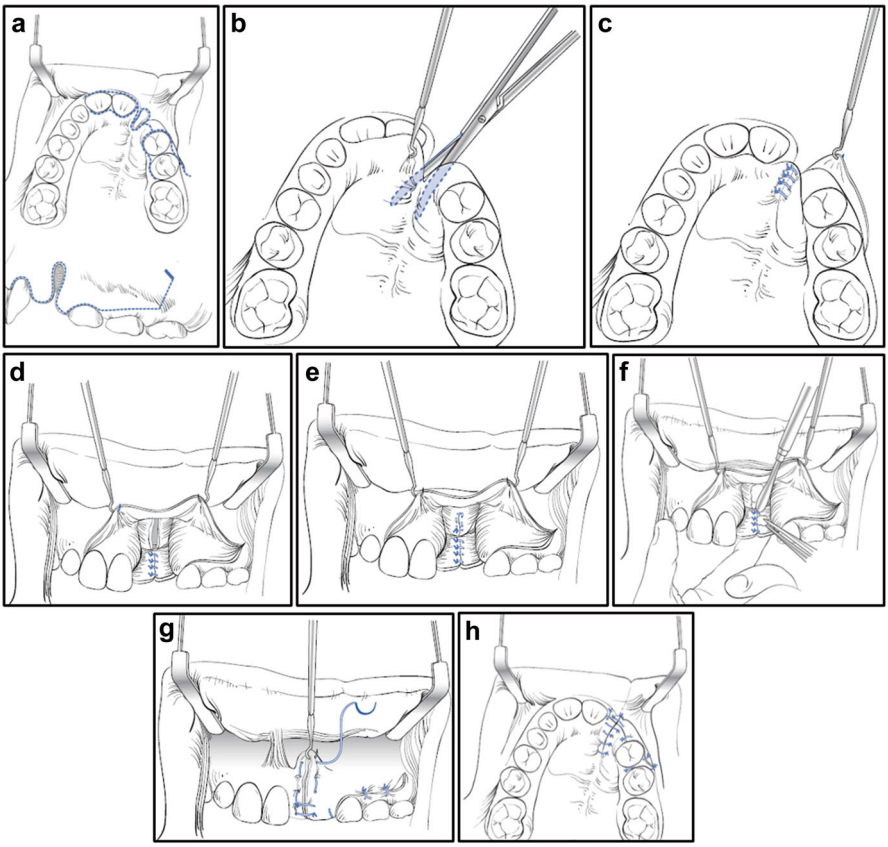


Fig. 4 (**a**) Sulcular incision, (**b**) palatal flaps are developed, (**c**) palatal closure, (**d**) reflection of nasal mucosa from the bony margins of the cleft, (**e**) approximation of nasal flaps with burying to obtain a watertight closure of nasal floor, (**f**) packing of defect with harvested bone, and (**g, h**) the labial flaps are mobilized and closure done. This ensures attached keratinized tissue. Exposed areas at distal region are left to granulate

- For oblique sliding flap, incision is given through the mucosa over the cleft extending over bone.
- There is no bony margin in the region of pyriform aperture. To aid nasal closure at this level, division of the adjacent soft tissue is carried out.
- A pericoronal incision is given along the deciduous teeth and over permanent dentition incision is given 3–4 mm above gingivodental junction.
- Mucoperiosteum within the fistula has to be turned in superior direction with the help of an elevator.
- Trim excess tissue judiciously.
- Suture nasal flaps to reduce the incidence of an oronasal fistula.

- Burying sutures used to approximate nasal flaps when possible.
- Move the palatal flap towards the cleft.
- The harvested bone is condensed compactly into the exposed bony cleft.
- The labial flaps are then mobilized over the graft.
- Releasing incisions in the periosteum are given perpendicular to the direction of the advancement.
- Approximation should be done keeping attached gingival over the reconstructed alveolus (McCarthy 1990).

Complications

- Incidence of exposure of bone graft due to dehiscence, flap necrosis, or infection is rare.
- Presence of foul smell or unpleasant taste may indicate exposure which could be managed by extensive debridement.
- Bone resorption tend to occur following exposure or infection.
- External tooth root resorption occurs as a result of proximity of osseous tissue to the cementum of the teeth.

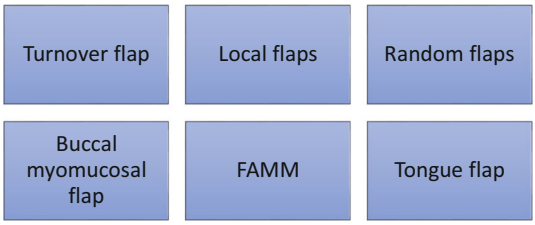
Long-Term Stability of Alveolar Bone Grafting

- Toscano et al. (2012) concluded in his study that the severity of cleft had no statistical correlation with success of bone grafting.
- Advocating for and applying a robust surgical-orthodontic protocol at the earliest possible opportunity was recommended by Toscano et al.; postoperative bone resorption can be prevented and guarantees proper placement of the teeth.
- Bergland and Witherow-derived scales are commonly used to assess (Witherow et al. 2002).

Cleft Palate Deformities

Cleft repair helps achieve an airtight closure to aid in speech production and creates a barrier between the two cavities, i.e., oral and nasal. Contributing factors may include the patient's nutritional health, the characteristics of the palate, the manner of surgery, and the operating surgeon's experience. The greatest risk of fistula is related with complete clefts bilaterally. Determining the size of the fistula, measuring palate length and motion, assessing speech hypernasality, and determining dependable and well-vascularized tissue transposition choices are all necessary for an adequate examination of a fistula. To lessen the likelihood of recurrence, nasal fistula repair should always involve restoring both layers (nasal and oral mucosal) (Marcks et al. 1958).

Key Techniques for Fistula Repair



What Causes a Palatal Fistula?

1. One layered hard palate closure (only oral closure without nasal floor closure).
2. If nasal mucosa is not released properly owing to inadequate dissection of nasal mucosa from hard palate bone, nasal layer develops tension on approximation which in turn can cause fistula.
3. Difficulty in approximation of unilateral or bilateral mucoperiosteal flaps due to inadequate dissection of flap causing tension in the midline after final closure.
4. Handling of tissues roughly like the medial edges of flap.
5. Overuse of cautery.
6. Tension in approximation from insufficient dissection or extra tight suture knots.
7. Severe infection after surgery.
8. Habits like thumb or finger sucking can cause injury to palate after surgery.

Key Preventive Postoperative Measures

- Minimizing manipulation of tissue, and ensuring gentleness when indicated.
- Minimally sufficient dissection and tension free closure.
- Postoperative hand restraints in case of thumb sucking habits.
- Surgical site should be kept clean and free of debris.
- Use of natural honey after surgery three times every day with a soft plastic spoon and do not give water after that. This may improve healing or help in reducing the fistula size, as suggested by several small studies from Indonesia.

Fistula Evaluation

Symptoms and Examination

- Illumination test using a nasopharyngoscope to:
  1. Inspect the fistula.
  2. Check soft palate mobility.
  3. Evaluate velopharyngeal (VP) competence.

A palatal plate or readily accessible dental wax can be used to temporarily seal the fistula in order to study potential influences during speaking. It is possible to record speech samples both before and after the fistula is plugged. If, upon plugging, hypernasality diminishes, the fistula is mostly to blame for the nasality. To unlearn VP dysfunction, however, speech therapy may be required in addition to fistula healing. Conversely, if the hypernasality continues following plugging, then a second VP operation ought to be taken into consideration. Using this easy technique, decision-making becomes more algorithmic and efficient. During initial phases after primary palate repair, fistula to be managed conservatively. A gap of minimum 6–9 months is needed before scheduling revision surgery.

Management of Palatal Fistula

Excessive stress on the suture line causes the majority of fistulae to form in the midline. The location, size, and degree of the fistula are factors that determine the success of its closure, extensive scarring on the palate (caused by prior surgery).

For oronasal fistulas, surgical intervention is usually the best course of action when properly thought out and carried out. Even yet, in really challenging situations or in patients with severe cardiac abnormalities, the fistula can be properly sealed using good obturators (if necessary, with speech bulbs).The rationale behind fistula surgery is to close the wound in numerous layers to reduce stress on any one layer and to provide redundancy in the event that one layer does not heal sufficiently. Two layers in the hard palate and three in the soft palate generally suffice (Sittah et al. 2017).

Description of Techniques for Fistula Repair

Turnover Flap: Used for nasal mucosal reconstruction. These flaps are developed from margins surrounding the edges of palatal fistulas, mostly 3–5 mm development and sometimes more. Outer margins lie on the bone or cartilage such that the periosteum or perichondrium is reflected along with it. Tissues should be handled gently and flaps need to be mobilized enough to ensure tension free closure. This nasal layer formed needs an oral layer cover from various flaps like buccinator flaps, local buccal sulcus flaps, or free flaps.

Mucoperiosteal Flap: They are either single, pedicled, or bipedicled based on greater palatine vessels. In case of wide or secondary repairs, radicle dissection is considered essential for achieving adequate mobilization of flaps. Greater palatine vessels need to be released. Ten–twenty millimeter dissection is advised. In case tension persists, soft tissues are incised medial to the pedicle resulting in complete release of the pedicle, and free medial movement of the flaps.

Levator Retropositioning: As per non-cleft palatal anatomy, levator veli palatini presents horizontally. The right and left sided muscles join each other forming a

sling, which is attributed to speech functions. In case of cleft palate, there is a change in the orientation of these muscles from horizontal to being directed obliquely upwards with bilateral aponeurotic insertion, causing faulty speech. This indicates a need for revision if not addressed primarily.

Ginvigobuccal Sulcus Flap: In previously operated lip/palate patients in which anterior palatal or nasal floor is not reconstructed, a fistula presents, with scanty tissue available. In case of an alveolar defect, nasal closure can use a medial vomer and lateral nasal flap. Oral closure can be performed by raising a superiorly based gingivobuccal sulcus mucosal flap from any side, turned postero-medially, thus covering the previously sutured nasal layer.

Premaxilla Gingival Flap: After a bilateral cleft repair, a defect develops next to the premaxilla. A premaxillary gingival lining flap can be designed for reconstruction of such defects.

Tongue Flap: These flaps are used in cases presenting with a fistula with larger dimensions, or a fistula positioned far too anteriorly. Tongue flaps are anteriorly based for the treatment of fistulae and can be raised either dorsally (common) or ventrally. It is traditionally done in two stages, with division being performed roughly 3 weeks following the first procedure.

FAMM or Buccal Myomucosal Flap: FAMM flaps based superiorly can be used for anterior hard palatal defects. Inferiorly based flaps are used for soft palate. This has been further modified by I. Jackson and R. Mann, who suggested a myomucosal or axial muscular flap using buccinator.

Case Scenarios

See (Fig. 5)

Cleft Maxillary Hypoplasia

Patients with cleft frequently exhibit maxillary growth retardation, which results in significant skeletal and/or dental problems in all three spatial planes. The most complex issue to address is the anteroposterior maxillary deficiency combined with unilateral/bilateral palatal collapse. Two main reasons causing this hypoplasia are—(1) less intrinsic growth potential leading to developmental deficiency and (2) scarring from primary surgical repair which inhibits maxilla from growing normally. These patients present with very specific features like concave facial profile with Class III skeletal tendencies, loss of contour in relation to cheek fullness being absent/diminished. Some people develop anterior and posterior crossbites with abnormalities in anterior and posterior facial heights and less commonly chin and mandibular asymmetry. Secondary maxillary hypoplastic defects can be managed

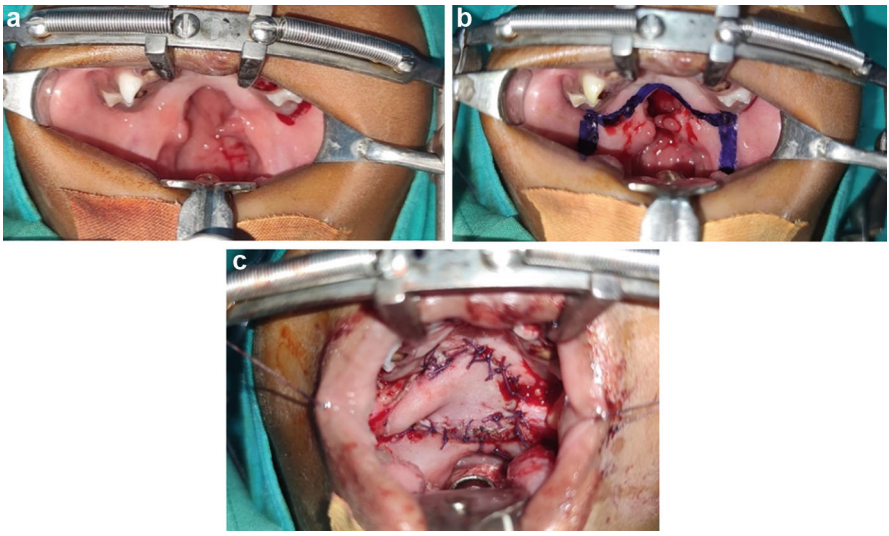


Fig. 5 (**a**) A case of mid palatine fistula, (**b**) surgical markings done, (**c**) correction done using bilateral FAMM flap

either surgically or conservatively with non-surgical orthodontics with or without protraction gears or face masks.

Management and Treatment Planning

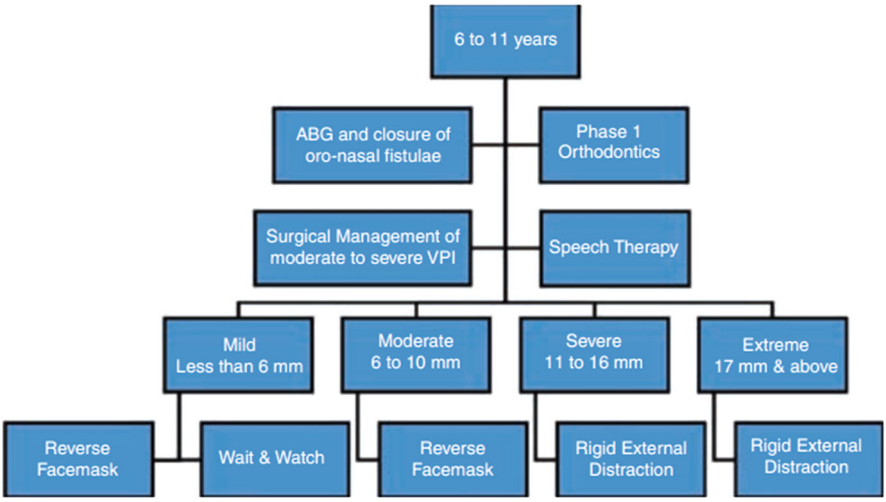
Management of maxillary hypoplasia secondary to cleft aims primarily to improve facial esthetics and to achieve adequate occlusion which in turn helps patients to masticate efficiently, improves dimensions of the airway, and phonation. Surgical treatment should be planned such that any deterioration of VPI is avoided and patient has an overall improvement in psychological health. The time of presentation, the severity of maxillary hypoplasia, alveolar status, and presence or absence of pre-existing fistulae all play an important role in surgical planning (Bonanthaya et al. 2021; Chow et al. 2003; Fatone et al. 2013; Fonseca 2000).

Surgical Methods for Advancing Hypoplastic Maxilla

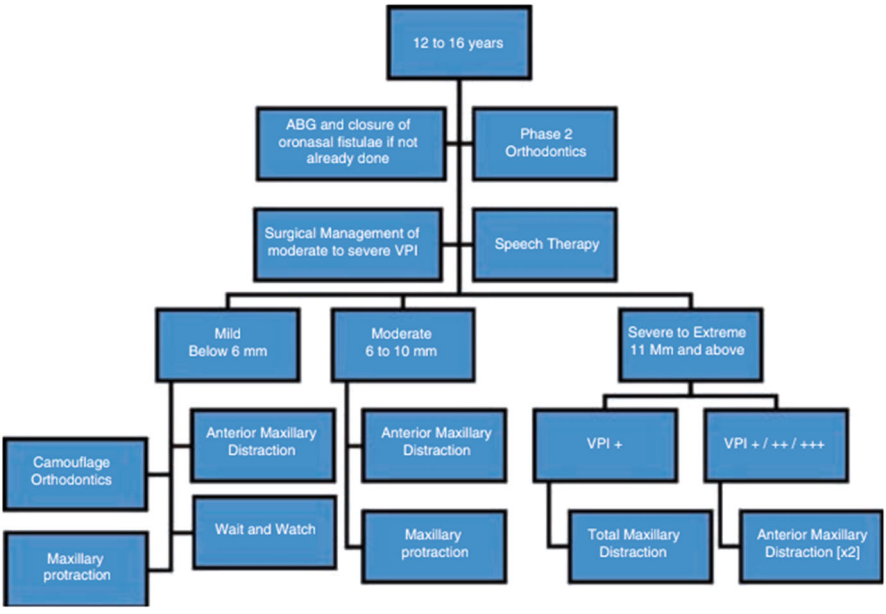
1. **Distraction Osteogenesis** can either be total distraction of maxilla or distraction only of the anterior maxilla.
2. **Conventional Orthognathic Surgery**

Treatment Plans

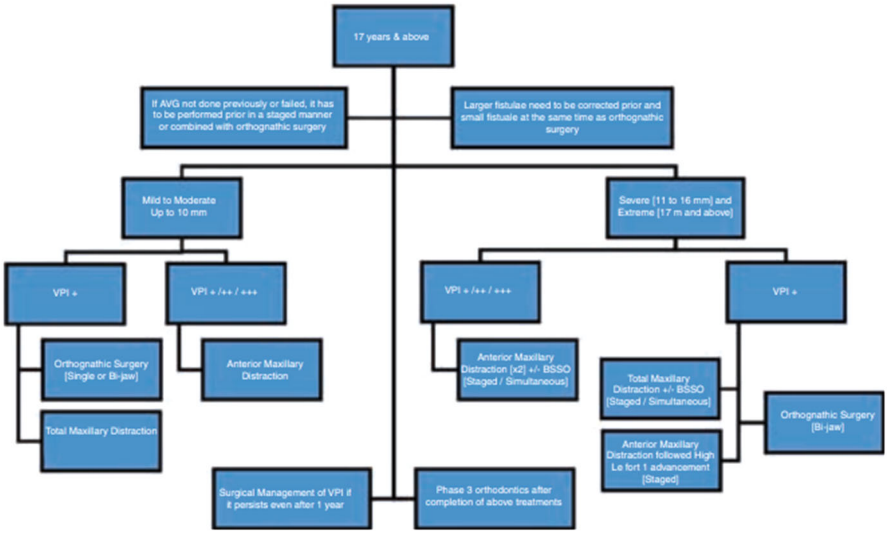
- For age 6–12 years



- For age 12–16



- For age 17 and above



- **LeFort I Procedure**

  In unilateral cleft cases, a molar-to-molar incision is placed in the vestibule, without violating any tissue distal to the mesial surface of the upper first molars.

  When there is bilateral CLP, the incision is staggered, i.e., first molar to lateral incisor on both sides of the midline. The nasal septal osteotome is placed by making an incision in the midline. This is to protect the pre-maxillary segment's blood supply, which is mostly derived from the upper lip.

  Before undergoing a LeFort I osteotomy, the nasal mucosa is usually raised off the nasal cavity's floor and walls. The nasal mucosa in the cleft maxilla is united to the palatal mucosa as a result of the prior palatoplasty. With extreme caution to prevent piercing the palatal mucosa, this tissue should be carefully dissected in the vicinity of the nasal floor. Additionally, this phase facilitates the mobility and down-fracture of the maxilla. There is typically paranasal hollowing in the cleft maxilla. For this reason, instead of the osteotomy a rather high LeFort I osteotomy is employed. Using a pterygoid chisel, also known as a curved osteotome, one must position it downward, medially, and anteriorly to perform dysjunction. To achieve adequate separation of thicker pterygomaxillary junction, a greater magnitude of force is required. The choice between a pterygomaxillary disjunction and a third molar vertical osteotomy cut is another crucial factor. Following pterygomaxillary disjunction, the maxilla is down fractured with the aid of leverage or specialized tools such as the Smith's spreader. For the nasal lining to remain continuous after the down-fracture, it is crucial to suture the separated nasal mucosa's edges. After the down-fracture, maxilla is mobilized

gradually with careful manipulation of scarred tissues. Once mobilization is achieved, the maxilla is advanced passively to its planned position. If surgeons face any resistance, the steps mentioned should be revisited and if it still does not remove resistance, then maxillary distraction or mandibular setback should be considered to avoid any relapse after surgery. Once desired movement is achieved, maxilla should be secured in new position with rigid fixation methods. In cleft cases, a short tight lip is commonly encountered which worsens with maxillary advancement, so a V-Y lip lengthening is advocated by some surgeons.

- **Distraction Osteogenesis**

  Distraction osteogenesis is being used in cleft maxillary advancements for correction of large anteroposterior discrepancies.

Part I: External Distraction
Face Mask Distraction: used to advance maxilla that has been osteotomized previously, usually a Le Fort I osteotomy. Distraction forces are usually provided by elastic bands placed between the anterior hooks which are soldered on an intraoral arch, with the hooks on the mask's arch supported by the chin and forehead.

Part II: Internal Anteroposterior Distraction
The use of internal micro distractors for maxillary distraction in cleft patients was initially documented by Cohen et al. in 1997. Since then, ongoing efforts have been focused on creating miniature internal multidirectional devices that can replicate the optimal and appropriate three-dimensional movement of the maxilla as precisely and consistently as possible. The primary benefit of internal distractors over exterior ones is their reduced heaviness, which makes them more socially and psychologically acceptable. The documented disadvantages of this method are:

1. Within the distraction phase, unable to alter vectors
2. Difficulty in achieving parallelism of bilateral distractors
3. Stretching of tissue leading to discomfort
4. Narrow therapeutic range
5. Need for surgery upon completion of consolidation for removal of hardware

Case Scenarios

See (Fig. 6)

Cleft Nose Deformity

Huffman and Lierle classified Cleft nasal deformities as follow:

1. The tip of the nose is deviated to non-cleft side.
2. Dome of the nose is displaced dorsally.

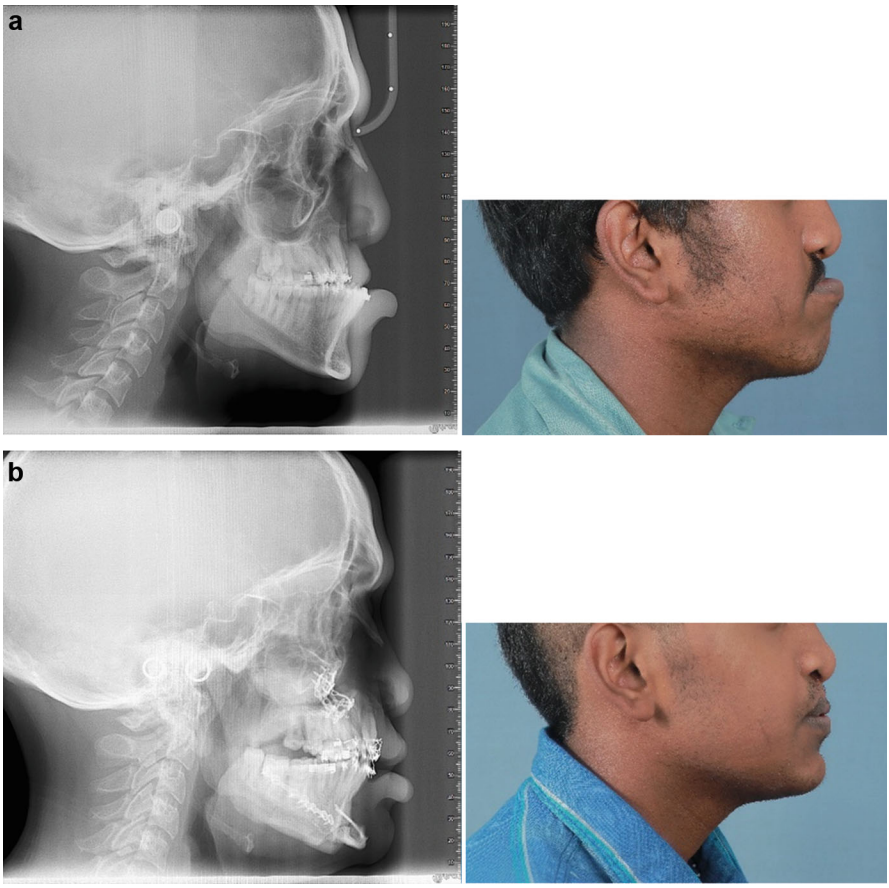


Fig. 6 A case of severe maxillary hypoplasia secondary to cleft. (**a**) Preop Lateral cephalogram and profile, (**b**) post-surgical correction. Lefort I advancement + BSSO setback with advancement genioplasty was done for this patient

3. Alar cartilage tipped caudally.
4. Alar buckles inwards on the cleft side.
5. Alar facial groove on the cleft side is absent.
6. Alar facial attachment is at an obtuse angle.
7. Real or apparent bone deficiency of maxilla on cleft side.
8. Circumference of naris is greater on the cleft side.
9. Naris on the left side is retrodisplaced.
10. Columella is shorter on the anteroposterior dimension on the cleft side.
11. Medial crus is displaced on cleft side.
12. Columella is positioned obliquely with dorsal ends slanted towards non-cleft side (Cuzalina and Jung 2016; Fatone et al. 2013).

Objectives of Surgical Correction

Functional Requirements

- Patent airway
- Proper position of maxilla to achieve Class 1 occlusion
- Normal speech

Esthetic Requirements

- Nasal symmetry with improved nasofacial and nasolabial relationship

Timing of Surgery

- Primary Rhinoplasty: 3 months of age (with lip repair)
- Intermediate Rhinoplasty: 4–6 years of age
- Secondary or Definitive Rhinoplasty Correction:
  14–16 years in females
  16–18 years in males

Preoperative Evaluation

Physical examination	External examination Internal nasal examination
Radiographic assessment	Cephalometric analysis—Evaluates the position of maxilla and its possible implications on nose of the patient Computed tomography of paranasal sinuses
Facial casts	Evaluate nasal morphology by direct anthropometry
Functional assessment	Rhinomanometry

Surgical Approaches

- Open approach (primary/definitive)
- Closed approach (primary only)

Surgical Technique

Primary Rhinoplasty

Unilateral Cleft Nasal Deformity

- Releasing the soft tissue attachments from the maxilla and pyriform aperture to allow for alar base repositioning

- Tip rhinoplasty, if indicated
- Repositioning of antero-inferior septum
- Alar basal levelling utilizing V-Y advancement
- Repositioning of lower lateral cartilage (LLC) using nasal bolsters/trans-nasal sutures

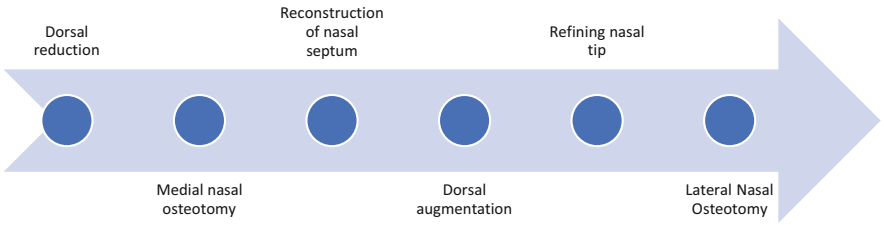
Bilateral Cleft Nasal Deformity

Secure nasalis to the septum on both sides, allowing for symmetrical repositioning of alar base.

Definitive Rhinoplasty

Approach: A combination of marginal incisions placed on both sides is used alongside an inverted V incision placed in the middle of the columella (Sykes et al. 1993; Tajima and Maruyama 1977).

Sequence of Steps



Dorsal Reduction: Performed for asymmetry correction and contouring.

- Dorsal Hump Correction: rasping for bone reduction, trimming of upper lateral cartilage using fine dissecting scissors (Gunter et al. 1997)
- Paramedian osteotomies to reduce width

Nasal Dorsal Augmentation

- Septal cartilage is a good candidate for use in augmentation.
- Autogenous cartilage graft is the first choice in rhinoplasty because of its resistance to resorption and infection, such as septal, auricular, or rib (costal) cartilage (Witherow et al. 2002; Tosun et al. 2008).
- Silicone and porous polyethylene have also been used.

Septal Reconstruction

Medial crura should be separated from the septum. A 1 cm L-shaped strut should be left behind during harvest from the septum.

For severe deviation, caudal septal extension graft serve well as dorsal marginal struts. Sutures are used to anchor the caudal L-strut to the anterior nasal spine.

Nasal Tip

Usually needs rotation and projection.

- Columellar strut grafts
- Septal extension grafts
- Shield graft

Alar Rim (the Lateral Crus of Lower Left Cartilage)

- Horizontal mattress sutures
- Alar batten graft (underlay or onlay)

Nasal Alae

Weir procedure for lateral symmetry and V-Y advancement

Postoperative Care

Intra nasal splint can be utilized for younger patients for up to 2 months. External nasal splints, preferred for definitive rhinoplasty, can be used for up to 2 weeks.

Case Scenarios

See (Fig. 7)

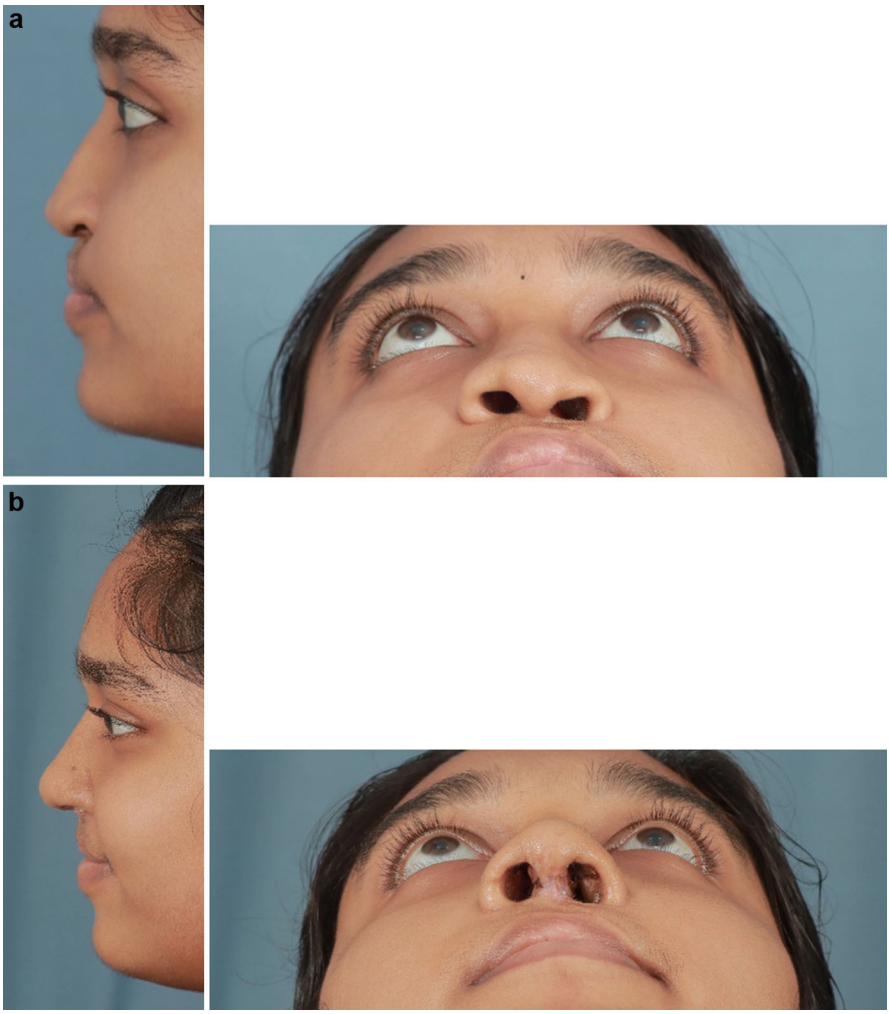


Fig. 7 Nasal Deformity secondary to cleft. (**a**) Figure shows depressed dorsum and no nasal tip support, depressed ala on the cleft side, reduced columellar height and support. (**b**) Photo post surgical correction. Rhinoplasty done using Rib Graft. Strips of graft used for dorsal augmentation with septal extensive and columellar struts for tip and columellar correction. Cleft side lateral crus shaped using graft

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