9. Bilateral Cleft Lip Repair

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ANATOMY

- In the bilateral cleft lip, the orbicularis oris muscle inserts on both alar margins, and no muscle fibers invade the prolabium.
- Unrestrained growth of the vomer and nasal septum result in protrusion of the premaxilla.
- The prolabial skin is flat, lacking philtral ridges, philtral dimple, & Cupid's bow.
- The columella is very short, and both lateral crura are flattened, resulting in alar flaring.

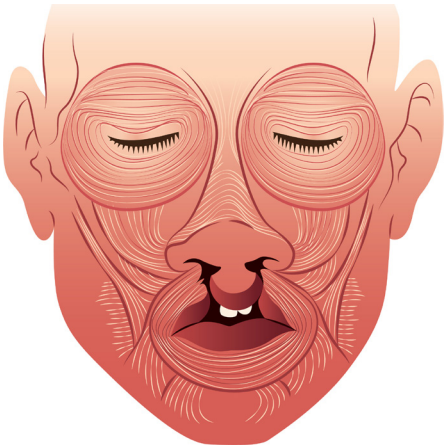


Figure 9-1. Anatomy of bilateral cleft lip. © 2017 A Campbell, C Restrepo

- The advantage of the bilateral cleft lip is symmetry.

INCOMPLETE BILATERAL CLEFT LIP

- Occasionally, bilateral clefts are incomplete with a near-normal nose, a normally positioned premaxilla, Simonart bands across the nasal floors, and clefts involving only the lip.
- Many patients have complete clefts on one side and incomplete clefts on the other.

Figure 9-2. Bilateral incompletecleft lip (A) and bilateral complete/incomplete cleft lip (B). © 2017 A Campbell, C Restrepo

- These cases have both the nasal deformity of a unilateral complete cleft lip and the paucity of lip tissue of a bilateral cleft.

COMPLETE BILATERAL CLEFT LIP

Figure 9-3. Bilateral cleft lip. © 2017 A Campbell, C Restrepo

- The most obvious aspect of a complete bilateral cleft is the protruding premaxilla. Because of the lack of connection of the premaxilla with the lateral palatal shelves, the premaxilla has not been held back into alignment with the lateral arch segments during fetal development. Uncontrolled growth at the premaxillary suture results in overprojection of the premaxilla, with or without rotation and angulation of the segment.
- The lateral palatal shelves are not pulled forward by their attachment to the premaxilla. Without the intervening premaxilla to maintain arch width, the lateral palatal shelves collapse toward the midline.
- The severity of this disruption of arch morphology varies, and will dictate the tension on the repair, the degree of dissection required, and, ultimately, the final aesthetic result unless it is corrected with presurgical orthopedics.
- The anterior nasal spine is poorly formed or absent in the bilateral cleft lip deformity, resulting in a retruded area under the base of the septal cartilage and recession of the footplates of the medial crura.
- The footplates of the lower lateral cartilages are displaced posteriorly and laterally, which in turn pulls the normal junction (genu) of the medial and lateral crura apart resulting in a broad, flat nasal tip. The recession of the medial crural footplates, along with lateralization of the domes, and deficient skin, produces the typical "absent columella."
- The most anterior and inferior extent of the frontonasal process,

which normally contributes to the skin between the philtral columns of the lip, forms a wide, short disk, called a prolabium, that appears to hang directly from the nasal tip skin.

COMPLETE BILATERAL CLEFT LIP, ALVEOLUS, & PALATE

- Most common of the bilateral clefts and is usually asymmetric
- Premaxilla protrudes and deviates to one side, and may also be rotated and displaced in the vertical dimension.
- The palatal cleft is usually symmetric.
- The size of the premaxilla varies greatly and is rarely found in the alveolar arch.
- Because the premaxilla is protruded, the prolabium seems to be attached to the tip of the nose.
- Variation in the complete bilateral cleft may be related to asymmetry of the cleft thath include complete cleft lip, alveolus and palate on one side and and cleft of the lip only in the other. Bilateral cleft lip and bilateral cleft of the palate can occur without cleft in the alveolus. The cleft may also include an asymmetric bilateral cleft lip, unilateral cleft alveolus, and bilateral cleft palate.

Figure 9-4. Bilateral complete cleft lip involving skin, muscle, mucosa, alveolus, primary and secondary palate. This patient also has a protrusive premaxilla.

NASAL DEFORMITY IN BILATERAL CLEFT LIP

- The nasal deformity associated with complete bilateral cleft lip, alveolus, and palate is related to the position of the premaxilla, prolabium, and maxillary segments and to symmetry of the cleft.
- Severe protrusion of the premaxilla adds to the complexity of the nasal deformity with the prolabium attached almost to the nasal tip with little or no columella.
- Each alar base is pulled laterally with abnormally wide position of the bases of the alae that results in deformity of the lower lateral cartilages with the lateral crus elongated and the medial crus shorter than normal.
- The nasal tip is flat and the distance between the alar domes is too wide. The simmetric nasal deformity is easier to correct than the asymmetric deformity.
- In the symetric one the septum is usually at midline and in the asymmetric clefts with a protruded premaxilla, the septum is deviated.

PRESURGICAL ORTHOPEDICS

Nasoalveolar Molding: (NAM)

- NAM utilizes wire and acrylic nasal stents attached to an intraoral denture.
- NAM prepares the lip and nose before the initial cleft lip and nasal repair, and can be used with unilateral and bilateral cleft lip. In effect, this presurgical management of the cleft infant is intended to reduce severity of the oronasal deformity prior to surgery.

Figure 9-5. NAM device in place with tape and rubber bands. .© 2017 A Campbell, C Restrepo

- Molding is initiated within the first couple of weeks of life to restore the correct skeletal, cartilaginous, and soft tissue relationships presurgically.
- Molding continues for approximately three months in unilateral cleft lip and often for 6 months for bilateral cases.

- The NAM appliance is used to mold the nasal cartilages, premaxilla, and alveolar ridges into normal form and position during the neonatal period. NAM is labor-intensive, requiring weekly visits and adjustments by a dedicated and experienced orthodontist as well as equally dedicated and compliant parents .
- NAM is time consuming and expensive, and especially challenging for patients in developing regions.

Figure 9-6. NAM utilizes wire and acrylic nasal stents attached to an intraoral denture.

External Elastic Bands / Taping

- Alternative presurgical treatment to treat protrusive premaxilla when NAM unavailable.
- Requires periodic visits to treating professional (dentist, orthodontist, surgeon) for monitoring and adjustment.

Figure 9-7. External elastic bands / taping.

SURGICAL MANAGEMENT OF BILATERAL CLEFT LIP

One Stage versus Two Stage:

- One stage - Facilitates creation of a symmetric, balanced lip. With adequate mobilization of tissues one stage repair is possible in nearly all bilateral clefts.
- Two stage - One side is closed first, allowed to heal, and then the other side is repaired a short time later. Symmetry is difficult to achieve with a staged approach, and we advocate a single-stage procedure.

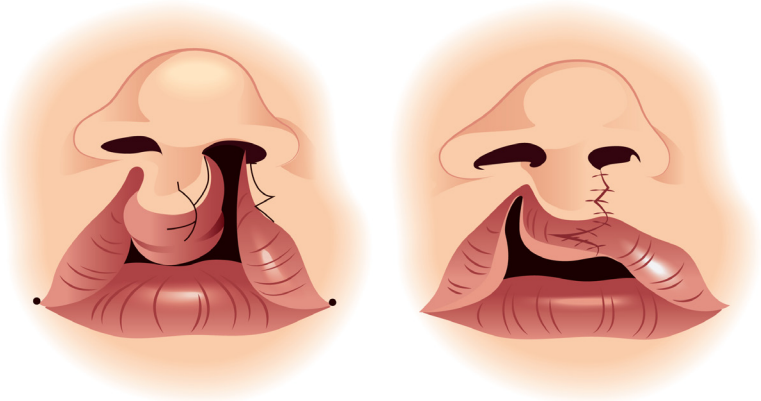


Figure 9-8. First stage in two-stage bilateral cleft repair. © 2017 A Campbell, C Restrepo

Surgical Techniques:

Modern principles that guide the repair of the bilateral cleft lip:

- Symmetry
- Primary muscular continuity
- Proper philtral size and shape
- Formation of the median tubercle from lateral lip elements
- Creation of adequate sulcus
- Preventing or minimizing tension is the key to successful bilateral cleft repair. Excessive tension causes tethering of the columella, increased scarring, widening of philtrum, and facial growth problems.

Lip Adhesion:

- Lip adhesion utilizes interdigitating flaps from the cleft margin from tissue normally discarded during lip repair.
- Designed to convert a complete cleft into an incomplete cleft, to mold the maxillary dental arch, and to reduce tension in the lip so that definitive repair can occur under more ideal conditions.

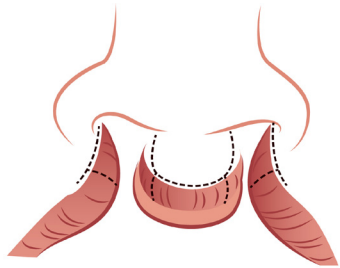


Figure 9-9. Lip adhesion markings.© 2017 A Campbell, C Restrepo

- Second stage definitive lip repair with repair of the muscle beneath the prolabium and narrowing of the prolabium after molding of

the premaxilla and lateral maxillary arches has occurred.

- Not widely used because there is an increase in scarring, with no real benefit. After the resection of the scar at second stage the tissue deficit could be the same or increased.
- Modern techniques allow for closure of nearly all bilateral cleft lip deformities without lip adhesion.

Manchester (Straight Line) Repair

- Uses the prolabium for the entire central portion of the lip, based on concept that there is adequate vermilion in the prolabium.
- Joins the medial aspect of the vermilion and white roll to the prolabium, thereby avoiding lateral segment sacrifice.
- Often results in an inadequate tubercle and central depression of the upper lip vermilion.
- Failure to reconstruct orbicularis muscle in midline results in philtral deformity most marked on animation.

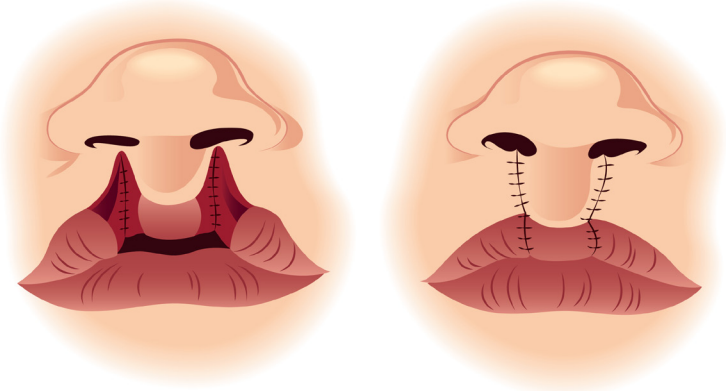


Figure 9-10. The Manchester technique for bilateral cleft lip repair. © 2017 A Campbell, C Restrepo

Millard Repair

- Based on concept that there is inadequate white roll and dry vermilion in the central prolabial segment, and these elements are thus recruited from the lateral lip segments.
- Technique advances lateral vermilion flaps beneath the prolabium and reconstructs the orbicularis muscle beneath the elevated prolabium in the midline. Reduction of the prolabium to a smaller philtral dimension.

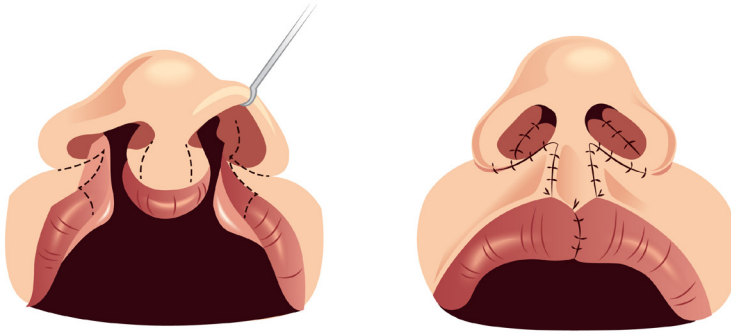


Figure 9-11. The Millard technique for bilateral cleft lip repair. © 2017 A Campbell, C Restrepo

Trott Repair

- Single stage repair. Design of prolabial flap with upper width the same width to the columellar base. The alar base flap forms the sill. The prolabial columellar flap is elevated and an open rhinoplasty is performed to reposition alar cartilages.

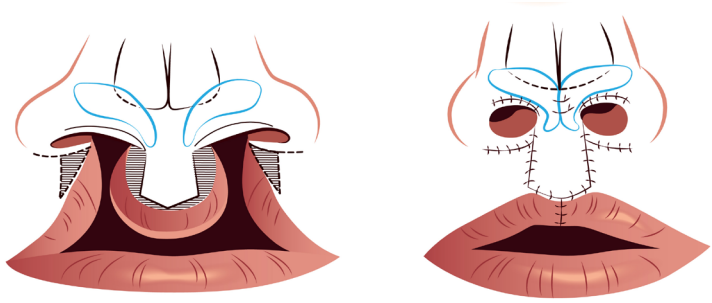


Figure 9-12. The Trott technique for bilateral cleft lip repair. © 2017 A Campbell, C Restrepo

Mulliken Repair

- One-stage lip and nose repair incorporating design of a narrow, concave prolabial flap. Based on author's documented prolabial widening to be as much as 250% over a 3-year period.
- Approach to nose based on observation that there is no deficiency of columellar skin and no need to recruit tissue from the lip or sills.
- The surgical strategy is symmetrical labial repair and synchronous anatomic positioning of the alar cartilages with sculpturing/ draping of the nasal soft tissues.

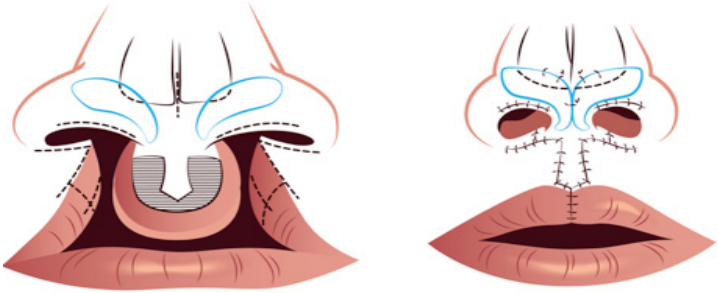


Figure 9-13. The Mulliken technique for bilateral cleft lip repair. © 2017 A Campbell, C Restrepo

Cutting

- Recommends presurgical orthopedic molding of the lip, alveolus, and nose combined with a one-stage lip, alveolus, and nose repair. Presurgical orthopedics aligns the protruding premaxilla with the maxillary arch and expands the tissues of the nasal columella and nasal lining with acrylic stents.
- Describes a prolabial unwinding flap for one-stage repair of the bilateral cleft lip, nose, and alveolus: "The columella of the nose and the central lip are produced by 'unwinding' the columellar and labial sections of the prolabium around a small central tab, which is used to center the junction between the lip and columella."
- Open approach to nasal-tip cartilages - Philtral-medial crural-columella complex is reflected by retrograde dissection to display the underside of the splayed middle crura and genu.

Figure 9-14. Cutting technique for bilateral cleftlip repair. © 2017 A Campbell, C Restrepo

Byrd

- Lateral nasal release, augmentation of nasal lining, and reconstruction of the external valve and muscle ring across the sill is performed as part of the lip repair. Columellar lengthening and domal

unification is delayed until 18 months of age.

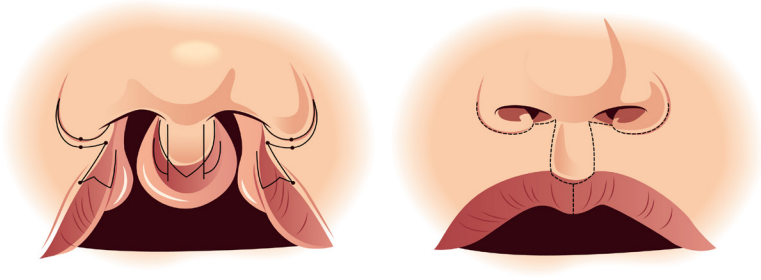


Figure 9-15. Cutting technique for bilateral cleft lip repair. © 2017 A Campbell, C Restrepo

Secondary Deformities After Bilateral Cleft Lip Repair

- Protruding/deviated/rotated premaxilla
- Collapsed maxillary segments
- Absence of gingivolabial sulcus
- Nasolabial fistulas
- V shaped whistle deformity
- Bulging of the orbicularis oris
- Wide prolabium
- Immobile prolabium
- Short columella
- Flat nasal tip
- Wide position alar base

Figure 9-16. Secondary deformities after bilateral cleft lip repair. © 2017 A Campbell, C Restrepo

SURGICAL REPAIR OF BILATERAL CLEFT LIP

Figure 9-17. Patient with complete bilateral cleft lip and palate with protruding premaxilla. © 2017 A Campbell, C Restrepo

Patient Position

- Head at end of table on doughnut
- ET tube taped down midline lower lip
- Neck extension, shoulder roll
- Mouth pack (silk suture tag)

Figure 9-18. Patient with complete bilateral cleft lip and palate intubated with oral RAE tube taped in the midline inferiorly. Surgeons should take standardized preoperative anterior-posterior (AP) and inferior (worm's eye) views. © 2017 A Campbell, C Restrepo

MARKINGS

- Toothpick, methylene blue, 5cc syringe, 27 gauge needle, caliber, alcohol wipes, gauze
- Precise marks with fine points
- Tattoo marks with Methylene blue before infiltration of local anesthetic
- Incisions designed along key points medial and lateral lips

Figure 9-19. Marking is performed with precision and tattooed with a fine needle. © 2017 A Campbell, C Restrepo

Marking the Prolabium:

1. Midline of the prolabium is marked with the center point at the mucocutaneous junction.
2. From this point 2.5 – 3 mm on each side , both peaks of the future cupid's bow 's are marked at the mucocutaneous junction. The width of the tubercle is determined by the width of the columellar base
3. At the base of the columella the midline is marked and from this point at each side is marked a point in each foot of the medial crus .
4. A symmetrical point is marked at each side from the foot of the medial crus to the junction of the skin of the columella with the vestibule over the columnela lip sulcus.
5. A line is drawn from each point of the future peak of the cupid's

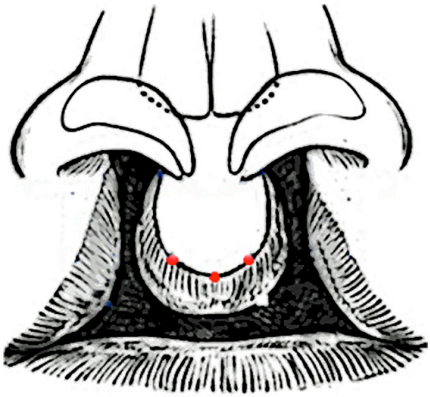


Figure 9-20. The midline and peaks of Cupid's bow are marked at the vermillion cutaneous junction. © 2017 A Campbell, C Restrepo

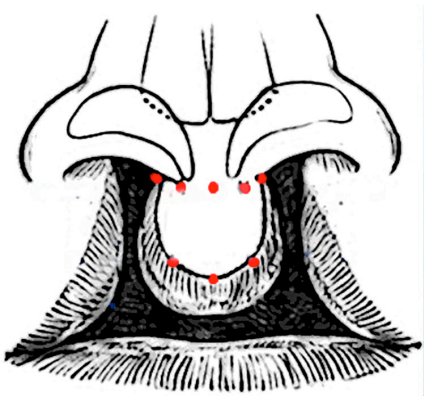


Figure 9-21. Points at the midline, columellar base, and skin/vermillion junction. © 2017 A Campbell, C Restrepo

bow to the lateral points at the base of the columnella on each side. The prolabium at the level of the cupid's bow is slightly wider than at the columella. The prolabium is design for the central portion of the upper lip which will be placed between vertical straight line scars that will simulate the philtral columns.

Marking the Lateral Lip:

1. On each lateral lip portion, the peak of the future cupid's bow is marked where the white roll disappears, measurements from this point to the alar base are adjusted to the same length marked on the prolabium. If there is a discrepancy between the lengths of those two lines the length of the lateral lip element must be adjusted to establish a proportional lip height.
2. From the point in the peak of the future cupid's bow, medially is marked a point in the mucocutaneous junction at the same distance than the middle point of the prolabium to the future peak of the cupid's bow at each side
3. Bilateral vermilion advancement flaps are then marked on the lateral lip segments. These areas of dry vermilion are recruited from the lateral lip to fill the central tubercle during the repair. The length of the vermilion advancement flaps should be slightly greater than one-half the prolabium width. The planned incisions for these flaps are marked on the dry vermilion in a medial direction, just parallel to and off the white roll.
4. "V" flaps are designed on bilateral lateral vermillion flaps to add more tissue for central closure and to prevent central whistle deformity. One side "V" flap designed superiorly and contralateral "V" flap designed inferiorly.
5. Marking at ala nasi external and internal. A point is marked in the mucocutaneos junction at the base of the nostril where the white line and the bulk of the orbicularis disappear: at the junction of the vestibule skin with the lip skin. The tissue above this point will form the nasal floor.
6. The distance between external ala nasi and the point at the junction of the nasal vestibule skin with the lip skin is compared in both sides to make the nasal floor symmetrical.
7. Marking of the junction of the vestibular skin and the intranasal

mucosa in the medial and lateral side. After the reconstruction of the floor these points must come together.

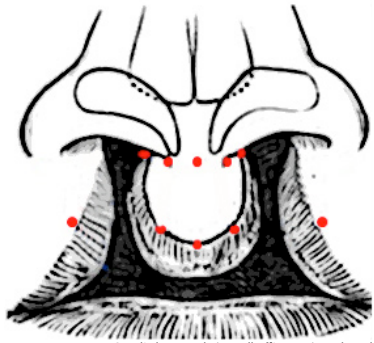


Figure 9-22. Future Cupid's bow peak (Noordhoff's point) on lateral lip elements. © 2017 A Campbell, C Restrepo

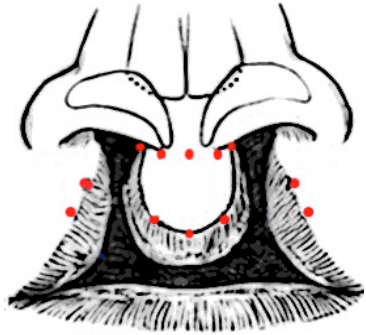


Figure 9-23. Vermillion flaps designed medial to Noordhoff's point. © 2017 A Campbell, C Restrepo

Figure 9-24. Red line marled at junction of dry and wet vermillion. © 2017 A Campbell, C Restrepo

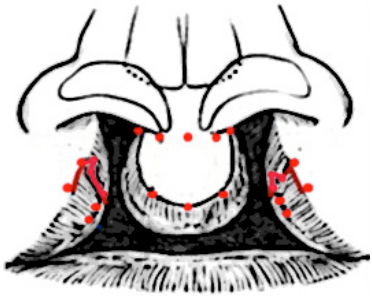


Figure 9-25. Lateral vermillion "V" flaps. © 2017 A Campbell, C Restrepo

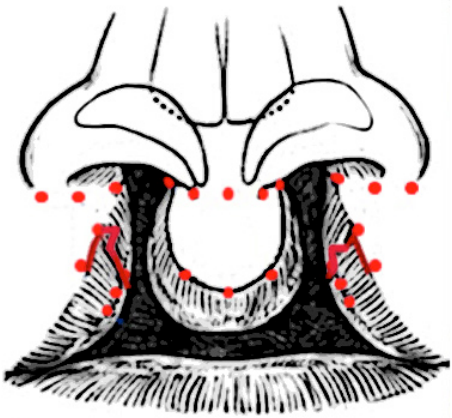


Figure 9-26. Alar markings lateral lip elements. © 2017 A Campbell, C Restrepo

Figure 9-27. Preoperative patient markings © 2017 A Campbell, C Restrepo

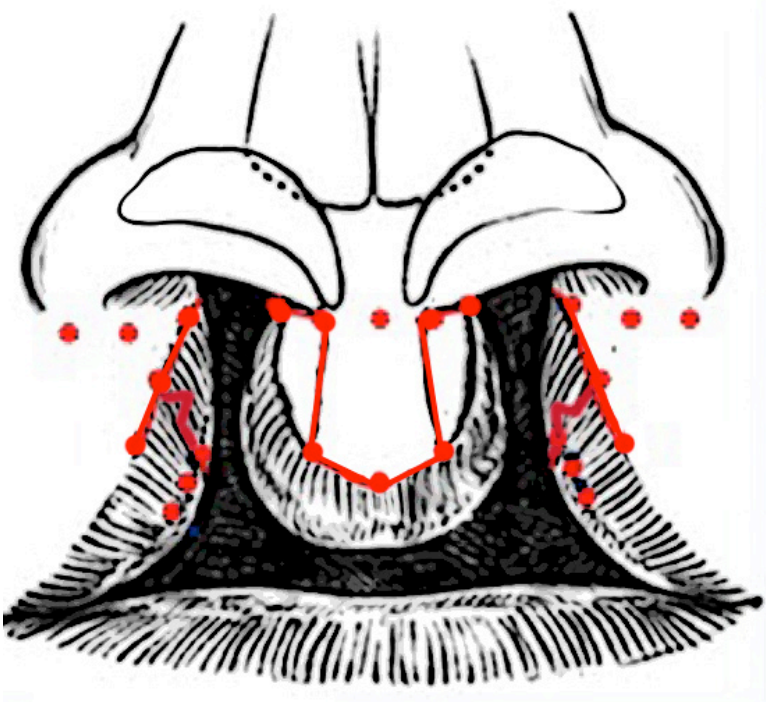


Figure 9-28. Final markings and incisions for bilateral cleft lip repair with author's technique.© 2017 A Campbell, C Restrepo

OPERATIVE SEQUENCE

Infiltration

- Wide and controlled infiltration of local anesthetic to dissection areas
- 0.25% Lidocaine, 0.125% Marcaine, 1:100,000 Epinephrine
- Always calculate maximum dosage prior to injection and confirm with anesthesiologist.
- Inject all areas of dissection adequately but without distending and Areas of injection:
    - Bilateral lip elements at cleft margin
    - Bilateral sulcus incisions
    - Bilateral maxillary and piriform regions
    - Prolabium
    - Bilateral nasal septum

- Bilateral infraorbital nerve block performed at end of case
- After injection, surgeon scrub and patient preparation for ample time for vasoconstriction

Figure 9-29. Wide and controlled infiltration of local anesthetic to dissection areas lateral lip elements (A-D), phitrum (E), and septum (F).

Lateral Lip Incisions

- 15 blade is used for londer cuts on the skin and mucosa.
- 11 blade is used for fine cuts in vermillion.
- Incisions made in lateral lip following markings in the skin, vermillion, and mucosa. The incision is made at the vermillion-cutaneous junction, dissecting the vermillion to the point where the peak of cupid's bow is marked.
- Full thickness cuts are made through the orbicularis oris and the oral mucosa of the upper lip.
- Lateral lip dissection is carried out so that existing vermillion is used to reconstruct the central vermillion element. Vermillion "V" flaps are elevated from each lateral lip element to maximize conservation of of vermillion in order to avoid a central deficiency and whistle deformity.

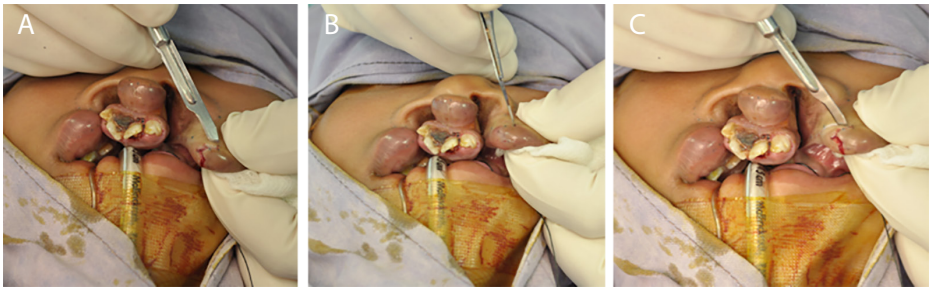
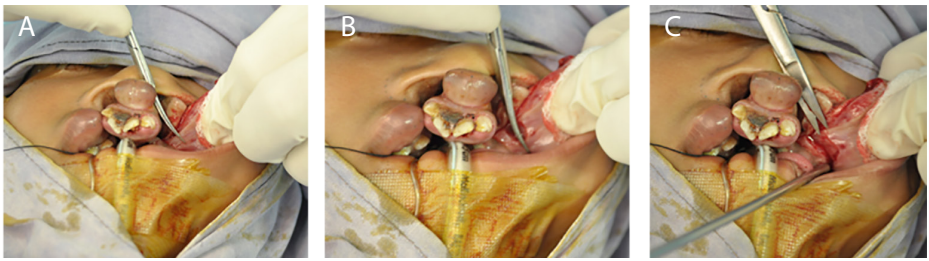


Figure 9-30. Incisions lateral lip element. © 2017 A Campbell, C Restrepo

Figure 9-31. Elevation of lateral "L" flaps at cleft margin. These are discarded or used for augmentation at the piriform. © 2017 A Campbell, C Restrepo

Lateral Lip Dissection

- Incision of the mucosa in the sulcus to detach and advance lateral lip element. Extent depends on cleft width.
- Mucosa elevated in plane between minor salivary glands and muscle.
- Muscle released from piriform aperature at maxilla.
- Minimal dissection (~1mm) between muscle and skin to allow for eversion of skin on final closure.



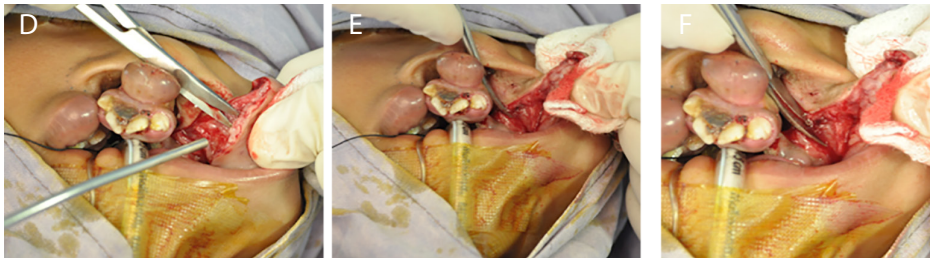


Figure 9-32. Retraction of lateral lip element (A), buccal sulcus incision (B), submucosal dissection in plane between muscle and minor salivary glands (C,D), and release of abnormal muscle insertion from piriform aperature (E, F). © 2017 A Campbell, C Restrepo

- Subperiosteal dissection to expose piriform aperature and to provide adequate mobilization and to adequately advance lateral lip elements to minimize tension.
- Subperiosteal dissection and release can be carried up to the inferior orbital nerve, allowing the lateral lip segment to be advanced to the midline.
- In wide clefts, a backcut at the lateral extent of the sulcus incision and periosteal scoring can be performed to maximize mobilization. Care of parotid (Stenson) duct laterally!

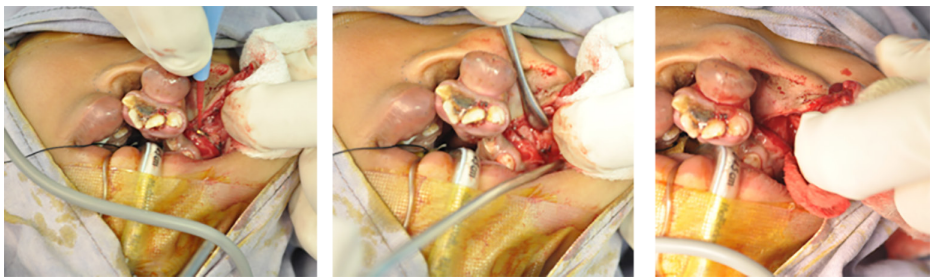


Figure 9-33. Subperiosteal dissection to expose piriform aperature and to provide adequate advancement lateral lip element. © 2017 A Campbell, C Restrepo

- The alar base is tethered to the piriform aperature by muscular, ligamentous, and lining attachments through fibrous attachments with the accessory chain of the lateral crus
- A full-thickness cut is made along the nasal lining at the pyriform aperture to release these attachments from the pyriform rim and maxilla up to the level of the nasal bones.
- This release creates a lining defect that will be filled with the septal flap and/or lateral "L" flaps.

- Dissection of mucoperiosteal flap from lateral nasal wall.

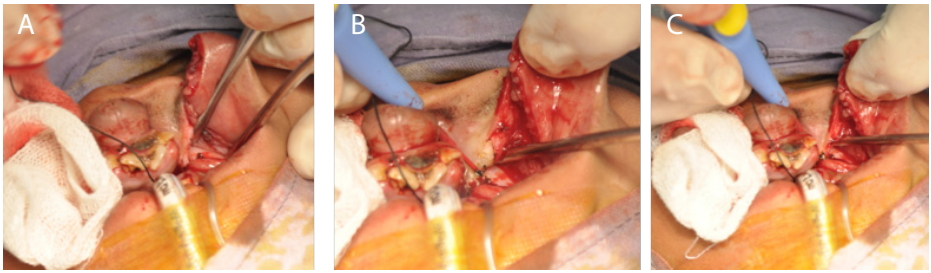


Figure 9-34. Subperiosteal exposure of piriform aperature (A) followed by a full-thickness, transmucosal cut along the nasal lining at the piriform aperture (B, C) to release abnormal attachments up to the level of the nasal bones. © 2017 A Campbell, C Restrepo

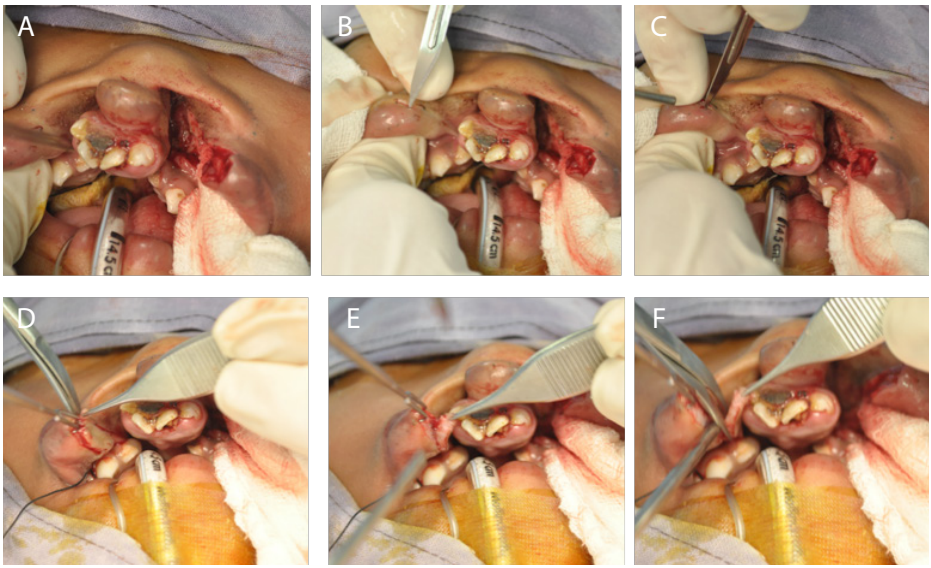


Figure 9-35. The dissected side is packed (A). Lateral lip incisions are made on contralateral side (B,C), and the lateral "L" flap is elevated (D-F). © 2017 A Campbell, C Restrepo

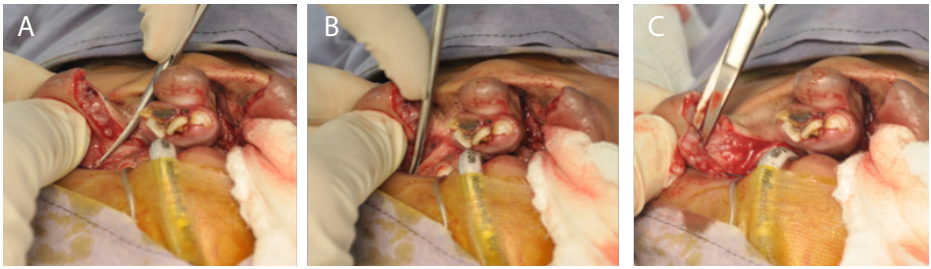


Figure 9-36. Buccal sulcus incision (A,B), submucosal dissection in plane between muscle and minor salivary glands (C). © 2017 A Campbell, C Restrepo

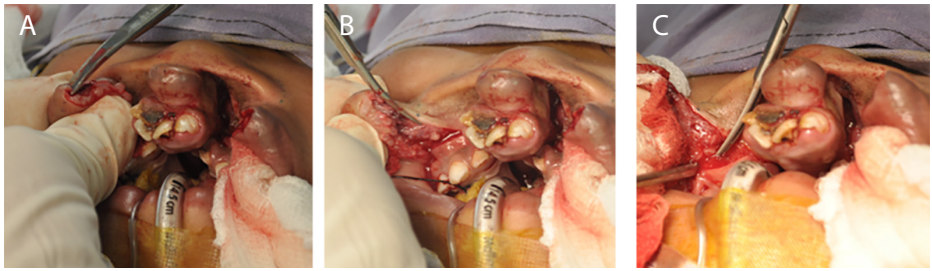


Figure 9-37. Release of "V" flap (A), minimal dissection (1-2mm) between skin and muscle (B), and release of abnormal insertions from piriform aperature (C).

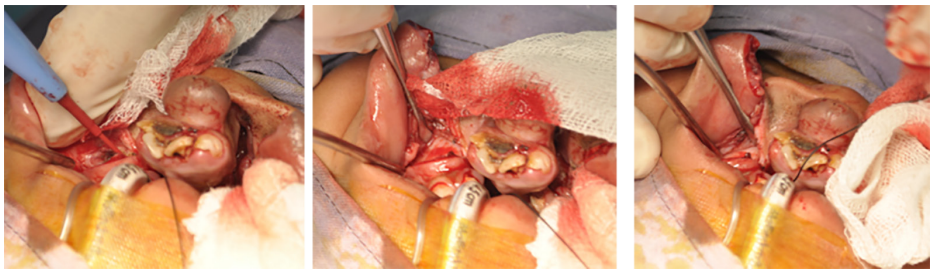


Figure 9-38. Subperiosteal dissection to expose piriform aperature and to provide adequate advancement lateral lip element.

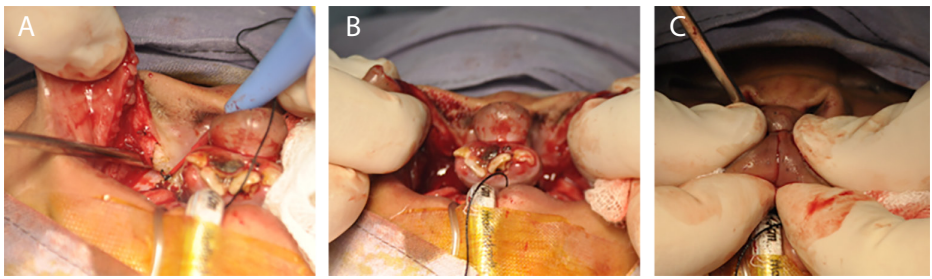


Figure 9-39. Full-thickness, transmucosal cut along the nasal lining at the piriform aperture to release abnormal attachments up to the level of the nasal bones (A). Bilateral lip elements mobilized adequately for closure without tension (B, C).

Prolabium Incisions

- Incision starts on the prolabium following the previously marked lines, creating a middle flap for philtrum reconstruction and two lateral segments that are are discarded.
- Philtral flap is dissected to the base of the columella, leaving a generous amount of deeper soft tissues attached.
- The central dermis of the philtral flap at the base of the columella can be sutured down to the underlying muscle repair to create columellar angle a.

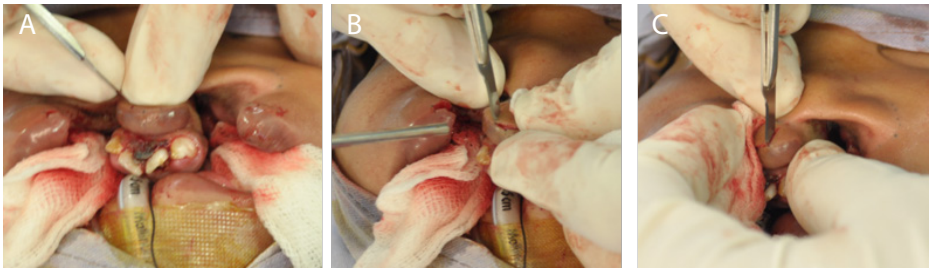


Figure 9-40. Incisions on the prolabium. © 2017 A Campbell, C Restrepo

Septal Flap Dissection

- Dissection of bilateral mucoperichondrial flaps from nasal septum for nasal floor reconstruction in complete clefts.

Sulcus Reconstruction

- The central mucosa segment is trimmed for sulcus reconstruction.

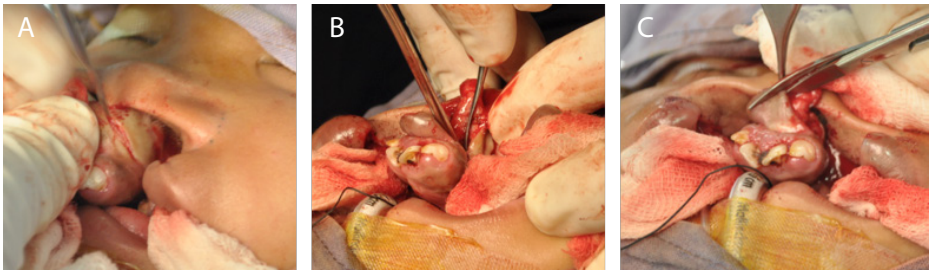


Figure 9-41. Bilateral mucoperichondrial septal flaps are elevated (A,B) for nasal floor reconstruction, and the central mucosa segment is trimmed for sulcus reconstruction (C). © 2017 A Campbell, C Restrepo

Reconstruction / Closure

<u>Nasal Floor</u>

- The floor of the nose is reconstructed to create a division between the oral and nasal cavities, and also facilitates repositioning of the maxillary segments consequently narrowing the palatal cleft.
- The floor is formed from the mucoperiosteal flap from lateral nasal wall and the mucoperichondrial flap from nasal septum.
- This also helps to bring the base of the ala medially to assist with creation of proper symmetry by narrowing the width of the nostril.
- The alar bases are repositioned medially to the premaxilla.
- Reconstruct the alar muscle ring with a three-point alar cinch su-

ture to secure the muscle both alar bases to the premaxilla in the midline.

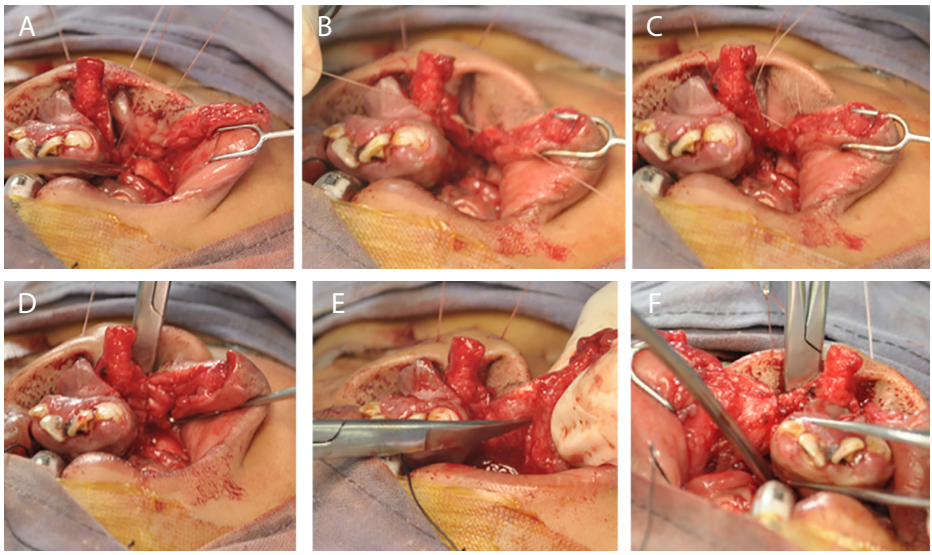


Figure 9-42. Nasal floor reconstruction. Placement of retraction sutures in bilateral soft triangles (A) of suture for exposure, and repair of septal flap to lateral nasal wall flap beginning at most posterior aspect (B) and continuing forward to nasal sill (C) for complete closure of nasal floor (D). Alar muscle ring reconstructed with a three-point alar cinch suture to secure the muscle at both alar bases (E) to the premaxilla in the midline (F).

Mucosa and Sulcus

- Reconstruction of the midline sulcus is performed by turning up the mucosa of the prolabium, establishing the lining of the labial sulcus and fixing it to the periosteum inferior to the nasal spine.
- Lateral lip element mucosal flaps are advanced medially and closed to the mucosal cuff above the gingiva
- Medial aspects of lateral lip element mucosal flaps are sutured to the mucosal flap of the prolabium, previously fixed, and sutured together in the midline from inferior to superior, starting in the red line (key stich).

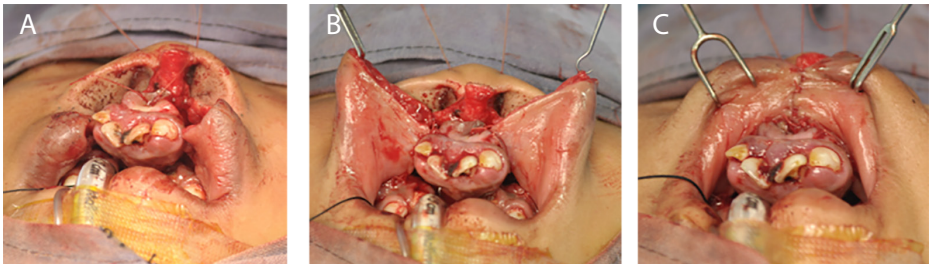


Figure 9-43. Reconstruction of the midline sulcus is performed by turning up the mucosa of the prolabium and fixing it to the nasal spine (A). The lateral lip element mucosal flaps are sutured to the mucosal flap of the prolabium and to each other in the midline to reconstruct the labial sulcus and lip mucosa. (B, C)

Muscle

- The ultimate goal of muscle repair is to bring the orbicularis oris into the region of the prolabium so the muscle can be sutured together in the midline under the skin of the prolabium.
- Bringing the muscle in to the region and freeing it from its abnormal attachment around the base of the nose is key in establishing a mobile animated lip with natural motion
- The musculocutaneous flaps from the lateral lips element are advanced medially and the muscle is brought across the midline below the prolabium.
- Apposition of the muscle is done from distal (key stich) to proximal.

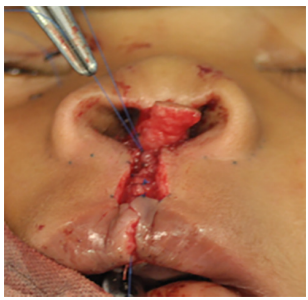


Figure 9-44. Muscle reconstruction beneath philtral flap.

Skin and Vermillion

- Preventing or minimizing lip tension is the key to successful bilateral cleft repair. Excessive tension gives rise to tethering of the columella and increased scarring and also contributes to the potential for subsequent facial growth problems.
- Attention is given to meticulous reconstruction of the Cupid's bow.
- Incisions along philtral columns are closed with running sutures to minimize inflammatory suture material
- The midportion of the vermilion is reconstructed by bringing lateral vermilion flaps below the philtral flap. Z plasty or lateral V-flaps can be incorporated to increase length of central vermillion and to break up vertical closure.

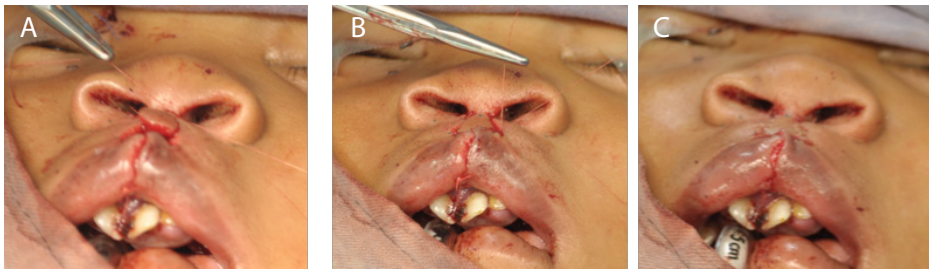


Figure 9-45. Key sutures are placed at the top and bottom of the philtral columns, and skin closure is accomplished with fine running sutures along the philtral columns. © 2017 A Campbell, C Restrepo

Figure 9-46. Bilateral infraorbital blocks are performed (A, B). Final closure (C). © 2017 A Campbell, C Restrepo

Figure 9-47. Preoperative and postoperative photographs of patient in demonstration. © 2017 A Campbell, C Restrepo

KEY READING

1. Chen P, Noordhoff S. (2009) Bilateral Cleft Lip and Nose Repair. In: Losse J, Kirschner R: Comprehensive Cleft Care (pp. 331-342). McGraw-Hill Companies.
2. Mulliken J. (2009) Mulliken Repair of Bilateral Cleft Lip and Nosal Deformity. In: Losse J, Kirschner R: Comprehensive Cleft Care (pp. 343-360). McGraw-Hill Companies.
3. Mulliken J. Principles and techniques of bilateral complete cleft lip repair. Plast Reconstr Surg. 1985 Apr;75(4):477-87.
4. Wolfe SA. A pastiche for the cleft lip nose. Plast Reconstr Surg. 2004 Jul;114(1):1-9.