Cleft Lip and Palate

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Pat Ricalde, Imran Ahson, and David E. Urbanek

Epidemiology/Embryology

- Cleft lip with or without cleft palate occurs in 1/940 live births [1].
- Cleft palate without cleft lip occurs in 1 in 1574 live births [1].
- Native Americans have the highest incidence at 1/500 live births [2].
- African Americans have the lowest incidence at 1/2500 live births [2].
- Cleft lip occurs more often in males (on the left side), and cleft palate occurs more in females. This is more likely to occur on the left side as there is a delay in the rotation of the palatal process of the maxillae on the left side.
- The incidence of isolated cleft palate is higher in females (3:2); 50% of isolated cleft palates are associated with a sequence or syndrome (e.g., Pierre Robin, Stickler, van der Woude, 22q deletion anomalies). Cleft lip with or without a cleft palate occurs with a 15% syndromic association [3].
- Formation of the lip, nose, and palate involves controlled proliferation, adhesion, apoptosis, and fusion of prominences. Failure of this process results in cleft formation.
  - 6 weeks gestation: Median nasal process fuses with maxillary process to form upper lip, philtrum of the lip, base of nose, and primary palate. Failure of fusion results in a clefting of the lip and/or alveolus.
  - 8–12 weeks gestation: Palatine shelves of maxillary processes merge in the midline to fuse with nasal septum/vomer to form secondary palate. This fuses from anterior to posterior. The degree of clefting is dependent on timing of disruption.
- The primary palate denotes the anatomy that is anterior to incisive foramen including the incisors, alveolus, and nasal spine. The secondary palate denotes all structures posterior to the incisive foramen including the soft/hard palates and the uvula.
- Complete cleft lips extend into the nares and the alveolus.
- Complete cleft palate involves the soft and hard palate and associated musculature.
- A submucosal palatal cleft is an incomplete cleft resulting from failure of the submucosal levator muscle to fuse completely in the midline (patients may clinically have a bifid uvula, zona pellucida, or absent posterior nasal spine).

P. Ricalde (✉)
Florida Craniofacial Institute, University of South Florida, St. Joseph's Cleft & Craniofacial Center, Tampa, FL, USA
e-mail: ricalde@flcranio.com

I. Ahson
Florida Craniofacial Institute, Tampa Bay, FL, USA

D. E. Urbanek
Missouri School of Dentistry and Oral Health, Mercy Hospital, St. Louis MO, Department of Oral and Maxillofacial Surgery, Chesterfield, MO, USA
e-mail: dxu2@case.edu

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- Factors associated with cleft include family history, maternal smoking and alcohol use, maternal zinc deficiency, advanced parental age, folate deficiency in the periconception period, exposure to alcohol, and certain medications (e.g., retinoids, corticosteroids, anticonvulsants including phenytoin and valproic acid).

Classification Systems

- The Veau system has been proposed for classification of palatal cleft. Group 1 includes soft palate only. Group 2 includes cleft of the soft and hard palate. Group 3 represents complete unilateral cleft lip and palate. Group 4 includes complete bilateral cleft lip and palate (Fig. 10.1).
- The striped Y of Kernahan and Stark classification is a symbolic representation of the untreated cleft lip and palatal deformity with the incisive foramen as the dividing point. Right and left are distinguished for clefts of the lip, alveolus, and premaxilla.
- Boxes 1 and 4 reflect clefts of the right and left lips.
- Boxes 2 and 5 represent clefts of the right and left alveolus.

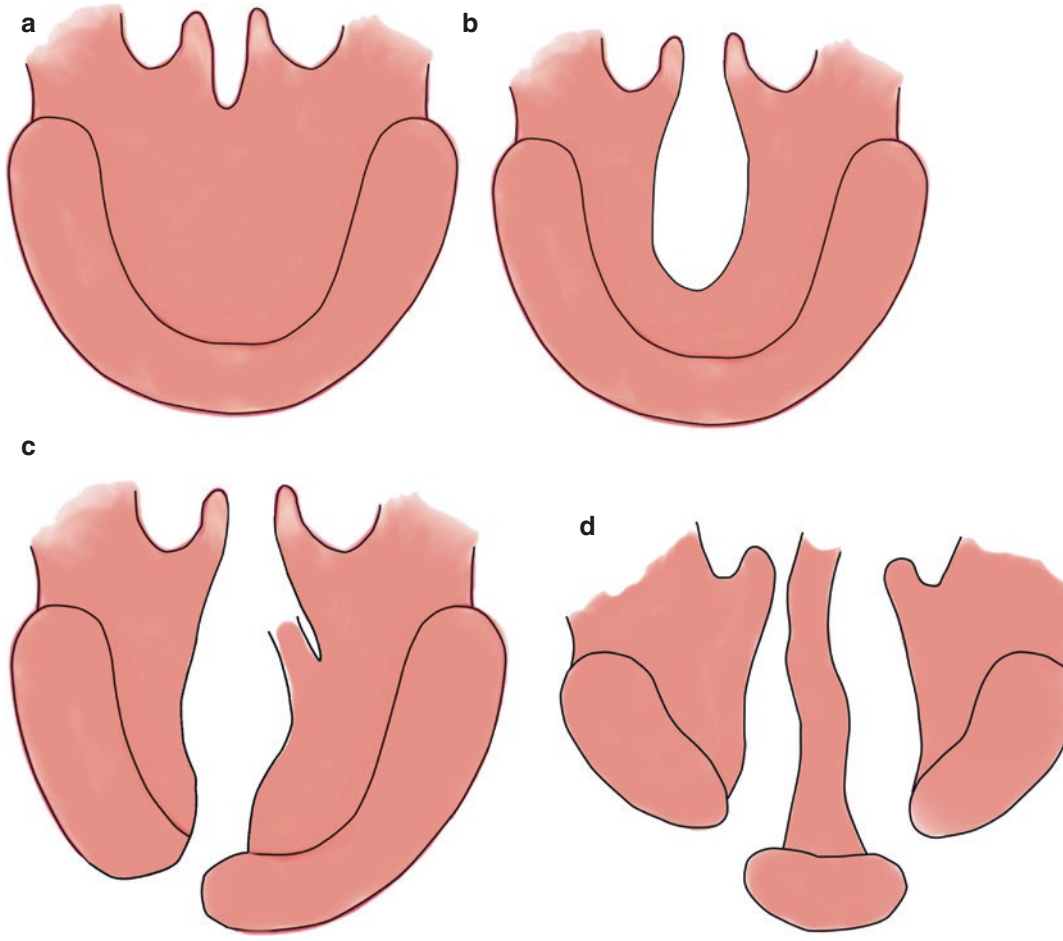

Fig. 10.1 Veau classification system. (**a**) Veau I cleft of the soft palate. (**b**) Veau II cleft of soft and hard palates. (**c**) Veau III unilateral cleft. (**d**) Veau IV bilateral cleft. (Modified from Butler CE. Head and Neck Reconstruction. Saunders; 2009.)

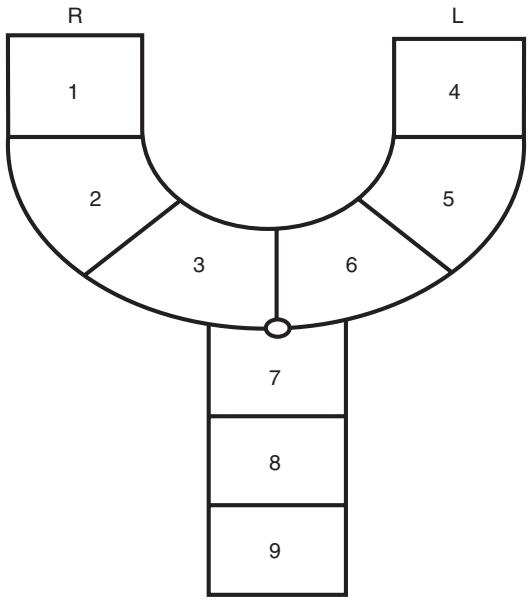

Fig. 10.2 Striped Y of Kernahan and Stark classification of clefts

- Boxes 3 and 6 represent clefts of the right and left premaxilla. The circle at the center reflects the incisive foramen.
- Box 7 reflects an isolated hard palate cleft.
- Box 8 represents a soft palate cleft.
- Box 9 represents a submucous cleft. It does not describe the degree of cleft (e.g., complete or incomplete cleft lip) or its functional impact (e.g., presence of velopharyngeal insufficiency) (Fig. 10.2).

Cleft Management

- A team approach at specialized cleft centers is the standard of care for patients with clefts to provide coordinated and continuous care through the child's growth. Members include surgeons (oral and maxillofacial, ENT, or plastic), pediatric dentists, orthodontists, speech pathologists, geneticists, audiologists, social workers, psychologists, and pediatricians.
- Ultrasound can identify clefting as early as 13–14 weeks of gestation. The sensitivity in detecting CL ± CP prior to 18–20 weeks is much lower [4]. This allows for anticipatory guidance and referral to a cleft team for interdisciplinary management.
- At birth, appropriate diagnostic testing includes echocardiogram and chromosomal array for isolated cleft palate patients in light of associated syndromes.

Sequence of Management

- At birth, a lactation consultation and/or speech therapy consultation is indicated. Bottles are available with customized nipples (e.g., pigeon bottle, Haberman with a one-way valve and reservoir spaces, Dr. Brown's cleft palate bottle) to reduce the child's work in feeding. Additionally, a Mead Johnson bottle is compressible to aid in forward flow of fluids.
- Try feeding promptly after birth. Keep child upright with frequent breaks. Feeds over 30–35 minutes will often fatigue the child and expend more calories than consumed.
- Preoperative orthopedics – During the first few weeks of life, lip and/or nasal taping or naso-alveolar molding appliances may be utilized to prepare the labial-naso-alveolar complex for surgery with a regularly adjusted appliance that helps guide growth and improve tension-free lip closure. The goal is to approximate the segments to within 5 mm. The need for frequent adjustments increases the risks of irritation and ulceration which are drawbacks of this therapy. Latham proposed appliances with screw activation and pin retention for better control of molding. Risk of growth restriction and the need for anesthesia are detractors from this therapy, as well as the baby having to undergo additional procedures.
- Lip adhesion procedure is a separate soft tissue closure used as part of a staged unilateral or bilateral cleft repair with the goal of narrowing the defect at the time of definitive closure. Normally carried out at 3–4 months of age. The major disadvantages is the need for an additional surgery with potential increase risk of scarring and questionable impact on the definitive repair.

- Primary lip repair (cheilorrhaphy) is usually performed around 10 weeks of age to reduce risks of anesthesia (rule of 10's). The timing of primary lip repair weighs the benefits of functional improvement (reestablishing muscular anatomy and function) with the risks of early surgery from anesthesia and growth limitation of surgical scarring.
- "Rule of 10's": 10 weeks old, 10 pounds in weight, and 10 mg/dL of hemoglobin.
- Audiology screening/ENT evaluation. Should be conducted before 6 months. Children with cleft palate have difficulty controlling middle ear pressure due to eustachian tube dysfunction as a result of abnormal insertions of the levator veli palatini and tensor veli palatini. With impaired ability to equalize middle ear pressure, infants with clefts usually have fluid in the middle ear space which can result in chronic otitis media, and conductive hearing loss. If left unmanaged, this can lead to permanent hearing loss. Treatment consists of myringotomy tubes or fluid evacuation.
- Primary cleft palate repair (palatoplasty in one or multiple stages) is generally performed between 9 and 18 months and timed with speech development to avoid compensatory misarticulations. Staged-soft palate includes soft palate closure at 6–10 months followed by hard palate at 1–3 years. Early palate repair (before 9 months) increases the risk of maxillary growth restriction.
- Correction of velopharyngeal insufficiency (including pharyngeal flap, sphincter pharyngoplasty, or mucosal augmentation procedures such as filler or fat grafting) is completed at 3–5 years following nasoendoscopy demonstrating poor adaptation of the soft palate to the posterior pharyngeal wall.
- Nasolabial revision is delayed until completion of nasal growth with anticipation that a definitive rhinoplasty may still be required. It is preferable to delay at least until the time of alveolar bone grafting so that concomitant treatment can be performed.
- Alveolar grafting is generally completed at 6–9 years, based on development of the dentition. Grafting is timed with formation of canine root (1/2 to 2/3 formation) to allow emergence into the cleft site. Some surgeons also time grafting to coincide with the development of the central or lateral incisor.
- Early orthodontics may be needed in the mixed dentition stage with the goal of maintaining transverse dimension, encouraging maxillary growth, and/or preparing for alveolar bone grafting. A second phase of orthodontics is often required with the permanent dentition.
- Orthognathic surgery (if required) is planned at 14 to 16 years of age in females, 16 to 18 years in males to address the maxillary hypoplasia, mandibular skeletal disharmony, and sleep-disordered breathing that is common in cleft patients.
- Rhinoplasty is often delayed at least 6 to 12 months following maxillary surgery due to changes in septum and tip support with movement of the anterior nasal spine.
- Cleft scar revision can be completed any time after 5 years of age but is ideally performed at the time of alveolar bone grafting.

Surgical Management of the Cleft Lip

Primary Lip Repair Techniques

Millard Rotational-Advancement Flap

(See Fig. 10.3)

- Millard proposed the rotation advancement flap. The goal of the Millard repair is the development of a three-layered closure following excision of hypoplastic tissue at the cleft margins. Orbicularis oris muscle continuity is reestablished and incision lines are designed to fall within the natural philtral ridges of the lip to promote symmetry. The repair also allows for columellar lengthening. Bilateral repair is complicated by lack of quality tissue, a short columella, and upward rotation of the premaxillary segment. The goal of the bilateral repair is reconstruction of the nasal floor, orbicularis continuity, and the maxillary vestibule.

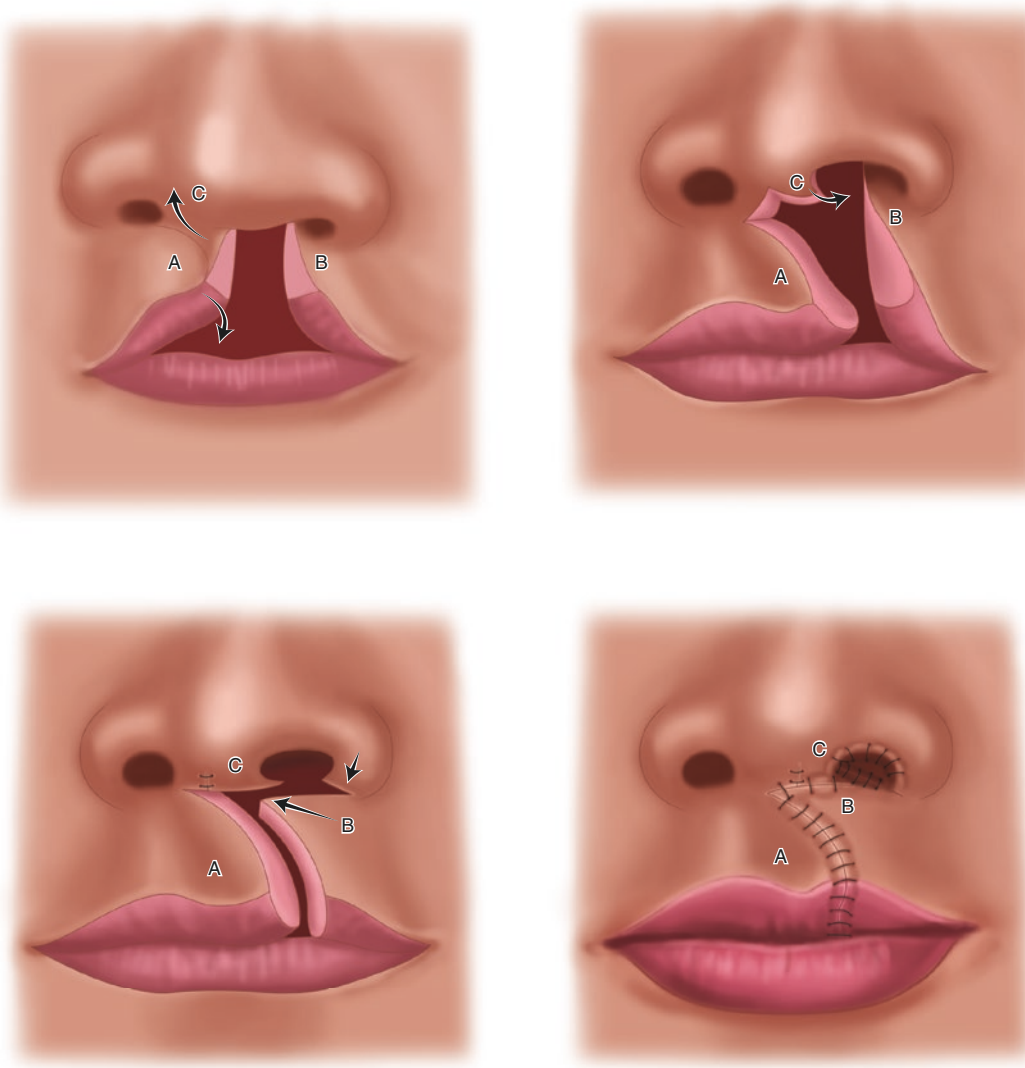

Fig. 10.3 Millard rotational-advancement flap

- The reason the technique is called a "rotational-advancement" flap is because the non-cleft side is cut in such a way that the tissue *rotates* (A) to create a longer vertical width and the cleft side *advances* (B) horizontally. This allows the scar to be hidden in a normal philtral line. This adds tissue to the deficient non-cleft side of the lip and the cleft side of the nasal columella. The premise for this and all techniques include a three-layered closure (skin, orbicularis oris, and mucosa); the excision of hypoplastic tissue from the cleft margins; and re-approximation of anatomic structures.

Delaire Technique (See Fig. 10.4)

- This technique is often used for bilateral cleft lips. Similar to the Millard technique, in that the cleft side advances while the non-cleft side rotates.
- In the case of a bilateral cleft, the technique is slightly modified. Both lateral sides are treated the same way, in that the design is the same and the muscles are dissected. The only

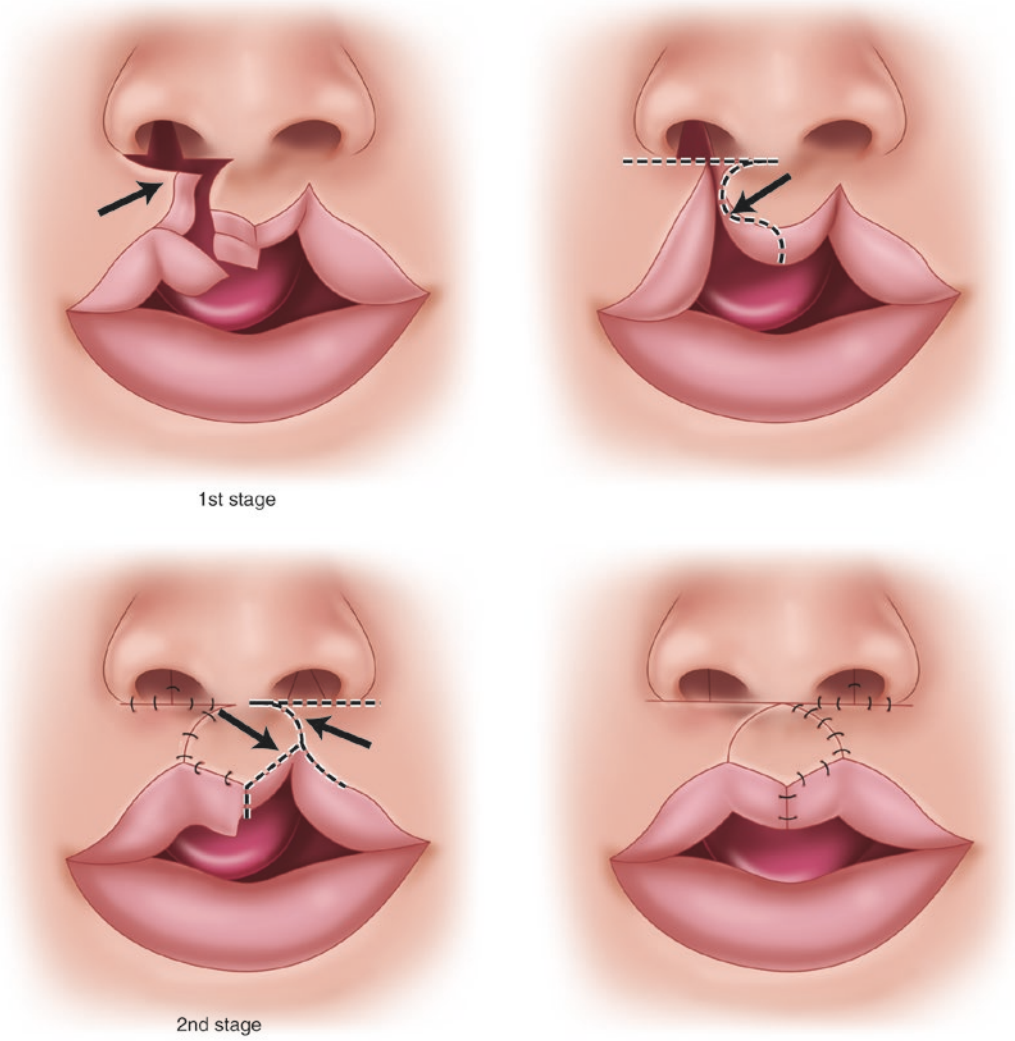

Fig. 10.4 The Delaire technique

difference is the prolabium, which is often proclined anteriorly. The prolabium is dissected with the following flap design seen in Fig. 10.4.

- The difference lies in the treatment of the nose, as the nasal tissue is gently dissected to create a laxity that allows for reapproximation in a symmetrical position. With the incisions open after the initial dissection of the Millard technique, a pair of tenotomy scissors are used to dissect over the lower lateral cartilages (still beneath the skin, cartilages not exposed) to free this tissue from its abnormal insertions. After the nasal tissue is dissected, the muscle and skin are reapproximated with additional sutures placed in the area of the nasal floor to create symmetry between the nostrils.
- A nasal bolster is often placed to maintain the nasal shape postoperatively.

Tennison-Randall (See Fig. 10.5)

- A Z-plasty technique that some feel is best used in wider clefts or if more vertical repositioning of Cupid's bow is needed. Proponents of this technique argue that the triangular

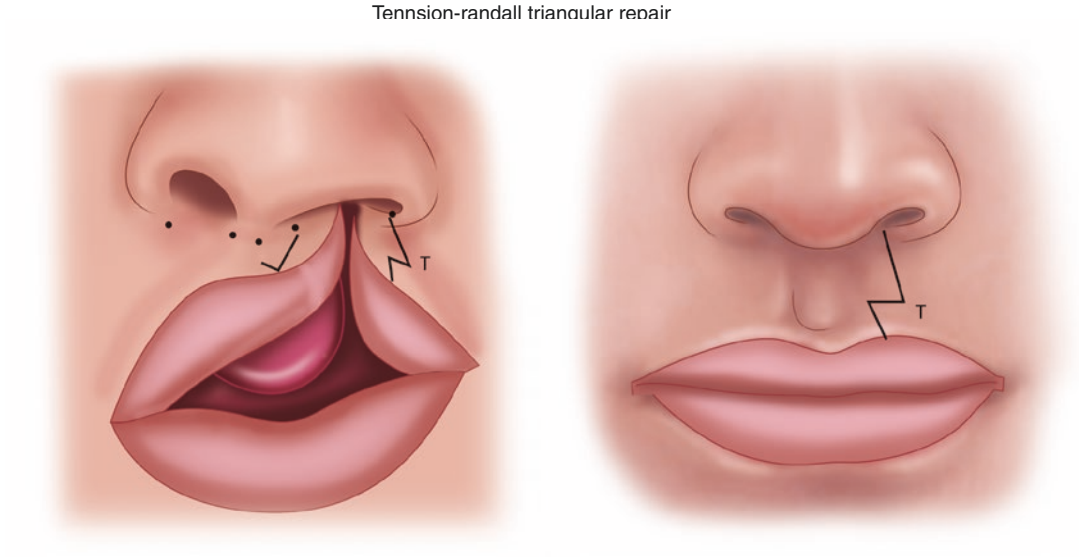

Fig. 10.5 Tennison-Randall technique

design avoids wound contracture and lip length shortening seen with the rotation and advancement flap operation. A major disadvantage of this technique is the scarring that crosses the philtral column which may contribute to the asymmetry.

Cleft Lip Repair Complications

Vermillion Deformities Vermillion deformities of the lip may occur secondary due to inadequate approximation of the marginal portion of the deep orbicularis or of the medial and lateral white roll at the primary repair, or excessive resection of the vermillion. Vermillion deficiencies are more common in bilateral clefts due to a dearth of tissue. May require a revision surgery.

Whistle Lip Deformity Inadequate release and advancement of mucosa and the vermillion can result in an indentation at the junction of the vermilion. The result is inadequate bulk of the lip with excessive show of the central incisors when the lips are in repose. Treatment includes nonkeratinized epithelium mucosal-submucosal flap procedures or fat grafting. Severe deformities may require an Abbe flap.

Hypertrophic Scar Can be managed by watchful waiting, scar revision, or steroid injection.

Nasal Asymmetry Can be managed at the time of nasolabial revisions or patient may require definitive rhinoplasty for management of asymmetry after orthognathic surgery.

Surgical Management of the Cleft Palate

Muscles of the Palate (See Fig. 10.6)

Muscles, innervation. and actions include the following:

- Tensor veli palatini – trigeminal nerve – tenses and depresses soft palate.
- Levator veli palatini – pharyngeal nerve – elevates the palate.
- Musculus uvulae – pharyngeal nerve – draws uvula upward and forward.
- Palatoglossal – pharyngeal nerve – draws palate down and narrows pharynx.
- Palatopharyngeal – pharyngeal nerve – draws palate down and narrows pharynx.

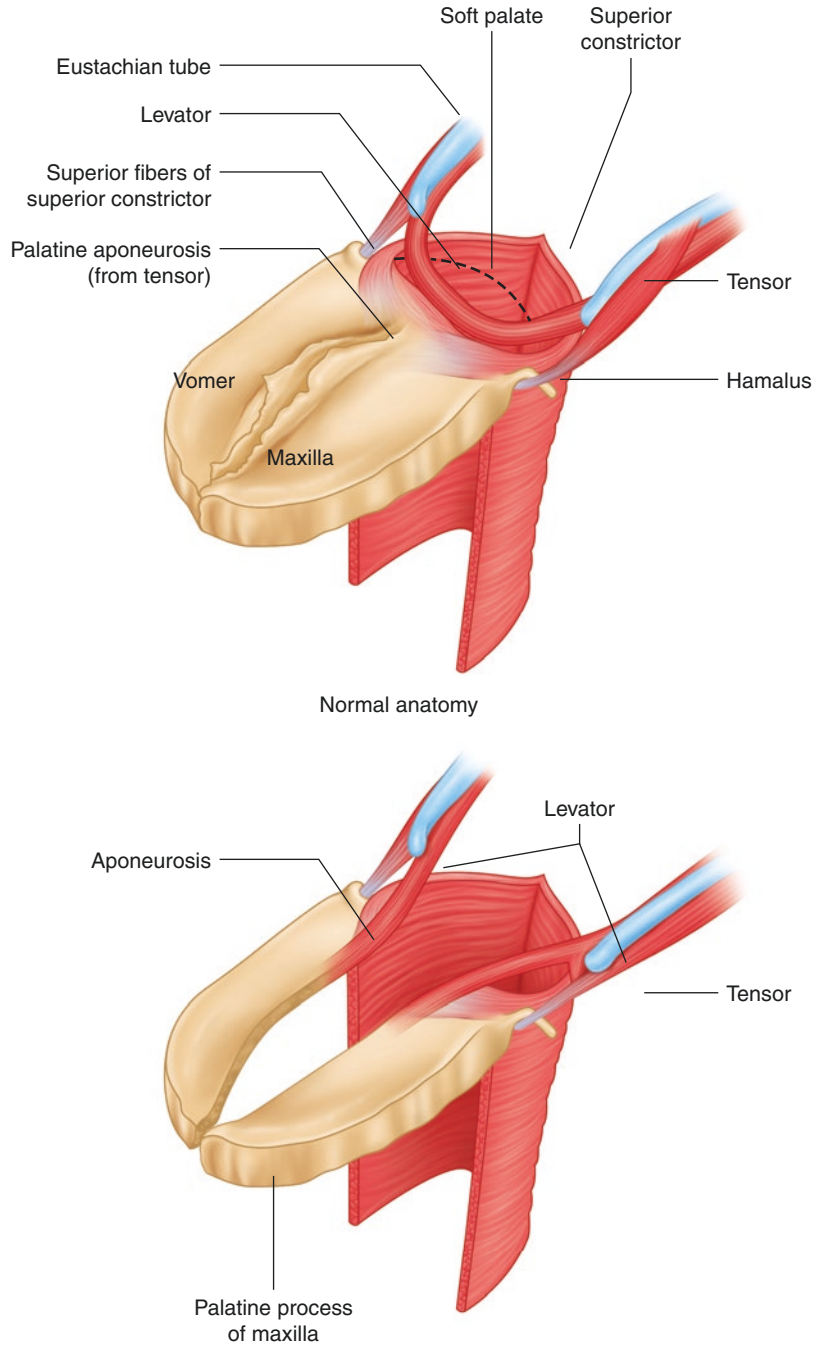

Fig. 10.6 Muscles of the palate (normal and cleft)

Palatal Repair Techniques

- Cleft palate repair has two goals, namely, the watertight closure of the oronasal communication and the anatomic repair of the musculature within the soft palate, which is critical for normal creation of speech.
- In the cleft patient, the tensor and levator veli palatini, as well as the palatoglossal and palatopharyngeal muscles, abnormally insert into the posterior hard palate in an AP orientation and have to be reoriented into their normal transverse alignment.

Modified von Langenbeck Procedure (See Fig. 10.7)

- In this procedure, two full thickness flaps are created and mobilized with layered dissection of the soft palatal tissues. The anterior and posterior portion remain connected to periosteum to

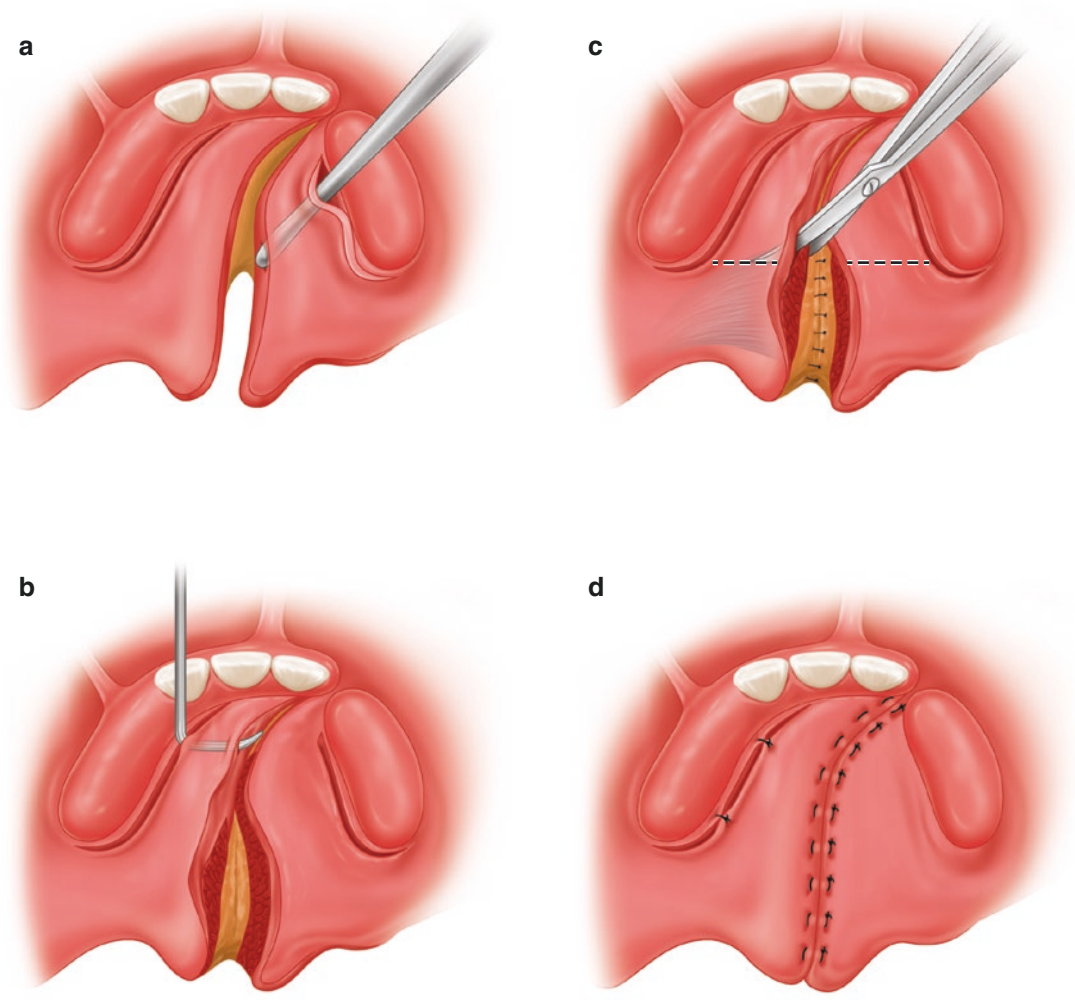

Fig. 10.7 Modified von Langenbeck procedure

increase circulation. Having an anterior attachment can decrease the visualization for nasal mucosa closure, and there is usually an area laterally that is allowed to granulate in. This healing through secondary intention can increase pain and create scar formation that restricts growth. For these reasons, it has fallen out of favor.

Bardach Two-Flap Technique (See Fig. 10.8)

- The most commonly used procedure, two flaps based off of the greater palatine artery are raised, dissecting either two layers (hard palate – nasal and oral mucosa) or three layers (soft palate – nasal mucosa, muscle, and oral mucosa). Dissecting the muscle off the posterior edge of the soft palate allows closure to reapproximate a more physiologic orientation.

Furlow Z-Plasty Technique (See Fig. 10.9)

- The Furlow Z-plasty was created to allow for lengthening of the hard palate by reorienting the muscles in a more physiologic reapproxi-

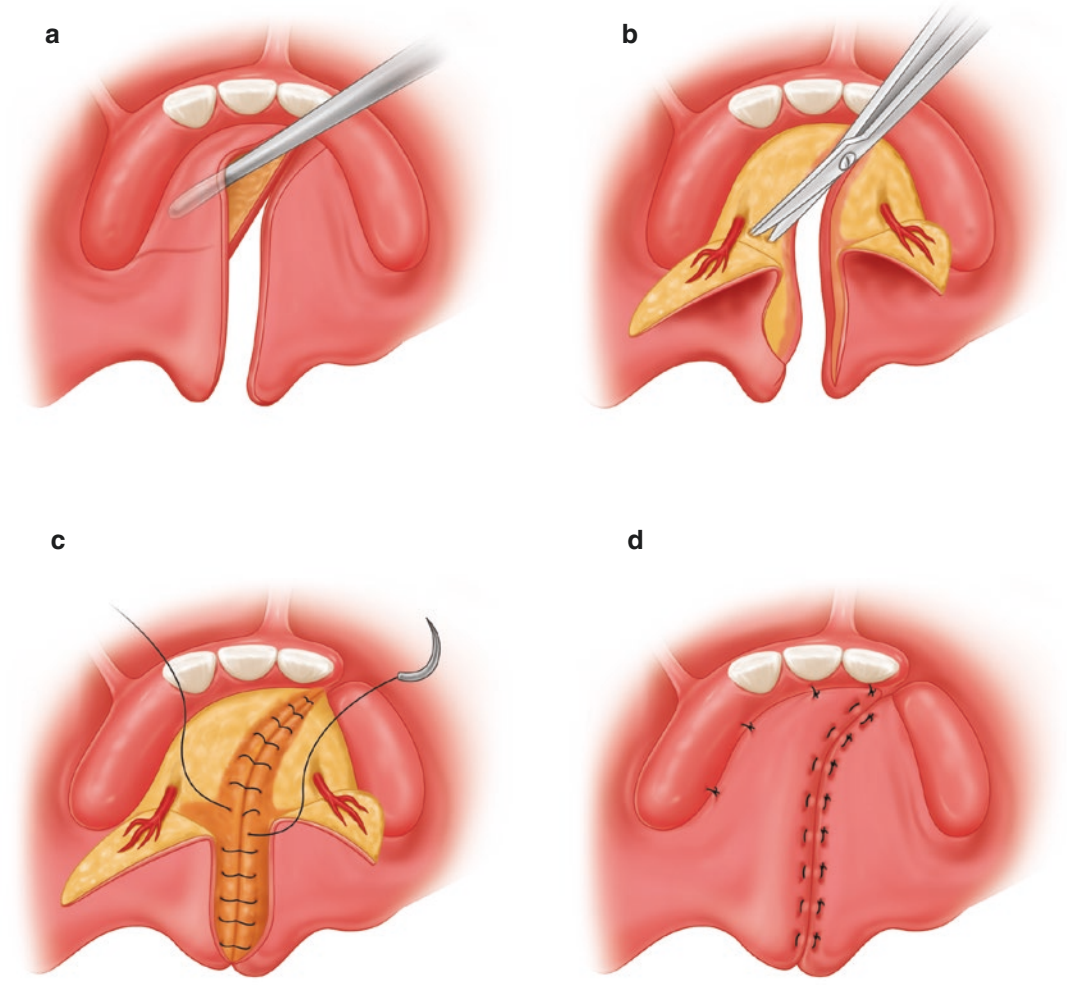

Fig. 10.8 Bardach two-flap technique

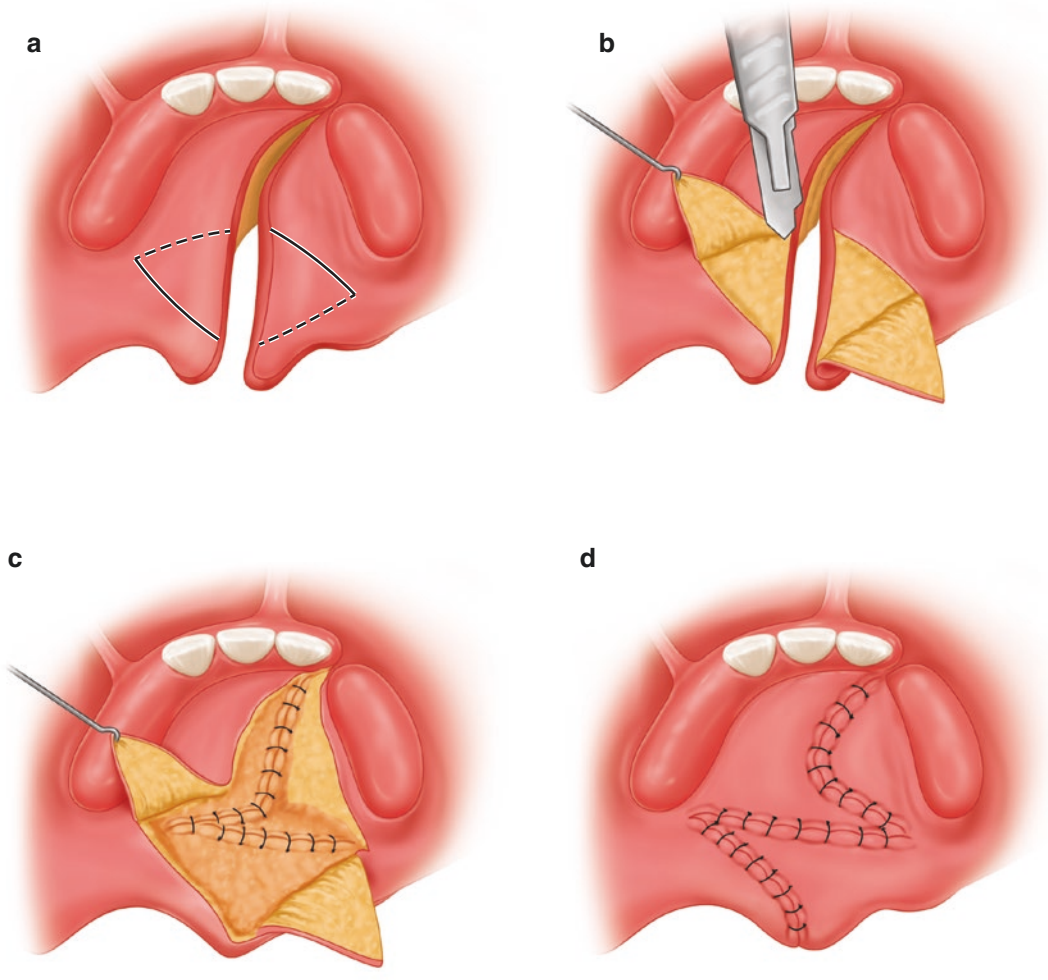

Fig. 10.9 Furlow Z-plasty technique

mation. One side is myomucosal and the other side is mucosal. This method is more technically difficult and has a higher rate of oronasal fistula formation.

V-Y Pushback Technique (See Fig. 10.10)

- Sometimes described as a modification of the von Langenbeck technique with anterior pedicle release, this technique involves the release of the muscles of the soft palate from the posterior edge of the hard palate and from the periosteum on the nasal side, allowing the creation of a more physiologic muscle sling and soft palate. There is an anterior two-layer closure and posterior three-layer closure, but still has an area that heals by secondary intention, which leads to severe growth restriction.
- A major advantage is maximization of palatal length to decrease postoperative VPI.

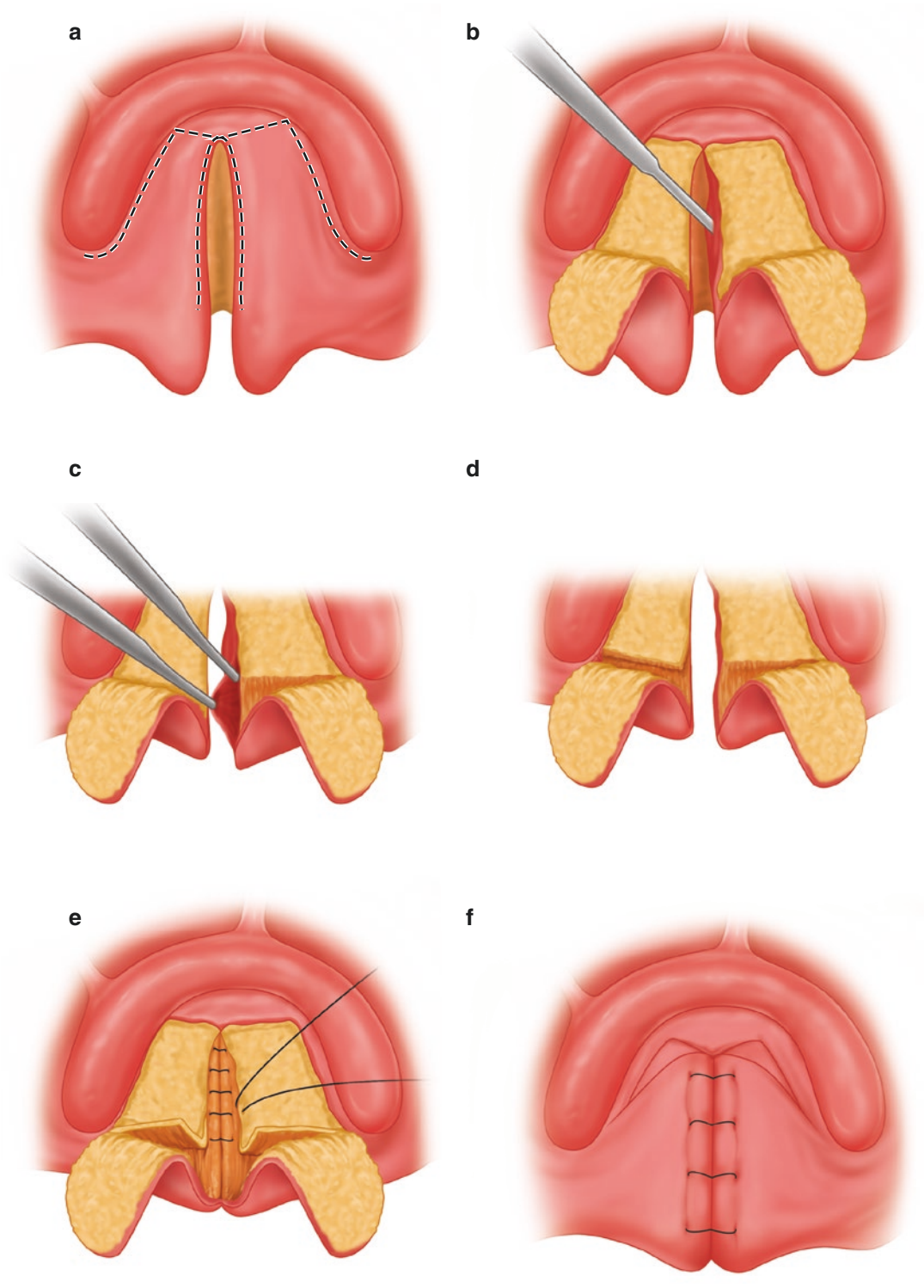

Fig. 10.10 V-Y pushback technique

Complications of Cleft Palate Repair

Oronasal Fistulae The most common sites for development of fistulae are at the junction of the hard/soft palate and the incisive foramen. Smaller fistulae may be addressed with regional palatal flap repair. Larger fistulae can be treated with robust regional palatal flaps, tongue flaps, facial artery myomucosal flaps (FAMM flaps), and temporalis flaps. Surgeons may consider waiting for further growth prior to repair to avoid maxillary growth restrictions secondary to surgery if speech or function not affected**.** Depending on age of patient, repair of fistulae may also be combined with the alveolar bone graft, or with orthognathic surgery so as to minimize the number of surgical interventions.

Velopharyngeal Incompetence VPI manifests as hypernasality and nasal emission. Diagnosis can be confirmed with nasoendoscopy with or without videofluoroscopy. The surgical management of VPI includes pharyngeal flap, pharyngoplasty, posterior pharyngeal wall augmentation, and palatal revision.

Velopharyngeal Insufficiency (VPI)

Velopharyngeal Insufficiency (VPI)

- Also called velopharyngeal incompetence, it is the incomplete closure of the velopharyngeal sphincter, and thus incomplete separation of the oral and nasal cavities during speech.
- It is the term of choice when diagnostic studies have clearly determined that a true physical limitation is present.
- The term velopharyngeal dysfunction (VPD), however, is used when there is some sort of malfunction, but the cause remains unclear.
- It is estimated that about 1/4 of patients will demonstrate some sort of VPD after cleft palate repair.
- Signs and symptoms of VPI:
  - **Hypernasal speech** – is the production of inordinate nasal resonance during the production of vowels produced by the inappropriate coupling of the nasal and oral cavities.
  - **Nasal emission** – nasal air escape during production of consonants requiring high oral pressure. Nasal emission may be audible or not.
  - **Nasal substitution** – incomplete closure of the velopharyngeal port causes sound to be produced as a nasal consonant rather than an oral consonant. For example, "B" becomes "M" and "D" becomes "N."
  - **Compensatory misarticulations.**
    - With VPI, the child cannot build up oral pressure to produce fricatives (e.g., S) or plosives (e.g., P). Instead, the child will attempt to create those pressures below the level of the velopharyngeal port. Such misarticulations may include glottal stops, pharyngeal stops, and pharyngeal fricatives.

Work Up and Diagnosis

Speech and Language Pathologist (SLP)

- An essential role of the SLP is to determine if the VPI is the result of true physical limitations, and therefore requires surgical management, or more related to learning or habituation of patterns, which would require speech therapy. In cases of severe and consistent VPI, it is easy to make the diagnosis and, therefore, treatment recommendations. However, in borderline cases, it is difficult to make this determination, and surgery may not be the proper treatment. In complex cases such as these, a period of speech therapy will provide additional and often necessary diagnostic information.
- Timing of evaluation – Evaluation for VPI is dependent on the development of speech in the child. It is impossible to accurately judge velopharyngeal function in infants and toddlers (up to 3 years of age). However, during the preschool years (3 to 5 years of age), as the child's sound repertoire, articulation, and

expressive language expand, velopharyngeal function can be assessed more accurately. A level of cooperation during the evaluation is also required on the part of the child.

Clinical Evaluation of VPI

- Observe speech for hypernasality, nasal substitutions, and compensatory articulation errors. These signs should serve only as a "red flag" for the surgeon. Formal speech testing and interpretation should be left to the SLP.

Tests for Nasal Emission

- Mirror test: a mirror is held under the patient's nares during production of pressure-sensitive sounds to observe for fogging.
- See Scape test: See Scape is a device that resembles an incentive spirometer. A flexible tube is placed into the patient's nare, and as the child produces pressure-sensitive sounds, a styrofoam stopper inside a connected vertical tube rises as a result of nasal emission.
- Nasal palpation: vibration from hypernasality can be palpated by placing one's fingers on the nasal sidewalls.
- Auscultation with a stethoscope (remove the drum and use the tube), flexible plastic listening tube, or straw.
- Nasopharyngoscopy:
  - Involves using a flexible fiberoptic endoscope to visualize velopharyngeal function. The nasal surface of the velum, and lateral and posterior pharyngeal walls can be visualized without interfering with speech production.
  - Is the primary instrumental assessment tool of most cleft palate-craniofacial teams because it does not expose the patient to radiation.
  - Patient cooperation is essential for a successful evaluation.
  - Is the most appropriate instrumental measure for diagnosing an occult submucous cleft of the palate, and for assessing velopharyngeal closure after a pharyngeal flap has been performed.
- Video Fluoroscopy:
  - Uses fluoroscopy to visualize the anteroposterior movement of the velum during a variety of speech tasks. Barium is instilled through the nose and provides visualization of the margins of the velopharyngeal structures.
  - Is performed by a radiologist.
  - Involves exposing the child to radiation, and because of this is a less frequently used test for VPI.

Treatment of VPI

Nonsurgical Management

- Prosthetics such as a speech bulb or palatal lift (if surgery is not an option).
- Speech therapy – for cases where velopharyngeal mislearning (faulty articulation) is the cause of hypernasality or nasal emission.

Surgical Management

- Timing of surgery – surgical management of VPI ideally occurs around age 5, however can be done at a later age as well.
- Indications of surgery – patients who have consistent VPI as determined by a SLP and experienced surgeon.

Contraindications of surgery:

- Patients who decline surgery by choice.
- Patient has known or suspected risk for airway obstruction.
- Patient has intermittent or inconsistent closure that responds well to speech therapy.
- Patient has incomplete diagnostic results.
- Patients who have an aberrant and medial position of the internal carotid artery, such as in those with velocardiofacial syndrome. Velocardiofacial syndrome (VCFS) – anomalous internal carotid arteries have been shown to be a frequent feature in VCFS and pose a potential risk of iatrogenic injury and hemorrhage during surgery. Preoperative cervical vascular imaging studies are recommended to define the course of these vessels.

Surgical Correction of VPI

- The patterns of velopharyngeal closure can be thought of as consisting of two main categories of movement: anteroposterior movement in which the velum contacts Passavant's ridge and lateral pharyngeal wall movement. Varying degrees of velum and pharyngeal wall motion contribute to many sphincteric closure patterns.
- Passavant's ridge – a bulge on the posterior pharynx above the arch of the atlas (C1), produced by forceful contraction of the superior pharyngeal constrictor. This ridge also may be associated with velopharyngeal incompetence as a compensatory mechanism to assist with velopharyngeal closure.

Pharyngeal Flap Surgery

- If the patient has undergone palatal repair and has been diagnosed with velopharyngeal incompetence, he/she may undergo pharyngeal flap surgery in order to correct hypernasality and nasal air escape.
- The goal of surgery is to develop a functional seal between the nasal cavity and the oral cavity. This is accomplished by taking tissue from the posterior pharynx and attaching it to the soft palate, effectively decreasing the opening between the nasal cavity and the oral cavity (also called the velopharyngeal port).
- There are multiple different methods, but the most commonly used techniques are either an inferiorly based or superiorly based pharyngeal flap. Below are descriptions of two superiorly based repairs.

Superiorly Based Pharyngeal Flap
(See Fig. 10.11)

- Indicated when there is adequate lateral pharyngeal wall movement. Tissue is taken from the posterior pharyngeal wall and attached to the soft palate, creating a midline subtotal obstruction of the oral and nasal cavities with two small lateral openings that ideally remain open during respiration and nasal consonant production and close for consonants.
- Involves elevating a myomucosal flap from the posterior pharyngeal wall and insetting it into the velum such that it converts the single velopharyngeal aperture into two lateral ports separated by the pharyngeal flap.
- It is the most commonly performed surgical technique used to treat VPI.
- For VCFS, preoperative cervical vascular imaging (CT/MRI angiogram) is obtained to rule out medial internal carotid arteries (ICA). Treatment options if aberrant vessel location identified:
- "Cautious" surgery.
- Alternate procedure such as a Furlow palatoplasty.
- No surgery and re-evaluate over time to see if things change with growth.
- Posterior pharyngeal veins are the most common site for bleeding and are visible on the prevertebral fascia.
- ICAs are located laterally and lateral dissection is avoided.

Sphincter Pharyngoplasty (Orticochea Dynamic Pharyngoplasty)

- Indicated for those with markedly impaired or absent lateral pharyngeal wall motion (when there is adequate posterior movement of the velum).
- Creates a smaller midline pharyngeal port.
- Scheduled for a minimum of 3 months after a tonsillectomy and adenoidectomy. These must be done carefully to preserve the posterior tonsillar pillars.
- Testing of VPI is repeated 6–8 weeks after T&A.

Alveolar Clefts

Alveolar clefts (Fig. 10.12) involve a separation of the bony alveolar ridge and can range from a simple notching in the buccal alveolus to a wide gap creating an oronasal fistula and disturbances in the eruption of teeth. The alveolar cleft is managed with bone grafting and rearrangement of the soft tissues.

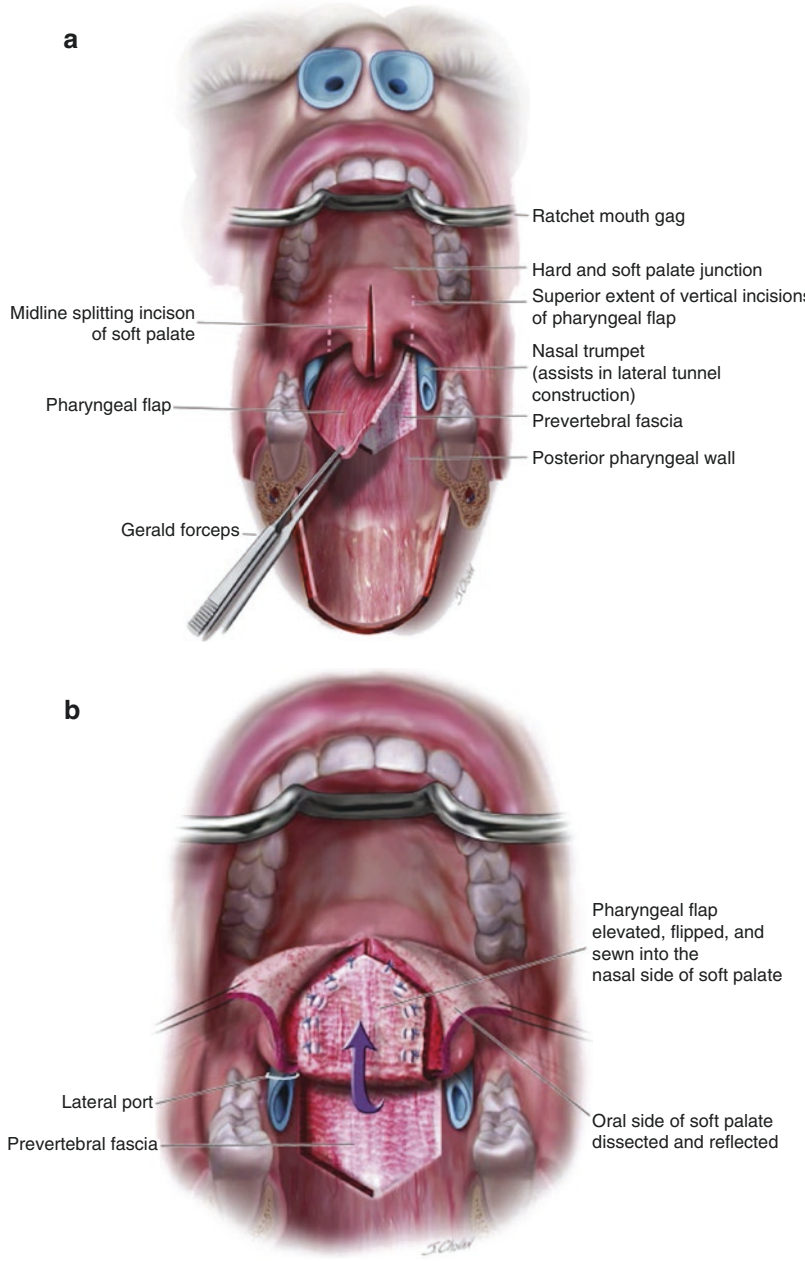

Fig. 10.11 Superiorly based pharyngeal flap. (Reproduced with permission from Posnick, JC (2000) The staging of cleft lip and palate reconstruction: infancy through adolescence. In: *Craniofacial and Maxillofacial Surgery in Children and Young Adults*, edited by JC Posnick. Philadelphia: WB Saunders)

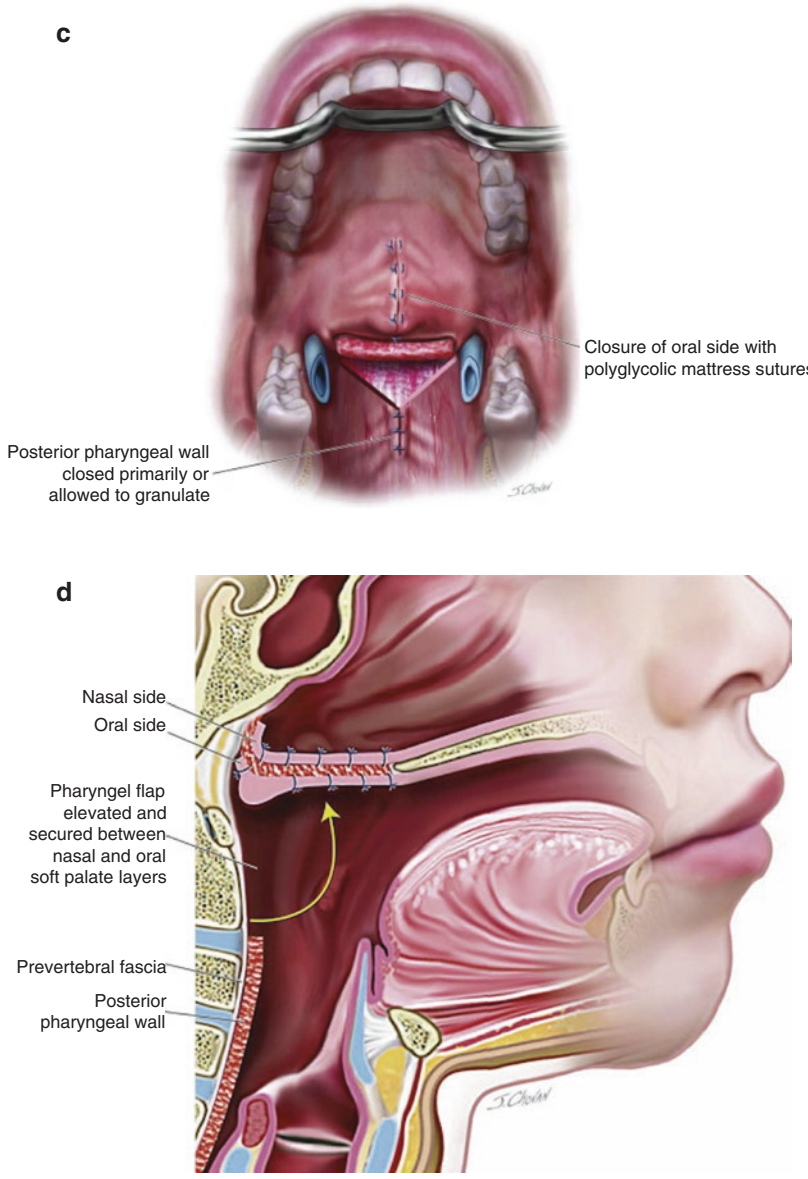

Fig. 10.11 (continued)

Presentation of a Cleft Patient

- The anatomy of a patient with an alveolar cleft can look very different from patient to patient, and therefore, an understanding of the different presentations and anatomical differences is important to treatment plan the patient appropriately.
- In unilateral clefts, the segment of the maxilla that is on the side of the cleft is known as the "lesser" segment (the "greater" segment being the non-cleft side).
- The cleft itself can range from a small notch to a wide gap with large oronasal fistula.
- Common presentations include medial collapse due to a lack of transverse stability, resulting in crossbite.

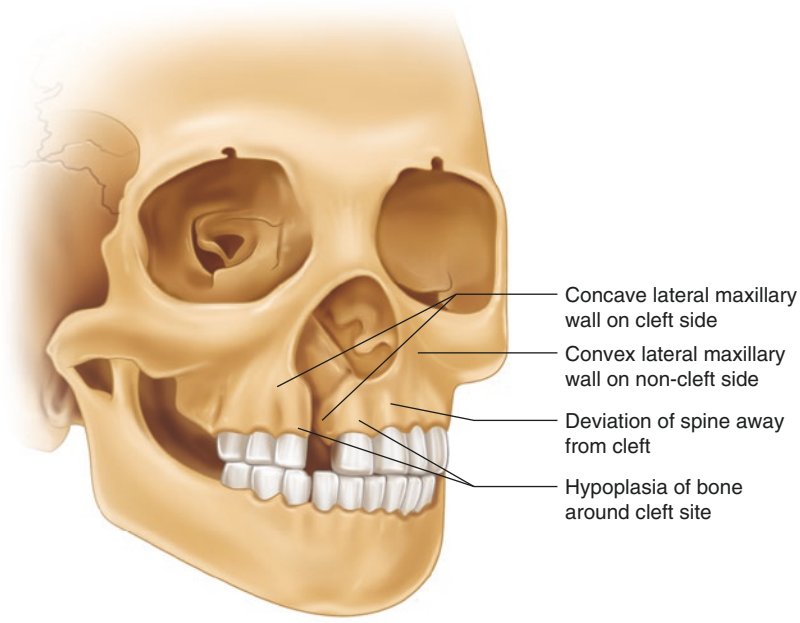

Fig. 10.12 Alveolar Cleft

- Teeth along the cleft side can be missing, hypoplastic, malrotated, or supernumerary. The periodontal support on the cleft side of the adjacent incisor and canine tend to be deficient.
- In bilateral clefts, the presentation can be varied as well. The premaxilla is commonly rotated anteriorly due to the unrestricted growth of the vomeropremaxillary suture. It also may be slightly mobile due to the flexibility of the pediatric bone. Occasionally the premaxilla may be missing due to the presence of a true midline cleft, or due to iatrogenic injury to the blood supply. If present, care must be taken at all times to preserve its vascularity due to a lack of collateral circulation from lateral anastomoses.
- Knowledge of the three-dimensional structure of the cleft will be important in helping you visualize the technique, as well as describing the information on the boards (Fig. 10.13).

Goals of Therapy

- Allow eruption of dentition.
- Provide support to the adjacent periodontium.
- Stabilization of maxillary segments.
- Closure of oronasal fistulae.
- Improve speech and language development.
- Provide appropriate tissue for dental health.
- Provide adequate bone for future dental implant therapy.
- Reconstruct nasal floor and lift the alar base.
- Allow for greater lip support.
- Cosmesis.
- Greater self-esteem.

Nomenclature

The nomenclature for alveolar bone grafting is based on the time of bone grafting in comparison to the lip repair and dentition. This is divided into:

- Primary (at or before the time of the cleft lip or palate repair).
- Secondary (later in life). Among secondary bone grafting, the timing can further be broken down into:
  - Early: 3–5 years.
  - Early mixed dentition: 6–8 years.
  - Late mixed dentition: 9–12 years.
  - Late: 13 years and above.

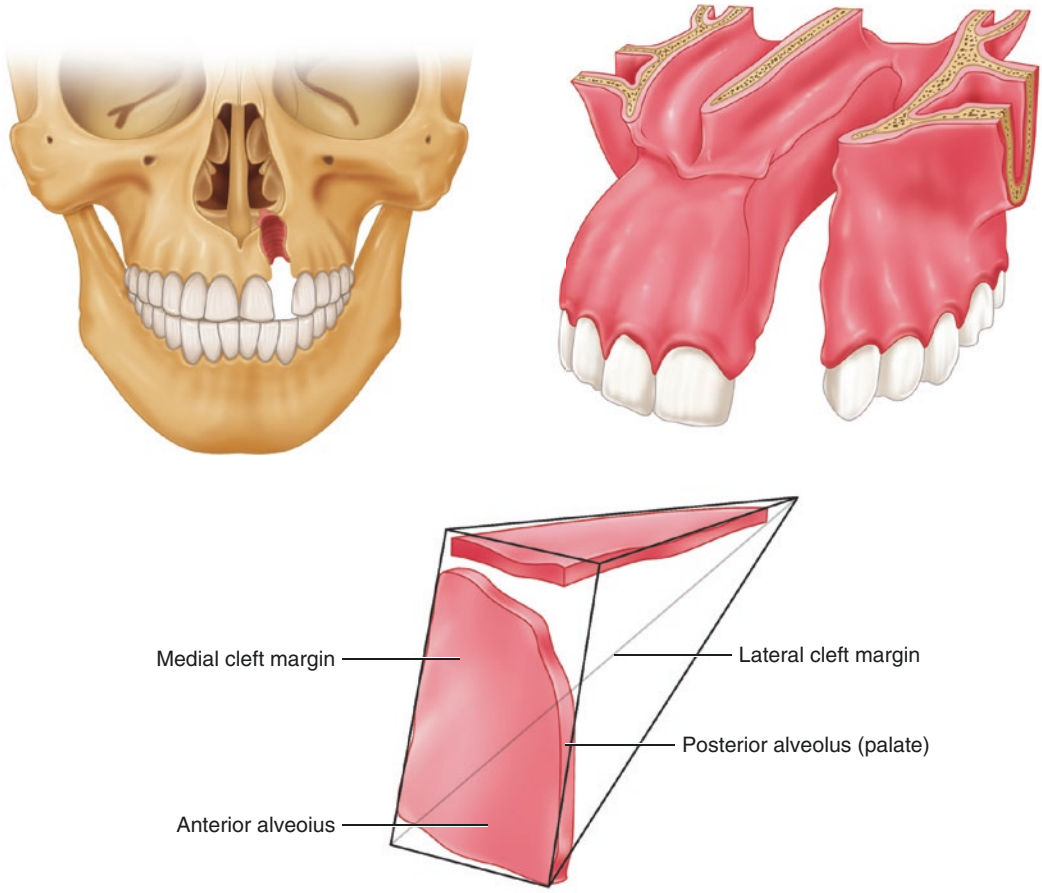

Fig. 10.13 3D structure of alveolar cleft

Gingivoperiosteoplasty Can be done with naso-alveolar molding techniques or at the time of primary lip repair. Gingivoperiosteoplasty approximates the alveolar cleft soft tissue resulting in a gingivoperiosteal tunnel. It is hoped that this will create bone healing so as to avoid the need for bone grafting. The procedure is not appropriate for wide clefts as the segments need to be within 2 mm of each other. The downside to this procedure is the potential for early scarring and restriction of maxillary growth, injury to the cleft and adjacent teeth blood supply, and increased costs/psychological burden to the child and family.

Primary Grafting The rationale behind grafting at the time of the initial lip repair is to prevent maxillary arch collapse and allow the teeth to migrate into the alveolar process. In bilateral cases, it can also stabilize the premaxilla at an earlier age. Unfortunately, these procedures have resulted in severe growth restrictions and have largely been abandoned.

Secondary Grafting

Early If done too early, it may lead to maxillary growth restriction.

Early Mixed Dentition Most commonly alveolar bone grafting is performed when the canine root is ½ to 2/3rds formed. There are many other factors to consider, and grafting based on development of central and lateral incisors is gaining favor among some cleft centers, but outcome

studies still lacking. Advantages include improved clinical crown height and periodontal support for the adjacent central incisor.

Late Mixed Dentition Grafting after the eruption of the canine has been shown to have a lower success rate due to the gradual loss of bone along the distal surface of the central incisor and the distal surface of the canine root. Once the cementum is exposed, the bone graft is unable to adhere to and make up for these losses. In addition, the positive effects of the tooth erupting into the bone graft does not occur leading to a decreased volume of remaining bone after grafting has been completed.

Late The late bone grafting requires extensive orthodontic forces to separate the collapsed arches, as well as having the same issues as grafting in the late mixed dentition. The success rate decreases as age increases. At this point, rather than attempting a procedure with a decreased success rate, it may be prudent to use a removable partial denture as an obturator. Another option is modified orthognathic surgery, during which time the arches are placed in the appropriate alignment, and the lesser segments are advanced to decrease the cleft dental gaps. With the intimate contact of the arches, the bone graft success is much higher.

Surgical Technique for Secondary Bone Grafting (Fig. 10.14)

- Although the technique may vary in timing, the main objective of the surgery is to fill in the pyramidal bone defect caused by the cleft (see below).
- The anterior flaps are raised first, to gain access to the underlying nasal mucosa.
- Next, the nasal floor must be recreated, forming the superior portion of the pyramid. The posterior portion of the pyramid is recreated by reapproximating the palatal tissue.
- Finally, bone graft is packed into the pocket of tissue that has now been created, and the anterior flaps are closed over the bone graft in a watertight seal.

Design of the Flaps

Split Thickness Component As the incision is carried superiorly, dissection remains anterior to the cleft to expose the lateral aspect of the anterior nasal spine and the lower pyriform rim. At the junction of the nasal cavity and vestibule, the flap should have a split thickness component (at the most anterior margin).

Full Thickness Component The anterior flaps abutting the alveolar bone are full thickness mucoperiosteal flaps, with their blood supply from the periosteum and lateral circulation. The design can range from a sulcular incision carried laterally or sparing the gingival sulcus. To get the tissue appropriately mobile, some surgeons use a back cut at the area of the first molar. The nasal flaps are created from the tissue adhering to the lateral and medial walls of the bony pyramid, extending posteriorly. These are raised in a full thickness fashion, except for the anterior component which is split thickness. The palatal flaps are full thickness mucoperiosteal flaps as well, with gingival sulcular incisions extending as far distal as necessary to ensure appropriate mobilization.

Bilateral Alveolar Clefts The main difference in the flap design of bilateral clefts is taking care to minimize dissection that would compromise the blood flow to the premaxilla (Fig. 10.15). The remaining blood flow to the premaxilla is from the midline branches of the posterior septal artery and to a lesser extent the lateral nasal and terminal branches of the anterior ethmoidal artery. These vessels pass from a superior to inferior direction, so care must be taken to have minimal dissection superiorly and focus on freeing tissue from the lateral aspect of the premaxilla as shown below.

Donor Site

- The gold standard for alveolar cleft repair is bone from the iliac crest that is harvested as a cancellous graft. The particulate structure of the graft allows for easy packing into the cleft site and conformity to the shape of the arch.

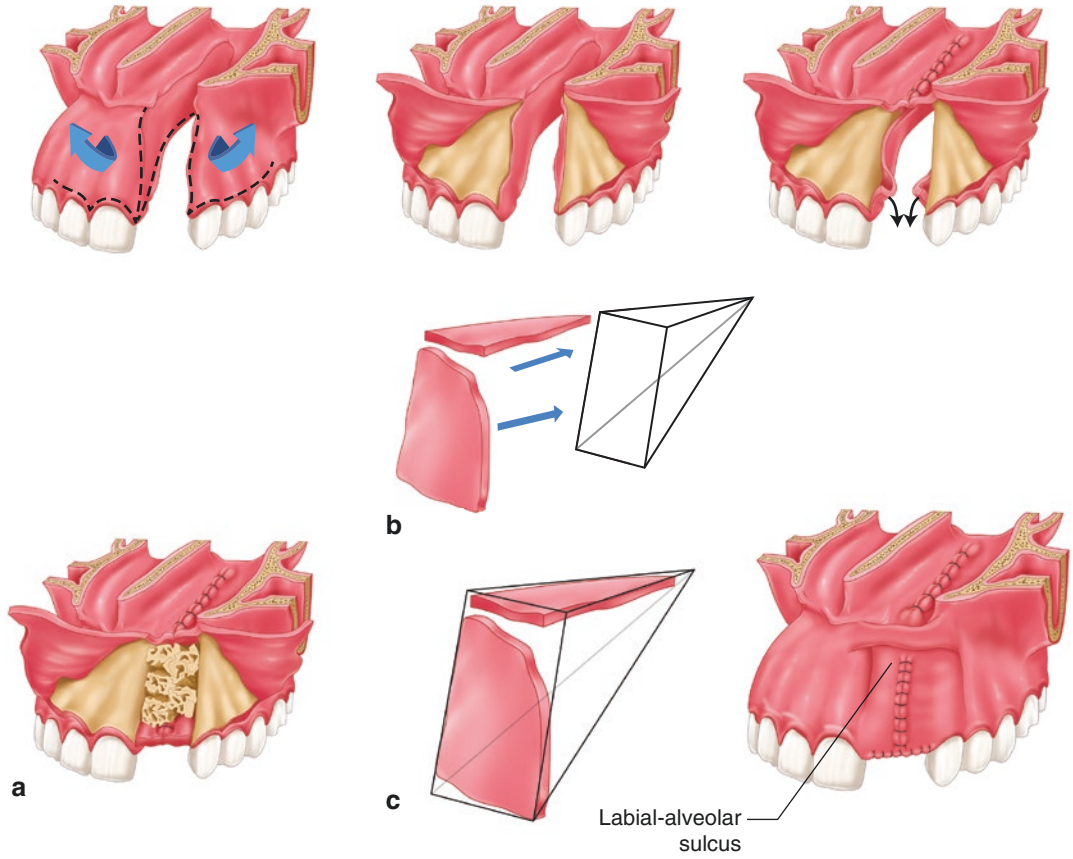

Fig. 10.14 Secondary bone grafting technique. (*Note: Some surgeons prefer to raise the palatal component second and retract the tissue to gain better visualization of the cleft to help in closing the nasal floor)

- Harvest from children is similar except the crest is cartilaginous. Cartilage cannot be harvested as it represents an ossification center. The cartilage is reflected to one side and bone below is harvested to prevent a contour deformity.

Other Sources of Bone Grafting for Alveolar Clefts

- Rib:

  Pros: Sole source of bone in primary grafting, large amount of bone.

  Cons: Morbidity (pain, scar, pneumothorax).

- Calvarial bone:

  Pros: Membranous bone, same field, hidden scar.

  Cons: Little cancellous, patient perception.

- Tibial bone grafting:

  Pros: Cancellous bone.

  Cons: Donor morbidity, moderate amount of bone.

- Mandible:

  Pros: Membranous bone, same field, hidden scar.

  Cons: Little bone, risk to dentition, mental nerve.

- Bone morphogenic protein:

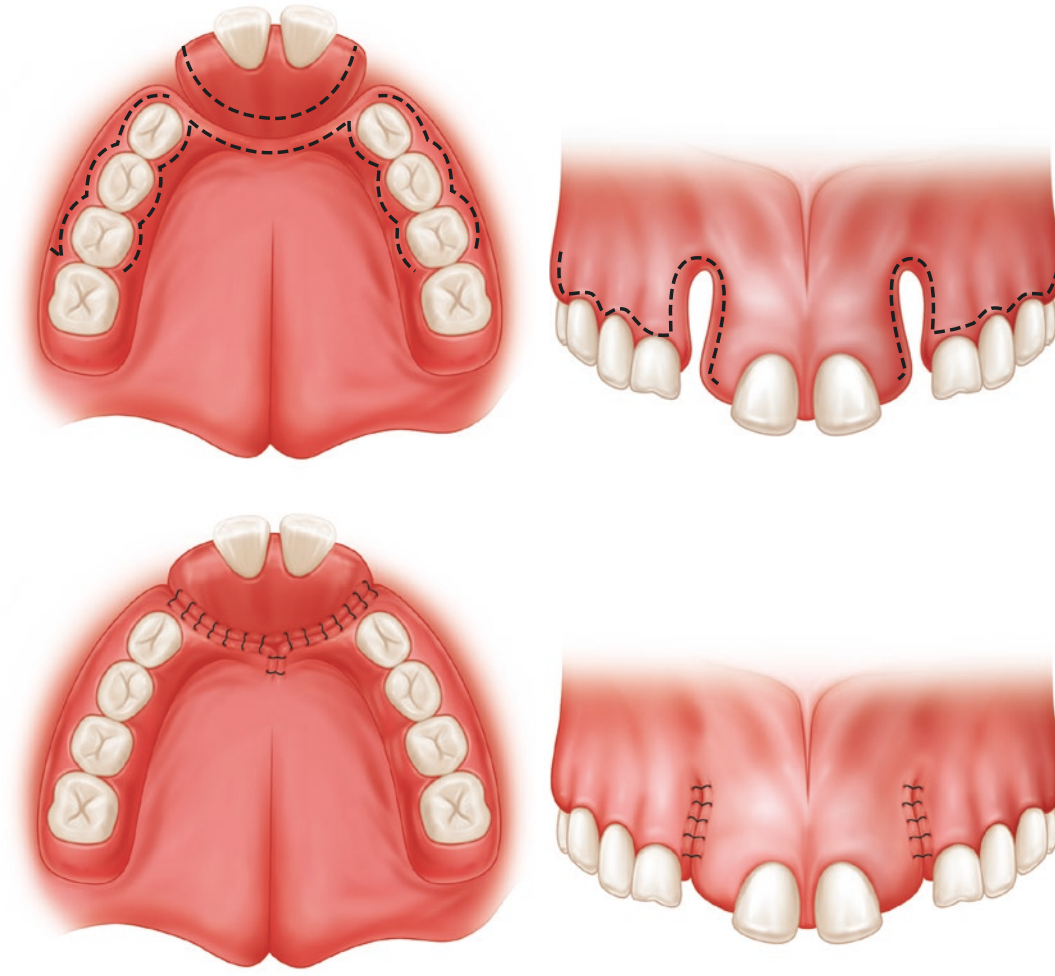

Fig. 10.15 Secondary bone grafting of a bilateral alveolar cleft

Pros: Reduce OR time, can be mixed with autologous or allograft, reduced risk of harvest site.

Cons: Higher expense, risk of ectopic bone formation, severe swelling/edema, and increased medico-legal risk. BMP is poorly studied in children and is contraindicated by the FDA for use in children.

Perioperative Dental Support:

- Highest success rates occur in a multidisciplinary setting. Consultation with other specialists and regular follow-up ensures that the patient is treated at the appropriate age and the appropriate steps are completed preoperatively to maximize success. Specifically, consultation with orthodontists and pediatric dentists preoperatively is mandatory for the following reasons:
- Orthodontic alignment of the arch form – Presurgical and postsurgical orthodontics are associated with superior secondary alveolar bone grafting outcomes. Presurgically, the teeth are repositioned into a more ordered alignment to facilitate surgery. Postsurgically, the functional stress on the grafted bone promotes alveolar remodeling.

- Approximately 4 to 6 months of orthodontic treatment should be anticipated in preparation for alveolar bone grafting.
- Pediatric dentists – Provide routine dental care. Pedodontists can also take occlusal radiographs which allow for preoperative visualization of the cleft site and allows for postoperative comparison.

Complications of Alveolar Grafting

Graft Exposure One of the most common complications is exposed bone. When the graft is exposed, the treatment is conservative therapy with antibiotics, chlorhexidine, removal of sequestrate, and optimizing hygiene. If secondary surgery is needed, a minimum of 3 months should be allowed between surgeries to allow for scar maturation.

Oronasal Fistulae The most common sites for development of fistulae are at the junction of the hard/soft palate and the incisive foramen. Smaller fistulae may be addressed with regional palatal flap repair. Larger fistulae can be treated also with regional palatal flaps, or with tongue flaps, facial artery myomucosal flaps (FAMM flaps), and temporalis flaps. May consider waiting for further growth prior to repair to avoid maxillary growth restrictions secondary to surgery if speech or function not affected.

Cleft Cases

Case 1

You have been consulted for a healthy 3-month-old male baby that presents to the craniofacial clinic with the following appearance (See Fig. 10.16):

Fig. 10.16 Cleft case image

- *What is your diagnosis?*

  This is a patient with unilateral complete cleft lip with associated complete cleft palate.

- *How would you treat this patient?*

  Once it has been determined that the patient has been deemed fit for surgery and has been evaluated by all members of the craniofacial team, I would plan for the primary lip repair. I would treat this patient with a primary lip repair using a modified Delaire technique (Use whatever technique you feel most comfortable with here. If you feel more comfortable using the description of the Millard technique, use that).

  After evaluation by anesthesia, I would recommend an oral RAE tube taped to the lower lip to confirm no tension on the upper lip. I would prep and drape the patient in a sterile fashion and make a mark with a surgical marker the edges of my flap design as well as making a mark with a syringe dipped in methylene blue. I would then inject the patient with lidocaine 1% with 1:100,000 epinephrine. Once 5 minutes have elapsed, I would I would then make an incision along these margins, removing the hypoplastic tissue at the cleft margins. Once the hypoplastic tissue is removed, I would dissect the orbicularis oris muscle from each side. Consistent with the Delaire technique, I would dissect bluntly the lateral crura and extend my dissection superiorly and laterally on the cleft side to achieve tension-free closure and to allow the deformed nose to be placed in a more symmetric position. I would first reapproximate the nasal floor by reapproximating the muscles on the lateral margins to the midline. This would reorient the perinasal musculature downward. Next, I would place on stitch in the fibrofatty tissue of the lateral nasal ala and subcutaneous tissue of the midline, and then tie to reapproximate the nostril opening on opposite side. I would then reapproximate the muscle layer with 4-0 polyglactin suture. This would reconstruct the orbicularis oris. Now that the muscle has been reapproximated, I would place one stitch to reapproximate the white line of the lip and then I would reapproximate the underlying mucosa. Lastly, I would reapproximate the skin flaps. After all suturing is complete, I would place a layer of topical skin adhesive in overtop the sutures.

- *Would you treat the palate?*

  No. I would wait until 12 months to treat the palate, as the normal time for treatment is 9–18 months. If completed during this time frame, the incidence of maxillary restriction is at its lowest point without affecting speech development (approx. 18 months).

- *Are cleft palates more common on the right or left side?*

  They are more common on the left.

- *Is there a proposed theoretical explanation?*

  In the embryological rodent model, it has been observed that the right palatal shelf reaches the horizontal position faster than the left during palatal shelf fusion. This delay is believed to occur also in humans which contributes to a higher incidence of left-sided palatal clefts.

- *The same patient returns to you at age 1 year old for palatal repair. How would you treat this patient?*

  I would close the palate at this point, using a Bardach two-flap technique. I would make incisions on either side of the cleft and dissect full thickness anteriorly until reaching the levator palatini muscles posteriorly. After this, my dissection would be split thickness and dissect the mucosa off of the muscle layer. I would then dissect the nasal floor and the muscle layer independently. Finally, I would reapproximate the tissue in layers, first with the nasal floor, then the muscle (reorienting them along the midline, which would reconstruct the palatoglossus, palatopharyngeus, levator veli palatini, and tensor veli palatini) and then the overlying mucosal layer.

- *During the palatal closure, you accidentally sever the greater palatine neurovascular bundle. What do you do?*

  Continue with the surgery. Although the known vascular supply has been severed, there

is still enough circulation from the palatal soft tissue pedicle. In essence, this flap changes from and "axial" pattern to a "random" pattern.

- *During the palatal closure, you have a tear in the mucosal component of your soft tissue flap. What do you do?*

  You can either excise the tear (usually done if a midline tear); or repair it; or place a layer of collagen membrane in the exposed area (the collagen membrane will only be sufficient, if there is an underlying muscle or nasal mucosal layer); or bring a buccal fat pad flap over as an additional tissue layer.

- *The patient follows up in 2 weeks and there is a fistula at the incision line. What do you do?*

  The size of the fistula would dictate treatment. Regardless of size, wait 6 months so that everything has scarred appropriately. Some fistulae resolve spontaneously. If the fistula is small and at midline, you can do a local closure or a redo of the palatoplasty. If it is large, you may need to do a revision palatoplasty. Other options include a buccal mucosal flap or a tongue flap.

- *The fistula resolves, but at age 3 the patient follows up and has been having speech problems, specifically a large amount of hypernasality. Why is this patient having these issues? How would you diagnose the patient?*

  The patient most likely has velopharyngeal insufficiency. This is an inability of the palate to create a separation between the nasal airway and the oral airway. The methods of detecting this include direct mechanism such as nasopharyngoscopy, multi-view video fluoroscopy, or indirect methods such as voice and resonance assessment by a speech pathologist. Considering the patient's age, a speech pathology evaluation is usually used due to less burden on the patient.

- *How would you treat the velopharyngeal insufficiency?*

  Pharyngeal flap surgery is indicated. Some surgeons will wait until age 5, but with a large degree of hypernasality verified by speech pathology, it is unlikely to resolve. If the patient has normal lateral movement of his pharynx, but difficulty in the midline, I would use a superiorly based pharyngeal flap (first dissecting to the prevertebral fascia, then splitting the uvula and insetting the flap into the mucosal pocket).

- *Preoperative work up reveals a diagnosis of Velocardiofacial syndrome. Are there any concerns with this type of patient?*

  Yes. Many of these patients have medially displaced carotid arteries. Dissection in the posterior pharynx could lead to vascular injury. This would require an MRA before surgery.

- *At what age would you treat the alveolar cleft and why?*

  Alveolar grafting is generally completed at 6–9 years of age based on development of the dentition. It has been described to graft once the formation of canine root is (1/2 to 2/3), but also important to carefully assess the development of the central and lateral incisor (if present).

- *Your orthodontist does not have a significant amount of experience in craniofacial patients and asks what you would like with presurgical orthodontics. What do you tell them?*
  - Expand collapsed segments.
  - Avoid displacement of the teeth into the cleft.
  - Remove traumatic crossbites with the mandible.
  - Position premaxilla for symmetry.

- *Where would you harvest the bone graft?*

  I would harvest a particulate bone graft from the iliac crest.

- *The patient's mother is extremely concerned about harvesting from the hip. What other sites could you harvest from?*

  Mandible, calvarium, tibia, or possibly rib.

- *What is BMP? Are there any side effects associated with using it?*

BMP is a protein complex belonging to a family of growth factors that promote osteoinduction. It is stored in the bone matrix and is available commercially for use in bone grafting. It can stimulate bone formation without the presence of bone graft and is usually placed in a carrier such as a collagen sponge.

Disadvantages of its use include a high cost and postoperative swelling at the graft site. Other disadvantages are ectopic bone formation and the theoretical risk of teratogenicity and carcinogenicity. In addition, it is FDA contraindicated for use in children. Considering that long-term studies have not been completed in pediatric patients and that the outcomes are equivalent to autogenous bone grafting in pediatric populations, BMP should be used only as a last resort.

- *How could you design/close your flaps?*

  Mucogingival flap designs include (lateral sliding flap was detailed above) the following:
  - Buccal Finger flap
    - Good blood supply.
    - Adequate soft tissue for closure.
    - Shortens the vestibule.
    - Provides nonkeratinized tissue in the area of the erupting canine.
  - Oblique Sliding Flap
    - Adjacent attached mucosa from lesser and greater segments brought to cleft site.
    - Adequate attached gingiva for tension-free closure of wide alveolar clefts.
    - Minor decrease in vestibular depth.
  - Lateral Sliding flap
    - Excellent blood supply.
    - Brings attached gingiva to area of cleft.
    - Tension-free closure.
    - Shortening of vestibule (although less).
    - Relies on secondary healing of mucosa on denuded area posterior to cleft.

  I would close the oronasal fistula first. This would create a floor to pack the bone graft. I would then close the palatal component creating a pocket to place the bone graft. I would then place the bone graft and then close the buccal flaps over top making sure to create a watertight closure. In addition, I would place a premade splint in the maxilla to stabilize the premaxilla during the bone grafting period. This splint also aids in patient compliance with the no chew diet.

- *What if you can't close the buccal flaps?*

  You can create a back cut at the posterior edge or make cuts in the periosteum to give additional laxity.

- *Why shouldn't you elevate medially in the premaxilla region to gain tissue laxity?*

  Elevation medially towards the premaxilla may compromise blood supply.

- *During the transfer of the bone graft to the mouth, the assistant drops the bone graft on the ground. What do you do?*
  - If you have prepped and consented for the other hip, you can discard the bone graft and harvest from the opposite hip.
  - If you would prefer to save the bone graft, you can wash the bone for 3 minutes with normal saline, place the bone in 10% betadine or 0.04% chlorhexidine for 20 minutes and then wash again for 3 minutes with normal saline.
  - You could harvest at another location after receiving parental consent.
  - You can close the wounds and return another time after the marrow regenerates.

- *The patient returns to the clinic and particles of bone are seen protruding from the bone graft site. The parents report that they have been spitting these particles out for the past few days. What do you do?*

  Place the patient on regimen of peridex and perform gentle superficial debridement with follow-up until the area is fully mucosalized. Although a portion of the graft has been lost, there may be some underlying osseoinduction. Take postoperative radiographs in 3 months and if there is no continuity, you can attempt the procedure again. The other option is orthognathic surgery when growth is complete.

- *The patient has a remaining oronasal fistula. When would you operate?*

  Any surgery should be delayed for 3 months to allow full integration of the graft. If the patient still has an oronasal fistula, a secondary procedure using local tissue rearrangement can be completed if necessary or can be delayed until orthognathic surgery.

Case 2

- *A 4-month-old male with an isolated cleft lip is brought by his mom to your office for consultation. Describe what you see (*Fig. 10.17*).*

  Complete right unilateral cleft lip with asymmetry of alar base of the nose.

- *Is cleft lip more common on the right or left? Is it more common in males or females?*

  Isolated cleft lips are more common on the left and is most common in males.

- *How would you proceed?*
  - Review medical history and pertinent perinatal history.
  - Review any other abnormalities/syndromic features. Ask if any other specialists are involved in his care. Ask about history of clefting in the family and issues with feeding.
  - Perform physical exam.
  - Explain to the mother the protracted course of treatment and the need for multidisciplinary care.

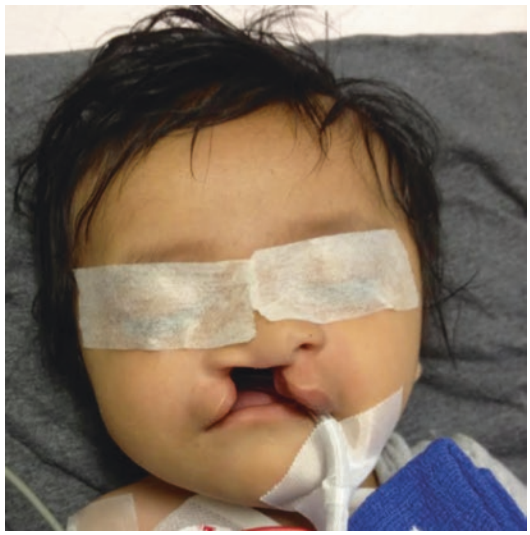

Fig. 10.17 Complete right unilateral cleft lip with asymmetry of alar base of the nose. (Image Courtesy of Dr. Damian Findlay)

- *At what week of gestation, could a cleft be identified?*

  Ultrasound can identify clefting as early as 13–14 weeks of gestation. The sensitivity in detecting CL ± CP prior to 18–20 weeks is much lower.

- *What are associated risks of cleft formation?*

  Chemical exposure (e.g., alcohol or cigarette smoke), radiation, viral infection, maternal hypoxia, teratogenic drugs (e.g., valproic acid and benzodiazepines), nutritional deficiencies, and genetics may play a role.

- *What can a mother do to reduce her chance of her having a child with a cleft?*

  Avoid exposure to known teratogens and take folic acid during pregnancy.

- *What is the difference between a complete and incomplete cleft of the lip?*

  An incomplete cleft only involves the part of the lip and/or alveolus. A complete cleft lip extends into the nares and the entire lip and associated musculature.

- *From an embryological standpoint how does of cleft lip form?*

  Lack of fusion of the median nasal processes and lateral maxillary processes.

- *What is the sequence of management you would expect for a patient with CL?*
  - Primary lip repair (cheilorhinoplasty), timing based on the rule of 10's.
  - Nasolabial revision is delayed until ages 3–5 years.
  - Alveolar grafting is generally completed at 6–9 years based on development of the dentition.

- Early orthodontics begins in the mixed dentition stage with the goal of improving arch form and width, allowing more space for eruption of teeth, and opening fistulae for surgical access.
- Orthognathic surgery if required is planned for 14 to 16 years of age in females, 16 to 18 years in males to address maxillary hypoplasia common in cleft patients.
- Rhinoplasty is often delayed at least 6 to 12 months following maxillary surgery due to changes in septum and tip support with movement of the anterior nasal spine.
- Cleft Scar Revision: Any time after 5 years of age, but preferably at the time of alveolar bone grafting.

- *What is the goal of cleft lip repair?*

  A three-layer closure of skin, muscle, and mucosa that approximates normal tissue and excises hypoplastic tissue at the cleft margins.

- *What are some surgical maneuvers to correct a cleft lip deformity?*
  - Tennison-Randal – a Z-plasty technique.
  - Delaire – rotation-advancement technique.
  - Millard – rotation-advancement technique.

- *Patient presents to your office and is unhappy with his cleft lip repair. What do you see?*

  This is a whistle lip deformity. It is caused either by scar retraction or inadequate reapproximation of the margin portion of the orbicularis oris and skin. This is usually treated with a V-Y plasty and ensuring correct reapproximation of the underlying muscle (Fig. 10.18).

Fig. 10.18 Whistle lip deformity

References

1. Parker SE, Mai CT, Canfield MA, Rickard R, Wang Y, Meyer RE, et al. Updated national birth prevalence estimates for selected birth defects in the United States, 2004-2006. Birth Defects Res A Clin Mol Teratol. 2010;88:1008–16.
2. Dixon MJ, Marazita ML, Beaty TH, Murray JC. Cleft lip and palate: understanding genetic and environmental influences. Nat Rev Genet. 2011;12:167–78.
3. Goldstein JA, Losee JE. 23 - pediatric plastic surgery. 7th ed. Elsevier Inc: Zitelli and Davis' Atlas of Pediatric Physical Diagnosis; 2018.
4. Campbell K. 23 - imaging of the face and neck. In: Creasy and Resnik's maternal-fetal medicine: principles and practice. 8th ed: Elsevier Inc; 2019.