Cleft Lip and Palate

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Introduction

Cleft lip with or without palate (CL ± P) and cleft palate in isolation (CPO) constitute the *most common congenital defects of the face* (second most common overall developmental defect following clubfoot). Despite similarities and overlapping characteristics, CL ± P and CPO should be considered as individual disorders with different embryologic, etiologic, and epidemiologic factors. Most current evidence points to CL ± P and CP being multifactorial in nature with innumerable contributions of both genetic and environmental factors. Additionally, patients with these conditions can have concomitant health issues including otologic disease (e.g., OME, ETD, rAOM, cholesteatoma), nutritional deficits, speech and language problems, dental deformities, sleep apnea, concurrent facial growth deformities, and psychosocial difficulties. Given the complexity inherent to these disorders and their treatment, a comprehensive management plan necessitates a multidisciplinary team approach including the otolaryngologist, plastic surgeon, speech-language pathologist, geneticist, pediatrician, nutritionist, orthodontist, and orthognathic specialist. Antepartum diagnosis is now a standard practice in most communities, and early diagnosis allows for parental counseling and enrollment into a cleft treatment program.

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Epidemiology

- Distribution of clefts: 50% cleft lip and palate (CL ± P); 30% isolated cleft palate (CPO) and 20% cleft lip (CL) alone.
- CL ± P—in North America 0.2–2.3 cases per 1000 population and varies by ethnic groupings—3.6/1000 in Native Americans; 2.1 per 1000 in Asians; 1 in 1000 in Caucasians; 0.41/1000 in African-Americans:
  - Males/females:1.5:1
- CPO—0.1–1.1 cases per 1000; females/males, 2:1; females more likely to develop cleft palate (due to later closure of palatal shelves during development).
- CL—distribution 6:3:1 for left-sided CL, right-sided CL, and bilateral CL, respectively:
  - Right-sided CL more commonly associated with syndromes
- CL ± P associated with syndromes (<20% of total cases)—most commonly associated syndrome is van der Woude (see below):

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  - Approximately 85% of bilateral cleft lips and 70% of unilateral CL associated with cleft palate
- CPO more commonly associated with syndromes than CL ± P—most commonly associated with autosomal dominant syndromes with microdeletion or addition at chromosome 22q11.2 (DiGeorge, velocardiofacial, conotruncal anomaly face syndrome):
  - 22q11.2 syndromes have incidence of 1 in 4000 live births.
  - CPO associated with syndromes 55% of the time.
- Approximately 30–65% of facial clefts appear to be associated with named syndromes.
- Recurrence risks: Table 13.1.

Etiology

Environmental

- Multifactorial for most non-syndromic cases of clefting.
- Maternal health—Both prepregnancy DM (though not gestational diabetes) and maternal obesity are known to increase risk. Meta-analysis has determined maternal age is not a factor:
  - Alcohol consumption during pregnancy, although definitive relationship with in utero alcohol consumption remains unclear.
  - Smoking, particularly high levels, are consistently associated with increased risk for clefting in numerous studies. Risk appears stronger for CL ± P than CPO.
  - Poor folic acid and vitamin intake, folate antagonists associated with increased risk of clefting (folate supplementation may play protective role though evidence more convincing for protective effect in neural tube defect).
- Use of valproic acid, phenytoin, retinoic acids, dioxin, thalidomide, corticosteroids.

Table 13.1 Risk of cleft lip with or without palate and cleft palate in isolation in subsequent children based on prior history of cleft disorders in relatives

Percentage likelihood of next child having cleft defect		
	CL ± P	Cleft palate
No family Hx of CL ± P or cleft palate	0.10%	0.04%
Parents normal, first child is affected		
No affected relatives	4%	2%
Affected relatives	4%	7%
Parents normal, two affected children	9%	1%
Parents normal, two affected relatives	9%	10%
One parent affected, no children affected	4%	6%
One parent affected, one child affected	17%	15%

Table adapted from *KJ Lee's Essential Otolaryngology*, and Bailey's 5th edition, Ch. 103, Comprehensive Cleft Care

Syndromes/Associated Disorders

- *Van der Woude Syndrome* (*VWS*)—most common syndrome associated with CL ± P; autosomal dominant (AD) inheritance, lower lip pits/sinus tracts, hypodontia; mutations of *interferon regulatory factor gene* (*IRF6*) felt to contribute to this disorder.
- *Pierre Robin Sequence* (*PRS*)—*micrognathia, glossoptosis, and CP*:
  - Micrognathia prevents tongue descent; retrodisplacement of the tongue → abnormal palatal development with clefting (characteristically U-shaped) or high-arched palate and respiratory distress especially in the supine position; most commonly *associated with Stickler syndrome* (14–34% of cases).
  - *Children with PRS should undergo routine ophthalmologic evaluation in the first year of life* to rule out concurrent eye issues.
- *Stickler syndrome*—autosomal dominant with variable expression; SNHL, characteristically contains Pierre Robin sequence, ocular abnormalities, and arthropathies. Ocular anomalies include early (first decade) myopia that can be severe, retinal abnormalities, and glaucoma. Arthropathies relate to connective tissue disorders—marfanoid features, hyperextensible joints, kyphosis, and scoliosis.

- *22q12 deletion*: most frequent interstitial deletion known to be causally associated with clefting in humans:
  - Associated with wide phenotypic spectrum including DiGeorge syndrome, velocardiofacial syndrome (VCFS).
  - Palatal anomalies; most commonly VPI (29–50% of cases).
  - Also associated with cardiovascular malformations, immune deficiencies, and neurodevelopmental disabilities.
  - Additional features: elongated, flattened midface with malar hypoplasia, broad nasal base, low-set ears, thickened helical rim, micrognathia, microcephaly, dysphagia, pharyngeal hypotonia, middle ear disease, CHL, and chronic suppurative OM.
  - Adenoidectomy and operative pharyngeal procedures should be performed carefully as incidence of VPI is high following surgery; *medial course of the carotid artery* may contribute to increased risk of carotid injury during pharyngeal surgery.
  - *Dx: FISH testing*.
- *Velocardiofacial syndrome* (*Shprintzen* syndrome)—deletion at 22q (same locus as *DiGeorge* syndrome and overlap exists between the two syndromes); affected children have clefts, cardiac anomalies, and characteristic facies (as above):
  - Patients with clefts and the following associated findings should be tested for DGS/VCF deletion: any cardiovascular malformation, short stature, microcephaly, developmental delay, immune deficiency, hx/Fhx of psychiatric disease, and facial dysmorphism.
- *Ectrodactyly-ectodermal dysplasia-clefting syndrome*: lobster claw anomaly of all four extremities, typically have bilateral CL with CP; but unilateral CLP and CP alone reported; ocular findings: related to absence of lacrimal puncta → epiphora, blepharitis, keratoconjunctivitis, corneal ulcers.
- *Popliteal pterygium syndrome—IRF-6* gene disturbance; similar to VWS with CL ± P and presence of popliteal pterygium/webbing, syndactyly, genital abnormalities, intraoral adhesions (syngnathia), pyramidal skin on hallux, ankyloblepharon.
- Other disorders associated w/ clefts: Down's (trisomy 21), oculo-auriculo-vertebral, hemifacial microsomia/Goldenhar (constellation of first, second branchial arch abnormalities), Kabuki (CP, arched eyebrows, long palpebral fissures, flat nose), Treacher Collins (AD, colobomas, low-set ears), orofacial-digital syndrome (x-linked, oral clefting, mandibular hypoplasia).

Embryology

The development of the lip and palate occurs sequentially rather than coincidentally and varying degrees of clefting can occur; the degree of clefting is based on the point in fetal development when the fusion process is interrupted.

Lip Development

Beginning at the end of the fourth embryological week, five structures surround the primitive stomodeum and will ultimately give rise to structures of the face, nose, and lips. These include the frontonasal prominence and the paired neural crest-derived facial prominences (maxillary and mandibular prominences), which appear from the first pair of pharyngeal arches.

Frontonasal Prominence (FNP) Mesenchymal proliferation ventral to developing forebrain:

- FNP → nasal dorsum, forehead, contributes partially to the septum (downgrowth from posterosuperior aspect of FNP; other contributions from fused medial nasal processes)

Nasal Placodes Ectodermal thickenings at ventrolateral aspect of FNP; placodes invaginate (at fifth week) to form nasal pits → primitive nasal cavity internally and medial (MNP) and lateral nasal processes (LNP):

- LNP—forms nasal alae and sidewall; nasolacrimal groove between LNP and maxillary process (failure of fusion → oblique cleft [rare]).
- MNP—right and left MNPs fuse to form *intermaxillary segment* (sixth week; failure of fusion → midline cleft, associated with holoprosencephaly); fusion forms the philtrum, medial upper lip, nasal tip, and columella:
  - Intermaxillary segment → primary palate (see below)

Maxillary Prominences (MP) First arch derivative; sixth to seventh weeks; MPs grow medially and ultimately fuse with MNPs (mid-fifth week) to give rise to the upper lip; MPs → lateral upper lip; also have significant contribution to secondary palate (see below):

- NOSE: Formed from five facial prominences FNP = bridge/dorsum; fused MNPs = tip and columella, and LNPs = nasal alae and sidewall
- UPPER LIP: MNPs = medial upper lip, philtrum; MPs = lateral upper lip (from philtral column laterally); NO contribution of LNPs to formation of the upper lip; development usually complete by the end of the seventh week
- *Lack of fusion between maxillary process and medial nasal process → CL.*

Palate Development

Weeks 5–12 (palatal fusion completed later in females) MPs grow and push the MNPs medially → MNPs fuse not only at surface giving rise to the lip but also at deeper levels to form the premaxilla, alveolus, and primary palate; once primary palate fully developed, the secondary palate begins to develop; during the sixth week of embryonic development, palatine shelves are directed obliquely downward on either side of the tongue; seventh week → palatine shelves migrate inferomedially to lie horizontally above the tongue; palatal fusion occurs from anterior to posterior and simultaneously fuses with nasal septum which descends from above (derived from FNP); at the eighth week, the tongue begins to withdraw/descend from its position between lateral maxillary prominences (which form secondary palate).

Primary Palate Derived from intermaxillary segment after fusion of MNPs = central lip (philtrum), columella, nasal tip, premaxilla, central maxillary alveolar arch, which accommodates four anterior teeth (medial and lateral incisors) and hard palate anterior to incisive foramen.

- *Lack of fusion between maxillary process and medial nasal processes/intermaxillary segment → primary CP*

Secondary Palate Part of the hard palate posterior to incisive foramen as well as lateral hard and soft palate; formed by fusion of palatine shelves (from maxillary prominences); makes up the majority of palate including the hard palate posterior to incisive foramen and all of the soft palate; development begins after completion of primary palate.

- *Lack of fusion in the midline between two palatine shelves (from lateral maxillary processes) → secondary CP*

Fusion of alveolus *between canine (cuspid) and lateral incisor*, i.e., *alveolar defect* between these teeth and clefts in this location can affect development and eruption of adult dentition.

Types of Clefts/Classification

Cleft Lip

- *Complete unilateral CL*—the entire vertical thickness of the upper lip and often associated with alveolar cleft because the lip and primary palate share the same embryologic origin; implies separation of medial and lateral lip with absence of all layers—skin, muscle, mucosa, and alveolar bone with extension to the nasal sill/floor.
- *Incomplete unilateral CL*—can contain muscle fibers within the cleft, residual fibers

known as *Simonart's band* (bridge or bar of lip tissue of variable size that usually consists of skin only, although some histological studies have shown that some muscle fibers lie within band). The band, which is more frequently observed in UCL than BCL, is located at the base of the nostril and associated with a higher frequency of maxillary lateral incisor development in the maxillary process.
- *Bilateral CL*—prolabium of complete bilateral clefts is devoid of muscular fibers and is attached at columella only (tenuous blood supply), although may have muscle fibers in incomplete clefts; the premaxilla can be horizontally oriented and may require molding/manipulation of cleft components prior to definitive repair.
- *Microform CL*—diastasis of orbicularis fibers with presence of alveolus, skin, and mucosa.

Cleft Palate

Unilateral or bilateral and extent may be classified as complete or incomplete. CP is classified according to location relative to the incisive foramen:

- *Clefts of primary palate* occur *anterior* to incisive foramen.
- *Clefts of secondary palate* occur *posterior* to incisive foramen:
  - Unilateral cleft of secondary palate is defined as one in which the palatal process of the maxilla on one side is fused with the nasal septum.
  - Bilateral cleft of the secondary palate has no point of fusion between the maxilla and the nasal septum.
  - Cleft palate in isolation (CPO) usually involves the secondary palate only and has varying degrees of severity.
- *Complete CP*—palate involves both the primary and secondary palate and includes one or both sides of the premaxilla/alveolar arch and frequently involves cleft lip.
- *Incomplete CP*—clefting of *secondary palate only*.
- *Submucous cleft palate* (SMCP)—least severe incomplete cleft; bifid uvula, thinning of central soft palate with translucent appearance (zona pellucida), and palpable notch in posterior aspect of the hard palate; patients at high risk for VPI as levator veli palatini has similar configuration to what is seen in true cleft palate with central diastasis and abnormal longitudinal orientation of muscle fibers → Furlow palatoplasty may be effective Tx (see below).
- *Veau classification of cleft palate*:
  - I—Soft palate only
  - II—Hard and soft palate
  - *III*—Unilateral complete cleft
  - *IV*—Bilateral complete cleft

Muscular and Vascular Anatomy

Lip Divided into red (convex) and white lip (concave) separated by mucocutaneous junction/vermilion border:

- *White roll*—epithelium just above vermilion border, reflects ambient light and is critical landmark in cleft lip reconstruction.
- *Vermilion*—dry portion of lip's red mucous membrane, lacks pilosebaceous units, salivary glands, eccrine glands.
- *Superior labial artery*—major blood supply to the upper lip; normally anastomoses in the midline; courses on undersurface of the orbicularis muscle:
  - In unilateral cleft—aberrant vascular supply on lateral aspect of cleft is better developed than on medial side; artery courses along margin of cleft, anastomosing with either angular or lateral nasal artery at base of the nose.
  - Incomplete cleft—thin, terminal branch of superior labial artery crosses bridge.
  - Bilateral clefts—artery is underdeveloped in prolabial segment which primarily derives its blood supply from the septal, columellar, and premaxillary vessels.
- *Orbicularis oris*—principal muscle of the lip; not a true sphincter as superficial and deep parts of muscle arise separately from the

modiolus; critical role in lip function and appearance; reestablishment of muscular sphincter is critical in achieving appropriate surgical outcome; superficial layer arises from the dermis and passes obliquely to insert into the mucous membrane lining the inner surface of the lips; deep fibers arise from the maxilla and mandible:
  - Orbicularis in CL: Muscle is hypoplastic and has abnormal attachments/insertion in CL—no muscle crosses the cleft in complete clefts; the skin bridge (Simonart's band) in incomplete clefts also contains no functional muscle (muscle fibers do not cross gap unless skin bridge at least 1/3 total lip height); muscle is more hypoplastic on medial side of cleft; abnormal insertion at alar base and anterior nasal spine; fibers parallel cleft margin.

Palate

Muscular Anatomy of the Palate and Pharynx

- *Tensor veli palatini* (TVP): *V3*; tenses palate and opens ET during swallowing; from base of internal pterygoid plate and lateral aspect of torus tubarius; terminates in tendon/aponeurosis after winding around hamulus.
- *Levator veli palatini* (*LVP*): *pharyngeal plexus* (derived from IX, X, XI) origin: petrous bone and medial torus, fibers occupy central 40–50% of the soft palate and fuse with contralateral LVP; palatal sling causes superior and posterior motion of the soft palate; causes elevation of the soft palate during deglutition.
- *Musculus uvulae*: pharyngeal plexus; adds bulk to dorsal surface of the soft palate.
- *Palatoglossus*: pharyngeal plexus; forms anterior pillar; lowers and positions velum, elevates the tongue.
- *Palatopharyngeus*: pharyngeal plexus; forms posterior tonsillar pillar; constricts pharyngeal isthmus *narrows VP orifice*, superior heads (2) clasp LVP as it enters the velum; fibers intermingle with SPC at posterolateral pharyngeal wall, action: retrodisplacement and downward motion of the velum; antagonist to LVP and position of the velum during normal function is the net result of action of LVP and palatopharyngeus; stretches posterior free margin of the velum; acts with SPC for medial movement of lateral pharyngeal wall below the level of the hard palate.
- *Superior pharyngeal constrictor* (*SPC*): pharyngeal plexus; *medial movement of lateral pharyngeal walls*; relative activity of SPC may contribute to different patterns of VP closure (coronal, minimal SPC fxn; sagittal, high degree of SPC activity during closure; circular, moderate); *main component of Passavant's ridge*.

In CP, VP muscles can be hypoplastic and generally have abnormal course and attachment:

- TVP—normally attached to posterior hard palate; in CP, aponeurosis is incomplete with abnormal attachment at posterior hard palate, results in ETD.
- LVP—in CP and SMCP has abnormal longitudinal orientation with *abnormal insertion to posterior aspect of hard palate, SPC, and TVP aponeurosis*; abnormal LVP appears to be primarily responsible for VP dysfunction.

Arterial Supply of Palate

- Hard palate—Greater palatine artery with some anterior contributions from arterial branches in incisive canal (anterior palatine artery)
- Soft palate—Ascending palatine (facial artery), greater palatine (internal maxillary), ascending pharyngeal branch, and contribution of the lesser palatine (branch of the greater palatine)

Treatment

Table 13.2 provides a general outline for timing of intervention in patients with cleft lip and palate.

Table 13.2 Timing of intervention in patients with CL ± P and CPO

Timing of intervention in patients with cleft deformities	
Presurgical orthopedics/ intervention	1–2 weeks
Speech eval—feeding	
Cleft lip repair	3 months
Primary rhinoplasty	
Tympanostomy tube placement	
Palatoplasty/palate repair	9–18 months
T-tube/long-term PE tube placement	
Speech evaluation	3–4 years old (as early as 2 years)
Velopharyngeal work-up/ surgery	4–6 years old
Repair of alveolus	6–8 years old
Intermediate rhinoplasty	7–10 years old
Definitive septorhinoplasty	16–19 years old
Orthognathic surgery	

Table adapted from Park Facial Plastic Surgery: Essential Guide

Rule of tens 10 weeks old, hemoglobin 10 g/dL, weight 10 lb; partially based on anesthetic safety; incomplete clefts have less urgency to repair as Simonart's band tends to hold alveolar segments in place as growth occurs.

Oral intake can be compromised as a result of inability to feed (sucking mechanism impaired—child is unable to form a seal due to incomplete muscular sphincter and/or escape of air through cleft palate); goal should be weight gain of 0.5–1 ounce/day; *Mead-Johnson, Haberman, or Pigeon* nipple can be used to facilitate feeding; feeds should be done in upright position, as cleft patients tend to swallow more air and require frequent burping.

Presurgical Infant Orthopedics (PSIO)

Preoperative manipulation of alveolar segments in complete CL ± P is performed particularly in wide clefts to facilitate closure; most important in bilateral complete cleft lip in which control of premaxillary segment is critical in achieving satisfactory surgical outcome; ideally, PSIO should be started in the first 1–2 weeks of life.

Techniques

- *Taping*—requires family compliance to properly apply tape across cleft: doesn't address nasal deformity.
- *Head bonnet*—applies pressure across the premaxilla; removable and easier than taping.
- *Latham* appliance—mold with pins used to create more favorable alveolar position.
- *NAM* (*nasoalveolar molding*)—molding and repositioning of alveolar processes, nasal cartilages, and lengthening of deficient columella to create lasting aesthetic outcome and reduce need or minimize the extent of secondary surgical revision procedures; significantly improves nasal symmetry over surgery alone; once alveolar segments in close proximity, nasal stent is added to mold distorted nasal cartilages; the alar cartilage is lifted by intranasal stent to achieve normal elevation and symmetry:
  - *NAM in bilateral cleft patients* used to lengthen columella, reposition nasal cartilages toward the tip, and align the alveolar segments; first stage in bilateral molding consists of retracting and straightening the everted premaxilla into space between two lateral alveolar segments; second stage involves nasal stent incorporated into anterior rim of molding plate.

Gingivoperiosteoplasty—closure of soft-tissue alveolar segments; possible if there is close approximation of the alveolar cleft segments; use of NAM and presurgical approximation of alveolar defect provides the surgeon with the option to perform GPP at the time of lip closure.

Lip adhesion procedure in which cleft segments are united via myomucosal flaps, essentially creating an incomplete cleft; contributes to molding of the alveolar segment and reducing cleft width; secondary procedure then converts the lip adhesion to a formal lip repair; approximation of the orbicularis oris creates muscle pull on the maxillary alveolar segments, which is critical for molding; alternative for those unable to tolerate NAM or other orthopedic devices.

Cleft Lip Repair

Goals: creation of symmetrical nasal tip and alar bases, Cupid's bow, and lip fullness without loss of normal philtral contour with fullness of labial mucosa that is equivalent on both sides.

Techniques

Goals: restoration/reconstruction of normal lip anatomy and function by reestablishing continuity of the orbicularis; secondary goals—closure of the nasal floor and correction of nasal tip asymmetry.

Straight-line repair: poor orbicularis closure may result in lip shortening, tissue waste, and unappealing scars with potential for scar contracture; largely a historical procedure.

Geometric closures, modified Z-plasties, quadrangular flaps, triangular flaps—designed to decrease the amount of lip shortening that may occur and to improve orbicularis oris muscle function.

Millard Rotation-Advancement Flap most commonly used technique in the USA:

- Incorporates two opposing flaps with the medial lip rotated downward and lateral lip advanced medially to restore the lip integrity:
  - Must be designed to correct vertical height discrepancy from Cupid's bow to the columella.
  - Difference accounts for length and design of rotation flap and back-cut.
- Technique (Fig. 13.1): Methylene blue used for skin marking as outlined below; rotation flap incision is made first (point 3 → 5 with back-cut to x as needed) and is taken through the entire thickness of the orbicularis and oral mucosa to allow for complete release; c-flap remains and may be used to add additional height to columella; the advancement flap starts with incision from point 8 → 9 and this incision is then continued along the alar base; the gingivolabial sulcus is incised and incisions are taken down to face of the maxilla; bilateral undermining assists in closure; first deep stitch reapproximates the orbicularis at back-cut and second stitch at vermilion; mucosa, muscle, and skin closure follows.
- *Advantages*: flexible—can be applied to a wide variety of cleft lips; allows continuous modifications during the design, incisions, and execution of the repair; minimal discarding of tissue; good nasal access; camouflage of suture line along philtral column.
- *Disadvantages*: requires experienced surgeon; possible excessive tension—may cause constriction of maxillary growth; extensive undermining required; vertical scar contracture with possibility of vermilion notching if improperly designed; tendency toward small nostril.

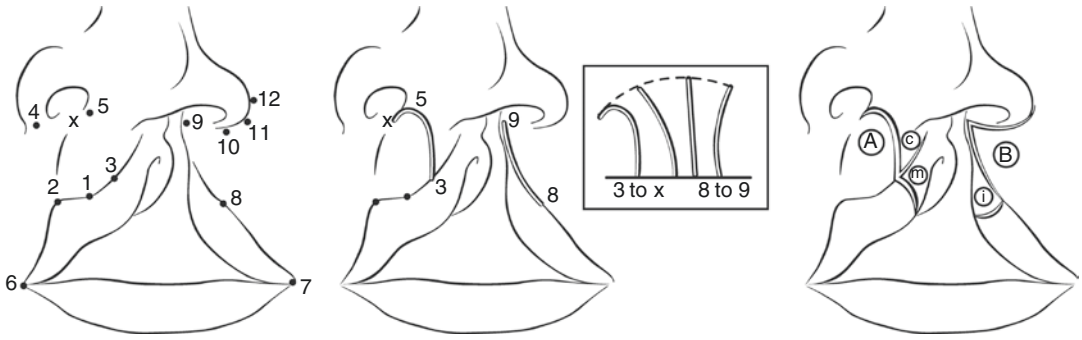

Fig. 13.1 *Left*: Anatomical landmarks used in the Millard rotation-advancement flap technique for repair of unilateral cleft lip deformity. *Middle*: The length of the rotation flap (non-cleft side) and advancement flap (cleft side) is equivalent in length (see *inset*). *Right*: Following incision, the flaps are advanced and rotated (A and B), while C contributes to repair of the nasal sill. (After Sykes and Tollefsen Management of Cleft Lip Deformity, depicting Millard Rotation-Advancement Flap)

Tennison-Randall Repair second most commonly used technique:

- Modified Z-plasty with a lateral, inferiorly based triangular flap
- *Advantages*—rebuilds the floor of the nostril; adds length to the medial lip element; preserves Cupid's bow; adds needed tissue volume in the lower one third of the lip
- *Disadvantages*—violation of normal philtral column on the non-cleft side, creates more prominent scarring across philtrum; requires exact presurgical measurement and lacks flexibility in surgical application

Bilateral Cleft Repair (Fig. 13.2)

The premaxilla can be quite protrusive and may be "locked out" from palatine segments requiring pre-op manipulation to achieve an appropriate repair via PSIO or lip adhesion → gives three segments best configuration to achieve subsequent repair; reestablishing continuity of the orbicularis is critical for optimal functional and aesthetic result; single-stage repair of bilateral deformity usually provides best chance of symmetrical outcome; staged repair of the bilateral cleft tends to create an asymmetrical result due to disparities of the two sides in terms of facial growth following

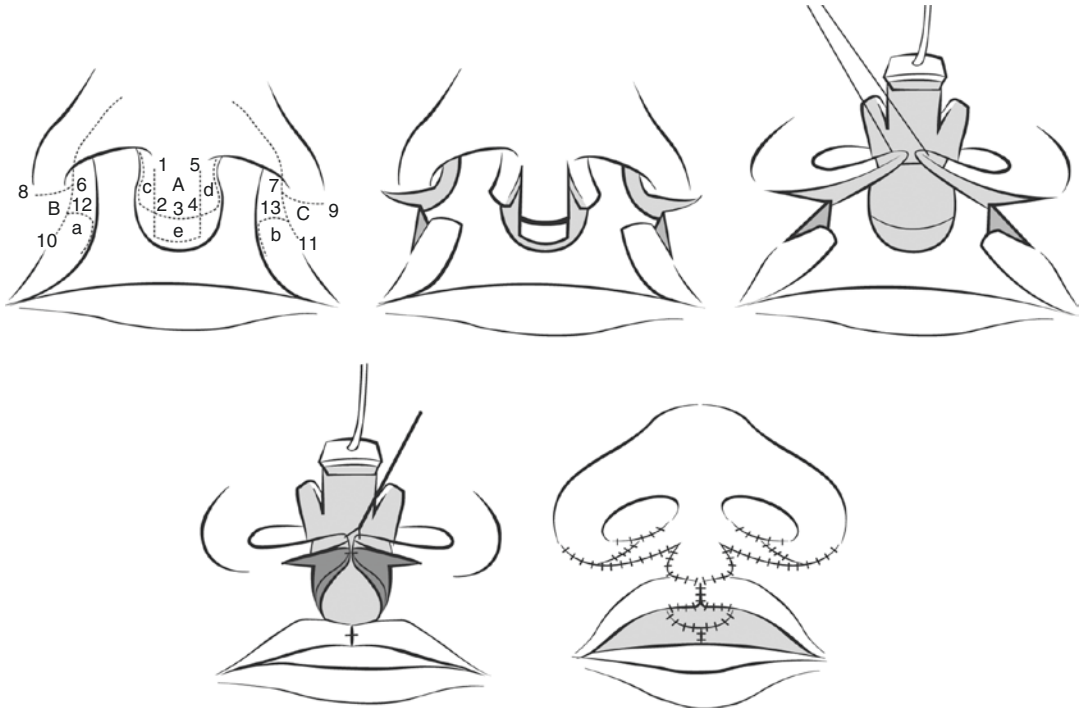

Fig. 13.2 *Top left*: Anatomical landmarks and incisions used for repair of the bilateral cleft lip deformity. *Top middle*: Incisions made in prolabial and lateral flaps in anticipation of flap advancement. *Top right*: De-epithelialized tips of lateral flaps advanced and sutured in place medially to the premaxillary periosteum. *Bottom left*: Tips of lateral flaps sutured medially and reconstruction of orbicularis muscle sling. *Bottom right*: Prolabium and fork flaps sutured in place. (After Seibert Surgical Repair of the Bilateral Cleft Lip Deformity)

"revascularization" of repaired lip; asymmetry is oftentimes difficult to correct at a later stage, and staged repair precludes muscle from being advanced across the prolabial segment.

Cleft Nasal Deformity

Unilateral Cleft Nasal Deformity (Fig. 13.3)

- Abnormal insertion of the orbicularis muscle at anterior nasal spine and alar base on cleft side.
- Cleft alar base displaced *inferiorly, laterally, and posteriorly* due to maxillary insufficiency and action of the orbicularis.
- Septum: caudal and inferior septum deviated to non-cleft side (away from the cleft); upper and posterior portions of the septum deviate to the cleft side.
- Columella is shortened on the cleft side and the nasal tip is deflected to the cleft side.
- LLC has normal size with a poorly projecting dome, alar flattening, and a horizontal nasal shape secondary to a short medial crus and long lateral crus.
- Deficiency and displacement of maxillary segment with lack of piriform aperture.
- Alar-facial groove is often absent and the ala attaches to the face at an obtuse angle. Absence of the nasal floor and sill commonly seen with relative stenosis of the internal nostril on the cleft side.
- Internal nasal valve collapse may occur due to the relative flaccidity of the ULC on the cleft side.
- External valve collapse also possible due to introversion of cleft ala.
- *Bilateral Cleft Nasal Deformity*—similar defects to those seen in unilateral CL nasal deformity, but present on both sides to varying degrees (if asymmetry of CL exists):
- Poorly projected tip with wide alar base.
- Alar bases positioned similarly to unilateral CL due to pull of abnormally inserting orbicularis oris and bilateral absence of maxillary segment → posterior, lateral, inferior displacement.
- Extremely short columella and the medial crura are displaced into the prolabium.
- Prolabium and premaxilla can be markedly displaced anteriorly with poor overall blood supply.
- Nasal tip is broad and flattened and the nasal domes are separated resulting in a bifid appearance.
- Depending on presence/absence of CP, the septum may not articulate with palatine shelves in bilateral complete CP; unilateral complete CP → the septum attaches to non-cleft side.

Fig. 13.3 Anatomical findings associated with unilateral cleft lip nasal deformity. (After Dutton, Bumsted Management of the cleft Lip Nasal deformity)

Treatment of Nasal Deformity

Correction begins as early as 1 week after birth with NAM (see above) → *positions CL and LLC for primary repair*; primary repair done at the time of lip repair; intermediate repair usually occurs at ages 7–11 years after alveolar bone grafting; definitive septorhinoplasty follows completion of growth after adolescence; staged nasal repair allows smaller corrections with each procedure:

- Goals: reconstruction of the nasal floor and sill, repositioning of alar base, columellar lengthening on cleft side, correction of

deformed ala, provision of adequate nasal tip support and symmetry, straightening of nasal dorsum, attainment of adequate nasal airway

Primary Rhinoplasty

- Does not affect nasal or midfacial growth
- Performed at the time of Millard rotation-advancement procedure
- Cleft LLC separated from overlying skin-soft-tissue envelope via alar base (laterally, the alar base is separated from its attachment to displaced maxilla and abnormal orbicularis insertion) and columellar incisions with intercrural dissection
- Alar base sutured in more medial position after reconstruction of the nasal floor (mucoperiosteal flaps from lateral nasal wall and mucoperichondrial flap from the septum)
- LLC sutured in a more anatomic position and secured via internal and external bolster

Intermediate Rhinoplasty

- Most secondary deformities can be repaired at age 7–8 years when lower nasal complex can be addressed.
- Alveolar bone grafting usually precedes intermediate rhinoplasty as eruption of lateral incisor and canine, following grafting, provides adequate base for nasal repair.
- Nasolabial fistula also corrected prior to intermediate rhinoplasty to release abnormal pull on alar base.
- Judicious septal surgery at the time of intermediate rhinoplasty does not appear to interfere with subsequent growth of the face or nose.
- Repositioning of the caudal septum in the midline and reconstruction of the nasal tip and alar cartilages are appropriate at this stage.
- Performed via open technique for adequate visualization and accurate repair.
- Technical points (although these may be delayed depending on severity of deformity at the time of intermediate rhinoplasty)—dissecting pocket between the medial crura; freeing the caudal septum from abnormal attachment with suture fixation to anterior nasal spine; repositioning abnormal dome in more medial and superior location (lateral crural steal); V-Y advancement from the upper lip to lengthen cleft-side columella.

Definitive Septorhinoplasty

- Definitive correction includes septoplasty, osteotomies, and dorsal revision performed after completion of nasal growth.
- Complete and aggressive restructuring of internal and external anatomy can be performed.
- Done via open approach with or without alotomy.
- Graft materials: usually from the septum, via separate hemitransfixion approach with correction of bony and cartilaginous deviation.
- Columellar strut with fixation of LLCs enhances nasal projection with more advancement of cleft-side LLC.
- Repositioning of LLC may be necessary with suturing to ULC.
- Tip graft can improve tip definition.
- Unilateral spreader on cleft side can be used to address INV collapse.

Cleft Palate Repair Techniques

Goals of CP repair are to restore the integrity of the levator sling and close and lengthen the palate. All techniques rely upon adequate flap mobilization, atraumatic tissue techniques, and multilayer closure of oral and nasal mucosa and musculature.

Most will proceed with repair once the child is 10 kg around 10–12 months of age. Development of normal speech is the primary concern and dictates timing of repair. Improved speech outcomes are seen with early closure of a palatal defect (repair before 18 months):

- *Von Langenbeck*—bipedicled flaps with anterior and posterior blood supply; incisions placed along cleft margin and adjacent to alveolus; useful in narrow clefts and incomplete clefts; Medial fracture of the hamulus is

sometimes needed to assist with closure. Disadvantages: no palate lengthening; leaving anterior pedicle → decreased flap mobility; vascular pedicle (~1 cm medial to upper second molar) not visualized (Fig. 13.4).

- *Two-Flap Palatoplasty* (*Bardach*)—commonly used for repair of complete unilateral and bilateral CP, incisions extend along cleft margin, posterior to alveolus, and around last molar tooth; great care taken not to damage tooth buds during flap elevation; posteriorly based pedicle (greater palatine artery); combined with vomer flaps (advanced laterally and sutured to lateral undermined nasal mucosa for nasal closure); does not lengthen soft palate; dissection in space of Ernst (posterior to vascular pedicle) or fracturing of the hamulus allows additional mobilization of flaps (Fig. 13.5).
- *Furlow* (*Double-Opposing Z-Plasty*)—used for closure of the soft palate and in SMCP, lengthens and thickens the soft palate while reorienting LVP; also useful in surgery for VPD (see below); one flap is muscle/mucosa, while the other is mucosa alone; can be used for soft palate closure in conjunction with two-flap hard palate defect although the soft palate should be addressed before undermining flaps (Fig. 13.6).
- *V-Y Pushback/Three-Flap*—used for clefts of secondary palate/complete clefts; incisions similar to bipedicled technique, three-layer closure of nasal mucosa, muscle, with V-Y palatal lengthening of palate; subperiosteal release of flap and release of muscle allow repositioning of musculature in transverse orientation; lengthens palate; preserves mucosa over primary palate with bilateral posteriorly based unipedicled flaps (Fig. 13.7).

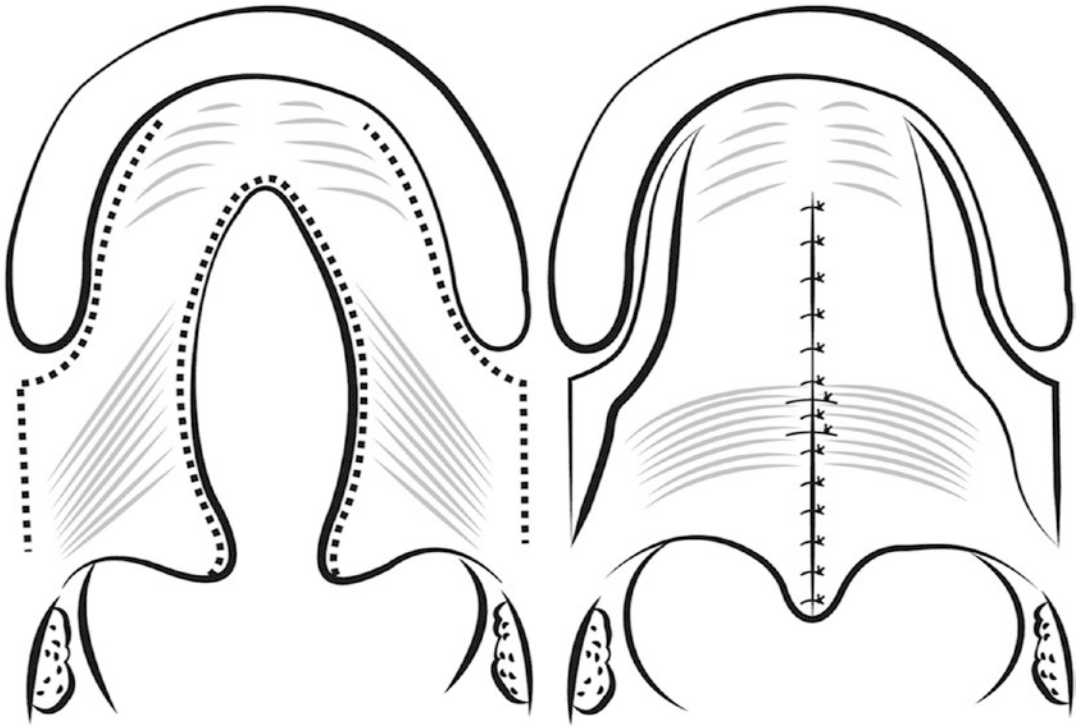

Fig. 13.4 Von Langenbeck palatoplasty. *Left image*: Incision design for right and left bipedicled flaps. *Right image*: Right and left flaps advanced for closure of cleft palate with residual exposed underlying hard palate laterally. Underlying levator sling is reconstituted with appropriate orientation of muscle fibers. (After Strong Management of the Cleft Palate Facial Plastic Surgery Clinics Feb 2001)

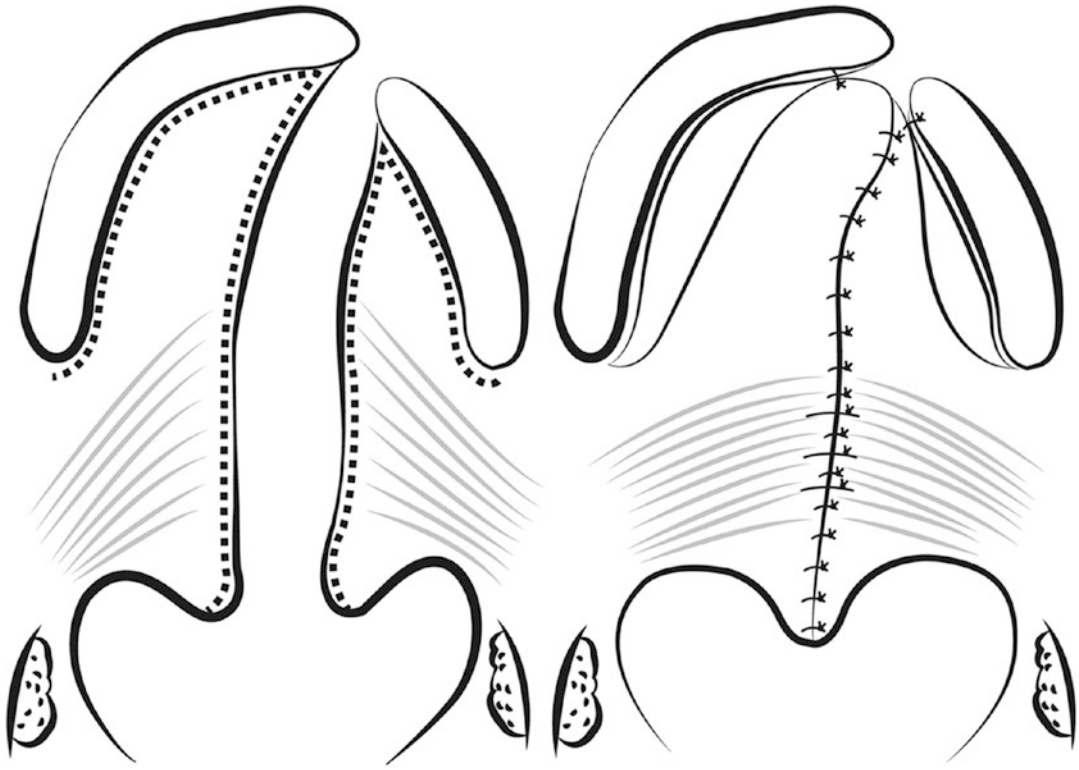

Fig. 13.5 Two-flap palatoplasty. *Left image*: Incision design for posteriorly based pedicle (off greater palatine vessels). *Right image*: Following elevation, mobilization and medialization of flaps for closure of cleft palate. Levator sling reconstituted in appropriate orientation. (After Strong Management of the Cleft Palate Facial Plastic Surgery Clinics Feb 2001)

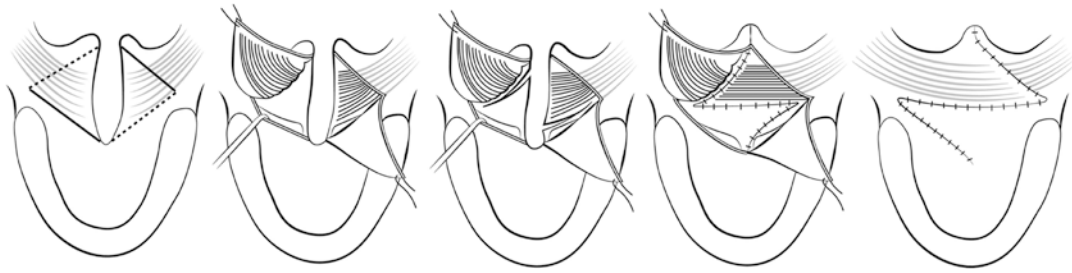

Fig. 13.6 Furlow (double-opposing Z-plasty) palatoplasty. *Leftmost image*: Incision design of opposing Z-plasties with cleft as central limb. Second image from *left*: Elevation of flaps—anteriorly based flaps are mucosa-only, while posteriorly based flaps are mucosa and palatal musculature. *Center image*: Incisions made through nasal mucosa. Second image from *Right*: Nasal flaps transposed. *Rightmost image*: Transposition and closure of oral flaps. (After Furlow Cleft Palate Repair by Double Opposing Z-plasty)

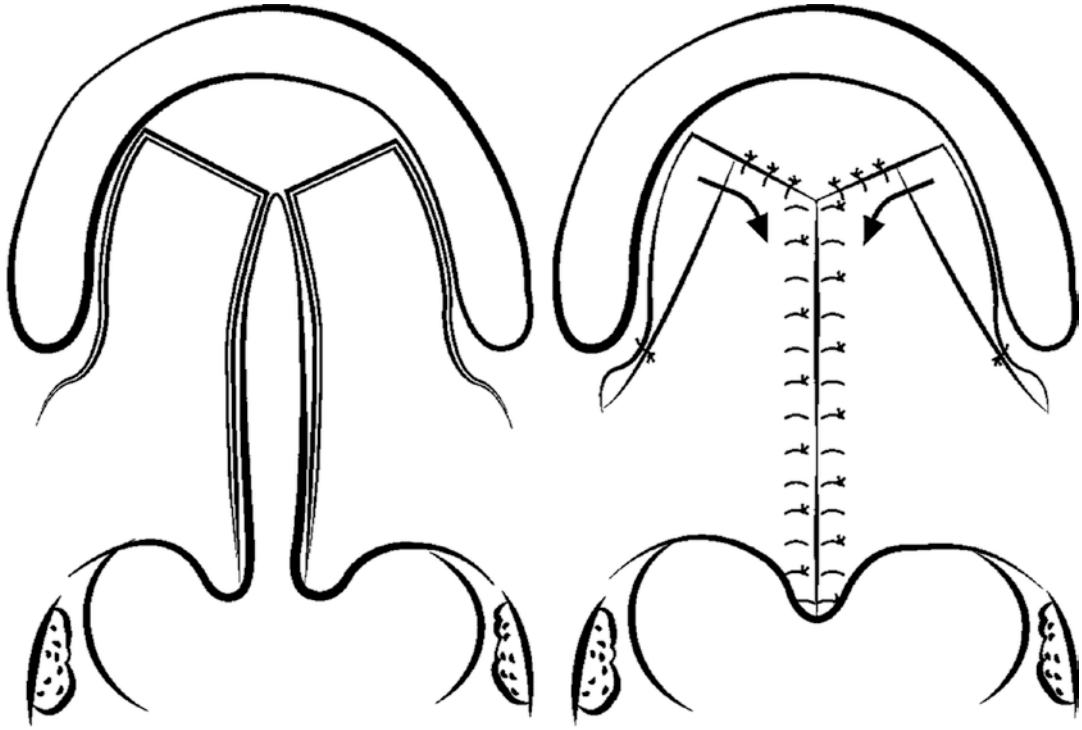

Fig. 13.7 V-Y (three-flap) palatoplasty. *Left image*: Incision design with right and left posteriorly pedicled flaps. *Right image*: Elevation, mobilization, medialization, and posterior advancement of flaps result in closure of defect and posterior lengthening of palate. (After Strong Management of the Cleft Palate Facial Plastic Surgery Clinics Feb 2001)

Complications of Palate Repair

- Velopharyngeal Dysfunction—most common complication following palatoplasty; lower rates seen with double-opposing Z-plasty than with other methods; abnormal orientation and insertion of LVP must be corrected to achieve appropriate functioning of the soft palate following CP repair.
- Fistulization can occur at varying rates depending on type and severity of cleft; sites of fistulization typically are the anterior hard palate and the junction of the hard and soft palate (*most common*).
- Airway problems following palatoplasty can occur as a result of tongue and FOM swelling (commonly due to relative hypoperfusion due to Dingman retractor or other retractor-exerted pressures → released every 30–45 min to decrease risk of post-op swelling); most likely to occur in children with Pierre Robin sequence.
- Bleeding—minimized with infiltration of epinephrine solution and appropriate tissue handling; exposed areas should be covered after repair with absorbable hemostatic material (e.g., Avitene, Surgicel) to promote healing and hemostasis.

Velopharyngeal Dysfunction

Velopharyngeal Inadequacy—inability to fully close VP valve during speech or swallowing:

- VP *Insufficiency*—structural, seen with cleft palate
- VP *Incompetence*—neurologic dysfunction
- Causes: previously repaired (inadequate lengthening of the velum at the time of primary palatoplasty and abnormal function of the levator musculature and cicatricial contracture of the velum) or unrepaired CP is the

most common cause of VPI; SMCP, neurogenic VPI, iatrogenic VPI following maxillary resection, UPPP, or adenoidectomy.
- Dx: speech evaluation/assessment of nasal/non-nasal phonemes; mirror exam—fogging during speech when held under nares; nasal endoscopy during phonation provides visualization of the velum, assessment of lateral pharyngeal walls and posterior pharynx during attempted VP closure; video fluoroscopy.
- Configuration of VP closure: can impact choice of surgical intervention; coronal (most common) > sagittal > circular:
  - Coronal—closure mainly due to action of the velum in anterior-to-posterior direction.
  - Sagittal—closure due mainly to medial motion of lateral pharyngeal walls.
  - Circular (±contribution by Passavant's ridge)—closure relies on combination of both A-P and medial/lateral motion for complete closure.

Treatment of VPI

Treatment decision made jointly by surgeon, SLP, and dentist/prosthodontist.

Speech Therapy

- Initial treatment indicated for children with mild VPD → aggressive speech Tx for 6 months, failure to improve with 6–12 months of speech therapy may indicate need for surgical intervention; with improvement → long-term aggressive speech therapy focused mainly on articulation.
- Not successful in remediating structural defects and surgical tx should not be delayed in these cases, e.g., children with VPD and nasal regurgitation secondary to obvious palatal abnormality such as a partial soft palate cleft or SMCP.
- Children with good articulation may be candidates for surgery if no VP closure demonstrated on nasal endoscopy with appropriately articulated phonemes.

Surgical Therapy

Surgery preferred for long-term management in children. Patients with structural causes of VPD or persistent VPD after speech therapy should be considered for surgical intervention:

- *Intravelar veloplasty*—reorientation of inappropriately oriented LVP sling with three-layer closure and reapproximation of (attenuated) muscle fibers in the midline.
- *Furlow* (*double-opposing Z-plasty*) *palatoplasty*—increasingly popular, reorients LVP, lengthens and thickens palate, and provides bulk in posterior VP; if VPD persists, sphincter pharyngoplasty can be performed; also used for primary closure of CP, repair of SMCP, or as a secondary procedure if the LVP longitudinally oriented; *sagittal* closure with VP gap of greater than 9 mm.
- *Pharyngeal flap*: permanent, central, passive obturator; brings posterior wall tissue to center of VP; relies on adequate function of lateral pharyngeal port for full velopharyngeal closure; superiorly based flap more common. Indicated for poor A-P velar motion with good lateral pharyngeal wall motion:
  - Most common complications: bleeding, airway obstruction (first 24 h following surgery); late complication: OSA, cor pulmonale, flap breakdown, nasal obstruction, and aspiration (associated with post-op hemorrhage). Consider pre-op sleep study in patients at risk for OSA.
  - VCFS—*pre-op angiography* should be obtained to evaluate course of carotid arteries as these patients can have a medial retropharyngeal course leading to significant intraoperative hemorrhage if unrecognized.
- *Sphincter pharyngoplasty* (dynamic sphincter pharyngoplasty, orticochea): narrows VP valve, decreased risk of postoperative airway obstruction; bilateral myomucosal flaps elevated from lateral pharyngeal wall/posterior tonsillar pillar wall and sutured to posterior NP wall to obturate the posterior and lateral portions of the VP; presence of adenoids may limit superior extent of flap placement → low

adenoidectomy ~6 weeks prior to sphincter pharyngoplasty; inset of flaps should be as high as possible (at or above the level of anticipated closure) to maximize function and minimize residual VPD and passive obstruction while in supine position (snoring common after sphincter pharyngoplasty); appropriate for correction of *coronal* and *circular* patterns of closure as long as the soft palate has adequate length.
- *Posterior wall augmentation*: filler or graft provides bulk near Passavant's ridge to aid in VP closure; suitable technique in child with small posterior midline gap.

Prosthetic Therapy

- Obturators—can be used if patient is poor candidate for surgical repair:
  - Palatal lift prosthesis pushes palate superiorly and posteriorly to contact pharyngeal wall.
  - Speech bulb prosthesis—with movement, walls of VP contact obturator.

Treatment of Alveolus/Bone Grafting

Alveolar bone grafting should occur during the period of mixed dentition; performed to restore normal architecture of maxillary arch and closure of oronasal fistula, if present (done prior to eruption of adult canine); secondary bone grafting allows eruption of permanent teeth (specifically lateral incisor and the canine) and provides base for subsequent correction of nasal deformity, orthognathic and orthodontic tx (including endosteal dental implantation).

Fresh, autogenous, cancellous bone is an ideal source because it supplies living, immunocompatible osteocytes that integrate fully with maxillary bone and are indispensable for osteogenesis. Grafting of alveolar bone defect has become standard of care at most cleft centers. Presence of bone in cleft creates odontotrophic milieu facilitating eruption and movement of teeth into cleft → dental aesthetics are enhanced by improvement of dental and gingival alignment.

Timing of Alveolar Repair

- Dental age/stage of dentofacial development more important than chronological age.
- Done as early as possible without causing deleterious effects on growth of the maxilla or damage to tooth buds.
- Root of permanent canine provides a guide to timing of therapy: should be formed 1/3–1/2 of definitive length at the time of graft placement (usually occurs sometime btw age 7 and 11 years old).
- Primary grafting: usually done before or during palatoplasty; has potential to inhibit midface growth, less common.
- Secondary grafting: performed after CP repair; *early* (if done <2 years old, may cause growth disturbances), *intermediate* (favored, during mixed dentition), or *late*:
  - Late grafting performed after exposure of cementum can lead to root resorption and ankylosis of teeth, predisposing to tooth loss and complicating later orthodontic management; graft resorption is also increased in grafts placed after eruption of teeth adjacent to the cleft.

Source of Grafting Material

- Allogeneic bone (treated homologous bone).
- Alloplastic materials—calcium phosphate based, eliminates donor-site morbidity.
- Autologous bone grafts considered gold standard because it provides osteogenic cells and osteoinductive factors; rapid incorporation, better dental support, resistance to infection; cancellous bone is preferred over cortical bone as it is more resistant to infection and undergoes less resorption.
- Iliac crest: major source of grafting material but postoperative gait disturbances can be seen.

- Alternative sources: cranium/calvarium, tibia, mandibular symphysis, rib (used for primary grafting in Rosenstein protocol).
- Bone morphogenetic proteins (BMPs)—recombinant BMP-2 in a soluble collagen matrix applied to alveolar cleft defect:
  - May be useful in skeletally mature patients with unilateral alveolar clefting; improved bone growth as compared to control group in this population
  - When performed in mixed dentition results are similar to autologous grafts

Correction of Secondary Defects

Secondary defects following correction of unilateral or bilateral cleft lip and palate are generally the rule rather than the exception. Age at the time of correction is key in decision-making process and largely based on severity of deformity; psychosocial effects must be considered; secondary procedures to correct speech or swallowing/feeding difficulty should be performed regardless of age; most commonly—preschool age (4–5 years old) or early adolescence; cessation of facial growth is another critical factor in timing of repair.

Lip—assessment in repose and during dynamic function; secondary defects classified as major/minor based on anticipated repair, i.e., if primary repair needs to be taken down to correct secondary defect; *secondary deformity of vermilion and Cupid's bow are most common* following primary CL repair.

Deformities of Vermilion—misalignment or peaked appearance of vermilion may be due to short vertical dimension of the lip; mild deformities common in early period following rotation advancement; massage may be useful early on; if persistent for >1 year → surgical tx:

- *1–2 mm vermilion misalignment → diamond-shaped excision* of scar and closure which increases length of scar and brings vermilion border down.
- >3 *mm*—problem is likely related to *inadequate rotation at the time of initial repair*: in these cases the repair should be taken down and rotation advancement should again be performed.
- Misalignment of vermilion-white roll is noticeable even if less than 1 mm: in these cases, a small Z-plasty can be designed to correctly align these structures.
- Deficient vermilion may be due to overaggressive mucosal resection at the time of initial repair or due to inadequate muscle alignment at the time of primary repair.
- *Whistle deformity*—mild deformities corrected with V-Y advancement (V apex toward sulcus carried through vermilion and orbicularis, incision carried to mucocutaneous junction, V-shaped flap then advanced to augment vermilion); vermilion augmentation with grafting materials (e.g., autologous fat) also an option but may be limited in patients with significant scar contracture.
- Vermilion excess: imprecise alignment at initial repair, treated by direct excision and closure.

Other Secondary Deformities

- Mucosal deficiencies—can consider buccal mucosal graft, especially in sulcus.
- Short lip—Z-plasty for mild defects; if orbicularis discontinuity exists (dx—have patient pucker his/her lips → visible bulge adjacent to cleft repair), must take lip down and reapproximate muscle; medial dissection should not extend past midline to avoid obscuring natural philtral dimple.
- Tight lip—relative underprojection of the upper lip; especially following closure of wide cleft; if severe can consider *Abbe cross-lip flap*.
- Wide lip—results from excessive width of initial design and more common in bilateral cleft lip repair; corrected by excising excessive portion along prior scar; reconstructed philtrum has tendency to stretch over time and should not be made wider than 4–5 mm initially to avoid *post-cleft repair wide philtrum*.

Questions

1. What ethnic group is most at risk of developing cleft lip and palate? Least at risk?
2. How does gender affect the likelihood of cleft lip with or without palate? Cleft palate in isolation?
3. What is the likelihood of cleft palate in the next child in normal parents with one affected child (assuming no other relatives are affected)?
4. What are some of the issues facing children with Pierre Robin sequence? Why is ophthalmology referral warranted in these patients?
5. Why are children with Shprintzen syndrome at particular risk during pharyngeal surgery, especially during a pharyngeal flap? What should be ordered prior to surgery?
6. What gene plays a role in van der Woude syndrome and popliteal pterygium syndrome? How do these two syndromes differ?
7. Describe the adult derivatives of the frontonasal prominence, nasal placode, and maxillary prominence. Failure of fusion between what structures causes cleft lip? What is the cause of primary cleft palate? Secondary cleft palate?
8. How does the anatomy of the orbicularis muscle and superior labial artery differ from normal in a unilateral and bilateral cleft lip deformity?
9. Describe the unilateral and bilateral cleft nasal deformity.
10. What are the stages of rhinoplasty to correct the unilateral cleft nasal deformity?
11. Describe the steps in the Millard rotation-advancement repair of the unilateral cleft lip deformity.
12. What is the treatment for unilateral complete CP? Bilateral CP? Secondary cleft palate? Submucous cleft palate?
13. What is the most common complication following cleft palate repair?
14. How does velopharyngeal closure affect the choice of surgical repair in velopharyngeal dysfunction?
15. What is the purpose of alveolar bone grafting in the cleft deformity? Should this be done before or after intermediate rhinoplasty to correct the nasal deformity?
16. What are the three abnormal attachments of the levator veli palatini muscle in the cleft palate deformity?
17. When should primary repair of the cleft lip be performed? What is the "rule of 10s"?
18. When should repair of the cleft palate be performed? Why is the timing of repair important?
19. Describe nasoalveolar molding? What are alternatives to NAM?
20. Describe the arterial supply of the soft palate.
21. Why do infants with cleft lip and palate have difficulty feeding? What can be done to overcome some of these difficulties?
22. How is the secondary tight lip treated? What predisposes to this secondary defect?
23. What is the whistle deformity? How is it treated?
24. What developmental pathology occurs in the condition of bilateral cleft lip and palate?
25. To what structure do the tensor veli palatini muscles originate from?
26. An adolescent patient has undergone early repair of a unilateral cleft lip and palate without correction of the alveolar defect. What type of malocclusion will likely be demonstrated?
27. What are the most common speech problems manifesting through VPI?
28. What anatomic derivatives develop from the embryonic lateral nasal processes?
29. Most surgeons today rely on a superiorly based pharyngeal flap for correction of VPI. What are the main disadvantages of the inferiorly based pharyngeal flap also utilized in the correction of VPI?
30. What pharyngeal motion is ideal for correction of VPI with a pharyngeal flap?
31. Patients with poor posterior pharyngeal wall motion may benefit from a sphincter pharyngoplasty for correction of velopharyngeal dysfunction. What muscles are transpositioned to accomplish tightening of the central port in this procedure?

32. What percentage best represents the incidence of associated malformations in neonates with isolated cleft palate?
33. What side of the face is a cleft lip most common and what side is more often associated with syndromes?
34. In secondary palatal development, when does elevation of the palatal shelves begin and when is fusion complete?
35. What is the purpose of the C-flap in the Millard rotation flap for the repair of unilateral cleft lip?
36. Describe the purpose of the lip adhesion procedure and define the optimal timing of the technique.
37. What is the most common otological manifestation of cleft palate and what is the proposed cause of the condition?
38. What percentage of patients will develop velopharyngeal dysfunction following surgical repair of the cleft palate?
39. At what age is definitive treatment carried out following a lip adhesion procedure?
40. When performing surgical correction of a cleft palate, what muscle sling repair is fundamentally important to velopharyngeal function?
41. In syndromic CLP, specific mutations of candidate genes have been identified. Describe three of the genes that are known to be responsible in a number of cleft-related syndromes.
42. Describe the importance of orbicularis oris muscle approximation in the lip adhesion procedure.
43. What is a submucous cleft and what is the underlying cause of this abnormality?
44. With respect to bone grafting of the deficient alveolar segment, what is the most important guide for timing?
45. How is VPI initially managed prior to determining the need for surgical correction?
46. What are the three main signs for the existence of a submucous cleft palate?
47. What is the purpose of presurgical infant orthopedics (PSIO) and when should such management be initiated?
48. Describe the actions of the velopharyngeal muscles.
49. What is the main cosmetic disadvantage of the Tennison-Randall repair versus the Millard rotation-advancement flap?
50. What method of palatoplasty confers the lowest rates of velopharyngeal dysfunction?
51. Where is the most common site of postsurgical palatal fistualization?
52. What is the most common configuration of VP closure?

Answers

1. Native Americans are the ethnic population at highest risk for the development of cleft lip and palate. African-Americans have the least risk.

   Overall incidence: 3.6/1000 in Native Americans, 2.1 per 1000 in Asians, 1 in 1000 in Caucasians, 0.41/1000 in African-Americans.
2. Males are more likely to develop cleft lip with or without palate (1.5:1 males/females).

   Females are more commonly affected by isolated cleft palate (2:1 females/males). This is thought to be due to later fusion of the palatal shelves in females.
3. 2%; see Table 13.1.
4. Management of the airway is critical in patients with Pierre Robin sequence. Due to the retro-positioned nature of the tongue, airway collapse at the pharyngeal level is common. This can oftentimes be managed by prone or lateral positioning; however these children may need further surgical intervention that may include glossopexy, tracheotomy, or mandibular distraction osteogenesis.

   Due to the high occurrence/association of Pierre Robin sequence with Stickler syndrome, these children should be evaluated at an early age by ophthalmology to rule out any retinal or ocular pathology.
5. Shprintzen syndrome or velocardiofacial syndrome results from an interstitial deletion of chromosome 22. It is associated with a medial course of the carotid artery,

and significant bleeding can result during pharyngeal surgery if this goes unrecognized prior to surgery. Preoperative angiography or alternative radiographic means of identifying the course of the carotid artery should be sought prior to proceeding with surgery in this setting. Additionally, patients with VCF tend to be at higher risk of developing VPI following adenoidectomy.

6. Interferon regulatory factor gene (IRF6) is the gene thought to play a role in both syndromes.

   Van der Woude—most common syndrome associated with CL ± P; findings include lower lip pits/sinus tracts, hypodontia, and autosomal dominant inheritance.

   Popliteal pterygium syndrome—findings similar to van der Woude syndrome with lower lip pits and absent or missing teeth. Additional findings include syndactyly, genital abnormalities, intraoral adhesions (syngnathia), pyramidal skin on hallux, and ankyloblepharon.

7. Frontonasal prominence derivatives: nasal dorsum, forehead, and septum.

   Nasal placode: gives rise to the medial and lateral nasal processes. Structures derived from the lateral nasal processes include the nasal alae and sidewalls. The medial nasal processes fuse and form the intermaxillary segment that forms the primary palate. Failure of fusion of the medial nasal processes results in a midline cleft lip. Additional structures derived from the medial nasal processes are the philtrum, medial upper lip, nasal tip, and columella. The maxillary prominences give rise to the lateral upper lip.

   Failure of fusion between the medial nasal processes and maxillary processes gives rise to cleft lip. Failure of fusion between the intermaxillary segment and the maxillary processes causes primary cleft palate. Secondary cleft palate results from failure of fusion of the palatal shelves (derived from lateral maxillary processes) in the midline, posterior to the incisive foramen.

8. In the unilateral cleft lip deformity, the orbicularis muscle is abnormally attached to the alar base and anterior nasal spine resulting in abnormal displacement of these structures. In the bilateral deformity, the orbicularis muscle is absent in the prolabial segment, while the lateral insertion onto the alar base is similar to that seen in the unilateral cleft lip deformity.

   The anatomy of the superior labial artery in unilateral cleft lip deformity is altered in that it travels parallel to the lateral aspect of the cleft and arborizes with branches of the angular artery. The arterial supply in the central prolabial segment is tenuous and stems largely from the columellar, septal, and palatine blood supply.

9. Unilateral cleft nasal deformity: The cleft alar base is displaced *inferiorly, laterally, and posteriorly* due to maxillary insufficiency and action of the orbicularis (as above), the caudal and inferior septum is deviated to the non-cleft side, and the upper and posterior portions of the septum deviate to the cleft side. Additionally, the cleft side may have inferior turbinate hypertrophy.

   Bilateral cleft nasal deformity: characterized by a poorly projected nasal tip with a wide alar base, alar bases that are positioned similarly to unilateral CL due to pull of abnormally inserting orbicularis oris and bilateral absence of maxillary segment → posterior, lateral, inferior displacement; extremely short columella and the medial crura are displaced into the prolabium; the prolabium and premaxilla can be significantly anteriorly displaced with poor overall blood supply; nasal tip is broad and flattened and the nasal domes are separated resulting in a bifid appearance.

10. Primary, intermediate, and definitive—primary involves separation of LLC from overlying skin with advancement and positioning closer to non-cleft dome. This is usually secured by bolstering the LLC in position following undermining and repositioning. The intermediate stage is performed prior to completion of growth and leads to further refinements in anticipation of definitive septorhinoplasty, usually performed

after growth is complete to correct residual defects of the septum, and nasal configuration. See text above for further details.

11. Methylene blue is used for skin marking. The rotation flap incision is made first (point 3 → 5 with back-cut to x as needed) and is taken through the entire thickness of the orbicularis and oral mucosa to allow for complete release. The c-flap remains and may be used to add additional height to the columella. The advancement flap starts with incision from point 8 → 9 and this incision is then continued along the alar base. The gingivolabial sulcus is also incised and incisions are taken down to face of the maxilla. Bilateral undermining assists in closure, and the first deep stitch reapproximates the orbicularis muscle at the back-cut. The second stitch is placed at the vermilion. The mucosal, muscle, and skin closure follows.
12. See text for full discussion of these techniques.

    Unilateral and bilateral complete: two-flap palatoplasty.

    Secondary cleft palate: V-Y pushback/three-flap.

    Submucous cleft palate: Furlow double-opposing Z-plasty.
13. Velopharyngeal dysfunction
14. See text for full discussion of each technique.

    Coronal—sphincter pharyngoplasty is generally preferred.

    Sagittal closure—pharyngeal flap vs Furlow double-opposing Z-plasty is the preferred method.

    Circular—sphincter pharyngoplasty preferred method.
15. Alveolar bone grafting is performed to restore normal architecture of the maxillary arch and closure of oronasal fistulae (if present, repair should be done prior to eruption of adult canine tooth); secondary bone grafting allows eruption of permanent teeth (specifically the lateral incisor and canine) and provides a base for subsequent correction of nasal deformity, orthognathic and orthodontic treatment.
16. The levator veli palatini has abnormal insertions to the posterior aspect of the hard palate, superior pharyngeal constrictor, and tensor veli palatini aponeurosis. This is seen in clefts of the secondary palate and in patients with submucous clefting.
17. Cleft lip repair is usually done at approximately 3 months of age. It follows the rule of 10s—10 weeks old, hemoglobin 10 g/dL, weight 10 lb; partially based on anesthetic safety.
18. Development of normal speech is a primary concern that dictates the timing of repair (usually done at ~12 months). Repair before 18 months of age has resulted in improved speech outcomes.
19. Nasoalveolar molding (NAM) is a process that involves molding and repositioning of the alveolar processes, nasal cartilages, and lengthening of deficient columella to create lasting aesthetic outcome and reduces need or minimizes the extent of secondary surgical revision procedures.

    Alternatives include lip adhesion procedures, taping, and the Latham appliance.
20. The soft palate is supplied by the ascending palatine (facial artery), greater palatine (internal maxillary), ascending pharyngeal branch, and contribution of the lesser palatine (branch of the greater palatine).
21. Oral intake can be compromised as a result of inability to feed (sucking mechanism impaired—child is unable to form a seal due to incomplete muscular sphincter and/or escape of air through cleft palate). A variety of feeding devices are available to assist in feeding a patient with cleft lip and palate such as the Mead-Johnson, Haberman, or Pigeon to facilitate feeding. Feeding can also be done in the upright position. Additionally, cleft patients tend to swallow more air and may require frequent burping.
22. The secondary tight lip often results from correction of wide cleft lips, and during the healing process, the premorbid absence of tissue leads to an underprojected lip after repair. In severe cases, this can be treated with an Abbe cross-lip flap.
23. The whistle deformity is a secondary deformity that may result following the repair

of unilateral or bilateral cleft lip and palate repair. It is characterized by central deficiency of the vermilion that leads to a shortened vertical height and the characteristic appearance of the upper lip seen with the deformity. Mild deformities may be corrected with V-Y advancement (V apex toward sulcus carried through vermilion and orbicularis, incision carried to mucocutaneous junction, V-shaped flap then advanced to augment vermilion); vermilion augmentation with grafting materials (e.g., autologous fat) is also an option but may be limited in patients with significant scar contracture.

24. Hypoplasia of the bilateral maxillary processes results in insufficient tissue to fuse with the fused medial nasal process/prominence.
25. The tensor veli palatini muscles arise from the membranous wall of the Eustachian tube. Their tendons pass around the hamular processes of the medial pterygoid plate of the sphenoid and insert into the palatine aponeurosis.
26. The arch is deficient in all dimensions: anteroposterior, transverse, and vertical. This child will demonstrate a posterior crossbite of the maxillary dentition relative to the mandibular dentition.
27. Hypernasality and articulation errors (glottal stops and pharyngeal fricatives).
28. Nasal ala and sidewall. Also, the nasolacrimal groove is formed between the LNP and maxillary process.
29. Length limitation and inferior tethering below the plane of the velum. This tethering is contradictory to the motion required to accomplish sufficient closure of the central port.
30. Lateral wall motion is imperative in valve closure in those patients with whom a pharyngeal flap is considered. An ideal patient has effective sagittal or circular closure patterns.
31. The paired palatopharyngeus muscles. The myomucosal flaps are elevated from the posterior tonsillar pillars and rearranged to achieve partial closure of a large central port without creating lateral ports. This arrangement is contrary to that accomplished with a pharyngeal flap.
32. Approximately 55%. Though CPO occurs less commonly than cleft lip and palate, it has a higher incidence of associated anomalies due to its higher association with various syndromes (DiGeorge, velocardiofacial, conotruncal anomaly face syndrome).
33. The left side is the most common (6:3:1—left/right/bilateral). The right side is more frequently associated with genetic syndromes.
34. In normal human development of the secondary palate, initiation begins in the sixth week with elevation of the shelves. The process is complete at week 12 with fusion, which occurs in an anterior-to-posterior direction from the incisive foramen. Additionally, the secondary palate fuses anteriorly with the primary palate and dorsally with the nasal septum, structures that are both derived from the medial nasal processes.
35. One characteristic finding of UCL includes columellar shortening. The C-flap is intended to lengthen the columella.
36. The lip adhesion procedure involves approximating the direct edges of the cleft without changing landmarks or disturbing the orofacial tissues, which will inevitably be required for definite treatment. A lip adhesion procedure should be considered for wide complete and wide near-complete clefts when a definitive lip repair is technically difficult due to tension. It essentially converts a complete cleft into a partial cleft. It is most commonly performed at 2–4 weeks.
37. Persistent OME is extremely common in the cleft palate population, and the prevalence has been estimated to be as high as 80–95%. The main cause is believed to be due to the abnormal insertion of tensor veli palatini, which prevents normal opening of the Eustachian tube.
38. Ten to twenty percent will manifest VPI after surgical correction. Hypernasality and articulation errors are the common manifestations of this condition, which is surgically

repaired by pharyngeal flap or sphincter pharyngoplasty.

39. Although the timing remains controversial with multiple time frames followed (6–12 weeks, 3–4 months, and 5–6 months), the majority of cases are performed at an average of 5 months of age.
40. Approximation and repair of the levator veli palatini sling (intravelar veloplasty) is critical for optimum VP valve functioning.
41. T-box transcription factor-22 (TBX22), poliovirus receptor-like-1 (PVRL1), and interferon regulatory factor-6 (IRF6). These gene mutations are accountable for X-linked cleft palate, cleft lip/palate-ectodermal dysplasia syndrome, and van der Woude and popliteal pterygium syndrome, respectively. The genetic complexity of non-syndromic CLP has also been recognized by a large number of candidate genes and loci (including the above mentioned); however, mutations have been identified only in a small fraction of cases.
42. Muscle pull is essential for the molding effect on maxillary arch segments.
43. A submucous cleft of the soft palate is characterized by a midline deficiency and/or lack of muscle tissue. The defect is lined by overlying oral mucosa. This type of cleft is caused by an anomalous insertion of the levator veli palatini muscle. Instead of muscle fibers being oriented in a transverse manner and decussating in the midline, they instead insert onto the posterior aspect of the hard palate.
44. Dentofacial development is more important than chronological timing with care taken to graft early enough without affecting maxillary growth or the developing permanent dentition. Optimal timing occurs during the mixed dentition stage. Generally, the length of the developing root of the permanent canine tooth is a good indicator. It should be approximately 1/3–1/2 of the permanent root length (17 mm on average). Typically, this would occur between the ages of 7 and 9.
45. In patients without structural defects and/or functional issues (i.e., nasal regurgitation), initial management includes aggressive speech therapy for 6 months. Failure of articulation improvement within 6–12 months should indicate the need for surgical intervention.
46. (1) Zona pellucida—a blue discoloration due to levator veli palatini muscle diastasis, (2) a bifid uvula, and (3) a palpable bony notch at the edge of the hard palate.
47. PSIO involves nonoperative reshaping of the alveolar and nasal segments prior to surgery. The concept, in particular NAM, works on the principle that the nasal cartilage could be molded due to increased plasticity concurrent to increased levels of maternal estrogen, if treatment is initiated within 6 weeks of life. Ideally, treatment should begin in the first 1–2 weeks.
48. *Levator Veli Palatini*: palatal sling causes superior and posterior motion of the soft palate; elevation of the soft palate during deglutition.

    Tensor Veli Palatini: tenses palate and opens Eustachian tube.

    Palatopharyngeus: retrodisplacement and downward motion of the velum; antagonist to LVP.

    Superior Pharyngeal Constrictor: main component of Passavant's ridge; contributes to different patterns of VP closure, specifically sagittal.
49. The Z in the lip crosses the philtral column creating more prominent scarring. The scar is better camouflaged with the Millard technique.
50. Furlow (double-opposing Z-plasty). Technique lengthens the soft palate and reorients LVP. Also, commonly utilized in SMCP repair.
51. Junction of the hard and soft palate.
52. Coronal (most common) > sagittal > circular. Coronal closure pattern is due to action of the velum in an anterior-posterior direction. Choice of VPI repair with coronal closure is a sphincter pharyngoplasty.

Additional Resources

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