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EVALUATION AND MANAGEMENT OF PATIENTS WITH CLEFT LIP AND PALATE

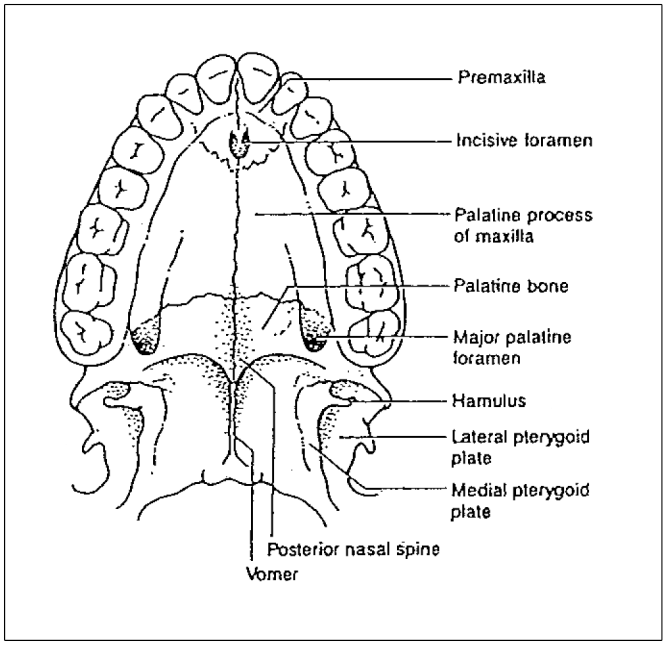
DEFINING TERMS

PRIMARY PALATE-

Structures anterior to the incisive foramen
Includes the nose, lip alveolus, and hard palate back to the incisive foramen
Clefts of the primary palate can occur on the left, right, or both sides
Complete clefts of the primary palate include all structures as mentioned above.
Lips can be described as complete (into the nostril), or incomplete (ending below the nostril)
"Simonart Band" is a small bridge of tissue joining both sides of the cleft just below the nostril.

SECONDARY PALATE-

Structures posterior to the incisive foramen
Clefts of secondary palate only occur in the midline
Complete clefts of the secondary palate travel from the incisive foramen through the uvula
Incomplete clefts of the secondary palate may start more posteriorly on the hard palate, at the junction of the hard and soft palate, or within the soft palate only submucous cleft palate involve intact mucosa, nothing of the hard palate "zona pellucida," abnormal orientation of the soft palate musculature, and bifed uvula.



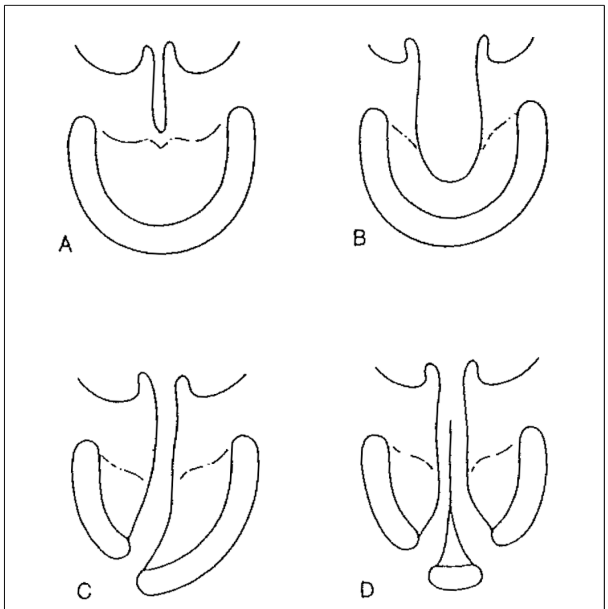


Figure 54–8. Classification of frequently seen clefts (after Veau). *A*, A cleft of the soft palate only. One can usually palpate a notch in the posterior hard palate and there is generally a submucosal extension into the hard palate. *B*, A complete palatal cleft extending anteriorly to the incisive foramen. This cleft may be narrow but often has a wide horseshoe-shaped defect. *C*, A unilateral palatal and prepalatal cleft. Note that the vomer is attached to the maxilla on the noncleft side. *D*, Bilateral complete cleft of the palate and prepalate. The premaxilla without any restraining force from the lip can protrude markedly and is stabilized only by the vomer. (After Veau, V.: Division Palatine. Paris, Masson et Cie, 1931.)

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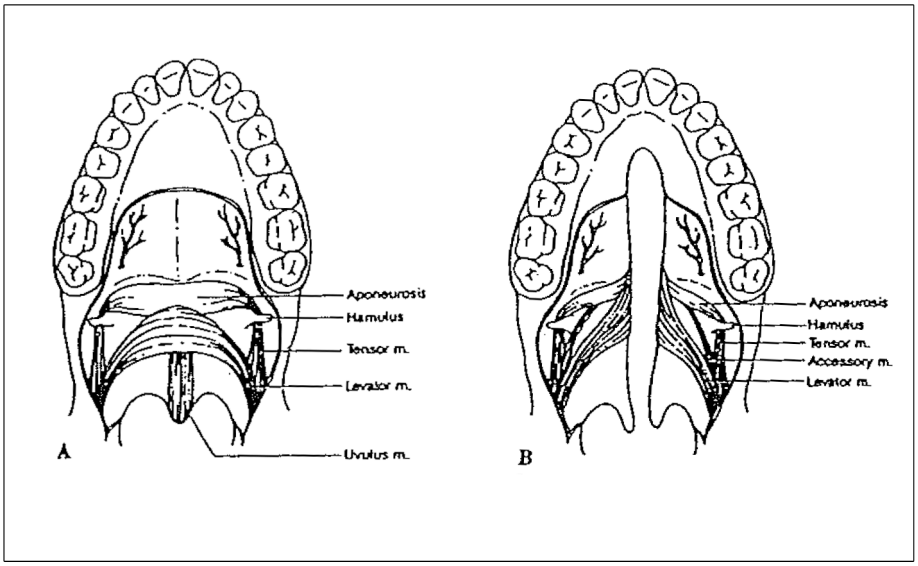

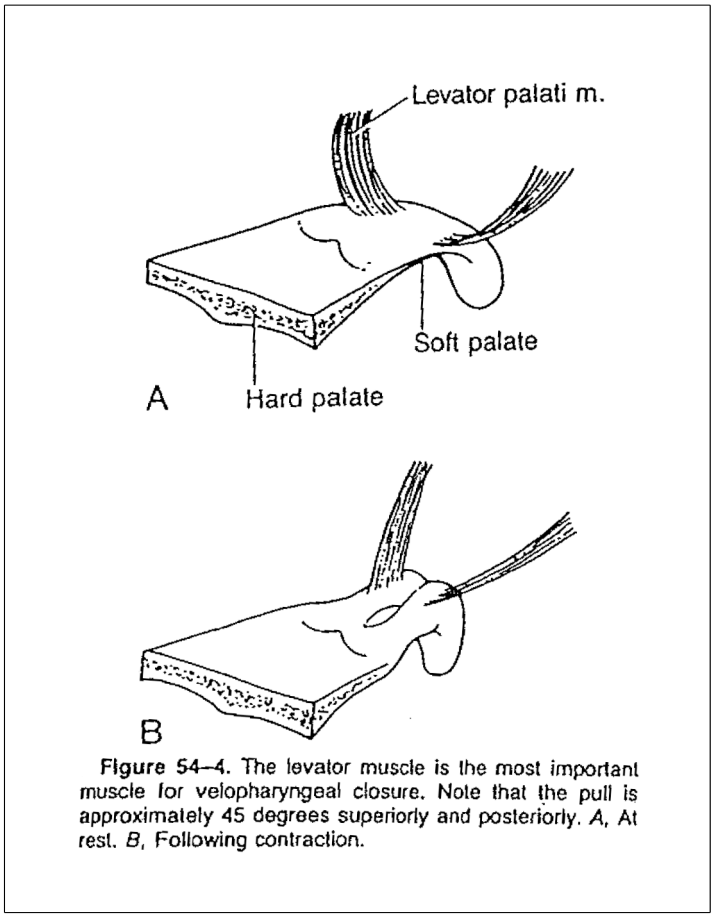

Figure 54–4. The levator muscle is the most important muscle for velopharyngeal closure. Note that the pull is approximately 45 degrees superiorly and posteriorly. *A*, At rest. *B*, Following contraction.

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PREMAXILLA-

Mobile bony segment located between bilateral clefts of the primary palate which includes the nasal spine and houses the four front teeth (incisors).

Often too far forward in unrepaired children, too retruded following completion of facial growth.

PROLABIUM-

Contracted soft tissues attracted to the premaxilla in patients with bilateral clefts.

Stretches significantly following lip adhesion or repair.

COLUMELLA-

Tissue located below nasal tip and between nostrils.

Asymmetric in unilateral clefts, deficient in bilateral clefts.

VELUM-

Soft palate

VELOPHARYNGEAL INSUFFICIENCY (VPI)-

Dysfunction of the soft palate and pharyngeal walls resulting in inappropriate airflow through the nose during speech.

RESONANCE-

A quality of speech that involves the balance of air passing through the nose and mouth. If too much air comes out of the nose they are *hypernasal* and may sound like the Nanny. If too little air comes out of the hose they are *hyponasal* and sound like they have a cold.

SECONDARY PALATAL MANAGEMENT-

Surgery done to help decrease the air coming out of the nose when speech therapy is not enough.

ARTICULATION-

The way sounds are made using the lips, teeth, and tongue. When people make sounds the wrong way they need speech therapy to learn how to make them correctly. Surgery does not correct articulation errors.

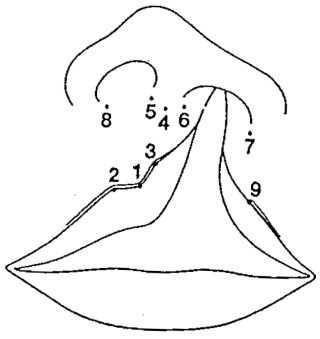
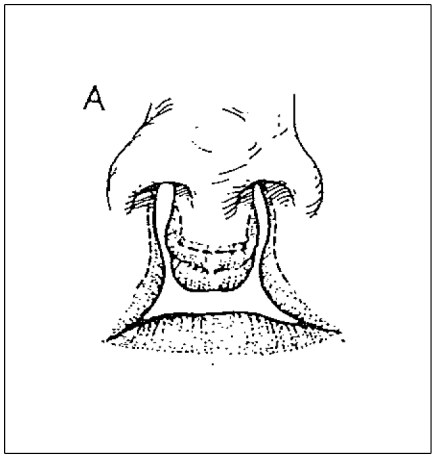


FIGURE 28.1. Anatomy of the normal and unrepaired unilateral cleft lip indicate "key points" used for planning repair: (1) lowest point in arch Cupid's bow, midline of the lip; (2) peak of Cupid's bow on the noncleft side; (3) proposed peak of Cupid's bow; (4) midpoint of the columella; (5) and (6) base of columella; (7) and (8) inset of alar base into nostril sill; (9) point of disappearance of white roll of the vermilion cutaneous junction (at the same level as point 2 and where the roll is well developed)



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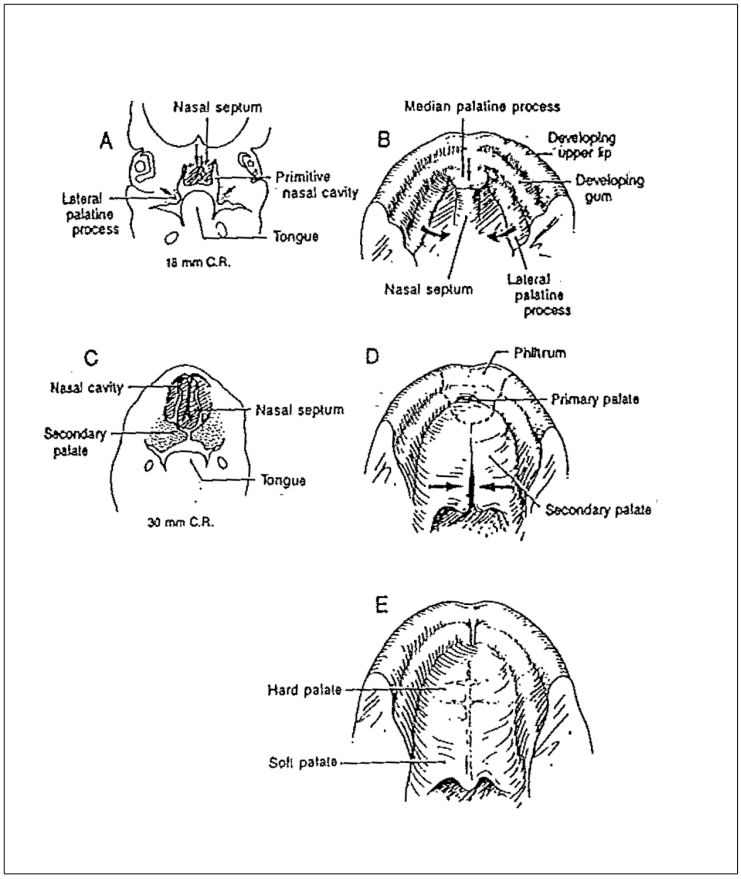
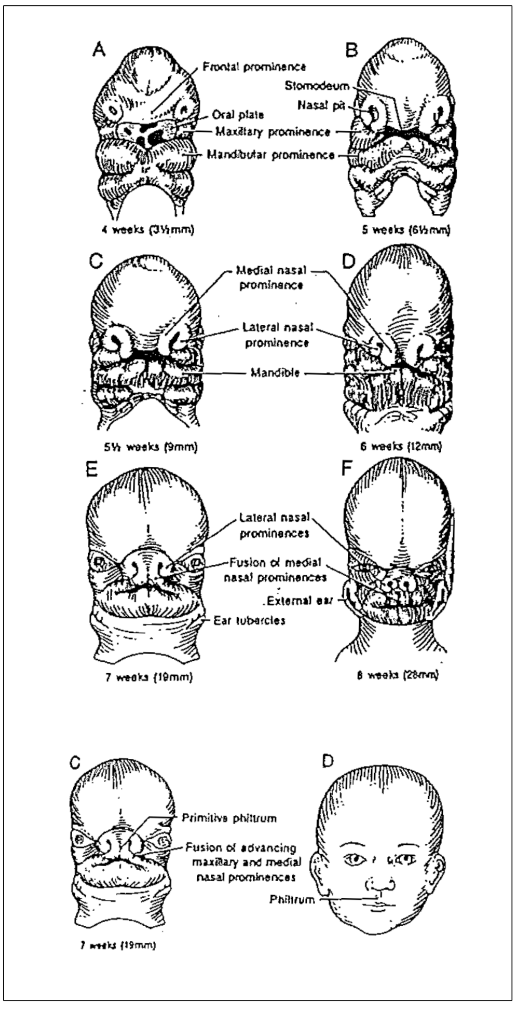
ETIOLOGY

Multifactorial and for the most part unclear.
Genetic, viral, teratogenic, nutritional, "sequence"
Occurs in the first trimester of pregnancy.

PRIMARY PALATE-
Early in the first trimester of pregnancy (4-7 weeks) the facial elements (frontonasal, two lateral maxillary, and two mandibular segments) fuse. Mesenchymal tissue migrates into these points of fusion. Lack of mesenchymal development in these areas of fusion leads to clefting.

SECONDARY PALATE-
Paired maxillary process (palatal shelves) are initially oriented in a vertical fasion alongside the developing tongue. As the mandible grows, the tongue lowers, and the shelves raise to a horizontal position. The epithelial lining breaks down, and the palate begins to anteriorly at about the 8th week of gestation. The fusion is completed by the 17th week. Tongue positioning (due to the cleft of the primary palate, small mandible, lack of neck positioning), deficiencies of the palatal shelves, or lack of epithelial breakdown may allo lead to clefting of the secondary palate.





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EPIDEMIOLOGY

Cleft lip or cleft lip and palate is NOT the same as an isolated cleft palate.
Some degree of clefting occurs in 1:600 to 1:1,000 live births
CL alone – 21%
CL/P – 46%
CP – 33%

CLEFT LIP AND PALATE
- Second most frequent major congenital anomaly
- Males > Females (2:1)
- Left: Right: Bilat = 6:3:1
- Positive family history twice common as with CP only (about 17-25%)
- Majority are isolated deformities

CLEFT PALATE
- Females > Males (2:1)
- Less likely to have positive family history (3-12%)
- More likely to have associated anomalies (approximately 29%)

INHERITANCE
- Normal parents + one or more children with CL+/-CP = 4% chance of additional children with cleft
- Parent with CL+/-CP = 4% chance of child with cleft
- Parent with CL+/-CP + one child with CL+/-CP=14-17% chance of additional children with cleft
- Normal parent + one child with CP = 2% chance of additional children with cleft
- The worse the cleft, the higher the chance of future children with cleft.
- The stronger the family history, the higher the chance of future children with a cleft.
- The stronger the family history, the higher the chance of future children with clefts

SYNDROMES

Over 150 syndromes involving clefts have been described
1% of newborns have multiple abnormalies or syndromes and only 40% of these are regovnizable.
Syndrome vs. Sequence

PIERRE-ROBIN or ROBIN SEQUENCE
- Micro or retrognathia, glossoptosis, cleft palate (wide, U-shaped)
- May have airway complications in first 3-6 months (prone positioning, Lip-tongue adhesion, tracheostomies)
- May occur as a component of sundrome or isolatred (nonsyndromic).

STICKLER SYNDROME
- Autosomal Dominant
- Often seen with Robin sequence
- Retinal detachment occurs in 20% , blindness in 15%
- Arthritis

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VAN DER WOUDE SYNDROME-
- Autosomal Dominant (50% penetrance)
- Lower lip pits or mucous cysts associated with CL+/-CP
- Variable phenotypic expression

VELOCARDIOFACIAL or SHPRINTZEN SYNDROME
- Autosomal Dominant, through most are sporadic (deletion AT22q11)
- 1:2000 of general population
- Anomolies include behavior, palate/speech, facies, cardiac, and vascular
- Comprises 11% of Robin cases, 8% of all isolated CPs

PEDIATRIC CONCERNS

Full history and physical examination
If associated anomalies identified, consult geneticist and specialists as indicated (ENT, ophthalmologist, cardiologist, ortho).
Helpful for parents to meet plastic surgeon as early as possible (prenatally if identified on ultrasound).
Careful monitoring of feeding, nutrition, and weight gain. (Breast feeding controversy, Haberman nipple, cross cut nipple, don't flood, burp frequently, utilize feeding specialist)
High frequency of ear infections (PETs usually placed at time of CP repair)
Dental hygiene
Speech therapy

COUNSELING

Relieve parents guilt
Refer to Multidisciplinary Cleft Team
Ensure adequate genetic counseling if indicated
Stress that they have a *normal* child who happens to have cleft lip or palate
Be supportive to parents excess time, expense, and stress with multiple surgeries, appointments, orthodontics
Support groups (AboutFace 1-800-665-FACE), contacting other families dealing with clefts

SURGERY TIMELINE

Remember that every child and surgeon are a little different
There is usually no true right and wrong (more a matter of preference)

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CLEFT LIP ADHESION / REPAIR

TIMING-

Some doctors like to repair lips in one stage, some in two, or case by case based on the width of the cleft.
Usually done from 6 weeks to 3 months of age
If two stages, second at about 6 months after the first surgery

SURGERY-

1-2 hours
General anesthesia
One night stay

RECOVERY-

Sutures removed in 3-7 days
Protect lip (car seat, arm splints, no macifier) 2-3 weeks

COMPLICATIONS-

Anesthesia
Infection
Dehiscence (splitting apart)
Poor cosmetic result

CLEFT PALATE REPAIR

TIMING-

6-12 months of age
Ideally before speech develops, but late enough to allow for maximal facial growth

SURGERY-

1.5-2 hours
General anesthesia
Tongue stitch possible
1-2 night stay

RECOVERY-

Stitches dissolve
Liquid or soft diet for three weeks
Protect palate 3 weeks (arm splints)
No spoons, straws, pacifiers in mouth three weeks

COMPLICATIONS-

Bleeding
Airway
Fistula
VPI
Facial growth stunting

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FISTULA REPAIR

TIMING-
- Based on severity of symptoms (nasal regurgitation, hypernasality)

SURGERY-
- Very similar to cleft palate repair
- General anesthesia
- 2-3 hours
- 1-2 night hospital stay
- May involve cartilage graft of tongue flap

RECOVERY-
- Protect palate three weeks
- Liquids or soft foods 3 weeks

COMPLICATIONS-
- Infection
- Dehiscence
- Poor cosmetic result

COLUMELLAR LENGTHENING/LIP REVISION/NOSE REVISION

TIMING-
- Elective
- Usually age 4, prior to starting school
- Nasal tip may need cartilage graft from ear

SURGERY-
- 1 to 4 hours
- General anesthesia
- Inpatient or outpatient

RECOVERY-
- Stitches out in 3-7 days
- May have nasal packing or head wrap for ear
- Protect for first few weeks

COMPLICATIONS-
- Infection
- Dehiscence
- Poor cosmetic result

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ALVEOLAR BONE GRAFT/ILLIAC CREST BONE GRAFT

TIMING-

- Usually 7 to 11 years of age based on tooth root development
- Follows palate expansion
- Orthodontist usually determines timing

SURGERY-

- May be done by oral surgeon and/or plastic surgeon
- Bone is taken from the upper hip bone and placed into alveolar and maxillary cleft
- This surgery completes maxillary arch, allows for adult tooth to come in, or implant
- Treats nasolabial fistulae
- Palate fistulae are often repaired at the same time
- Surgery takes 2-4 hours
- General anesthesia
- 1-2 night hospital stay

RECOVERY-

- Liquids or soft foods 3 weeks
- Hip may be sore for over a month
- Follow-up includes x-rays

COMPLICATIONS-

- Infection
- Bleeding (hematoma)
- Resorption (dissolving) of bone graft
- Injury of teeth or guns
- Numbness of upper thigh

SECONDARY PALATAL MANAGEMENT

TIMING-

- Following adequate trial of speech therapy
- Work-up includes speech evaluation, endoscopy (camera down nose) or xray study
- Rule out palatal fistula
- Can be done at any age

SURGERY-

- Pharyngeal flap, Sphincter pharyngoplasty, Furlow Z-Plasty
- 2 hours
- General anesthesia
- Tongue stitch
- 1-2 night stay

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RECOVERY-
- Swelling first few weeks may cause snoring, muffled voice
- Liquids or soft diet 3 weeks
- Stitches dissolve

COMPLICATIONS-
- Bleeding
- Airway
- Obstructive sleep appnea
- Overcorrection (hyponasal voice)
- Undercorrection (continued hypernasal speech)

ORTHOGNATIC SURGERY

TIMING-
- Teen years, following orthodontics

SURGERY-
- Usually performed by Oromaxillofacial surgeon
- May involve maxilla, mandible, or both
- May involve distraction (stretching the bone)
- 2-4 hours
- General Anesthesia

RECOVERY-
- May have jaws wired 2-6 weeks
- Liquids or soft foods 3 weeks
- 1-2 night hospital stay

COMPLICATIONS-
- Airway
- Infection
- Relapse of position
- Injury to teeth of gums
- Numbness

RHINOPLASTY

TIMING-
- Females 13-16 years
- Males 15-17 years
- Best to be done after orthognathic surgery

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SURGERY-

- 2-5 hours
- Usually general anesthesia
- "Open" rhinoplasty with cartilage grafts
- Can be done as outpatient
- Improves appearance and/or breathing

RECOVERY-

- Nasal packing and splint first week
- Bruising and swelling 2-3 weeks
- Final swelling may take 1 year to go away
- Protect from trauma 6 weeks

COMPLICATIONS

- Bleeding
- Infection
- Persistent deformity
- Persistent nasal obstruction
- Poor cosmetic result