

Chapter 12: Cleft Lip and Palate

Anatomy

Anatomy of the Lip

The lip is composed primarily of muscles, covered by skin on the outer surface and mucosa on the inner surface. The lip edge or vermillion is covered by nonkeratinizing epithelium made red by numerous highly vascular connective tissue papillae. The junction between the vermillion and skin is called the mucocutaneous ridge or white line. This line forms a gentle arch in the upper lip and is depressed lightly in the middle to form the center of "cupid's bow". Lateral extension of the white line completes the cupid's bow. From the ends of this central depression of the white line, small ridges extend upward to the base of the columella enclosing a small depressed skin area called the philtrum. A slight protrusion of the vermillion below the cupid's bow is called the tubercle. These anatomic landmarks are used for orientation in repairing clefts of the upper lip (Fig. 12-1).

The lip is a movable muscular curtain composed primarily of the orbicularis muscle which creates a sphincter and is formed by significant contributions from paired muscles converging on the mouth (Fig. 12-2). The muscle substance arches around the lips and interlaces at the angle of the mouth. The muscle lies between the skin and mucous membranes of the lips, limited superiorly by the nose and inferiorly by the chin.

An almost infinite variety of movements can be obtained by the lips by the individual action, inaction, or antagonistic action of these various paired muscles.

The main arterial blood supply to the upper lip is provided by the paired superior labial arteries which are located near the mucous membrane deep to the muscle. Sensation to the upper lip is supplied mainly by fibers from the inferior orbital branches of the fifth cranial nerve. Motor fibers are derived from the labial branches of the seventh cranial nerve.

Anatomy of the Palate

The palate is composed of a bony anterior portion and a soft muscular posterior portion. The tooth-bearing alveolar ridges surround the hard palate. An anterior wedge-shaped portion of the alveolar ridge carrying the four incisor teeth and a triangular section behind constitute the premaxilla or primary palate. The remaining hard palate is made up of palatine processes of the maxillary bone, and to a lesser degree by the palatine processes of the palatine bone. The maxillary and palatine portion of the hard palate plus the soft palate constitute the secondary palate.

The oral surface of the hard palate is covered by firm mucoperiosteum of variable thickness. Anteriorly this membrane has raised ridges called palatine rugae. The nasal surface of the hard palate is divided into two portions by the nasal septum and is lined by a thin mucoperiosteum surfaced anteriorly by stratified squamous and posteriorly by respiratory epithelium. Blood supply to the hard palate is provided chiefly by the anterior palatine artery

which is derived from the internal maxillary via the greater palatine foramen. The nerve supply is chiefly from the anterior palatine and nasopalatine branch from the sphenopalatine ganglion. The nerve supply follows the arterial blood supply in distribution.

The soft palate is movable muscular curtain covered on its oral surface by stratified squamous epithelium and on its nasal surface by pseudostratified columnar epithelium. This structure contains numerous muscles and glands. The palate has five muscles as shown in Table 12-1.

Table 12-1. Nerve Supply and Muscles of the Palate

Tensor veli palatini	Fifth nerve	Tense and depress soft palate
Levator veli palatini	Pharyngeal plx	Elevate the palate
Musculus uvulae	Pharyngeal plx	Draw uvula upward and forward
Glossopalatine	Pharyngeal plx	Draw palate down and narrow pharynx
Palatopharyngeus	Pharyngeal plx	Draw palate down and narrow pharynx

The arterial blood supply to the soft palate is the descending palatine branch of the internal maxillary, the ascending palatine branch of the external maxillary, the palatine branch of the ascending pharyngeal, the twigs from the tonsillar branch of the dorsalis linguae. The sensory nerve supply is mainly from the lesser and middle palatine branches from the sphenopalatine ganglion.

Embryology of the Cleft Lip

The central upper lip is formed by growth of the nasomedian process in a downward medial and forward direction. This process provides the central lip consisting of philtrum, labial tubercle, the central alveolar ridge containing the paired central and lateral incisor teeth, the anterior nasal septum, and an anterior palatal triangle (primary palate). Paired lateral maxillary processes grow medially toward the descending nasomedian process. These processes provide the lateral upper lip elements.

Failure of the structure to unite on one side, or both, provides clefts of the lip, extending into the nose and through the alveolus between the lateral incisor and cuspid teeth. Paired processes or shelves arise from the maxillary processes. They converge upon the primary palate, on each other, and the septum, to fuse (from front to back) to provide the secondary palate. The lip and hard palate formation is completed by the eight week of embryonic life and the soft palate and uvula are completed by the twelfth week of embryonic life.

Note: The genesis of harelip has not been agreed upon by various investigators. Some believe it is the result of failure of fusion between the maxillary and frontonasal processes. Others believe that it is the lack of "filling up" between the premaxillary and maxillary centers that results in a furrow and hence a harelip.

Types of Cleft Lip and Palate (Fig. 12-3)

Types of cleft lip and palate include:

1. Unilateral cleft lip: occurs in varying degrees and may involve the alveolus in a like manner.
2. Unilateral cleft lip with cleft palate.
3. Bilateral clefts of the lip: usually involve complete clefts of the secondary palate but can occur (rarely) without the presence of a cleft palate.
4. Bilateral clefts of the lip, alveolus, and palate: both nasal cavities are exposed to the oral cavity.
5. Clefts of the secondary palate: occur in varying degrees such as bifid uvula, submucous cleft, and complete division up to the primary palate.
6. Median cleft of the upper lip: very rare.

Incidence of Cleft Lip and Cleft Palate

Cleft lip with cleft palate is the most common; cleft palate alone is next; cleft lip alone is the least common. The incidence of cleft lip with or without cleft palate varies from 0.8-1.6:1000 births. The incidence of combined cleft lip and cleft palate occurs 1.5-3 times more frequently than cleft lip alone. The incidence of cleft lip with cleft palate is greater in males. The incidence of cleft palate alone is greater in females. The frequency of single cleft lip is greater on the left than on the right side. The incidence of cleft lip is three times greater in Caucasians than in Negroes. Genetic factors in cleft lip with or without cleft palate are:

1. Mutant genes
2. Chromosomal aberration
3. Environmental teratogens
4. Multifactorial inheritance.

Incidence of Cleft Palate

Cleft palate is embryologically and genetically different from cleft lip alone or combined cleft lip and cleft palate. The rate is 0.45:1000 Caucasian births. The cleft palate is more common in females. Genetic factors in cleft palate are:

1. Mutant genes
2. Chromosomal aberration
3. Environmental teratogens
4. Multifactorial inheritance.

Counseling Parents Regarding Lip and/or Palate Clefts

Situation A

The parents are normal, the first child is affected with cleft lip with or without cleft palate.

Question 1: What are the chances for the next child if there are no affected relatives?

Answer: 4%.

Question 2: What are the chances for the next child if there is an affected relative?

Answer: 4%.

Question 3: What is the chance for the next child if the affected child has another malformation?

Answer: 2%.

Question 4: What is the chance for the next child if the parents are related?

Answer: 4%.

Situation B

Both parents are normal and have two affected children with cleft lip with or without cleft palate.

Question: What are the chances for the next child to have the same defect?

Answer: 9%.

Situation C

One parent is affected with cleft lip or cleft palate and there are no affected children.

Question: What is the chance for the next child of having a defect?

Answer: 4%.

Situation D

One parent is affected and there is one affected child.

Question: What is the chance that the next child will be affected?

Answer: 17%.

Counseling for Cleft Palate Alone

Situation A

The parents are normal, one child is affected with cleft palate.

Question 1: What is the chance for the next baby to have cleft palate if there are no affected relatives?

Answer: 2%.

Question 2: What is the chance for the next baby to have cleft palate if there is an affected relative?

Answer: 7%.

Question 3: What is the chance for the next baby to have cleft palate if the affected child has other malformations?

Answer: 2%.

Situation B

The parents are normal but have two children, both with cleft palate.

Question 1: What is the chance for the next baby to have a cleft palate?

Answer: 1%.

Situation C

One of the parents has a cleft palate.

Question 1: With no affected children, what is the chance of the next baby having a cleft palate?

Answer: 6%.

Question 2: What is the chance for the next baby with one child having a cleft palate?

Answer: 15%.

Timing and Techniques

Lips

There is no set time for repair of cleft lip. Since cleft lip is a congenital deformity, adequate time should be allowed to properly observe and examine the infant to determine the possibility of other associated congenital defects. Closure usually is performed within 3 months; however, social pressures often dictate that the defect be closed before the infant leaves the hospital. The rule of over 10 is advocated, over 10 weeks in age, over 10 lb in weight, and hemoglobin over 10 g.

Single Cleft Lip

Defect

1. Floor of nose communicates freely with oral cavity.
2. Maxilla is hypoplastic on the cleft side.
3. Lower lateral nasal cartilages lower on cleft side in all respects.
4. Nasal columella is displaced to the normal side.
5. Cleft side of the nasal ala base is deformed.
6. Cartilaginous nasal septum is deflected.
7. Alveolar defect passes through developing dentition.

Repair

1. Millard's rotation advancement - technique is accepted by many as the standard for repair of the unilateral cleft lip. It is a cut-as-you-go technique which places the scar in a normal philtral line adding tissue to the deficient noncleft side of the lip and the cleft side of the nasal columella (Fig. 12-4).

2. Tennison's triangular flap - is designed to add tissue to the deficient noncleft side of the lip low in the lip. It is advanced as a simple to teach method with a predictable result (Fig. 12-5).

3. Le Mesurier's quadrilateral flap - was originally designed with the incision in the very midline of the lip but later was shifted toward the cleft side of Cupid's bow. This technique is perhaps the easiest to understand, teach, and obtain a satisfactory result for the low volume surgeon (Fig. 12-6).

4. Skoog's double triangular technique - recognizes the deficiency in the noncleft side of the lip both low and high at the columella base. This technique, although difficult to conceive, provides real improvement in the nasal portion of the maxillary cleft (Fig. 12-7).

Bilateral Cleft Lip

Defect

1. Floor of the nose is missing on both sides and the nasal and oral cavities communicate freely.
2. Central portion of the alveolar arch is rotated forward and upward out of the area.
3. Prolabium skin for the lip is underdeveloped.
4. Columella is short.
5. Central portion of the lip contains no lip muscle or lip vermillion.

Repair

1. Single stage repair is often greatly appreciated by the family since a horrendous defect is converted to a continuous lip with a much improved nose (Fig. 12-8). The single stage bilateral lip closure is often the cause of tremendous secondary defects which require major secondary repair.

2. Staged bilateral lip closure 3 months apart using techniques for single cleft lip repair have come into favor since the premaxilla is more readily controlled by using the lip as an orthodontic device and the lip repair can add tissue to the columella, hopefully avoiding serious secondary defects of the lip and nose. Some surgeons prefer to do a lip adhesion technique first to help control the alveolar arch segment and then proceed with the primary lip repair.

Alveolar Arch

The alveolar arch is completely separated in the single cleft lip defect. To obtain osseous continuity, the epithelial tissue must be removed from the opposing alveolar surfaces and a force provided to bring them together. Epithelial tissue is removed and hinged upward to form the floor of the nose. The lip closure provides the necessary orthodontic force. With proper timing of the lip surgery, preoperative dental orthodontic appliances have been found unnecessary by most surgeons.

Maxillary growth and dental arch malalignment, secondary to scar contracture following primary closure, are factors which have influenced timing of the procedures. Simple closure of the lip without muscle approximation plus closure of the soft palate portion of the complete cleft palate have been advocated. Dental appliances adjusted frequently to accommodate the growth serve to cover the osseous portion of the remaining cleft defect. The defect of the hard palate is closed after optimum maxillary and alveolar arch growth has occurred. Redirection of the fibers of the orbicularis oris muscle displaced by the cleft has received increased attention in repair of primary clefts.

Hard and Soft Palate Cleft Repairs

Following repair of the cleft lip, including the nose, the patient with a complete cleft is left with a midline cleft of the hard palate extending from about the canine area all the way back including the uvula. This is true for both single and double clefts. The only major

variable is the position of the vomer. Techniques designed to correct clefts of the palate only are used for closure of the primary cleft and those present following lip repair.

The timing for closure of the cleft palate is less variable than that for the lips. The most popular time is 18 months. The time of cleft palate closure is very important in the development of speech, hearing, swallowing, dental occlusion, and facial growth.

Techniques

1. Von Langenbeck's: Bilateral relaxing incisions are performed with freshening of the cleft margins. The bipedicle oral mucoperiosteal flaps are elevated. The nasal mucoperiosteal layer is elevated and the cleft is closed in two layers with a third layer in the soft palate muscles.

2. V-Y repositioning (Wardill-Kilner): Bilateral relaxing incisions are performed. The anterior part of each relaxing incision is angled backward toward the midline to form a W. The cleft edges are incised. The oral mucoperiosteal flaps are elevated to the posterior border of the hard palate. The nasal layer is elevated and the cleft is closed in two layers, with a third layer in the soft palate muscles. The oral mucoperiosteum is anchored to the unelevated anterior tip of the oral mucoperiosteum.

3. Dorrance's: Bilateral relaxing incisions are performed and continued around behind around behind the anterior teeth to meet each other. The entire oral mucoperiosteum is elevated back to the posterior border of the hard palate. Margins of the cleft are incised between the nasal and oral layers. The oral mucoperiosteum is returned to the hard palate about 1-2 cm distal to its original position and anchored by passing sutures through holes drilled in the hard palate. This oral mucoperiosteal flap is best used when the cleft does not extend too far forward in the bony hard palate.

4. Vomer flap: In both single and bilateral clefts of the hard palate, flaps from the vomer may be elevated laterally to be joined with flaps from the lateral nasal aspect of the cleft. Closure of the hard palate defect in complete clefts may be performed with the lip surgery. The usual time is age 10-18 months as a separate procedure and the soft palate is closed shortly thereafter. Great controversy exists regarding timing of closure of the hard palate defect, its relation to the maxillary arch growth, and development of orthodontic problems. Some surgeons close the soft palate early and delay closure of the hard palate to avoid affecting maxillary bone growth.

Various Flaps Associated with Treatment of Cleft Lip and Palate Problems

Including Velopharyngeal Insufficiency

Abbe's

Abbe's technique is a lip switch flap based on the coronal (labial) artery and used to correct defects of up to 50% of either lip. It is used mostly for late correction of defects of the upper lip following bilateral cleft lip repair.

Millard's Forked Flap

The Millard's forked flap technique utilizes scars of the upper lip to lengthen the columella and raise the tip of the nose in cases of previously repaired bilateral cleft lip.

Ecker's Buccal Flaps

This technique is used in primary cleft lip repair and in secondary lengthening procedures to reduce the side-to-side tightness of the soft palate by bringing tissue in from the buccal area.

Dorrance's Palatal Flap

This is an incision along the palatal border of the maxillary teeth for raising the mucoperiosteum of the entire palate in both primary and secondary palate procedures.

Wardill's V-Y Flaps

This incision is similar to the Dorrance incision but angles backward to the midline in the cuspid region.

Langenbeck's Bipedicle Flap

This is similar to the Wardill incision but stops at the cuspid area. Is used as a primary relaxing incision for cleft palate repair.

Pharyngeal Flap

1. An inferiorly based flap may be used for additional tissue in closure of primary clefts of the soft palate. It may be used to decrease hypernasal speech in short or paralyzed palates.

2. A superiorly based flap is used in conjunction with palatal lengthening procedures requiring placement of the mucosal lining or nasal resurfacing of a retropositioned palate. The flap also can be used after a midline splitting of the soft palate, and using the superiorly based pharyngeal flap as a means of closing the nasal portion of the split palate it provides skin lining for the undersurface. The oral surface of the soft palate is closed over the pharyngeal flap. This provides bilateral ports in the area leading to the nasopharynx.

3. Hynes' pharyngoplasty has crossed vertical flaps on the posterior pharyngeal wall to build up a contact pad for the soft palate on phonation.

4. A transverse pharyngeal flap, designed to preserve the superior constrictor muscle innervation and prevent atrophy which has been observed when the Hynes' pharyngoplasty has been performed. The technique includes making a split in the soft palate for visualization and utilization of the crossed flaps for lining the nasal surface of the split soft palate.

Cronin's Nasal Flap

The mucoperiosteum covering the nasal surface of the hard palate posteriorly is carried backwards along the oral mucoperiosteum in palatal lengthening procedures. This tissue provides nasal epithelial covering for the repositioned soft palate.

Island Flap (Millard's)

This is a cut section of the anterior hard palate mucoperiosteum still attached to the greater palatine artery. This flap is turned over and sutured to the nasal raw area created by the palatal pushback procedure.

Smith's Nasopharyngeal Pushback

The lateral wall of the nasopharynx directly behind the hard palate overlying the medial surface of the medial pterygoid plate is elevated widely and incised up to the base of the skull to provide accommodation in the pharynx for repositioning of the soft palate making repeat pushback procedures possible.

Smith's Double Palatal Pushback

The Smith's double palatal pushback was designed for those patients in whom a one-stage operative procedure selected to provide maximum benefit was deemed inadequate. Operation 1 consisted of either a Dorrance's or Wardill's palatal pushback combined with Cronin's nasal flap and a nasopharyngeal pushback. Operation 2 6-9 months later, consisted of a repeat Dorrance's or Wardill's palatal pushback with repeat nasopharyngeal pushback combined with a superiorly based pharyngeal flap. In operation 1 the greater palatine arteries were freed from the undersurface of the oral mucoperiosteal flap for a distance adequate to allow 2 cm of repositioning of the soft palate. The arteries were transected at the time of operation 2.

Smith's Nasopharyngoplasty (Third Faucial Arch) Technique

The Smith's nasopharyngoplasty was designed to improve velopharyngeal competence in patients with palatal paralysis and those unsuccessfully repaired by other techniques or those patients in whom pharyngeal flaps were unsuccessful or found inadequate to reach the shrunken and scarred soft palate remnant. The procedure consists of a transpalatal approach to reach the superior nasopharynx and detach the entire superior portion of the superior constrictor muscle from the base of the skull and pull it down to the lower border of the soft palate. As the muscle is mobilized it can be pulled downward in a draping manner so that the upper central portion anteriorly remains attached only to the soft palate. The muscle is closed upon itself starting inferiorly leaving a central opening at the upper undersurface of the soft palate less than 1 cm in diameter. The closure in effect produces a third faucial arch and nasalizes the eustachian tubes. The potential space laterally and posterior to the displaced muscle cone granulates in.