Cleft Lip

History of the Procedure. Chinese physicians were the first to describe the technique of repairing cleft lip. The early techniques involved simply excising the cleft margins and suturing the segments together. The evolution of surgical techniques during the mid-17th century resulted in the use of local flaps for cleft lip repair. These early descriptions of local flaps for the treatment of cleft lip form the foundation of surgical principles used today.

Tennison introduced the triangular flap technique of unilateral cleft lip repair, which preserved the Cupid's bow in 1952. The geometry of the triangular flap was described by Randall, who popularized this method of lip repair. Millard described the technique of rotating the medial segment and advancing the lateral flap; thus, preserving the Cupid's bow with the philtrum. This technique has resulted in improved outcomes in cleft lip repair.

Problem. Cleft lip is among the most common of congenital deformities. The condition is due to insufficient mesenchymal migration during primary palate formation in the fourth through seventh week of intrauterine life. This results in disfigurement and distortion of the upper lip and nose. Cleft lip may be associated with syndromes that include anomalies involving multiple organs. Patients may have impaired facial growth, dental anomalies, and speech disorders (if a cleft palate is present), and they may experience late psychosocial difficulties.

Frequency. The incidence of cleft lip in the white population is approximately 1 in 1000 live births. The incidence in the Asian population is twice as great, whereas that in the black population is less than half as great. Male children are affected more often than female children. Isolated unilateral clefts occur twice as frequently on the left side as on the right and are 9 times more common than bilateral clefts. Combined cleft lip and palate is the most common presentation (50%), followed by isolated cleft palate (30%), and isolated cleft lip or cleft lip and alveolus (20%). Fewer than 10% of clefts are bilateral.

For parents with cleft lip and palate or for a child with cleft lip and palate, the risk of having a subsequent affected child is 4%. The risk increases to 9% with 2 previously affected children. In general, the risk to subsequent siblings increases with the severity of the cleft.

Etiology. Little evidence exists that links isolated clefts to exposure to any single teratogenic agent. The exception is the anticonvulsant drug, phenytoin. The use of phenytoin during pregnancy is associated with a 10-fold increase in the incidence of cleft lip. The incidence of cleft lip in infants born to mothers who smoke during pregnancy is twice that of those born to nonsmoking mothers. Syndromic clefts are those associated with malformations in other developmental regions, with reported frequencies ranging from 5-14%.

The most commonly recognized syndrome associated with clefts of the lip and palate is Van der Woude syndrome. This syndrome is an autosomal dominant disorder characterized by clefts of the lip and/or palate and blind sinuses, or pits, of the lower lip. Clefts of the secondary palate alone are far more likely to be associated with syndromes than are clefts involving the lip alone or the lip and palate. Most cases of lip clefts are nonsyndromic and believed to be either multifactorial in origin or the result of changes at a major single-gene locus.

Pathophysiology. Development of the upper lip is characterized by fusion of the maxillary prominences with the lateral and medial nasal prominences. This process starts during the fourth week of gestation and is completed by the seventh week. Failure of mesenchymal migration to unite one or both of the maxillary prominences with the medial nasal prominences results in a unilateral or bilateral cleft of the lip, respectively.

Classification. No universally accepted classification scheme exists for clefts of the lip and palate. Veau categorized clefts into 4 classes, as follows:

1. Clefts of the soft plate alone
2. Clefts of the soft and hard palate
3. Complete unilateral clefts of the lip and palate
4. Complete bilateral clefts of the lip and palate

This classification scheme does not provide a means of classifying clefts of the lip alone and ignores incomplete clefts. Kernohan stripped–Y classification allows the description of the lip, the alveolus, and the palate. In this classification, the incisive foramen defines the boundary between clefts of the primary palate (lip and premaxilla) and those of the secondary palate.

Presentation. Clefts of the lip may manifest as microform, incomplete, or complete clefts. Microform clefts are characterized by a vertical groove and vermilion notching with varying degrees of lip shortening. Unilateral incomplete lips manifest varying degrees of lip disruption associated with an intact nasal sill or Simonart band (a band of fibrous tissue from the edge of the red lip to the nostril floor). Complete clefts of the lip are characterized by disruption of the lip, alveolus, and nasal sill.

Bilateral clefts are almost always associated with cleft palate, with 86% of patients with such clefts of the lip presenting with palatal clefts. Unilateral clefts of the lip are associated with palatal clefts in 68% of cases. Nasal regurgitation during suckling may indicate an associated cleft of the palate. All infants with clefts of the lip should have a complete head and neck examination, including careful examination of the palate as far as the tip of the uvula. The presence of a bifid uvula, a translucent central zone in the velum, and a detectable notch of the posterior border of the hard palate indicate submucosal palatal cleft.

All patients with clefts are best referred to multidisciplinary cleft lip and palate centers. Persistent otitis media and middle ear effusions are associated with palatal clefts and warrant regular follow-up care. Depending on the

preference of the surgical centers, the otolaryngologist may elect to perform myringotomy before or after definitive cleft lip and palate repair.

Most cases of lip clefts are nonsyndromic. Parents should be reassured and advised sensitively. At the initial visit, review feeding techniques carefully. For the infant, breastfeeding and the capacity to suck are difficult. However, breastfeeding may be possible with isolated clefts of the lip and the alveolus. For infants with palatal clefts, a variety of special bottles and nipples are available. Crosscut soft nipples made for premature infants facilitate feeding of infants with cleft palate. At the conclusion of the initial consultation, the parents and the infant should be comfortable with the feeding method.

Indications. Clefts of the lip are usually repaired in early infancy. Reassure and advise the parents that operative intervention is best carried out at age 2-3 months. The rule of 10 serves as a safe guideline, ie, body weight should be approximately 10 lb, the hemoglobin concentration 10 g/dL, and age greater than 10 weeks.

Relevant Anatomy. The typical unilateral complete cleft lip deformity results from both a deficiency and a displacement of the soft tissues, the underlying bony structures, and cartilaginous structures. An imbalance of the normal muscular forces acting upon the maxilla results in an outward rotation of the premaxillary-bearing medial segment and posterolateral displacement of the smaller lateral segment.

The inferior edge of the anterior nasal septum is displaced out of the vomerine groove into the noncleft nostril, and the anterior septum leans laterally over the cleft. The overlying columella invariably is short on the cleft side and distorted by the displaced caudal septum. In the nasal tip, the alar cartilage is characteristically deformed, and the medial crus is displaced posteriorly. The dome is separated from that of the noncleft side, and the lateral crus is flattened and stretched across the cleft. The axis of the nostril on the cleft side is characteristically oriented in the horizontal plane. This position is in contrast to the normal vertical axis of the nostril on the opposite side.

The muscular fibers of the orbicularis oris do not decussate transversely as in the normal lip; rather, they course obliquely upward, paralleling the cleft margin toward the alar base on the lateral side of the cleft and toward the base of the columella medially. The philtrum on the cleft side is short, and the presumptive Cupid's bow peak is displaced superiorly. The vermilion is deficient on the cleft side of the medial element.

Complete bilateral clefts of the lip result from failure of the premaxillary segment to fuse with the lateral maxillary segments. Subsequent forward growth of the premaxilla, attached only to the vomer above, leads to its projection beyond the lateral segments. Within the isolated prolabium, the skin is foreshortened vertically, the white roll is underdeveloped, and the vermilion is deficient. The prolabium lacks muscle fibers, and the philtral ridges, the central philtral dimple, and Cupid's bow are absent. The bilateral cleft nasal deformity is

characterized by flaring of the alar bases and wide separation of the domal segments of the alar cartilages. The columella is markedly shortened, causing the nasal tip to be depressed.

The orbicularis oris muscle is the primary muscle of the lip and can be divided functionally and anatomically into 2 parts. The deep component, in concert with other oropharyngeal muscles, works in swallowing and serves as a sphincter. The superficial component is a muscle of facial expression and inserts into the anterior nasal spine, sill, alar base, and skin to form the philtral ridges.

In a complete cleft lip (CL), the deep fibers of the orbicularis oris muscle are interrupted by the cleft and end on either side of the defect instead of making their way around the mouth. In addition, the superficial component of the orbicularis oris turns upward, along the margins of the cleft and ends beneath the ala or columella. Incomplete cleft lip (CL) behaves in a similar manner, except when the cleft is less than two thirds of the height of the lip. In this case, the fibers of the muscle run along the margins of the cleft, then change direction and run horizontally over the top of the cleft. These muscle fibers are interspersed with connective tissue.

The blood vessels parallel the course of the muscle fibers and run along the margins of the cleft toward the columella or alar base, where they form anastomoses with nearby vessels.

In the bilateral deformity, the anatomic characteristics are determined by the degree of completeness of the cleft and its symmetry. The cleft may involve the primary palate alone or in conjunction with the secondary palate. Although the prolabium varies in size, it is usually retracted and lacks muscle fibers. In addition, the columella is absent and the prolabium appears attached to the top of the nose in some cases. The size and position of the premaxilla vary and effectively can be excluded with a collapse of the alveolar arch.

The extent of nasal deformity associated with cleft lip (CL) varies from patient to patient.

Contraindications. Coexisting medical conditions that would result in cardiopulmonary complications, bleeding disorders, infection, and/or malnutrition are all contraindications to surgery.

Treatment. Surgical Therapy. Depending on the width of the cleft, some centers may perform presurgical orthopedic management to reduce the width of the bony cleft and to align the maxillary arch prior to definitive lip repair. This may be accomplished by using external traction devices or passive orthodontic plates. Surgical lip adhesion may be used for wide clefts as an alternative to presurgical orthopedics. In lip adhesion, the soft tissues of the superior lip are united, essentially converting a wide complete cleft to an incomplete cleft.

Because each cleft is unique, definitive repair of the cleft lip should be individualized. Mirault was among the first to describe the technique of increasing lip length by using a small flap taken from the cleft side. However, this technique did not reconstruct the Cupid's bow. Le Mesurier later described

the inset of a rectangular flap from the cleft side into a releasing incision on the noncleft side to create an artificial Cupid's bow. Tennison described the triangular flap to preserve the Cupid's bow. Randall later worked out the geometry of this flap, adding precision and reproducibility to the triangular flap technique. The technique creates an unnatural scar across the philtral column and flattens the philtral dimple. In addition, the triangular flap method does not address the nasal deformity.

Millard introduced the rotation-advancement technique in 1955, which overcomes many of the pitfalls of earlier techniques. The rotation-advancement technique is the most commonly used method today for the repair of unilateral clefts. The technique preserves the Cupid's bow and the philtral dimple and improves nasal tip symmetry. The rotation-advancement lengthens the lip by means of a rotation incision that releases the medial lip element, allowing the Cupid's bow to rotate downwards into normal position. A small backcut may be used to further increase the length, if needed. The lateral lip element is advanced into the gap created by rotation of the medial element, thus completing reconstruction of the upper lip.

Preoperative Details.

Administer general endotracheal anesthesia.

Monitor pulse, respiration, blood pressure (BP), ECG, and temperature.

Gently hyperextend the neck to provide optimal exposure.

Ensure appropriate padding of pressure points.

Ensure corneal protection (eg, lubricant, taping of the eyelids).

Place the patient on a warming blanket.

Intraoperative Details. Unilateral cleft repair. Skin markings.

Center of Cupid's bow;

Cupid's bow peak at the vermilion-cutaneous junction, noncleft side of the medial element;

Proposed Cupid's bow peak, cleft side of medial element;

Midline of columella;

Alar base, noncleft side;

Alar base, cleft side;

Proposed Cupid's bow peak, lateral element;

Tip of advancement flap;

End of backcut.

Key points: 1) The position of point 3 may be determined by transposing the distance between 1 and 2, such that the distance between 1 and 2 is equal to the distance between 1 and 3. 2) The distance between the alar base and Cupid's bow peak on the noncleft side should equal that on the cleft side, ie, the distance from 2-5 is equal to the distance from 7-6. 3) The difference between the distance from the columellar base to points 2 and 3 represents the deficiency in vertical length that must be gained to level the Cupid's bow. Although the rotation incision allows point 3 to drop inferiorly, some vertical deficiency of

the cleft side may remain. The added length may be gained by making a small backcut medial to the philtral column on the noncleft side. The advancement flap derived from the lateral element fills the opening created by the rotation incision and any backcut in the medial element; hence, the distance from 3-5 plus the added length gained by the backcut equals the distance from 6-7. Introduction of a small triangular flap from the lateral element into a small transverse incision in the lower part of the lip may also serve to lengthen the cleft side of the medial element and to improve the contour of the lip. The base width of this flap is equal to the height of the vermilion-cutaneous roll. 4) The rotation incision curves gently from point 3 to the columellar base, hugging the columellar-lip junction, and stops medial to the philtral column on the noncleft side. Crossing the normal philtral column results in an undesirable elongation of the lip on the noncleft side. In the infant with a rectangular philtrum, this incision may be modified as described by Mohler. 5) Place point 7 on the lateral element at a point level with the Cupid's bow peak on the noncleft side (point 2) and where the white roll remains well developed. Placing this point too far laterally produces an unnatural shortening of the lateral lip element, which results in a noticeable imbalance. Placing this point too far medially, where the white roll is poorly developed, results in a noticeable irregularity of the white roll. To gain some extra vertical height, if needed, point 7 may be moved 1 mm laterally and point 3 moved 1 mm medially. 6) The advancement incision curves from point 7 to point 8, then a variable distance to point 9, depending on the amount of rotation needed to correct the flare of the displaced alar base. 7) When possible, line up the point at the junction of the wet and dry vermilion; this point is also called the red line.

Repair of the orbicularis oris. Reorientation and repair of the orbicularis oris muscle bundles are essential for normal lip function and eversion of the lip border. Failure to adequately address the muscle at the time of lip repair results in abnormal motion or contour when pursing the lips and in a characteristic bulge in the lateral lip element. A variety of techniques for reorienting the orbicularis oris muscle fibers in unilateral clefts have been described, although the optimal method of muscle repair remains to be determined. Park advocates careful identification and precise reapproximation of the superficial and deep components of the muscle.

Primary nasal correction. In every case, reconstruct both the lip and the nose at the primary operation. Repair of the lip in infancy while delaying nasal repair until later in childhood is no longer appropriate. Reconstruction of the cleft nasal deformity remains the most challenging aspect of cleft surgery. Principles of primary nasal correction include the following: 1. Wide undermining of the nasal skin on the cleft side, freeing the skin from the underlying nasal skeleton. 2. Elevation of the slumped alar cartilage on the cleft side to the normal level using internal or external suspension sutures. 3. Medial advancement of the lateral crus and alar base on the cleft side.

Bilateral cleft lip repair. Prior to surgical repair, the use of presurgical orthopedic appliances can reduce significant premaxillary protrusion. The premaxillary segment varies considerably in size and in the extent of its protrusion. In incomplete clefts, attachment of the premaxilla to one or both lateral maxillary segments limits premaxillary protrusion. In complete clefts, retroposition of the premaxilla prior to definitive lip repair often is necessary. This procedure may be accomplished through presurgical orthopedics, using external traction devices or passive orthodontic plates. Retroposition of the premaxilla may also be accomplished through surgical lip adhesion. Surgical setback of the premaxilla, a technique popular in the 19th century, is associated with subsequent midfacial growth impairment and should be avoided.

Modifications of the Millard straight-line, banked, forked-flap technique currently are the most widely used methods for repair of bilateral cleft lip. These techniques work well for the repair of complete bilateral clefts and may be modified for the repair of incomplete and/or asymmetrical clefts.

Key points. 1. The prolabium is always used in reconstruction of the philtrum, even if severely deficient. 2. The prolabial vermilion nearly always is deficient, and the prolabial white roll usually is indistinct. Therefore, in most cases, the prolabial mucosa is turned down to line the buccal alveolar sulcus; the tubercle and white roll are reconstructed using paired white roll–vermilion–orbicularis marginalis flaps from the lateral lip elements brought beneath the prolabium. 3. The prolabium must not be left too wide and should rarely exceed 5-6 mm in width. 4. The orbicularis peripheralis muscle is freed from its abnormal attachments at the alar bases and from the overlying dermis. This allows the muscle to be mobilized and reconstructed over the premaxilla. 5. Anatomic positioning of the alar cartilages is performed at the time of primary lip repair.

Postoperative Details. 1. After surgery, feeding is resumed using a soft crosscut nipple. 2. Infants remain hospitalized for intravenous hydration until oral intake is sufficient (usually 24 h). 3. The suture lines are kept clean by gentle application of a dilute hydrogen peroxide solution, and a small amount of antibiotic ointment is applied to the repair 3 times daily and after feeding. 4. If nonresorbing suture material is used, the sutures are removed by the fifth postoperative day. 5. Soft elbow restraints are used for 2-3 weeks to keep the infant from manually disrupting the repair.

Follow-up. Arrangements for suture removal are made prior to or immediately after discharge. All patients require long-term follow-up care. A dedicated multidisciplinary team approach and evaluation in different stages of the patient's life is important. Assess speech, language, hearing, somatic growth, and development regularly. Appropriately assess general dental health. Orthodontic management and secondary surgical procedures, such as bone grafting, are carried out during the school years. Patients with significant midface retrusion may require treatment. Secondary procedures to correct the tip

in nasal asymmetry may be performed at school age; however, if the reconstruction involves osteotomy, delay the procedure until the completion of nasal growth (age 16-17 y). Emotional difficulties may emerge because of poor self-esteem during adolescence and should be recognized and addressed early.

Complications. Complications following cleft lip repair are unusual. Wound infections following surgery are uncommon and are treated with appropriate antibiotic therapy. Although immediate wound dehiscence is best repaired prior to discharge, treat delayed dehiscence after the scar has settled.