

Chapter 66: Cleft Lip and Palate

Rober W. Seibert, Robert M. Bumsted

Cleft lip and palate deformities are common congenital disorders whose management requires a long-term comprehensive program of care involving many medical and dental disciplines. Patients with a cleft lip or palate have numerous associated problems, such as the following:

1. Basic anatomic deformity
2. Deficient facial growth
3. Dental problems
 - a. Missing, malformed, and supernumerary teeth
 - b. Malocclusion
4. Speech problems
 - a. Velopharyngeal incompetence
 - b. Secondary articulation disorders
5. Otologic problems
 - a. Eustachian tube dysfunction
 - b. Chronic ear disease
 - c. Hearing loss
6. Psychologic disorders
7. Additional congenital anomalies.

These problems are discussed in detail, with special emphasis on the timing and management techniques necessary to correct them. The numerous disciplines needed for the management of these patients have various therapeutic approaches; one must develop an overall program of care to maximize the benefits of each therapy for the patient.

Classification

Although numerous systems for classification of cleft lip and palate exists, there is no universally accepted standard classification of these deformities. The following is a basic classification of cleft lips and palates.

- I. Cleft lip
 - A. Unilateral or bilateral
 - B. Complete or incomplete
- II. Cleft palate
 - A. Location in reference to incisive foramen
 - 1. Primary - anterior
 - 2. Secondary - posterior
 - B. Unilateral or bilateral
 - C. Complete or incomplete
- III. Submucous cleft palate.

In addition, Fig. 66-1 illustrates the classification system used by the University of Iowa Cleft Palate Team. These systems are not proposed as the best method of classification; however, they are functional and useful classifications of these congenital abnormalities.

Clefts of the lip can be classified as unilateral or bilateral; the extent of the cleft is also classified, as incomplete or complete. A complete cleft involves the entire lip and is usually associated with involvement of the underlying alveolar arch (premaxilla). An incomplete cleft involves a portion of the lip and may vary from a muscular diastasis with intact overlying skin to a large cleft with only a small band of residual tissue connecting the two sides of the lip. This small band, frequently called Simonart's bar or band, is demonstrated in Fig. 66-2.

Clefts of the palate can be classified according to the side (unilateral or bilateral) and the degree of involvement (complete or incomplete). Congenital clefts can also be classified by the area of involvement, which is based on embryologic development. The incisive foramen is the dividing line. The primary palate is formed initially and consists of the portion of the palate located anterior to this foramen, which also includes the alveolar arch. The secondary palate, which forms later in embryonic development, is located posterior to the incisive foramen. A unilateral cleft of the secondary palate occurs in those clefts in which the palatal process of the maxilla of one side is fused with the nasal septum. A bilateral cleft of the secondary palate has no connection between either palatal process and the nasal septum. Figs. 66-3 and 66-4 illustrate clefts of the palate. A complete cleft of the entire palate

involves all of the palate and one or both sides of the premaxilla (alveolar arch). This type of cleft is usually associated with a cleft of the lip. An incomplete cleft of the palate is usually considered to involve only the secondary palate. The degree of involvement may vary, from submucous cleft to a cleft palate to a cleft extending to the incisive foramen. A submucous cleft palate, which is caused by a mesodermal deficiency of the muscles of the palate with intact overlying oral and nasal mucous membranes, is characterized by the presence of a bifid uvula, muscular diastasis of the muscles of the soft palate, resulting in a bluish midline streak, the zona pellucida, and loss of the posterior nasal spine or frank notching of the posterior hard palate (Fig. 66-5).

Embryology

The embryologic development of the lip and palate may be subdivided into two phases: the primary palate and lip, and the secondary palate. The lip and primary palate normally develop at 4 to 7 weeks of developmental age. At no time during normal development of the lip and primary palate does a cleft exist (Streeter, 1948). Clefts of this area probably result from a failure of a mesodermal delivery or proliferation from the midline frontonasal process and lateral maxillary processes. It is unlikely that a failure of fusion of these processes causes clefts of the lip (Patten, 1971). The primary palate develops during the same period of time that the lip does. It is formed from a block of mesoderm that lies beneath the nasal pits. Normally the ectoderm in this area forms a continuous layer from side to side, and, again, at no time does a cleft exist (Davies, 1973).

The secondary palate develops at 7 to 12 weeks of age. It is formed by medial growth of the palatal shelves of the maxilla, which eventually fuse together during normal development. Initially the tongue is located between these palatal shelves and prevents their fusion. However, as development proceeds, the tongue is shifted downward and allows contact between the medially growing palatal shelves and the nasal septum, forming the secondary palate. Fusion of these shelves and septum begins at the area of the incisive foramen and proceeds posteriorly to the uvula. Thus a natural cleft occurs during normal development of the secondary palate.

This embryologic development results in two basic types of clefts. One is a cleft involving the secondary palate, which is always located posterior to the incisive foramen. The other is a cleft involving the primary palate (the alveolar arch and lip), located anterior to the incisive foramen. These basic clefts may exist separately or in combination. In addition, a cleft involving only the lip may occur as an isolated entity (in up to 25% of the cases); however, a cleft of the alveolus is always associated with a cleft lip.

Incidence and etiology

A cleft deformity occurs in approximately 1 of every 680 births. In general, 10% to 30% are isolated cleft lips, 35% to 55% involve both the primary and secondary palate (cleft lip and palate), and 30% to 45% involve only the secondary palate (Drillien et al, 1966).

Several factors influence the incidence of cleft lip and palate. The etiology of cleft lip and palate is multifactorial. Numerous conditions have been shown to contribute to the development of these congenital deformities. They consist of the patient's sex, race, and

parent's age, as well as genetic disorders and environmental factors.

Sex of the patient is important because isolated cleft lip and cleft lip and palate have a greater incidence in males, with a ratio to females of approximately 2:1. Isolated cleft palate is more common in females, with a similar 2:1 ratio to males (Fogh-Anderson, 1942).

Increased parental age appears to result in a slightly increased incidence of cleft disorders. The most important factor appears to be increased paternal rather than maternal age (Fraser and Calnan, 1961).

Race appears to play a significant role in the incidence of cleft lip and palate; however, it does not affect the incidence of isolated cleft palate (Chung and Myrianthopolus, 1968). In previous studies the highest incidence occurred in native Americans, followed by Orientals and whites, with the lowest incidence in blacks (Converse et al, 1977).

Isolated clefts of the lip and combinations of cleft lip and palate appear to be genetically different from isolated cleft palates. The incidence of isolated cleft palate is not increased in relatives of patients with either isolated cleft lip or combined cleft lip and palate. Conversely, patients with isolated cleft palate also fail to have an increased incidence of isolated cleft lip or combined cleft lip and palate in their relatives (Fogh-Anderson, 1942).

Classic mendelian inheritance of congenital clefts is rare. Occasionally a cleft lip and palate are seen in syndromes that demonstrate both autosomal dominant and recessive inheritance. More common syndromes include lobster claw defect, cleft lip with lip pits (Van der Woude's syndrome), and cleft lip and palate associated with ectodermal dysplasia. In addition, cleft lip and palate are also occasionally associated with chromosomal abnormalities, such as trisomy D. Multifactorial inheritance causes the vast majority of cases of congenital cleft lip and palate; the frequent occurrence of clefts in families over several generations without a definitive mendelian pattern of inheritance demonstrates this fact. Genetic counseling is extremely valuable for parents of patients born with a cleft disorder to determine the possibility of a chromosomal abnormality or mendelian inheritance. Thus the value of consultation with a pediatric geneticist must be emphasized. Table 66-1 presents the rate of recurrence in subsequent children, after chromosomal or genetic causes of cleft disorders are excluded.

Table 66-1. Risk factors for cleft recurrence

Affected relatives	Predicted recurrence (%)	
	Cleft lip and palate	Cleft palate
General population	0.1	0.04
One sibling	4.4	2.50
One parent	3.2	6.80
One sibling, one parent	15.8	14.90
Two siblings	9.0	1.00

Data from Curtis E et al: Am J Dis Child 102:853, 1961.

Several environmental teratogens have been demonstrated to produce cleft deformities in laboratory animals. However, in humans, only rubella virus, thalidomide, and aminopterin have been demonstrated to produce clefts (Cahen, 1964). The incidence of other major congenital malformations in patients with cleft lip and palate has been shown to be increased when compared to the normal population. The most common associated anomaly of cleft lip and palate is the Pierre Robin syndrome, which consists of cleft palate, micrognathia, and glossoptosis.

General Management

Team approach

The multitude of complex clinical problems present in cleft patients necessitates a team approach to provide optimal management. Because of the problems' variation and complexity, no single specialist can be expected to provide satisfactory treatment of the entire disorder. The team concept is an interdisciplinary approach that maximizes diagnostic and therapeutic procedures in patients with clefts. Each team member contributes in a particular area of expertise, interacting with team members of other disciplines to facilitate a proper sequence and timing of therapeutic procedures in the comprehensive care plan. The following outline presents the composition of the ideal cleft team.

1. Reconstructive surgeon
2. Otologist
3. Dentist
 - a. Pedodontist
 - b. Orthodontist
 - c. Prosthodontist
 - d. Oral surgeon
4. Speech pathologist
5. Audiologist
6. Pediatric geneticist
7. Psychologist/psychiatrist
8. Social worker
9. Nurse
10. Nutritionist.

These professionals makeup the ideal membership of a cleft team, but not all are absolutely necessary. A minimal cleft team includes a surgeon, a dental specialist, and a speech pathologist. However, this team requires occasional additional consultation from other, nonrepresented specialists for proper management of certain cleft problems.

The cleft team should meet on a regular basis to discuss and plan the overall management of each patient with a cleft disorder. These meetings allow a free interchange of ideas and an open discussion of proper timing and techniques of patient care. This provides an individualized approach to the management of each cleft patient based on physical and psychosocial abnormalities and development. *The primary advantage of the team approach is that it is patient oriented rather than specialty oriented.*

Table 66-2 presents the usual age at which various procedures are performed. However, this is an overview; the timing and sequence of these procedures may vary significantly in individual patients.

Table 66-2. Usual age of cleft patient for various therapeutic procedures

Procedure	Initial procedure	Second procedure
Lip adhesion	2-4 wk	3 mo
Lip repair		
Primary	2-3 mo	4-5 mo
After adhesion	5-7 mo	7-10 mo
Palate repair	12-30 mo	
Correction of velopharyngeal incompetence	4 yr and older	
Orthodontic therapy	4 yr and older	
Lip revision	4 yr and older	
Premaxillary recession	5 yr	
Nasal reconstruction		
Tip	6-10 yr	
Dorsum and septum	15-17 yr.	

Psychologic issues

The birth of a child with a cleft deformity is often a devastating event for the parents, who may react with disappointment, guilt, resentment, grief, and feelings of inadequacy (Tisza and Gumpertz, 1962). These feelings may have an adverse affect on the parent's relationship with the child, which in turn complicates the child's character and personality development (Ross, 1964). During the initial parental counseling it is vital that they be allowed to verbalize these feelings and be reassured they are not to blame for the cleft deformity. In addition, a general outline for the child's long-term care should be presented at this time. This information increases their understanding of the deformity and methods available for correcting it.

Numerous conditions exist in cleft patients that may adversely affect their intellectual, social, and personal development. These factors include care and feeding problems in infancy,

the parental reaction to the cleft deformity, the frequent surgical procedures that may be required during early childhood, and possible defective speech, hearing loss, and facial disfigurement, which are associated with the cleft. However, the frequent occurrence of these factors in cleft patients has not been clinically found to produce a typical "cleft palate personality", that is, a personality significantly different from noncleft children (Wirks, 1971).

Speech intelligibility problems are found in the majority of cleft palate patients, and in 25% of them these problems are so severe that their speech is difficult to understand (Westlake and Rutherford, 1966). Hearing loss of varying severity is also more common in cleft palate patients. Although its incidence decreases with increasing age, it may also affect the speech, learning, and psychologic development of cleft palate patients (Levine, 1962). Other congenital malformations are present in 14% of patients with cleft lip and palate (Greene et al, 1965). In addition, intelligence tests have demonstrated a higher proportion of children with less than average intelligence or retardation in the cleft palate population (Goldstein, 1961; Ruess, 1965). Usually the deficit is greater in verbal, as compared to nonverbal, intelligence. This may be related to the delayed speech development of cleft patients rather than to a basic underlying neurologic problem.

Nursing care: feeding

Special problems in the nursing care of cleft patients include feeding difficulties, the necessity of frequent surgical procedures, which require specialized nursing care to prevent airway and bleeding complications, and special diets. In addition, parents often initially verbalize guilt feelings to the nursing staff; and for this reason, the staff's knowledge and education in the management of psychologic problems associated with cleft disorder is extremely beneficial. The staff also frequently provides available instructions to the parents on the day-to-day care of the cleft child.

Neonates with cleft palate frequently have feeding problems, and initially, devices such as an Asepto syringe with rubber tubing or a bulb syringe may be required. We have found a compressible feeder such as the Mead Johnson feeder to be successful in many infants. Trial and error is required until the best feeding device and technique are found. In infants with very wide complete clefts, a palatal feeding prosthesis may be of benefit. This device is constructed by a prosthodontist or an orthodontist and is well tolerated by infants when placed in the first few days to weeks of life. Parents are instructed how to remove and clean the prosthesis once each day. A nutritionist evaluates the patient's feeding regimen for optimal growth. Often concentrating the formula to provide additional calories is necessary. During the feeding the infant should be placed in a slightly upright position, with the nipple or bulb placed in the buccal area of the cheek opposite the cleft. Aerophagia is likely to occur, requiring more frequent burping than noncleft children need. After the feeding the oral cavity should be cleaned by being rinsed with water and the mouth should be wiped with a damp cloth to prevent the accumulation of food and mucus in the area of the cleft. Breast feeding is rarely successful for children with cleft palates but may be attempted for children with clefts involving only the lip.

Experienced and well-trained nurses providing postoperative care after primary and secondary surgical procedures on cleft patients are invaluable. Postoperative airway obstruction and bleeding may result in a life-threatening situation, and early detection and

proper management are essential. In addition, the use of a mist tent may prevent intraoral crusting, which can also contribute to airway obstruction. Suture lines should be cleaned as necessary with hydrogen peroxide to remove crusts and an antibiotic ointment is applied two to three times each day. The nursing staff initially performs this task; they show the parents how to perform this care after discharge.

Anatomy

Cleft lip deformity

The anatomic deformity associated with a cleft lip usually involves the bony skeleton and the soft tissue of the lip and nose. The degree of deformity often depends on the severity of the cleft lip.

In normal patients the orbicular mouth muscle forms a complete sphincter around the oral cavity. Patients with an incomplete unilateral cleft lip that does not involve more than two thirds of the height of the lip have intact upper fibers of this muscle, which pass over the top of the cleft and connect the medial and lateral lip segments to form a partial oral sphincter. In these patients the lower muscle fibers are directed upward and usually attach to the subcutaneous tissue of the cleft margins or to the underlying bone (Fig. 66-6). In a complete unilateral cleft lip the muscle fibers are directed upward, parallel to the margins of the cleft, and terminate beneath the ala nasi (laterally) and the base of the columella (medially). Frequently, excessive muscle exists in the lateral segment. This may be detected visually or by palpation. In addition, the musculature of the medial segment is often underdeveloped, and muscle fibers may be disorganized and fail to extend to the cleft margin (Fara, 1977. Fig. 66-7 illustrates the muscle deformity of the complete unilateral cleft lip.

The muscle deformity of bilateral incomplete cleft lip is similar to that described for incomplete unilateral cleft lip. In the complete bilateral cleft lip the lateral segments are similar to those of the unilateral cleft. However, muscle fibers are absent in the prolabial (medial) segment, which is composed of collagenous connective tissue (Fig. 66-8).

The arterial supply of various cleft lip deformities is illustrated in Figs. 66-7 and 66-8. Frequently the arterial supply is smaller than normal on the medial portion of the unilateral cleft defect and markedly reduced in the prolabium with a bilateral cleft lip.

Cleft palate deformity

Clefts of the secondary palate have varying deficiencies of bone (hard palate) and muscle (soft palate). Mucosal deficiency is always present except in a submucous cleft palate. Normal structure and function of the soft palate involve numerous muscles, including the tensor veli palatini, levator veli palatini, palatopharyngeal, palatoglossus, and uvula muscles. The attachments of these palatal muscles occur in a midline raphe normally, forming the so-called levator sling in the middle third of the soft palate; the anterior third is an aponeurotic segment. In cleft patients, these muscles are directed anteriorly and have an abnormal insertion into the posterior margin of the bony palate and are usually hypoplastic (Fig. 66-9). This abnormal muscle insertion often prevents adequate palatal function, which in turn leads to additional retardation of the palatal muscular development. In general, the levator and

tensor veli palatini and uvular muscles are more hypoplastic and demonstrate greater functional abnormalities than the other palatal muscles (Fara, 1977).

Facial skeleton

The facial skeleton is frequently deformed in patients with a cleft of the lip and palate (Latham, 1977). The severity of the skeletal deformity is often directly related to the severity and degree (unilateral or bilateral) of the cleft.

Common deformities of the unilateral cleft include lateral displacement of the premaxilla on the noncleft side, frequent upward tilting of the premaxilla into the cleft defect, malformation of the lower nose, distortion of the nasal septum, and frequent underdevelopment of the maxilla on the side of the cleft. There may also be a marked malalignment of the alveolar arches.

In patients with a bilateral cleft lip and palate, the premaxilla is frequently protruding (located anterior to the maxillary alveolar arches) and often grossly deficient in bone. The overlying midportion of the lip commonly attaches directly to the nasal tip with a nearly total absence of the columella. The maxillary alveolar arches are often underdeveloped and are frequently collapsed in an upward and inward direction. Fig. 66-10 shows these deformities.

Facial growth in patients with cleft lip and/or palate is often abnormal. Both the cleft deformity and the surgical procedures performed to correct this deformity appear to influence it. The precise extent of each factors' contribution to the normal facial growth is still unknown. Previously reported studies (Bishara et al, 1976, 1978; Krause et al, 1975; Ortiz-Monasterio et al, 1966) have demonstrated that the majority of clefts are capable of developing an essentially normal facial skeleton except in the area of the cleft defect. Thus the surgical procedures performed to correct the deformity may play a greater role than the actual cleft deformity in the abnormal facial growth frequently observed in patients with cleft lip and palate. Patients with repaired clefts of the lip and palate may demonstrate impaired maxillary growth with collapse of the maxillary arches, producing midface retrusion (Ross, 1977; Fig. 66-11).

Palate repair may affect facial growth (Ross, 1977, 1987), but well-controlled studies on the extent of the effect are not available at this time. In general, facial growth appears to be adversely influenced by the age at the palate repair (there is greater inhibition of repair at an earlier age), the type of repair used (there is better growth with a less traumatic procedure), and the tension of the repaired palate (increased tension produces greater inhibition of growth). The interrelationship of all these variables precludes any definitive statement regarding the precise effect of each on facial growth, but their total combined effect does frequently appear to cause a clinically significant inhibition of facial growth.

The effect of repair of an isolated cleft lip on maxillary growth in humans is uncertain. Some authors (Ross, 1977, 1987) consider lip repair to have no significant effect on facial growth, but others (Eisbach and Bardach, 1978) demonstrated in animal experiments that excessive tension of the repaired lip often contributes to decreased anterior growth of the maxilla.

In summary, much of the abnormal facial growth observed in cleft patients appears to be related to surgical procedures performed to correct the cleft deformity. Careful consideration therefore must be given to the possible effect on future facial growth of all procedures performed on cleft patients. This appears to be most significant for the surgical procedures performed on a young patient.

Cleft Lip Repair

I. Lip adhesion

A. Performed at 1 month

B. Advantages

1. Converts complete to incomplete cleft
2. Improves alveolar arch alignment
3. Possible prolabial growth
4. Improves nasal symmetry
5. Assists with feeding
6. Psychologic benefit to parents

C. Disadvantages

1. Additional operations (one for unilateral, one or two for bilateral)
2. Increased scar tissue

II. Lip repair

A. Timing

1. Traditional - 10 weeks (rule of 10s (see explanation below))
2. Early repair - 4 to 6 weeks
3. Delayed if lip repair

B. Techniques

1. Straight line
 - a. Advantage: easy repair

- b. Disadvantage: limited indications
- c. Indications: incomplete and narrow clefts
- 2. Rotation advancement flaps (Millard, 1958)
 - a. Advantages
 - (1) Re-creation of philtrum
 - (2) Modification during repair
 - b. Disadvantages
 - (1) Difficult in wide-cleft or short lateral lips
 - (2) Requires experienced surgeon for excellent results
 - (3) May narrow nostril
- 3. Triangular flap (Z-plasty technique) (Bardach, 1967; Randall, 1959; Tennison, 1959)
 - a. Advantages
 - (1) "Measured" technique
 - (2) More easily taught
 - (3) Possible wide cleft
 - b. Disadvantages
 - (1) Scar interrupts philtrum line
 - (2) Difficult to modify during procedure
- 4. Reconstruction of orbicular mouth muscle is vital.

Basic goals of primary lip repair include lip reconstruction with correct alignment of the orbicularis oris muscle and Cupid's bow, symmetric reconstruction of the vermilion, creation of a nasal floor and sill, and symmetric placement of the base of the ala and columella.

Timing

The timing of lip repair has marked variation in previously reported series. In some countries the lip is repaired within 48 hours after birth (Cannon, 1967; Lewin, 1964). In the USA most surgeons use the "rule of 10s", in which surgical repair is performed when the

infant is at least 10 weeks old, weighs 10 pounds, and has a hemoglobin level of 10 g. The advantages of immediate repair are that the possibility of maximal healing potential exists, the surgical procedure prevents a separate hospitalization and is thus less traumatic, and the parents leave the hospital with a relatively normal-appearing child. Advantages of the delayed repair of the lip include larger tissue for the repair, ample time to allow complete pediatric evaluation of the patient, and an increased understanding of the congenital defect by the parents, often producing increased parental tolerance of any residual deformity after surgical reconstruction. Recently repair of the lip at 4 to 6 weeks has been found to be highly satisfactory in healthy infants. A surgical procedure in such small infants may require special pediatric anesthesia expertise, the use of magnification during the operation, and the absence of other significant congenital problems

Lip adhesion

Lip adhesion may be used as a preliminary procedure in the therapy of cleft lip. This procedure is usually performed within the first month of life, with definitive lip repair performed 4 to 6 months later. It may be performed with laterally based flaps obtained from the lip itself (Randall, 1965) or inferiorly based flaps of vermilion (Walker et al, 1966) to convert a complete cleft of the lip into an incomplete cleft. During the time after the lip adhesion, the lip can act as an orthodontic appliance, improving the alignment of the maxillary arches and narrowing the cleft lip defect. Fig. 66-12 illustrates one technique of unilateral lip adhesion (Seibert, 1990).

The primary advantages of lip adhesion are conversion of a complete cleft into an incomplete cleft of the lip, improvement of the alignment of the maxillary arches, and occasional occurrence of considerable growth of the prolabium in patients with bilateral clefts (Randall and Graham, 1971). Disadvantages of lip adhesion include the additional operation in the unilateral cleft or symmetric bilateral cleft, increased formation of scar tissue produced by the adhesion, and the possible dehiscence of the adhesion. Staged adhesions may be indicated in a bilateral cleft with a deviated premaxilla as an orthopedic maneuver.

No universally accepted indications currently exist for the use of lip adhesion. Some surgeons never use this technique, whereas others utilize it in all complete clefts. In general, it appears to be most beneficial in wide clefts with marked malalignment of the alveolar arches that may prevent obtaining a satisfactory result in primary lip repair (Fig. 66-13).

Definitive cleft lip repair procedures

Although numerous methods of definitive lip repair have been described in the literature, three basic types of procedures are available: straight-line closure, rotation advancement flaps, and interdigitation of triangular or rectangular flaps. The straight-line repair is rarely used and is not discussed in this chapter.

Rotation advancement technique

Millar (1958) originally described the rotation advancement method, which is commonly known as Millard's repair (Fig. 66-14). This technique utilizes downward rotation of the superiorly displaced medial lip segment with advancement of the lateral lip segment

medially into the defect created below the nose. Advantages of the rotation advancement method include minimal discarding of tissue, creation of a suture line that re-creates the philtrum on the side of the cleft, construction of a nasal sill, repositioning of the base of the ala nasi to a more normal position during the repair, and adjustability of the repair during the surgical procedure. Disadvantages of this technique include difficulty in closing wide clefts without extensive undermining of soft tissue overlying the face of the maxilla, especially without a lip adhesion, occasional sacrifice of excessive mucosa and even lip tissue when there is inadequate height of the lateral lip segment, and excessive manipulation of the nostril, which occasionally results in an abnormally small nostril. In addition, a significant amount of surgical judgment and experience with this repair may be necessary to obtain excellent results. Also, this technique of lip repair does not lend itself to the use of exacting measurements to reconstruct the lip. Currently it is probably the most frequently used repair in the USA and appears to be the method of choice for the repair of most clefts of the lip (Musgrave and Garrett, 1977).

Triangular flap techniques

The second most common method of lip repair is the interdigitation of triangular flaps (basically a Z-plasty technique) for reconstruction of the lip. The Tennison-Randall technique (Randall, 1959; Tennison, 1959) utilizes a single inferiorly based triangular flap on the lateral side of the flap, which is inserted into an incision on the medial side of the lower portion of the lip (Fig. 66-15). Other surgeons (Skood, Bardach) employed two triangular flaps, superior and inferior, to obtain sufficient lip length and break up the scar line. These techniques are based on exact and careful measurements and definitive landmarks; they can be more easily used on wide clefts, require minimal discarding of tissue, and preserve the Cupid's bow (Fig. 66-16). Disadvantages of the transposition flaps include that a zigzag scar is created on the lip rather than following the line of the philtrum and that the repair cannot be modified significantly once it is designed and incisions are performed.

Repair of bilateral cleft lip

The repair of a bilateral cleft lip is frequently more complex and challenging than repair of the unilateral cleft because of two major problems: the prolabium and premaxilla.

The prolabium is frequently extremely small, deficient in muscle and vermilion, and attached to the nasal tip or based on an extremely short nasal columella. This markedly deficient prolabial segment must be used to reconstruct the entire central portion of the lip. Growth of this segment before the definitive cleft procedure can sometimes be obtained with daily massage by the parents or by performing a preliminary lip adhesion. If the lip repairs are staged (two procedures), significant growth of this segment frequently follows the reconstruction of one side of the cleft. The improved blood supply to the area created by the initial reconstruction may produce this growth.

The premaxilla frequently demonstrates a marked variation in size and position. It usually protrudes forward from the maxillary arch and is often deviated. In addition, the cleft maxillary arches are often collapsed, with insufficient space between them for the premaxillary segment. The abnormal position of the premaxilla and maxillary arches in combination with the markedly deficient prolabium frequently results in an extremely difficult

surgical reconstruction of the bilateral cleft lip.

Bilateral clefts of the lip may be repaired in one or two stages. The two-stage method allows closure of the cleft with reduced tension, converts a bilateral cleft into a unilateral cleft, and may promote significant growth of the prolabium between the two stages of repair. The initial repair is usually performed at 3 months of age, and the second repair is performed 2 to 3 months later. Although a staged repair of bilateral cleft lip requires an additional hospitalization and surgical procedure, it may be the most prudent course for the relatively inexperienced cleft surgeon. Any of the basic techniques of surgical repair of unilateral cleft lip may be utilized in staged bilateral cleft repair.

Applying the principle of maintaining or creating bilateral symmetry in these severe deformities, excellent results may be obtained by a single-stage definitive repair of the bilateral cleft lip. The technique is best performed by an experienced cleft surgeon when the bilateral deformity is relatively mild or following a bilateral adhesion (Seibert, 1983). It provides lip reconstruction in a single procedure, eliminating the need for an additional operation and hospitalization.

Surgical technique

Rotation advancement technique

After a preliminary lip adhesion has been performed, the Millard rotation advancement technique is indicated for the entire spectrum of unilateral lip deformities, from the microform or minimal clefts to wide complete clefts. Although a few measurements are made (described later in this chapter), this is basically a "cut as you go" technique. Fine adjustments depend on skills that can be gained only from experience. Because the goal is to create a lip that is functionally and cosmetically symmetric, it is one of the most difficult challenges in reconstructive surgery. The following is a summary of the principal steps of the technique, with particular emphasis on those features found to be most helpful.

As for the lip adhesion, definitive cheiloplasty is performed with the patient under general inhalation anesthesia via an endotracheal tube that is secured to the midline of the lower lip and chin without distorting the lip. The surgeon sits at the patient's head, and loupe magnification is used. Lip landmarks are marked out with gentian violet (Fig. 66-17). Point 1 represents the foot of the nasal ala at the lip on the normal side. Point 3 is the lowest point of Cupid's bow. Point 4 is the high point of Cupid's bow on the medial side of the cleft. This point may be determined by marking off a distance equal to the distance between points 2 and 3. Point 5 represents the peak of Cupid's bow on the lateral side of the cleft, usually at the point where the white roll begins to attenuate on the lateral lip. Point 6 is the upper end of the advancement flap and is not determined until the length of the rotation incision from points 4 to 8 has been placed. Point 7 is located along the alar crease so that the distance between points 5 and 7 equals the distance between points 1 and 2. This determines the height of the lip on the cleft side. Point 8 is the lateral limit of the rotation incision, which ascends the vermilion cutaneous junction to swing across the lip where columella meets lip. It should not extend into the normal philtral column. Point 9 is at the end of the back cut of the rotation incision. In most cases the back cut is necessary to achieve sufficient downward rotation of the medial flap.

Incisions. Skin incisions are made with a 6300 Beaver blade. The incisions are scored first, and the cleft edge mucosal flaps are elevated. The major flaps may be cut through-and-through with the Beaver blade or with a No. 11 Bard-Parker blade. Next, the rotation incision is made and carried across the base of the columella far enough to allow point 4 to drop down to a position symmetric with point 2, the normal Cupid's bow peak. This usually requires a short back cut through skin and often through muscle. A small, triangular-shaped piece of tissue remains attached to the columella (Millard's "C" flap). It is used later in lengthening the shortened cleft side columella, or it contributes to the medial portion of the nasal sill.

Length of the advancement flap medial edge (points 5 to 6) is not determined finally until adequate downward rotation of point 4 is achieved - that is, when both cleft edges (points 4 to 9 and points 5 to 6) are of equal length. It is permissible to place point 6 slightly higher into the nasal vestibule or to place point 5, 1 to 2 mm more laterally to gain additional length.

Along with the circumalar incision, an incision in the intercartilaginous area of the nasal vestibule down to the piriform aperture frees the nasal ala to advance medially independently of the lip. A triangular white roll flap is cut to match the height of the white roll on the normal side. Incision for the interdigitation of this flap medially just above the vermilion cutaneous junction is not made until satisfactory approximation of the major flaps has been accomplished. The lateral and medial lip segments are freed by sharp dissection from the underlying maxilla in a supraperiosteal plane. Only enough dissection to allow approximation of the lip segments without undue tension is performed (Fig. 66-18).

Suturing. Cleft edge mucosal flaps may be excised when the alveolar ridge is intact; however, in patients with wider clefts, they may be rotated upward and used to help with closure of the alveolar cleft and anterior floor of the nose. The lateral mucosal flap may also be rotated into the intercartilaginous incision in the nose to avoid a raw surface at this point. The first suture is a 3-0 or 4-0 Vicryl that grasps the lip muscle back along the upper edge of the advancement flap as well as along the depths of the back cut of the rotation incision (F. 66-19). This suture bears a great deal of tension and creates a more normal convex lip contour. It helps lengthen an often deficient vertical lip dimension (points 5 to 7), a difficult problem in patients with more severe unilateral clefts, by facilitating lip advancement medially. The orbicularis muscles are then approximated with 4-0 Vicryl sutures, with care taken not to shorten the lip. The "C" flap may be advanced on itself to lengthen the columella. This is done by placing a single-prong skin hook in the midline of the columella, retracting the "C" flap superiorly so it folds on itself. Sutures of 7-0 nylon on a P-6 needle are placed. No trimming of the "C" flap is done until it is ascertained that this tissue will not be needed for nasal sill reconstruction. The nasal ala is then sutured medially with a deep 3-0 or 4-0 Vicryl suture. Little or no trimming of the alar foot should be done. At this point, the rotation advancement flaps should be snugly together, with the white roll flap overlapping the medial side of the cleft. If there is a tendency toward peaking of the cleft side Cupid's bow because of either inadequate rotation or inadequate length of the edge of the advancement flap, the repair is taken down and adjustments are made. Finally, skin edges are trimmed and slightly undermined. The skin is approximated with interrupted 7-0 nylon sutures, and the skin edges are slightly everted. The white roll flap is then inserted along the medial side of the cleft either in an incision, which creates slightly more rotation, or after excision of an area

of skin of the same size and dimensions as the white roll flap. At the vermilion, the lip should be snugly together with deep sutures of 4-0 or 5-0 Vicryl. Cutaneous sutures of 7-0 nylon are placed, beginning at the vermilion cutaneous junction and advancing toward the mucosa (Fig. 66-20). Any tendency toward a notch or "whistle deformity" along the free border of the lip can be corrected by careful suturing or transposition of a mucosal flap from the fuller side to the deficient side to balance the lip. A Z-plasty of mucosa may also be performed posteriorly to break up a straight-line closure and to minimize any tendency for contracture of the lip with healing.

It cannot be overemphasized that the primary goal of cheiloplasty is to create symmetric lip contour in all three dimensions. The form achieved on the operating table will be the final result after healing and scar maturation. Slight discrepancies will not improve with time. In some patients who are hypertrophic scar formers, contracture and shortening may occur at the time of initial healing. With scar maturation, the lip assumes the configuration achieved during the operation.

Triangular flap techniques

Once the infant has been prepared and draped, important anatomic landmarks can be determined and marked with gentian violet, which is applied with a cotton-tipped applicator stick broken to create a very fine point. These landmarks (Fig. 66-21) include the high point of the Cupid's bow on the noncleft side, the base of the columella on the cleft and noncleft sides, and the low or middle point of the central portion of the lip.

If the single triangular flap technique is utilized, an equilateral triangular flap is created on the lateral cleft segment, with the sides of the triangle equal to the previously measured difference in the lip length plus 2 mm (Fig. 66.2). The initial portion of this triangle is drawn, starting at the high point of the Cupid's bow on the lateral segment and continuing the predetermined distance upward on the vermilion cutaneous junction. After this, an equilateral triangle is constructed (Fig. 66-22). This triangle is inserted into a defect created by an equally long incision drawn perpendicular to the vermilion cutaneous junction from the high point of the Cupid's bow on the medial segment of the cleft.

If the two-triangle, or Bardach, technique is used, the previously measured difference in lip length of the cleft and noncleft sides plus 2 mm is divided in half, and then an equilateral triangle is constructed in the lower portion of the lip, similar to that previously described. The upper equilateral triangular flap is based on the medial cleft segment. This incision is started at the base of the columella and meets the vermilion cutaneous junction at the previously determined length necessary for construction of a side for the triangle. A superiorly based equilateral triangle is then constructed based on this line, which is inserted into a defect created by an incision of equal length located in the crease of the nasal sill on the lateral segment. Fig 66-23 illustrates these incisions. When the cleft lip is associated with a cleft palate, the incisions along the vermilion cutaneous junction are extended into the nose to create flaps that are used to reconstruct the nasal floor. On the medial segment this incision is carried along the base of the septum, so that a flap can be created by upward elevation. The lateral incision is also carried posteriorly beneath the inferior turbinate so that a similar flap may be developed. These flaps are then sutured together to close the floor of the nose.

Once the drawing of the repair is complete, the incisions are performed. The incision along the vermilion cutaneous junction of the lateral segment extends completely through the lip tissue at a right angle to the surface incised, creating a laterally based vermilion flap, which may be used to augment the vermilion of the medial segment of the lip. On the medial segment of the lip, the vermilion cutaneous junction incision extends to (but not through) the mucosal layer. This creates a medially based flap of vermilion on the mucosal surface of the lip to provide additional mucosal lining for lip reconstruction.

The triangular skin flaps are created by incisions through the skin, leaving the underlying muscle intact. Similar flaps are created in the mucosa opposite the skin incisions. Following this, a blunt and sharp dissection is made to elevate the overlying skin and underlying mucosa from the abnormally directed cleft musculature for approximately 1 cm from the cleft margin along the entire length of the lip (Fig. 66-24). The upper portion of the abnormally directed muscle fibers are transected from the base of the columella medially and the ana nasi laterally (Fig. 66-25). This creates two flaps of muscle, which may then be rotated downward to reconstruct the orbicular mouth muscle completely.

When a cleft of the alveolus and palate is associated with a cleft lip, the previously described flaps from the septum and lateral nasal wall are elevated and closed with interrupted No. 4-0 chromic sutures. After this the muscular flaps from the margins of the cleft are rotated downward and the orbicular mouth muscle is reconstructed by suturing the ends of the muscular flaps together with two or three No. 4-0 Vicryl or chromic sutures. These muscular flaps must be adequately separated from the underlying and overlying mucosa and skin to allow adequate length of the reconstructed lip (Fig. 66-26). The skin flaps are then transposed and sutured in place with No. 7-0 synthetic sutures. The vermilion cutaneous junction of both segments is accurately approximate. These are the high points of the Cupid's bow - which were permanently marked with a 25-gauge needle, dipped in gentian violet, and inserted through these points before the initiation of any incision for surgical repair - to provide accurate approximation. This repair is illustrated in Figs. 66-27 and 66-28.

Once the lip length is determined to be satisfactory, the underlying mucosa is closed with interrupted No. 5-0 chromic sutures. Following any necessary modification, the inner layer of the vermilion is also reconstructed with the same suture; No. 6-0 synthetic sutures are used on the external surface.

Both the triangular flap and rotation advancement methods of lip repair may produce satisfactory results. The selection of the specific surgical method depends on the severity of the cleft and the experience and ability of the surgeon.

One-stage bilateral cleft lip repair

Figs. 66-29 to 66-34 show the technique for the one-stage bilateral cleft lip repair. This technique, described by Millard in *Cleft Craft*, Vol. III, *Bilateral and Rare Deformities*, has a number of advantages: symmetric lip and nasal reconstruction with orbicularis oris muscle continuity, mucosal-lined labial sulcus, symmetric Cupid's bow, midline lip tubercle, occasionally a philtral dimple, and flaps saved for a columella lengthening at a second stage. Fig. 66-29 shows a typical bilateral complete symmetric cleft lip with landmarks and incisions indicated. The height of the lip is determined by the vertical height of the prolabium as

indicated by incisions 1 to 3 and 2 to 4. The philtral flap (A) should be made as narrow as feasible because of its tendency to stretch and widen with healing. The small quadrilateral shaded flap (e) just below the vermilion cutaneous junction may be used to reinforce a midline vermilion tubercle. Both lateral lip elements are handled in the same way with the cleft edge incisions 6 to 10 and 7 to 11 matching the prolabial height. The small flaps (a and b) of length 10 to 12 and 11 to 13 consist of vermilion/mucosa and submucosa and will be brought beneath the prolabial flap to reconstruct the midline vermilion. Lip and nose are freed by circumalar incisions (6 to 8 and 7 to 9), and the lip is freed from the maxilla as is necessary to allow medial advancement without tension. The nasal ala may be freed by an incision in the intercartilaginous area of the nose for medial advancement. The forked flaps (c and d) are full-thickness flaps that will be saved and banked later in the floor of the nose. Fig. 66-30 shows that all incisions have been made. The vermilion flap (e) remains attached to the infero/posterior aspect of the prolabial flap (A), which has been thinned posteriorly. Note that incisions 10 to 12 and 11 to 13 are placed just at the vermilion cutaneous junction or at most 0.5 to 1 mm above it. Including more skin in this flap results in an often conspicuous scar across the philtrum above the vermilion cutaneous junction, especially with the growth of the lip. Fig. 66-31 shows the prolabial flap retracted superiorly. The tips of the nasal ala have been brought medially and sutured to periosteum in the region of the anterior nasal spine. In order to achieve the best symmetry, suturing each flap separately may be required. Excess vermilion from the prolabium is trimmed and sutured onto the anterior surface of the premaxilla to provide labial sulcus lining. Fig. 66-32 shows that the superior portions of the lateral lip flaps have been secured medially in the same fashion as the alar flaps. Again, separate suturing may be required for the best symmetry. Orbicularis oris muscles are then approximated (Fig. 66-33). Skin edges are trimmed and adjusted as necessary, and a philtral dimpling stitch is placed from the undersurface of the prolabial flap to the reconstructed muscle. Final suturing with subcutaneous and skin stitches is applied (Fig. 66-34). When there is a tendency for vermilion deficiency (whistle deformity) in the midline, the flap (e) attached to the prolabium may be tucked behind the vermilion flaps (a and b) to help create a midline tubercle.

Postoperative care

A tongue stitch is applied to help control the airway until the patient is fully awake. The patient is kept in arm restraints, and feedings via an Asepto syringe are begun the day after surgery. The patient is discharged when taking feedings well and when the parents are comfortable with feeding technique and the care of the incision. Antibiotic ointment is applied to the sutures twice per day. Sutures are removed at 5 or 6 days on an outpatient basis with the patient under sedation. Arm restraints and specialized feedings are continued for 10 days after operation.

Cleft Palate Repair

The basic goals of palate repair include separation of the nasal and oral cavities, construction of a watertight and airtight velopharyngeal valve, preservation of facial growth, and development of aesthetic dentition and functional occlusion. An overview of the timing and techniques of palate repair is outlined as follows:

1. Timing - controversial

A. Speech: early

B. Dental: late

C. Common age for procedure

1. Soft palate only: 6 to 8 months

2. Hard palate only: much variation

a. Repair performed before 1 year

(1) Increasingly popular

(2) No long-term studies to show improved speech versus possible effect on facial growth

b. Most repairs performed at 12 to 24 months

(1) Number and occlusion of teeth

(2) Width and extent of defect

II. Techniques

A. Primary veloplasty (Schweckendiek, 1978)

1. Soft palate closure at early age (6 to 12 months)

2. Delayed hard palate closure (several years later than soft palate closure)

3. Currently appears to be rarely indicated

a. Severe speech problems

b. Requires additional procedure, technically difficult

B. Von Langenbeck's palatoplasty

1. Bipedicle mucoperiosteal flaps

2. Effective only for soft palate clefts

C. Oxford palatoplasty (V to Y push-back) (Kilner, 1937; Wardill, 1937)

1. Posteriorly and anteriorly based unipedicle mucoperiosteal flaps

2. V-Y retrodisplacement advantages

- a. Lengthens palate
- b. Better speech than with Von Langenbeck's palatoplasty
- 3. V-Y retrodisplacement disadvantages
 - a. Deficient mucosal coverage of nasal surface, leading to scar contracture and shortened palate
 - b. Failure to close alveolar portion of cleft
 - c. Fistulas occurs at hard/soft palate junction
- D. Two-flap technique
 - 1. Only for posteriorly based unipedicle flaps
 - 2. Advantage: closes alveolar cleft
 - 3. Disadvantage: no palatal lengthening
- E. Reconstruction of levator musculature possibly most important factor in final result; however, not supported by literature (Marsh et al, 1989; Jarvis and Trier, 1988).

The timing of surgical closure of the palate is controversial; currently the optimal age for this procedure has not been settled. In considering the optimal age one must balance the desirability of creating an adequate mechanism for normal speech production with the possible effect of the surgical procedure on facial growth and dental occlusion. These factors appear to be in complete opposition to each other. An adequate velopharyngeal valve is necessary for normal speech production. Studies tend to indicate that the earlier the palate is repaired and velopharyngeal competence established, the better the resultant speech will be (Grabb, 1971). Conversely, early surgical repair may increase abnormalities in facial growth, increase maxillary arch collapse, and exacerbate orthodontic problems. Palatoplasties performed before deciduous molars are in occlusion have been shown to produce a greater alteration in facial growth than those performed at an older age (Bernstein, 1968).

Palate repair is most commonly performed on children between 12 and 24 months of age (Grabb, 1971). However, the extent and width of the palatal cleft, the alignment of the maxillary arches, and the number and occlusion of deciduous teeth frequently play vital roles in determining the actual timing of palate repair. Clefts involving only the soft palate are frequently closed at 6 to 18 months of age. A marked variation in the age of patients exists with closure of complete clefts of the palate. In some centers repair is commonly performed before 12 months of age; other centers may delay closure until 24 to 30 months of age, especially in wider complete clefts.

Currently, nearly all cleft palates are successfully repaired with various surgical techniques. This includes even massive clefts, in which the size of the cleft is larger than the size of the remaining palatal tissue (Bumsted, 1982). However, equally effective therapeutic

results may be obtained by dental obturation of the palatal defect without the associated trauma and morbidity of a surgical procedure and its possible subsequent secondary effects on facial growth. Dental obturation of palatal clefts cannot be performed as early in life as palatoplasty because of the necessity for patient cooperation and adequate dentition to secure the appliance. At the age of 3 to 4 years, obturation is usually highly successful.

Palatoplasty techniques

The basic surgical techniques of palate repair include Schweckendiek's procedure or primary veloplasty (early closure of the soft palate and delayed closure of the hard palate), Von Langenbeck's palatoplasty, the V-Y push-back palatoplasty (Kilner, 1937; Wardill, 1937), reversing Z-plasty (Furlow, 1986), and the two-flap palatoplasty. In addition, a combination of palatoplasty and pharyngeal flap may be used for the repair of extremely large clefts (Bumsted, 1982) and occasionally as a method of primary palate repair (Stark and DeHann, 1960). The following sections describe these procedures in detail.

Basic considerations in the repair of any cleft palate must include the previously described pathologic anatomy (Fig. 66-9). Although the following methods of palate repair do not emphasize the reconstruction of the muscular sling of the palate, such reconstruction appears to play a significant role in subsequent palatal and eustachian tube function. The technique of reconstruction of muscular sling (Fig. 66-35) requires dissection of the oral and nasal mucosa from the remaining hard palate. This muscular insertion is then transected, and each muscle bundle is rotated to the midline and sutured together, thereby reconstructing the muscular sling (Fig. 66-36).

Schweckendiek's technique

Schweckendiek's procedure (primary veloplasty consists of closure of only the soft palate cleft (Fig. 66-37). This initial repair is usually performed at a relatively early age (from 3 to 12 months), with delayed closure of the hard palate at a later date. A dental prosthesis is usually necessary to close the remaining defect in the hard palate until the secondary procedure is performed (usually at 4 to 5 years of age). Advantages of this technique include the construction of a velopharyngeal valve at an early age, minimal disturbance in future facial growth, and the frequent occurrence of significant narrowing of the width of the remaining palatal cleft with increasing age. However, disadvantages include the necessity of an additional operative procedure and hospitalization, the requirement for a dental prosthesis that must be changed frequently (time and expense), and significant speech disorders that are hard to treat with speech therapy (University of Iowa Cleft Palate Team, 1962). In addition, no significant improvement in eustachian tube function was detected clinically in patients with primary veloplasty compared to those without surgical closure of the soft palate defect (Bumsted et al, 1984). Currently it appears that this technique is rarely indicated.

Von Langenbeck's palatoplasty

Von Langenbeck's palatoplasty (1861) utilizes bipedicle mucoperiosteal flaps of the hard and soft palate for repair of the defect (Fig. 66-38). These anteriorly and posteriorly based bipedicle flaps are advanced medially to close the palatal cleft. The advantage of this method is that it is simply performed, requires less dissection, and results in decreased

denuded palatal bone. The disadvantages include failure to provide any additional palatal length, inability to close complete clefts of the primary and secondary palate, and poorer speech results in more extensive palatal clefts (Krause et al, 1975). Currently this technique is used infrequently, primarily for clefts involving only the soft palate.

V-Y push-back

The Oxford method (V-Y push-back) involves the creation of two posteriorly based unipedicle flaps with a single or double (depending on the extent of the cleft), anteriorly based, unipedicle, palatal flaps (Fig. 66-39). The anterior flaps, if present, are advanced or rotated medially, and the posterior flaps are retrodisplaced with a V-to-Y technique, increasing the length of the reconstructed palate. Advantages of this technique include lengthening of the reconstructed palate, avoidance of a straight-line closure (presumably decreasing both subsequent scar contracture and shortening of the reconstructed palate), and improved speech results in more extensive palatal clefts than that obtained by Von Langenbeck's method. Disadvantages include failure to provide mucosal coverage of the retrodisplaced palate's nasal surface, which reduces the palatal lengthening obtained in the final result secondary to scar contracture of the raw nasal surface, and the difficulty of closing the cleft's alveolar portion. Also, fistulas tend to occur in the thin mucoperiosteal membrane near the hard/soft palate junction. Currently this technique appears to be the best procedure for most incomplete clefts of the palate; however, it is used infrequently in clefts extending into the maxillary alveolus.

Two-flap palatoplasty

The two-flap palatoplasty involves the creation of two posteriorly unipedicle-based flaps that extend all the way to the alveolar portion of the cleft (Fig. 66-40). These flaps are then rotated and advanced medially to reconstruct the defect. This method allows closure of the alveolar cleft but does not lengthen the palate unless a Z-plasty is used in the soft palate. It is used primarily in complete clefts of the palate and alveolus.

Palatoplasty and pharyngeal flap combination

The combination of a palatoplasty and pharyngeal flap is rarely used as a method of primary palate closure. This technique may be indicated in patients with extremely wide palatal clefts; however, it would appear that it should be performed at an older age rather than at the time of customary palate repair. The main disadvantage of this combined technique for primary repair is the inability to identify the patients who fail to develop adequate velopharyngeal closure after palatoplasty and who would therefore require a pharyngeal flap as a secondary procedure. The use of this technique thus subjects the majority of patients to an unnecessary additional procedure at the time of palatoplasty, with a resultant increase in morbidity, especially postoperative upper airway obstruction, and blood loss. In the repair of extremely wide palatal clefts at an older age, this combined technique appears to offer an advantage by providing a complete two-layer closure of the defect.

Surgical technique

At the time of palatoplasty the patient is positioned with the head in the seated surgeon's lap, with exposure of the palate obtained by a self-retaining mouth gag. A

hypopharyngeal pack is placed to prevent aspiration of blood during the procedure. Decreased blood loss and easier elevation of the mucoperiosteal flaps overlying the remaining hard palate can be obtained by the injection of 1% lidocaine (Xylocaine) with 1:100,000 epinephrine before creation of the palatal flaps.

Two-flap palatoplasty. The two-flap palatoplasty is a technique in which a single mucoperiosteal flap is elevated from each palatal shelf. The flap is based posteriorly on the descending palatine artery. This technique allows excellent exposure for dissection and levator sling reconstruction. The incidence of fistulization is extremely low. A disadvantage of the technique is that if a neurovascular bundle is transected, part of the distal flap may be lost.

The procedure is performed with the patient under general inhalation anesthesia via an endotracheal tube taped to the midline. A towel roll is placed under the patient's shoulders. The Dingman self-retaining mouth gag is used for exposure of the oral cavity and pharynx as well as for holding the endotracheal tube in place. The mouth gag is not suspended from a Mayo stand; however, a folded towel is often placed beneath the handle of the gag.

Incisions. The cleft margins, adjacent hard palate, and retromolar areas are infiltrated with 1% lidocaine and epinephrine 1:100,000. The cleft margins are incised with a #30 Beaver or No. 11 Bard-Parker blade, and the relaxing incisions are made laterally with a No. 15 Bard-Parker blade (Fig. 66-41). At the hard palate cleft the incision is made 2 to 3 mm lateral to the cleft edge. To avoid injury to a developing tooth, care is taken not to bring the relaxing incisions too far laterally over the alveolar ridge. The relaxing incisions are carried posteriorly just into the medial retromolar area and just through the mucosa at this point.

Mucoperiosteal flaps are then elevated from the hard palate using a Freer elevator (Fig. 66-42). This elevation is quite easy to accomplish once the subperiosteal plane is entered against bare bone. The neurovascular bundle coming from the greater palatine foramen is identified and preserved. On the noncleft side, mucoperiosteum is elevated from the vomer as well. This vomer flap is kept continuous posteriorly with the nasal mucosa layer of the soft palate. Care is taken in dissecting around the vomeropremaxillary suture. Velar muscle fibers turn anteriorly and insert along the medial and posterior edge of the hard palate (Veau's muscle). It is necessary to dissect these fibers sharply from bone using a dental type of elevators. Muscle fibers are dissected from the soft palate nasal mucosa for 1 to 1.5 cm. The medial edges of the soft palate are prepared for suturing by freeing mucosa from muscle for 2 mm all around.

Medial mobility of the soft palate is enhanced by blunt dissection through the posterior extension of the relaxing incision in the plane between the pterygoid and superior constrictor muscles. This plane is almost avascular, and blunt dissection is performed with little bleeding. In patients with wider clefts, it may be necessary to infracture the hamulus processus medially and sometimes to strip the levator veli palatini muscle tendon from the hamulus. Mucoperiosteum is tightly bound down to the maxillary palatine suture and must be freed up from the suture to provide sufficient medial mobility of the flaps in most clefts. Finally, nasal mucoperiosteum is elevated from the nasal surface of the hard palate on the cleft side using right-angle dental type of elevators.

Suturing. The palate is closed in three layers. Fig. 66-43 shows the closure of the

nasal layer by a vomer mucoperiosteal flap to a palatal mucoperiosteal flap and velar mucosa posteriorly. This closure is performed with a simple No. 4-0 or 5-0 Vicryl suture with the knots placed on the nasal side. In patients with very wide clefts, it may be impossible to approximate the nasal layer at the hard/soft palate junction. This closure may be facilitated by performing a Z-plasty of the nasal mucosa at this point or by simply leaving a small gap for secondary healing. Next, the palatal muscles are approximated with care with multiple buried horizontal sutures of No. 3-0 or 4-0 Vicryl so that the levator muscle sling can be reconstructed. This closure brings oral mucosa into close approximation. The mucoperiosteal flaps are sutured anteriorly with buried No. 4-0 Vicryl through the periosteum, which tends to evert the edges. The oral layer may be tacked down to the nasal layer to decrease dead space. The oral mucosa is approximated with simple everting sutures of No. 4-0 or 5-0 chromic catgut. Vertical mattress sutures may be placed if there is any gaping of the muscle. Two or three sutures are also placed from the tips of the flaps anteriorly to the alveolar ridge mucosa (Fig. 66-44). The relaxing incisions are then packed with microfibrillar collagen (Avitene). Hemostasis is ensured by electrocautery and pressure.

Complications

The most common complication of palatoplasty is hypernasal speech, which may occur in up to 30% of cleft palate patients (Grabb, 1971). The actual incidence of hypernasal speech varies with the severity of the cleft, the type of repair performed, the patient's age at repair, and the variation in the definition of adequate speech or velopharyngeal competence used in reporting the results of various cleft palate procedures. However, in general, the more severe the cleft defect, the poorer the speech result obtained.

Fistula formation has previously been reported to occur in up to 21% of patients after palatoplasty, but currently this incidence should be much lower: 10%. These fistulas are most commonly located anteriorly on the lingual surface of the maxillary alveolus and posteriorly at the junction of the hard and soft palates. Hemorrhage and excessive blood loss are uncommon, averaging 60 to 140 mL (Dingman et al, 1949; Tempest, 1958).

Airway impairment may occur intraoperatively and postoperatively. Bleeding and oral secretions are its most common intraoperative causes, and edema is its most common postoperative cause. An endotracheal tube and a hypopharyngeal pack protect against airway obstruction during the operation. Allowing the patient to awaken with the endotracheal tube in place is beneficial in preventing obstruction in the immediate postoperative period. Humidification in a mist tent and adequate suturing of the oral to the nasal layers of closure during the surgical repair diminish postoperative airway obstruction. Mortality in palatoplasties is less than 0.5% and is usually associated with inadvertent extubation, aspiration, pneumonia, and congenital heart disease.

Cleft Nose Repair

Most patients with a cleft lip have an associated nasal deformity with marked variation in its severity. In general, the cleft nose deformity becomes more apparent with increasing age. The unilateral cleft lip nose has abnormalities of the columella, tip, ala, and internal nose (Huffman and Lierle, 1949, Figs. 66-45 and 66-46). The columella is shortened on the side of the cleft, with its base and underlying anterior nasal spine directed into the noncleft side.

The nasal tip is deflected toward the side of the cleft by displacement of the medial crus of the lower lateral cartilage of the cleft side in an inferior or downward direction. In addition, the angle between the medial crus and the lateral crus of the lower nasal cartilage on the side of the cleft is less acute than normal, with the lateral crus displaced downward, causing hooding of the nostril and buckled inward on itself (producing an S-shaped fold of the lateral crus), and is frequently longer than the noncleft side. The frequent underdevelopment of the maxilla on the cleft side displaces the alar base laterally, posteriorly, and down - producing a widened naris on the cleft side with its usual orientation horizontal rather than vertical.

Internally, the caudal or anterior end of the cartilaginous septum is frequently deviated into the noncleft side, with an associated inferior septal deviation into the same side and the upper portion of the septum deviated into the cleft side. A bowstring contracture of the nostril from its apex continues along the upper border of the lateral crus to the piriform aperture on the side of the cleft, producing a relative stenosis of the internal nares. In addition, the inferior turbinate is often hypertrophied on the side of the cleft, and many patients have an inadequate nasal airway (Bumsted, 1981).

The bilateral cleft lip nose has deformities of the columella, ala, alar base, and internal nares similar to those described for the unilateral cleft; they are present bilaterally. This frequently results in an extremely short columella, a broad, flattened nasal tip, and horizontal orientation of the nares. The columella, caudal septum, and anterior nasal spine are frequently displaced forward and/or downward (Fig. 66-47). Nasal deformities associated with cleft lip are outlined as follows:

1. Columella
 - a. Shortened on cleft side
 - b. Base directed to noncleft side
2. Anterior nasal spine deviated to noncleft side
3. Nasal tip; lower lateral cartilage on cleft side
 - a. Medial crus displaced medially and downward
 - b. Angle between medial and lateral crus widened
 - c. Lateral crus displaced downward and has S-shaped fold
4. Maxilla underdeveloped
 - a. Displaces alar base laterally, posteriorly, and downward
 - b. Results in horizontal orientation of nares
5. Internal nose

- a. Caudal septal deviation to noncleft side
- b. Upper septum deviation to cleft side
- c. Contracture along upper border of lower lateral cartilage; relative stenosis on cleft side
- d. Inferior turbinate hypertrophy on cleft side
6. External nose often deviated toward cleft
7. Skin often thick
8. Bilateral cleft, similar (but bilateral) deformity of columella, ala, alar base, and internal nares.

The goals of surgical reconstruction of the cleft nasal deformity includes reconstruction of the nasal floor and sill, columellar lengthening, correction of the deformed ala, repositioning of the alar base, provision of adequate nasal tip support and symmetry, straightening of the nasal dorsum (if necessary), and obtaining an adequate intranasal airway. Often achievement of these goals requires one or more operations performed at different ages.

The previously described techniques of lip repair reconstruct the nasal floor and sill at the time of the primary lip repair. In addition, the base of the ala may be placed in a more symmetric position and the columella lengthened (in the Millar repair). Some surgeons also perform a partial correction of the nasal tip and alar cartilage during the primary lip repair. This provides an immediate result; however, long-term effects on facial and nasal growth are uncertain at this time.

Reconstruction of moderate to severe deformities of the nasal tip is frequently performed at 6 to 10 years of age, utilizing a tip rhinoplasty with insertion of an implant or graft under the alar base on the side of the cleft (if there is maxillary deficiency). A deficient maxillary platform may also be improved when an alveolar ridge bone graft is performed, the other indications being to provide bony support for erupting permanent teeth and to close fistulas. Minor deformities of the nasal tip, intranasal septum, and nasal bones are usually repaired at 14 to 18 years of age. This delay may allow the use of local anesthesia and minimizes possible disturbances of facial growth.

Available surgical techniques

Numerous authors have developed a multitude of surgical techniques for the correction of the cleft nasal deformity. Since Millard (1976) has already presented these thoroughly, they are not discussed in detail here. Standard intranasal rhinoplasty approaches for correction of the cleft nasal deformity are usually inadequate, often producing less than the desired results. Basic external approach appears to provide the best results. Basic external techniques for correcting the cleft nasal deformity include the approach described by Dibbell (1982) and Bardach (1967) (Fig. 66-48).

Dibbell technique

The surgical technique developed by Dibbell involves extensive surgical incisions of the nasal ala, the lateral alar margin, and the lip. The horizontally directed cleft ala and nares are rotated into a vertical orientation, excessive skin at the margin of the cleft ala is excised, the alar base is rotated and advanced into a correct anatomic location (requiring advancement of the lateral portion of the lip to correct the resultant deformity), and the misplaced and deformed lateral crus of the alar cartilage is inserted into proper position maintained by the use of sutures. The advantage of this technique is the ability to correct most of the cleft nose deformities. In addition, Dibbell has achieved excellent cosmetic results. However, this technique involves an extensive surgical procedure, and obtaining these results appears to require significant surgical ability and judgment. Although my experience with this technique is limited, it appears to provide a significant advancement in the correction of this difficult nasal deformity.

Bardach technique

The external approach described by Bardach (1967) is illustrated in Figs. 66-49 to 66-51. This method of correction lengthens the columella by the use of a triangular skin flap from the middle portion of the upper lip in both unilateral and bilateral cleft nasal deformities. A V-Y shaped advancement technique is used, with the apex of the V-shaped skin flap located beneath the middle of the columella in the bilateral cleft and at the lateral margin of the shortened side of the columella in the unilateral cleft. This incision in the upper lip skin is extended superiorly at the base of the columella on the lateral surface of the columella just caudal to the caudal margin of the medial crus. Superiorly the incision continues laterally as a marginal incision along the caudal margin of the lower lateral cartilage. Subcutaneous dissection and elevation of the lip and columellar skin as a superiorly based flap provide excellent exposure of the nasal tip. The lateral crura of the lower lateral cartilages are dissected from the underlying vestibular skin and trimmed so that they are symmetric, and the lateral crus of the cleft side is advanced medially and partially incorporated into the shortened medial crus on this side. This lengthens the medial crus. Tip support is obtained by suturing the apices of the medial crura together to create appropriate tip projection. If hooding of the alar margin on the side of the cleft is still present, further elevation of the lateral crus may be obtained by suturing it to the upper lateral cartilage. A V-Y closure of the skin of the upper lip provides elongation of the columellar skin and narrows the base of the ala. The malpositioned ala may also be corrected at this time by a V-Y advancement technique. If the maxilla on the side of the cleft is deficient, the alar base may be elevated by implanting irradiated cartilage or an iliac crest bone graft beneath the repositioned base. This procedure is often performed when the patient is between 6 and 10 years of age. A result obtained with this technique is demonstrated in Fig. 66-52.

The correction of the septum, inferior turbinate, and bony external nose is usually performed when the patient is 14 to 18 years of age, utilizing conventional septorhinoplasty techniques.

In summary, the correction of the external nasal deformity associated with cleft lip is frequently difficult. The timing, selection of surgical technique, and extent of repair usually depend on the severity of the deformity and the experience and ability of the surgeon. In

general, it appears that the Bardach approach is the easier technique to perform and master, whereas the Dibbell method may provide superior results but is difficult and requires significant experience in the surgical correction of the cleft nasal deformity.

Speech Disorders in Cleft Palate

The reported incidence of speech disorders in cleft patients varies widely in the literature; however, some abnormality of speech exists in most cleft palate patients. This marked variation in incidence appears to be related primarily to the definition of acceptable or normal speech that is used in evaluating these patients. The phrase *typical cleft palate speech* is sometimes criticized when it is used to describe the speech of patients who have a cleft palate, but these patients do tend to make certain types of articulation errors more frequently than do noncleft patients with speech disorders (Spriesterbach and Morris, 1977). Errors involving the affricatives and fricatives are the most common abnormalities of speech in patients with cleft palate (Spriesterbach et al, 1961). Other articulation errors heard frequently include stop, glides, and nasal semivowels.

Velopharyngeal incompetence is associated with an audible escape of air from the nose during the production of the pressure sounds and is termed *nasal emission* or *snort*. In an extensive review of the literature Morris (1973) reported an overall success rate of 75% in obtaining velopharyngeal competence following primary cleft palate surgery. Assuming adequate follow-up, 90% to 95% of cleft palate patients are eventually provided with the physical structure necessary to obtain velopharyngeal competence after primary and secondary procedures directed to this disorder. Adequate velopharyngeal function is the most important determinant of articulation performance and listener understanding of speech in cleft palate patients (Morris, 1973). Other factors influencing speech in cleft patients include dentition, the presence and severity of associated hearing loss, and muscular and neurologic defects. In addition, the abnormal compensatory patterns of speech production learned by cleft patients before closure of the palatal defect or obtaining velopharyngeal competence also appear to play a significant role in the speech obtained after secondary surgical procedures. These previously learned incorrect speech patterns usually continue, and speech therapy is needed to obtain normal speech.

A summary of diagnostic tests for the evaluation of speech disorders in cleft patients is presented in the following outline:

- I. Speech pathology
 - A. Articulation tests
 - 1. Isolated speech
 - 2. Conversational speech
 - B. Response to speech therapy
- II. Radiography

- A. Lateral radiographs of head (palate at rest and phonation)
- B. Cinefluorography during speech
- III. Manometry - maximal pressure with open and occluded nares
- IV. Clinical examination
 - A. Nasopharyngeal depth
 - B. Palatal length and mobility
 - C. Facial grimace
- V. Fiberoptic nasal endoscopy - palatal and pharyngeal wall motion.

The vital role of experienced speech pathologists in the diagnosis and therapy of speech disorders in cleft patients cannot be overemphasized. In addition to their expertise in diagnosis, their role in therapy is of the utmost importance. Tests performed by speech pathologists include articulation tests, which are performed both in isolated and connected speech, and evaluations of the patient's response to standard speech therapy. Production is an easier task with isolated words than in running conversation. This fact may result in a marked difference in articulation proficiency between isolated responses and connected speech. In addition to the results of the specific articulation tests, the observation of the patient's articulation in conversational speech provides an evaluation of the speaker's intelligibility level and the degree of the communication handicap. Additional clinical features of velopharyngeal incompetence include facial grimaces associated with the production of pressure sounds, audible nasal emission of air, and improvement in articulation when the nostrils are manually occluded. In some patients the response to speech therapy may also provide valuable diagnostic information.

Radiographic evaluation of patients with speech disorders may include cinefluorography and lateral radiographs of the head and palate performed during speech and at rest. Cinefluorography is best used for evaluation of the degree of palatal movement and velopharyngeal closure produced during conversational speech. Lateral radiographs of the head can be used to determine palatal position and motion during the production of a speech sound, such as a sustained /s/, which requires an essentially complete velopharyngeal closure. Lateral radiography techniques provide information on the degree of palatal elevation and posterior wall movement and the degree of closure obtained in an anteroposterior direction during the production of isolated words rather than the degree of closure actually obtained during conversational speech. The degree of lateral pharyngeal wall motion cannot be determined by this technique; it requires a basilar view, which is more difficult to obtain and evaluate clinically.

Clinical examination of the depth of the nasopharynx and palatal length and an estimation of the degree of palatal mobility may be obtained during phonation. However, it must be remembered that there may be a marked difference between palatal mobility in connected speech and that in phonation of a single sound. Additional information regarding

palatal and pharyngeal wall motion may be obtained by nasopharyngoscopy performed with a flexible fiberoptic endoscope. This technique allows direct visualization of palatal motion, pharyngeal wall motion (including both the lateral and posterior pharyngeal walls), and the degree of any velopharyngeal gap during connected speech (Croft et al, 1978). Manometry may also be used as a general method to compare the maximal pressures obtained with rapid, forced expirations with the nares open and with the nares manually occluded. The ratio of open to occluded pressures allows a gross quantitative measurement of velopharyngeal competence, and a value of 0.5 or less usually indicates significant velopharyngeal incompetence. The greater the degree of incompetence, the lower the ratio.

These tests provide a useful assessment of velopharyngeal competence. A controlled study comparing the results of the Iowa pressure articulation test (Morris et al, 1961) with judgments of velopharyngeal closure obtained on a lateral radiograph during phonation all demonstrated an accuracy greater than 90% in determining which patients would require secondary surgical procedures to obtain velopharyngeal competence. The combination of the lateral radiographs and the results of the articulation tests provided a 96% accurate prediction (Van Demark et al, 1975).

Management techniques

The basic techniques of management of speech disorders in cleft patients are as follows:

1. Speech therapy (with speech pathologist)
2. Dental
 - a. Obturator
 - b. Palatal lift
3. Surgery
 - a. Posterior pharyngeal wall implants
 - b. Pharyngoplasty (for a narrow pharynx)
 - c. Pharyngeal flap.

Each of these techniques is discussed in detail in the following sections.

Speech therapy

Speech therapy is the initial method of choice in all patients with speech disorders. The speech therapist conducts initial parental counseling, preferably before the child is 6 months old. The importance of talking and playing with the child is reinforced to the parents, since they frequently assume that speech stimulation is unnecessary because the infant has an abnormal mechanism for speech production. Formal speech therapy is usually not initiated

before 4 years of age and is used only after the speech pathologist can determine which aspects of the speech abnormality can be improved by therapy. The failure to respond to speech therapy may also play an important role in selecting patients who need secondary surgical procedures for the correction of velopharyngeal incompetence.

In addition to the diagnostic role of the speech therapist, correction of the anatomic defect by dental or surgical techniques frequently requires additional speech therapy after the procedure. Prior speech therapy familiarizes both the patient and parents with this approach and is more readily accepted and effective after the secondary procedure. Once a program of therapy has been established by an experienced speech pathologist, further therapy may often be obtained in the local community or school system.

Failure to obtain significant improvement in velopharyngeal competence following a 6- or 12-month period of programmed speech therapy is usually an indication for consideration of secondary methods of management. Articulation errors may require correction of dental abnormalities, but once dental problems are corrected, the only available method of correction of articulation errors is speech therapy.

The basic goal of secondary procedures for the correction of velopharyngeal incompetence is the development of a clinically competent velopharyngeal sphincter to produce normal speech. This need not be a completely airtight velopharyngeal port because, as Bjork (1961) has demonstrated, speech is usually not affected if the size of the velopharyngeal port during speech production requiring port closure is less than 20 mm². A trained speech pathologist can frequently detect mild velopharyngeal incompetence in patients who have a port size ranging from 10 to 20 mm²; "however, the nontrained listener cannot detect this" (Bjork, 1961).

Dental prostheses

Dental prostheses that may be used for the correction of velopharyngeal incompetence include palatal lifts and obturators. In patients with cleft palate and velopharyngeal incompetence a palatal lift is rarely used. A palatal lift is indicated only for patients with a poorly mobile palate and with adequate palatal length to close the velopharyngeal port on elevation of the palate. In most cleft palate patients a dental obturator is necessary. Dental obturators are prosthetic devices used to occlude the nasopharynx so that the patient may obtain velopharyngeal competence during speech. These obturators are best used as a primary method of management in an unoperated cleft palate, since it is difficult to obtain satisfactory results in patients with prior attempts at surgical repair of the palatal defect.

The use of a dental prosthesis that an experienced and qualified prosthodontist has constructed is frequently a highly effective method of obtaining velopharyngeal competence. Patients for whom use of a dental prosthesis is indicated include those who refuse a surgical procedure, certain patients for whom surgery has been a failure, and high-risk patients for whom a surgical procedure is contraindicated. Advantages of this method are avoidance of a surgical procedure with its associated morbidity and mortality and a success rate equal to or greater than that obtained by surgical therapy. Disadvantages include the necessity of wearing a prosthesis; the needs for modification of the prosthesis as the patient grows and for repairs as it wears out; and the frequent failure of insurance plans to reimburse the patient for

the cost of constructing, modifying, and repairing the prosthesis.

Patients who successfully utilize a dental prosthesis for speech production should be persuaded if at all possible to continue its use since their speech production after surgical correction may be less satisfactory than their presurgical speech. In addition, the construction and successful use of a dental prosthesis may be extremely difficult or impossible following a failed surgical procedure. Thus the patient could have permanently poorer speech following the surgical procedure. This situation must be repeatedly stressed to the patient and relatives before undertaking any attempt at surgical correction.

Surgical correction

Three basic methods of surgical correction of velopharyngeal incompetence are available. They include the insertion of an implant into the posterior pharyngeal wall to project this wall anteriorly, the use of pharyngeal tissue flaps to provide both anterior projection of the posterior pharyngeal wall and narrowing of the valve area of the pharynx (pharyngoplasty), and the use of a pharyngeal flap. They are discussed in the following sections.

Implant insertion. Numerous materials have been implanted into the posterior pharyngeal wall for anterior advancement of the posterior pharynx. Patients who have mobile soft palates and a velopharyngeal gap of 1 cm or less (5 mm or less is preferable) on lateral radiographs during isolated and connected speech obtain the best results either from implants into the posterior pharyngeal wall or from pharyngoplasty procedures. The best implant appears to be Teflon past injected submucosally into the posterior pharyngeal wall; 60% of patients (with the above indications) obtain complete elimination with this implant, and 20% have a significant decrease in hypernasality (Smith and McCabe, 1977). However, the US Food and Drug Administration has currently rescinded approval for using this material as a posterior pharyngeal wall implant for the correction of velopharyngeal incompetence. Although synthetic materials, such as Silastic or Proplast, have been utilized in the management of these patients, the incidence of extrusion, infection, and delayed failure is high enough that their desirability at this time is open to debate.

Pharyngoplasty techniques. The use of various flaps of pharyngeal tissue to narrow the pharynx or produce a forward projection of the posterior pharyngeal wall is known as a pharyngoplasty. These techniques basically involve the transposition of local flaps of posterolateral pharyngeal wall tissue, which are sutured into an incision in the posterior pharyngeal wall. Hynes (1967) created superiorly based flaps consisting of the salpingopharyngeal muscle and a significant portion of the lateral pharyngeal wall musculature. They were transposed and sutured together into a horizontal incision across the posterior pharyngeal wall. In addition to creating an anteriorly projecting ridge in the posterior pharynx, closure of the flap donor sites narrows the entire velopharyngeal port. The pharyngoplasty Orticochea (1970) described attempts to produce a dynamic sphincter in the velopharyngeal port area by use of bilateral superiorly based flaps of the posterior tonsillar pillars (palatopharyngeal muscles), which are sutured into an inferiorly based pharyngeal flap. It was his belief that after a healing period, this procedure would create a diaphragm-like sphincter, which would produce a circular, contractible hole in the center of the velopharyngeal port. This hole could be closed during speech production and swallowing,

thereby providing velopharyngeal competence. He reports excellent results, but no other accurate evaluation of this procedure is currently available in the literature.

In general, flap techniques of pharyngoplasty appear to be useful in only a limited number of cases. They are not the most commonly used procedures for correcting velopharyngeal incompetence, they appear to be indicated for patients in whom an implant in the posterior pharyngeal wall could also be utilized. Thus these patients should have a very small velopharyngeal gap during connected speech with a relatively mobile soft palate. Most other cases obtain better results from a pharyngeal flap.

Pharyngeal flap. A pharyngeal flap is created by elevating mucosa and its underlying muscle from the posterior pharyngeal wall; it is then sutured into the palate. This produces tissue obturation of most of the area of the velopharyngeal valve with the only remaining openings located between the lateral margin of the flap and the lateral pharyngeal wall. Thus during speech production only these lateral port areas must be occluded to obtain velopharyngeal competence (Fig. 66-53). A pharyngeal flap is the most common surgical technique for correcting velopharyngeal incompetence.

Significant improvements in the surgical construction of pharyngeal laps include the creation of as wide a based flap as possible (Webster et al, 1956), mucosal coverage of the raw surface of the flap to minimize flap contracture during healing (Blackfield et al, 1963), control of the surgically constructed lateral pharyngeal port's size (Hogan, 1973), and variable lateral port size based on the degree of lateral pharyngeal wall motion (Johns and Salyer, 1977). Hogan designed an operation (lateral port control) to limit the combined size of both lateral ports to less than 20 mm², which is reported by Bjork (1961) to be the maximal velopharyngeal port opening during clinically normal speech. This was accomplished by suturing the lateral margin of a superiorly based pharyngeal flap to an incision in the lateral pharyngeal wall so that it was snug around a No. 14 French catheter. This provided 97% success in obtaining velopharyngeal competence. However, this high rate of success is also associated with a high incidence of significant nasal airway obstruction.

The use of a variable approach to lateral port size not only produces a high rate of velopharyngeal competence but also reduces the incidence of nasal airway obstruction in patients in whom a larger port can be utilized. Patients with little or no lateral pharyngeal wall motion require lateral ports of less than 20 mm² to obtain satisfactory velopharyngeal competence. A high incidence of nasal airway obstruction results; patients should be informed of this before the procedure. Patients with moderate or large degrees of lateral pharyngeal wall motion can maintain velopharyngeal competence with larger port sizes and have a low incidence of postoperative nasal airway obstruction.

Currently most surgeons prefer a superiorly rather than inferiorly based pharyngeal flap; however, studies have failed to show any significant difference in speech obtained from these flaps (Whitaker et al, 1972). The advantages of the superiorly based flap include the ability to construct a flap of any length, a more normal situation because the flap tends to elevate rather than depress the soft palate, and postoperatively the raw surface is accessible compared to the inaccessibility of an inferiorly based flap if bleeding occurs.

The indications for a prepharyngeal flap adenoidectomy include an extremely large

adenoid mass that may partially occlude the lateral ports or make creation of the pharyngeal flap difficult, chronic eustachian tube dysfunction that has not responded to prior insertion of ventilating tubes, recurrent respiratory infections or chronic nasal drainage associated with clinical evidence of recurrent adenoiditis, and chronic sinus disease in a child who needs a pharyngeal flap. The pharyngeal flap prevents access to the nasopharynx for adenoidectomy, which is usually performed 6 weeks before the flap procedure. Staging of these operations is necessary, since postoperative bleeding from the nasopharynx is inaccessible after the flap is created, and the adenoidectomy may result in diminished vascularity at the base of a superiorly based flap.

Operative technique. A pharyngeal flap is performed on a patient under general anesthesia and in a position similar to that assumed for palatoplasty, with endotracheal intubation and hypopharyngeal pack insertion. The injection of 0.5% lidocaine (Xylocaine) with 1:200,000 epinephrine into the posterior pharyngeal wall and palate decreases operative bleeding. Vertical incisions in the posterior pharyngeal wall should be made just medial to the junction of the lateral and posterior pharyngeal walls to create as wide a flap as possible (Fig. 66-54). These incisions extend through the superior constrictor muscle to the prevertebral fascia, on which blunt and sharp dissection is used to elevate a superiorly based flap. The inferior margin is then transected and flap elevation continued until the base of the flap is level with the plane of the hard palate. Electrocautery is used to control bleeding.

The opening in the palate may be created either by a vertical midline incision that transects the soft palate or by a horizontal incision on the nasal surface of the soft palate that extends into the midportion of the palate (Fig. 66-55). Both techniques use the mucosa of the nasal surface of the palate as a turnover flap based on the posterior margin of the palate to provide mucosal coverage of the raw surface of the superiorly based pharyngeal flap. A pocket is created in the entire width of the remaining palatal musculature into which the pharyngeal flap is inserted and secured with interrupted No. 3-0 chronic vertical mattress sutures.

Before suturing the flap in place, the surgeon obtains hemostasis by electrocautery, and pediatric endotracheal tubes of varying size (3 to 5 mm if a variable port control technique is used) are inserted through the nose into the oropharynx to provide lateral port control. After the suturing of the pharyngeal flap into the entire width of the palate, the surgeon makes an incision in the posterior tonsillar pillar or lateral pharyngeal wall at the level of the lateral margin of the flap. A single absorbable suture between the lateral margin of the flap and the incision in the lateral pharyngeal wall or posterior tonsillar pillar is placed so that the flap is snug around the tube used to control the size of the lateral port. Figs. 66-56 to 66-59 show the lateral port control technique.

The flap donor site in the posterior pharyngeal wall may be left open to heal by secondary intention, approximated with a single absorbable suture, which must be tacked to the prevertebral fascia in the midline of the posterior pharyngeal wall to prevent the formation of dead space - or it may be closely approximated. Although approximation of the remaining pharyngeal tissue over the defect does narrow the pharynx and provide a pharyngoplasty effect, it occasionally results in sleep apnea.

Postoperative care includes intravenous and oral antibiotics for 5 to 7 days and a mist

tent for 24 to 48 hours. A tongue suture or even retention of the nasal tubes controlling the size of the lateral ports for 12 to 24 hours is employed when there is special concern regarding postoperative upper airway obstruction.

Complications. The primary complication of a pharyngeal flap is a hemorrhage, which usually occurs within the first 24 hours. Control must be obtained, and if necessary the patient should be returned to the operating room to establish hemostasis. Control of the airway during and after the operation is also vital in these cases, since airway obstruction is the major cause of death in this procedure. Both the mist tent and the lateral port control tubes improve the maintenance of the postoperative airway.

Velopharyngeal competence is initially achieved in approximately 85% to 90% of patients after creation of a pharyngeal flap. The failure to obtain competence may be caused by flap disruption, shrinkage (which usually occurs secondary to inadequate mucosal coverage of the raw surface of the flap), inadequate control or inappropriate sizing of the lateral ports, and restriction of lateral pharyngeal wall motion secondary to scarring from the surgical procedure. Persistent velopharyngeal incompetence may be corrected by an augmentation of one or both of the pharyngeal ports by the use of an additional flap, a Teflon injection into the flap portion of the lateral pharyngeal port, or a prosthetic device. Occasionally stenosis of the lateral ports occurs, resulting in total nasal obstruction and denasal voice. When severe, it may be corrected by enlarging the lateral ports with a Z-plasty technique or by laser excision of the port margin followed by prolonged (4 to 6 weeks) endotracheal tube stenting of the enlarged ports.

Dental Management

Adequate dental care is necessary and desirable in all patients; however, cleft patients have more severe dental problems than the normal population has. The local family dentist or pedodontist can often deliver routine dental care. All teeth should be preserved if at all possible, since future occlusion and the ability to retain a dental prosthesis may depend on the presence of an adequate number of teeth. In addition to the basic dental problems, cleft children frequently have congenitally missing teeth (50%) and an abnormally formed tooth (27%) in the area of the cleft (Olsen, 1971). Supernumerary teeth also occur in 21% of cleft patients, compared to 1% of the normal population. Frequently cleft patients require additional specialized dental care, which only an experienced prosthodontist and orthodontist can deliver.

A maxillofacial prosthodontist is frequently involved in the lifelong management of cleft patients. Areas of prosthodontic cleft care include the use of presurgical orthopedics, fixed prosthodontics (crown and bridge), and removable prostheses.

Presurgical orthopedics involve the use of a prosthetic appliance to obtain a more favorable relationship of the maxillary arches before lip repair in unilateral cleft lip and palate. Improved alignment of the maxillary arches often facilitates lip repair by improving the relationship of the underlying bony structures in patients who have wide clefts or severe malalignment of the maxillary arches. The appliance is usually constructed during the second week of life and is designed so that pressure is applied to specific areas of the maxillary arch to move segments into a more favorable anatomic position. This appliance is usually fitted to the bottle nipple and pacifier and functions when the child sucks. It must be modified

every 2 weeks because the segments change position and growth occurs in this area. This appliance is usually utilized for 3 to 6 months before the closure of the lip, and the majority of patients undergo lip surgery after 4 months of use (Lavelle and Bardach, 1974).

A bridge is a fixed dental prosthesis that is permanently placed in the mouth, using two or more remaining teeth or an individual tooth. A crown is a fixed dental prosthesis that restores the normal contour and function of a damaged or malformed tooth; however, it uses the remaining tooth and root structure rather than adjacent teeth for retention. Cleft patients frequently require these types of fixed prostheses for the maintenance of teeth and for replacement of teeth lost secondary to disease or to the congenital defect.

Removable prosthetic appliances include dentures (either partial or complete) and dental appliances, such as palatal lifts or obturators. Removable partial dentures may be used instead of fixed bridges to restore missing or malformed anterior teeth in cleft patients. A dental obturator may provide velopharyngeal competence or be used to close a defect in the palate by physically occluding the opening between the oral cavity and nose, thereby improving speech and swallowing. Basic indications for the use of a dental prosthesis to occlude palatal defects include the requirement of a prosthesis to replace missing teeth, medical problems contraindicating surgical therapy, patient refusal of surgical therapy, and patients with prior failure of surgical closure who do not desire additional surgical therapy. The primary advantages of prostheses are the problems associated with wearing them and the necessity for their intermittent replacement.

The main goals of orthodontic care in cleft patients are to establish a functional dental occlusion, create an acceptable facial appearance, facilitate facial growth, and assist in speech production and development. Olin (1971) has divided orthodontic care of the cleft patient into three separate phases: phase 1 includes therapy delivered during primary dentition, which consists mainly of expansion of the maxillary arches; phase 2 consists of therapy during mixed dentition; and phase 3 involves the final corrections performed after the eruption of all permanent teeth.

The most common orthodontic deformity found in patients with cleft palates involves an anterior and/or posterior crossbite, which usually occurs on the side of the cleft. With this condition there is an abnormal occlusion with the teeth of the maxillary arch contacting the mandibular teeth medial to the normal anatomic position. These occlusion abnormalities may be corrected by orthodontic expansion of the maxillary arch to an appropriate anatomic position. Repair of any oronasal fistula in a patient with a crossbite should be delayed until the maxillary expansion is completed to prevent dehiscence or recurrence of the fistula secondary to this expansion. Rotated, malaligned, or missing teeth adjacent to the cleft defect may frequently be corrected by basic orthodontic techniques used to correct deformities involving only the teeth. If a significant bony defect exists in addition to the abnormal occlusion, a surgical procedure may be necessary for correction. This procedure can involve either a maxillary or a mandibular osteotomy, depending on the patient's appearance, the occlusion, and cephalometric analysis. With proper orthodontic care, most patients can eventually be provided with a functional and aesthetically normal dental occlusion.

This description of the dental care required by cleft patients demonstrates the vital role of dental specialists' basic management of dental problems, their interaction with other team

members in regard to the method and timing of various secondary surgical procedures is also very important. Without this input into the overall patient management, surgical correction of various deformities can make orthodontic therapy more difficult or result in recurrence of the original problem once the orthodontic therapy is completed. Dental specialists are thus an absolute necessity in the management of patients with cleft lip and palate.

Ear Disease

Patients who have clefts involving only the lip have an incidence of hearing loss similar to that in the general population. However, many authors have reported a greater incidence of hearing loss in patients with cleft palate (Graham, 1963; Linthicum et al, 1959; Masters et al, 1960; Miller, 1959a, 1959b; Sataloff and Fraser, 1952; Skolnik, 1958; Spriesterbach et al, 1962). Although these studies have demonstrated an increased incidence of hearing loss, there is disagreement regarding the type and degree of hearing loss. This variation appears to be related to both the definition of hearing loss employed in the studies and to the age of the cleft patient evaluated for hearing loss.

The abnormal insertion of the levator and tensor veli palatini muscles into the posterior margin of the hard palate and the muscular hypoplasia appear to be the primary cause of eustachian tube dysfunction in cleft patients. Severeid (1971) found evidence of eustachian tube dysfunction resulting in a conductive hearing loss or middle ear fluid in 84% of patients with cleft palate. The only significant variable found to correlate with the incidence of eustachian tube dysfunction was the age of the patient. With increasing patient age the incidence of eustachian tube dysfunction decreased, and in most cases normal eustachian tube function eventually developed with increasing patient age. During long-term follow-up of these patients, 7% developed cholesteatomas (Severeid, 1977), which occurred in the face of regular otolaryngologic examinations given every 3 to 6 months from birth.

Currently the goal of the management of ear disease in patients with cleft palate is to provide adequate hearing, maintain ossicular continuity, maintain an adequate middle ear space, and prevent deterioration of the tympanic membrane until the patient develops adequate eustachian tube function. All patients are carefully examined at a minimum of every 3 to 4 months until they develop adequate eustachian tube function. After this they are evaluated at maximal intervals of 1 year. Any patient with recurrence of eustachian tube dysfunction is evaluated at minimal intervals of 3 to 4 months until it resolves and is adequate on two consecutive interval examinations.

Indications for a myringotomy and insertion of ventilating tubes into the tympanic membrane include (1) the presence of a significant conductive hearing loss or chronic middle ear effusion if audiometric evaluation is not available because of the age of the patient, (2) recurrent episodes of otitis media, and (3) deterioration or retraction or pocket formation of the tympanic membrane. The use of ventilating tubes should improve the patient's hearing and may improve the acquisition of speech. In addition, this aggressive management and frequent evaluation may provide early detection of patients developing cholesteatoma, so that it can be prevented by myringotomy, tympanotomy, atticotomy, or mastoidectomy. The evaluations also allow maintenance of adequate hearing, ossicular continuity, an adequate middle ear space, and a functional tympanic membrane until the patient eventually attains satisfactory eustachian tube function.

Summary

This chapter has presented an overview of basic methods, surgical techniques, and timing of therapeutic procedures required in the management of the complex problems that patients with cleft lip and palate have. These patients have complex and diverse disorders. An absolute necessity exists for input from many specialists; for an optimal overall management of cleft disorders the team approach must be utilized.