Cleft Palate

Introduction. Cleft palate is a congenital deformity that causes a multitude of problems and represents a special challenge to the medical community. Special care is needed for patients with cleft palate. Speech production, feeding, maxillofacial growth, and dentition are just a few important developmental stages that may be affected.

Multiple specialists make up the team that works together to improve the quality of life for patients with cleft palate. Otolaryngologists, oromaxillofacial surgeons, plastic surgeons, nutritionists, and speech pathologists are just a few of the members of the team. Psychological effects on both the patient and the parents are important aspects that also need to be addressed. A significant number of patients with cleft palate have associated syndromes that may result in cardiac, limb, or other system defects. Although cleft palate deformity was described hundreds of years ago, to this day, no agreed-upon management algorithm exists for patients with cleft palate.

History of the Procedure. The first recorded operation on a palate was performed in 500 AD for inflammation of the uvula. For centuries, literature and interest in clefts were lacking because the deformity was thought to be due to syphilis.

Misconception also existed regarding the genetic predisposition. In 1887, the following passage was published in *Lancet:*

George Williamson stated that "[the fourth] law of heredity that [imposes] hideous physical impressions on the mind of a mother are [sic] capable of producing deformity and monstrosity in the offspring." As evidence, he introduced a report by Dr Child that described "a mother attending a penny show, where a trained horse pulled the trigger of a pistol, pretending to shoot a rabbit. A dummy was thrown out; back of its head was bleeding, having all the appearance been shot off." The woman bore a child resembling a rabbit.

Pare first described the use of obturators for palatal perforations in 1564. In 1552, Jacques Houllier proposed that the cleft edges be sutured together. However, it was not until 1764 that LeMonnier, a French dentist, performed the first successful repair of a cleft velum. Dieffenbach closed both the hard palate and the soft palate in 1834. von Langenbeck first described cleft palate closures with the use of mucoperiosteal flaps in 1861. In 1868, Billroth thought that fracturing the hamulus would enable better outcomes in surgery. Further modifications of the von Langenbeck technique came from Gillies, Fry, Kilner, Wardill, Veau, and Dorrance. The debate over the timing of closure led to a short break in early surgical repair. However, in 1944, Schweckendiek again began closing cleft defects in young patients.

Frequency. Cleft deformities of the palate are among the most common congenital malformations. A cleft palate can be diagnosed as early as the 17th week of gestation by means of ultrasonography. Although many studies exist,

the exact environmental and genetic factors that play a role are still largely unknown.

The overall incidence of cleft palate with or without cleft lip is 1 case in 1000 live births. The incidence of cleft palate varies by race, with the highest rate among American Indians, at 3.6 cases per 1000 live births, and the lowest rate among African Americans, with 0.3 cases per 1000 live births. Among the total number of clefts, 20% are an isolated cleft lip (18% unilateral, 2% bilateral), 50% are a cleft lip and palate (38% unilateral, 12% bilateral), and 30% are a cleft palate alone. The incidence of isolated cleft palate (without cleft lip) is 1 case in 2000 live births. Submucous cleft palate is more common, with an incidence of 1 case in 1200-2000 patients, depending on the study population. Bifid uvula occurs in 1 of 80 patients and often occurs in isolation, with no clefting of the palatal muscles.

No racial predilection exists for cleft palate, with an equal incidence among all races. Although cleft lip and palate together occur more commonly in males, isolated cleft palate is more common in females.

Etiology. Palate formation begins at the end of the fifth week of gestation. At this stage, the palate consists of 2 parts, namely, the anterior (primary) palate and the posterior (secondary) palate. The medial nasal prominences form the intermaxillary (premaxillary) segment, which comprises the primary palate and incisor teeth. The primary palate extends posteriorly to the incisive foramen.

The secondary palate, which is formed by the lateral palatal processes, begins at the incisive foramen and contains a bony section and a muscular section. The lateral palatine processes appear at about the sixth week of gestation. They comprise the deep portions of the maxillary prominence that form 2 horizontal structures or palatal shelves, which ultimately are derivatives of the first branchial arch. These shelves are originally on either side of the tongue. As the tongue moves downward in the seventh week of gestation, the lateral processes grow medially. Fusion of the hard palate begins anteriorly and continues posteriorly in the eighth week of gestation.

A number of processes are involved in the fusion of the 2 processes. Programmed cell death at the free edges and production of a sticky coat of glycoproteins and desmosomes provide an ideal bonding surface interface. The left side tends to lag behind the right side, leading to a propensity for left-sided clefts. The nasal septum subsequently grows downward into the newly formed palate. The process is completed between the 9th and 12th weeks of gestation.

Bone begins to form in the anterior palate first and extends posteriorly. The soft palate and the uvula, which make up the posterior portion of the secondary palate, develop during the eighth week of gestation. The tensor veli palatini develop, followed by the musculus uvula. These structures are completed by the 17th week of gestation.

The genetic basis of cleft deformity is most likely heterogeneous and multifactorial. Autosomal recessive, autosomal dominant, and X-linked

inheritance patterns have been described. For all parents, the odds of having a child with a cleft are 1 in 700. In families in which no first-degree relatives are affected, the recurrence rate for a cleft lip or palate in subsequent children is 2.5%. When one first-degree relative is affected, the rate of recurrence is 10%. Similar recurrence rates (10-12%) occur in offspring of persons born with cleft deformities. If the cleft is part of an autosomal dominant syndrome, the recurrence rate can be as high as 50%. A cleft deformity is associated with a syndrome in 30% of cases. More than 400 syndromes with a cleft deformity as one of the characteristics have been described.

As previously mentioned, the etiology of the cleft palate is not well understood; however, some evidence exists that external factors may play a role. Relatively few of the many recognized teratogens cause cleft palates. Alcohol consumption in the embryologic period does result in many infants with clefts. Other teratogens associated with cleft palates include phenytoin, retinoids, and illegal drugs (eg, cocaine). Mechanically induced clefts can occur in utero by means of direct impingement on the embryo.

Genetic mapping of families with inherited forms of cleft palate has resulted in the identification of genes involved in palate development. Cleft palate associated with ankyloglossia, an X-linked disorder, was shown to be caused by mutations of the *TBX22* gene. *TBX22* is a member of the T-box gene family, which are transcription factors in vertebrates involved with mesoderm direction. Specifically, *TBX22* is expressed in the palatal shelves just prior to their elevation above the tongue. Mutations in this gene result in cleft palate due to loss of *TBX22* function.

Presentation. Most overt clefts of the hard palate and/or the soft palate are discovered at birth and are often manifested by feeding difficulties. Suckling may be compromised by the loss of an oral seal on the nipple. Cleft palate, especially when associated with mandibular hypoplasia (as with a Pierre Robin sequence), may also cause airway difficulties because the tongue prolapses through the cleft into the nasal cavity and the posterior oropharynx.

Partial clefts of the soft palate or submucous clefts may be overlooked in neonates because they may be asymptomatic. Early manifestations include nasal reflux of liquids or food. Later, as speech develops, hypernasal speech or nasal emission may result.

Indications. Major clefts of the hard palate and/or the soft palate are repaired surgically before the patient is aged 1 year. Instances in which this does not occur include those with complicating medical conditions, such as congenital heart disease or airway compromise. Cleft repair is deferred for cardiac conditions that may be compromised by a change in upper airway resistance. When upper airway obstruction is a major problem, such as with a Pierre Robin sequence, a tracheotomy may be necessary. Cleft repair can then be accomplished with a secure airway.

When a submucous cleft is present, the indications for surgery concern velar competence. Often, the decision to repair a submucous cleft palate is deferred until the patient is aged 4-5 years, when speech development is sufficient to determine the degree of hypernasality and the effect of the cleft on intelligibility. Cleft repair at this age may involve a pharyngeal flap, depending on the amount of velopharyngeal incompetence present.

Relevant Anatomy. The role of the palate is to provide a barrier between the nasal and oral portions of the respiratory tract. Velar actions with deglutition, respiration, and phonation are similar to those of a sphincter; hence, the velopharyngeal mechanism is often termed the velopharyngeal sphincter.

Familiarity with the anatomy of the palate is essential in understanding functional and surgical repair. Blood is supplied to the hard palate by the greater palatine artery, which enters via the greater palatine foramen. The lesser palatine artery and nerves pass through the lesser palatine foramen. Nerve supply originates from the maxillary branches from the trigeminal nerve, which forms a plexus that innervates the palatal muscles. Contributions from cranial nerves VII and IX enter posterior to the plexus.

The palatine aponeurosis is the principal structural element within the velopharynx. It provides an anchoring point for muscles, adding a degree of stiffness, and is continuous laterally around to the hamulus with the tensor veli palatini muscle. The aponeurosis is diamond shaped. More posterolaterally, the salpingopalatine ligament, the fascia of Tröltsch, and the internal fascia of the pharynx (which all form the membranous portion of the eustachian tube) contribute to the velopharynx.

The normal structure and function of the soft palate is dependent on the levator sling. This structure comprises portions of the tensor veli palatini, palatoglossal, palatopharyngeal, and uvular muscles. Functionally, the levator veli palatini, palatoglossus, and musculus uvulae muscles either elevate the soft palate or alter its shape. Other muscles, such as the superior constrictor, palatopharyngeus, palatothyroideus, and salpingopharyngeus muscles, are involved with movements of the lateral and posterior pharyngeal walls. The tensor veli palatini is involved mainly with middle ear aeration. In patients with cleft palate, the muscle attachments are directed anteriorly and attach onto the posterior portion of the bony palate. These fibers must be surgically reoriented to achieve proper palatal function.

Treatment. Medical Therapy. A cleft palate is primarily a surgical problem, so no particular medical therapy exists for the condition. However, complications of a cleft palate, including feeding problems, airway obstruction, and otitis media, may require medical management prior to repair.

Feeding problems. Feeding a baby with a cleft lip and palate can be a challenge. Usually, a team of professionals is available to help the family meet this challenge by providing information regarding feeding and nutritional needs.

A speech/language pathologist who specializes in feeding and swallowing disorders can provide the family with information regarding the most appropriate feeding position and equipment to use to make the feeding as normal as possible. An evaluation of the patient's swallowing skills for signs of aspiration or dysphagia is an important part of this assessment.

A nutritionist can help establish a feeding regimen that provides appropriate energy intake for optimal nutrition and growth. In general, a newborn needs 100-150 mL of breast milk or formula per kilogram of body weight per day. A prosthodontist or an orthodontist can construct an appliance to assist with feeding for infants who cannot tolerate prefabricated feeders. Since no separation exists between the oral and nasal cavities, children with a cleft palate (with or without a cleft lip) have difficulty obtaining adequate intraoral pressure for sucking and extracting liquid from the nipple. This can cause the baby to tire easily and to be unwilling or unable to suck long enough to obtain enough milk. In addition, food or liquid may back up and run out of the baby's nose and cause choking, coughing, or spitting up.

Families can make several modifications to help improve oral feeding. These modifications include using special cleft feeding equipment, properly positioning the infant during feeding, adjusting the placement of the nipple, supporting the cheek, and altering the rate of feeding.

Most babies with a cleft palate are fed with a bottle, although breastfeeding is not precluded and may be attempted in some cases. With bottle-feeding, the baby with a cleft palate typically feeds slower and needs help regulating the flow of liquid. Often, a chewing type of sucking is observed with use of the nipple. Various bottles and nipples can be used to assist with feeding.

A cleft feeding nipple must have a large enough opening to allow the formula to flow easily to prevent sucking fatigue, but it must not be so large as to cause choking. Nipples should be soft and compressible, allowing liquid to flow easily. Soft nipples designed for premature infants used with a regular bottle often work well. Occasionally, the hole in the nipple designed for a premature neonate may need to be enlarged to increase the flow of milk. This is best performed by creating an X-shaped opening to help regulate the flow of formula. Enlarging the opening too much may result in the free flow of milk, which can cause the baby to choke.

Several cleft palate feeders are available from several manufacturers. The Mead-Johnson Cleft Palate Nurser is the most commonly used bottle and nipple for babies with clefts. It includes a long soft nipple with a crosscut hole attached to a flexible plastic bottle that can be squeezed to increase the flow of liquid. The person feeding the infant can squeeze the bottle in conjunction with the baby's sucking/breathing rhythm to assist with swallowing. Squeezing timed with naturally occurring jaw compressions can also facilitate swallowing.

Other devices include the Ross Cleft Palate Nurser, manufactured by Ross Laboratories, that allows for a steady flow of liquid with minimal sucking. This

nipple can be shortened to individualize the speed of the flow. The Haberman Feeder, manufactured by Medela, provides 3 flow rates determined by the position of the nipple in the oral cavity. The Haberman nipple is larger and longer than most nipples, and it can be gently squeezed to help the baby extract the liquid. To facilitate flow, a 1-way valve separates the nipple from the bottle. Air is squeezed out of the nipple before the feeding begins, and the valve allows the nipple to refill with liquid as it is squeezed or sucked. Reduction of air in the nipple helps decrease the baby's overall intake of air.

Placement of the nipple within the oral cavity is important. Optimally, the nipple should be positioned to the back and along the side of the mouth on the noncleft side (in the case of unilateral cleft palate). Supporting the cheek by gently squeezing the cheeks together around the nipple may also improve oral suction.

With regard to positioning, a semiupright position, as upright as possible, is best. This position helps prevent food and liquid from entering the nasal cavity. Upright positioning may also decrease eustachian tube reflux, which may lead to middle ear inflammation (otitis media). Eustachian tube reflux can also cause otorrhea in children with ear grommets. An angle-necked bottle can make feeding in an upright position easier.

If a mother is interested, she should be encouraged to try breastfeeding. If the cleft affects only the lip and alveolar ridge and not the palate, breastfeeding may be successful. A baby with a cleft palate is not likely to obtain adequate suction to extract the milk. However, the difficulty varies and depends on how fast and easily the mother's milk flows; at the least, breastfeeding should be attempted to determine its success. The baby with a cleft lip and palate can be held with the cleft lip side next to the breast, as the soft breast may help create a lip seal not achievable with a regular nipple. With a good lip seal, the noncleft side may function more normally. Remember to position the baby as upright as possible.

Mothers who attempt breastfeeding must be aware of the signs of dehydration and the need to seek medical attention at the first sign of problems. Signs of dehydration in the baby include sleepiness and listlessness, urination fewer than 10 times per 24 hours, and urine that is strong smelling and/or dark and concentrated. In addition, the mother should carefully monitor the baby's weight and take into account the baby's frustration with feeding, sucking fatigue, and signs of hunger when deciding to continue or discontinue breastfeeding.

Frequent burping is important because babies with clefts tend to take in a lot of air with sucking. The gastroesophageal reflux may also be increased because of excessive air intake. Regular burping during feeding may help minimize spitting up. It may be beneficial to feed the baby smaller meals and to increase the number of feedings throughout the day, especially if the baby fatigues quickly with sucking. A nutritionist should carefully monitor these changes to ensure adequate energy intake for optimal growth. Ensuring that the

mouth and lip area are clean after feeding and prior to placing the baby in a reclined position is important to prevent choking.

Spoon-feeding and feeding of textured and table foods usually evolves in the same quantities and developmental sequence as with any baby or child, even if the cleft palate remains open. Things to consider with spoon-feeding include presenting the food slowly, allowing the baby to remove food from the spoon by using its lips, and allowing the baby to regulate the timing of the next mouthful. For textured and table foods, the feeder should continue with the slow rate of presentation and provide sauce or gravy with small textured foods, such as rice, that tend to spread throughout the mouth. Adding sauce or gravy helps the pieces of food stick together as they are transported through the oral cavity.

Most babies and children with a cleft lip or palate learn to eat orally, with modifications. They become adept at moving the bolus through the oral cavity around the cleft. If food falls from the nose or gets stuck in the palate, the feeder should not become alarmed because the food does not interfere with breathing or cause harm. Occasionally, the child may sneeze when food enters the nasal cavity. Food can be removed with a finger or a cotton swab without frightening the child. If the patient with cleft palate continues to have feeding difficulties even with the appropriate modifications, further consultation may be necessary to rule out such problems as dysphagia or sensory integration difficulties.

If the lack of weight gain due to feeding difficulties is a problem, use of a feeding tube should be considered. If problems with weight gain do not respond to feeding therapy, a gastrostomy may be necessary.

Airway obstruction. Airway obstruction may present in children with a cleft palate, especially those with mandibular hypoplasia (ie, a Pierre Robin sequence). Upper airway obstruction results from posterior positioning of the tongue, which is prone to prolapse into the pharynx with inspiration. Nasal obstruction can also result from the tongue protruding into the nasal cavity.

Airway obstruction is usually managed by placing the child in a prone position to prevent prolapse of the tongue. In severe cases in which the obstructed airway is not relieved with conservative measures, a tracheotomy may be necessary. In these instances, such measures as a lip-tongue adhesion are generally not as effective and not as well tolerated as a tracheotomy.

Otitis media. Otitis media is a common complication of a cleft palate and is present in nearly all children with unrepaired clefts. Although recurrent suppurative disease can be a problem, the primary complication is that of persistent middle ear effusion with resultant hearing loss. Medical management for this problem typically involves careful observation, which must be performed in light of the potential complications of prolonged hearing loss, especially in a child at risk for speech problems due to a cleft palate. In most instances, grommet insertion for middle ear ventilation is the preferred treatment to avert potential speech problems due to conductive hearing loss.

Surgical Therapy. General agreement exists that surgical correction of a cleft palate should be accomplished when patients are younger than 1 year, before significant speech development occurs. The potential benefits of an intact velum as a child begins to speak are believed to outweigh the possible complications of early closure, namely later collapse of the maxillary arch with a resultant crossbite.

How closure is accomplished is subject to some variation. Generally, 1-stage closure of the soft palate and/or the hard palate can be accomplished when the patient is aged 11-12 months. However, some advocate a 2-stage closure, with repair of the velum (soft palate) when the patient is aged 3-4 months. This procedure results in narrowing of the hard palate cleft, facilitating closure at a later date, usually when the patient is aged 18 months. Similar to a lip adhesion for a wide cleft lip, a 2-stage approach may be useful when the cleft palate is particularly wide.

When cleft palate repair is deferred to later childhood or adulthood, repair often involves a pharyngeal flap. Incorporating a pharyngeal flap into the repair can help close a large defect and compensate for velopharyngeal dysfunction and speech problems.

The goal of repair in patients with cleft palate is to separate the oral and nasal cavities; this separation involves the formation of a valve that is both watertight and airtight. The valve is necessary for normal speech. The repair also helps with the preservation of facial growth and the development of proper dentition. Three factors that are considered necessary for satisfactory function of the soft palate for speech are adequate length, adequate mobility, and conformity of the dorsal surface to the pharyngeal wall. Most surgeons include levator muscle complex reconstitution as part of palate repair. Reconstruction of the muscle sling appears more important than anatomical retropositioning in terms of obtaining a dynamic functioning levator sling. However, not all surgical teams have accepted intravelar veloplasty.

If cleft lip is present, its repair can precede palatoplasty. Although early repair seems to have an advantage in decreasing the chances of speech delays, the risk for facial growth abnormalities and other midface-related problems may be increased. In the past, several criteria have been listed for patients undergoing any of the procedures. Some of these criteria include a hemoglobin level higher than 10 g/dL, weight gain, the absence of infection, and a full preoperative evaluation by a pediatrician.

Preoperative Details. Repair of the hard palate is not always possible when the soft palate is repaired, especially with wide bilateral clefts. The cleft size can decrease as much as 7% with growth in patients aged 3-17 months. The size can be further reduced with early repair of the soft palate (in patients aged 3-4 mo) followed by closure of the hard palate in patients aged nearly 18 months. This fact should be taken into consideration in planning the time and the type of the repair. The defect is usually smaller than it was originally when

closure is performed after the soft palate defect has completely healed. The procedure can be performed in patients aged as young as 3 months, with a second procedure for closure of the defect when they are 6-12 months.

A great deal of debate exists regarding the timing of the repair. In the past, many surgeons believed that hard palate repair should be delayed until after eruption of the molar teeth. Currently, most centers focus on completion of the cleft palate before the patient is 12 months. Debate had existed over whether or not delayed closure of the hard palate was beneficial or harmful to facial growth, but the evidence for either side has not been conclusive.

Intraoperative Details. Investigators in a multicenter study involving surveys of more than 300 surgical teams attempted to establish the common ground for repairs of cleft defects. Although no single technique was used universally, a trend has been established toward the use of earlier palate closure over the last several years. Of the closure techniques surveyed, the Furlow procedure was the most common technique for cleft palate closure. The basic surgical techniques included the following: von Langenbeck, Schweckendiek, 2-flap, 3-flap (V-to-Y), and double reverse z-plasty (Furlow) palatoplasties. Although most of the repairs do not involve repairing the muscular sling, doing so allows better palatal and eustachian tube functions. Descriptions of the major techniques used for palatoplasty are outlined below.

von Langenbeck technique. First described in 1861, the von Langenbeck technique underscores the importance of separating the oral and nasal cavities. Virtually every repair performed today incorporates principles initially included in this technique. Bipedicle mucoperiosteal flaps of both the hard palate and the soft palate are used to repair the defect. After their elevation, the flaps are advanced medially to close the palatal cleft. Advantages of this technique include less dissection and its simplicity. A disadvantage of the von Langenbeck repair is that it does not increase the length of the palate, which results in an inability to close primary and secondary clefts. Other criticisms of this technique include the occurrence of anterior fistulas and the resultant inferior speech due to the short soft palate. Airway obstruction during sleep seems to be an insignificant problem with this repair. Because of the physical limitations in lengthening the palate with this technique, many modifications have been made over the years.

Schweckendiek technique. In the 1950s, Schweckendiek began to repair clefts in a staged fashion. In this technique, the soft palate is first repaired when the patient is young (typically 3-4 mo), and this is followed with hard palate closure when they are nearly 18 months. In the interim, an obturator is used to allow swallowing and speech. This technique has the advantages of achieving closure when the patients are young and causing minimal disturbance of facial growth. However, the disadvantages include the need for additional operations; the resultant speech disorders that cannot be easily managed; and the need for frequent changing of the dental prosthesis, which can be expensive.

As noted previously, the initial repair is usually performed in a patient aged 3-12 months. The second stage is usually performed when the patient is 18 months, but it may be delayed until the patient is 4-5 years. Longer delays (ie, until primary dentition is established) were believed to be advantageous in that they prevented lateral contraction of the palatal arch. Currently, speech and feeding difficulties with delayed closure are thought to outweigh the dental alignment problems, and the current trend is to use earlier closure. Collapse of the maxillary arch is now dealt with by means of palatal expansion when the patient is young.

The initial repair is accomplished by making incisions in the soft palate along the margins of the cleft. The levator muscle, which is abnormally attached to the posterior free edge of the bony palate, is dissected free and reoriented. A 3-layer closure of the nasal mucosa, the levator muscles, and the oral mucosa is then performed. The resultant hard palate fistula is closed at a later date.

Although many methods to close the hard palate exist, one technique is the use of the vomer flap. The mucoperiosteum of the vomer bone is elevated in an inferior-to-superior direction. This flap is then rotated laterally for attachment to a small palatal mucoperiosteal flap. This procedure can provide a watertight closure with minimal elevation of the palatal mucoperiosteum. The preferred method involves raising the mucoperiosteal flaps on the oral and nasal surfaces of the hard palate and closing them in 2 layers across the defect. The vomer flap is primarily useful with wide or bilateral clefts. Vomer flaps have the disadvantage of requiring closure of 2 suture lines on the nasal surface. When used with oral mucoperiosteal flaps, the vomer flaps are attached to the flaps raised from the nasal surface of the cleft.

Two-flap technique. The 2-flap technique involves 2 posteriorly based flaps that extend the length of the defect. The flaps are rotated medially to close the defect. This method is the most common technique used for closing complete clefts. No additional length is available for closure of any alveolar defect with this type of repair. An advantage of this method is that the incidence of posterior fistula is low.

After incisions along the cleft margins have been made, the levator veli palatini muscles are dissected away from the hard palate. Modifications that include infracture of the hamulus or stripping the levator veli palatini muscle from the hamulus can be made; these changes greatly improve medial rotation of the mucoperiosteal flaps. This maneuver also reduces closure tension at the junction of the hard and soft palates, helping to prevent fistula formation. Once the nasal mucosa is freed from the nasal surface of the hard palate, the palate can be closed in layers: the nasal and oral layers anteriorly and the nasal, muscular, and oral layers posteriorly.

Three-flap/V-Y (Wardill-Kilner-Veau) technique. In 1937, Kilner and Wardill independently described the V-Y repositioning technique. This technique is primarily used for repair of incomplete clefts or clefts of the

secondary palate. The incisive foramen is the anterior border of the repair, and the uvula is completely divided posteriorly. The theoretical advantage of this technique is that pushing back the flaps adds length to the palate. This length is difficult to achieve without incising the nasal layer of the repair.

Incisions are made along the free margins of the cleft and extended anteriorly from the apex of the cleft to where the canine teeth erupt. Dissection is then continued posteriorly along the oral side of the alveolar ridge to the retromolar trigone. Mucoperiosteal flaps are elevated from the nasal and oral surfaces of the bony palate. Dissection of the greater palatine vessels from the foramen lengthens the pedicle. In the event that the vessel is avulsed and injured, the collateral flow from the lesser palatine and posterior nasal septal arteries is usually sufficient. The bony foramen surrounding the vessel can be opened posteriorly to gain more length. The tensor veli palatini muscle is elevated off the hamulus to aid in relaxing the midline closure. Billroth had advocated infracture of the hamulus, but further study revealed that this structure returned to the prefracture position within 6 months.

As in other repairs, the nasal mucosa is freed from the bony palate and closed to either side or, if necessary, closed by using vomer flaps. The muscle and oral mucosa are closed in a second single layer, usually in a horizontal fashion. Anteriorly, the oral mucoperiosteal flaps are attached to the third flap, which is the mucosa overlying the primary palate. Posteriorly, the palate is closed in 3 layers: nasal mucosa, levator muscle (which was previously freed from the bony palate), and oral mucosa.

Double reverse z-plasty. In 1986, Furlow described a technique to lengthen the velum and to create a functioning levator muscle sling. This method is difficult to perform in wide clefts. However, it is considered a good method when the cleft is narrow or if a submucous cleft exists. The technique involves opposing z-plasties of the mucosa and the musculature of the soft palate. The goal is to separate the nonfunctioning attachments to the posterior border of the hard palate and to displace the mucosa and the musculature posteriorly.

The first z-plasty is created on the oral mucosa side, while the second z-plasty is inverted on the nasal mucosa side. The incisions are made, and the oral mucosa is dissected free from the underlying muscle. On the left side of the patient, the oral mucosa flap also contains the muscle. On the patient's right side, the muscle is kept with the underlying nasal mucosa. The 2 muscle-bearing flaps transpose posteriorly, while the thin nonmuscular flaps are placed anteriorly. This technique has the effect of rotating the muscular sling posteriorly and lengthening the soft palate. One potential problem with this technique is the formation of a fistula at the junction of the hard and soft palates.

Submucous clefts. In 1825, Roux described the most common posterior cleft: the submucous cleft. He stated that 3 factors were involved in this deformity: (1) the membranous portion of the soft palate is absent, (2) the palate

is short, and (3) the nasopharynx is abnormally expansive. In 1930, Dorrance found that the anatomical defect occurred with the anatomical position of the levator veli palatini. In 1956, Calnan described the classic triad of the submucous cleft palate, defined by a bifid uvula, a palatal muscle diastasis, and a notch in the posterior surface of the hard palate.

Controversy exists regarding whether the incidence of otitis media with effusion is increased in children with submucous clefts. Studies have revealed an improvement in effusions following repair of a submucous cleft. However, more recent studies have revealed no improved resolution of the effusion after surgery.

The degree of velopharyngeal insufficiency that can exist is based on the anterior displacement of the muscles. Surgery is indicated for patients with 2 categories of conditions: (1) an overt cleft of the soft palate with velopharyngeal insufficiency or (2) an overt defect, usually undetected at birth, with a presentation of hypernasal speech. Occasionally, a submucous cleft palate defect is discovered at the time of adenotonsillectomy, either intraoperatively or postoperatively, as a complication with hypernasal speech.

Techniques for closure of submucous clefts are the same as those described above. Alternatively, the surgeon can use a pharyngeal flap technique or a pharyngoplasty. Pharyngeal flaps are usually superiorly based pedicle flaps of mucosa and underlying constrictor muscle. The overall goal is to create lateral ports that can easily close. Use of a pharyngeal flap is best when a sagittal closure pattern exists (ie, when the greatest contribution to velar closure is lateral wall movement). A sagittal closure pattern most commonly occurs with a cleft palate.

Pharyngoplasty involves 2 flaps that are positioned on either side of the pharynx and rotated superiorly to create a smaller velar opening, thereby aiding in closure of the soft palate. This method is preferred when a circular or coronal closure pattern exists because it does not interfere with the posterior motion of the palate. The choice of technique depends on the preoperative velar closure pattern.

Alveolar bone grafting. Alveolar bone grafting is an integral part of repairing clefts that involve the anterior maxilla. Establishing a bony union can help to prevent maxillary segmental collapse, to close oronasal fistulas, and to encourage eruption of teeth. Regardless of whether the repair is early or late, the neonate should be fitted with an obturator within the first month after birth. Bone grafting in patients younger than 2 years is considered primary, and secondary grafting occurs afterward. Graft material can be obtained from the hip, the ribs, the extremities, or the outer table of the skull. Although morbidity can exist at the various donor sites, the benefit of closing the maxillary gap outweighs the potential risk.

The surgical procedure involves raising mucosal pedicles on either side of the maxillary defect. With the use of any of the described donor sites, the graft of cancellous bone is placed into the pocket. The mucosal flaps are closed in a simple fashion. Many times, the depression of the alar base is immediately corrected on completion of the procedure.

Postoperative Details. Immediate postoperative concerns in cleft palate repair include airway management and analgesia. Repairing the palate changes the nasal/oral airway dynamics and may present problems in the immediate postoperative period, especially in children with a Pierre Robin sequence. The lasting effect of narcotics used for anesthesia may also alter upper airway dynamics. Since placement of an oral airway may disrupt the palate repair, a ligature of 2-0 chromic (or silk) suture is placed through the anterior tongue to allow forward traction on the tongue while the patient is in the postanesthesia area. This suture is removed once the child is fully alert and able to maintain the upper airway.

Adequate analgesia is important in the postoperative period to allow patients to return to their activities as quickly as possible. However, the use of analgesics must be balanced with the risks of oversedation and subsequent airway compromise. Generally, acetaminophen with codeine is sufficient for this purpose. Analgesics may be continued as needed for as long as 7-10 days postoperatively with few problems; the most common adverse effect is constipation.

In infants and younger children, arm restraints or "no-no's" are used when the child is unattended to prevent the placement of fingers in the mouth because this may disrupt the repair.

Diet in the postoperative period is generally limited to liquids and soft foods that do not require chewing. The use of bottles is avoided because the nipples may interfere with the repair. The use of spoons is also avoided for similar reasons. Feeding is accomplished by using either a cup (not a sipping cup) or a Breck feeder (a red rubber catheter attached to a syringe). Normal diet and feeding may be resumed after 10-14 days, depending on the type of repair. At 3 weeks, all dietary and feeding restrictions are removed.

Oral hygiene is best performed by rinsing with clean water, with the patient taking care to remove all collected food particles. The use of hydrogen peroxide should be avoided because it may inhibit healing. After 5-7 days, careful toothbrushing may be resumed.

Follow-up. Once discharged from the hospital, the patient should have follow-up visits at 7-10 days and at 3 weeks. If a small fistula or a wound breakdown is noted in this period, waiting at least 6 months prior to attempting closure is advised. This delay allows for maximal wound contracture and for reestablishment of the blood supply to the tissues.

Complications. Airway obstruction. As mentioned previously, postoperative airway obstruction is the most important complication in the immediate postoperative period. This situation commonly results from prolapse of the tongue into the oropharynx while the patient remains sedated from anesthetics. Intraoperative placement of a tongue traction suture helps in the management of this situation. Airway obstruction can also be a protracted problem because of changes in airway dynamics, especially those in children with a small mandible. In some instances, placement and maintenance of a tracheotomy is necessary until palate repair is complete.

Bleeding. Intraoperative hemorrhage is a potential complication. Because of the rich blood supply to the palate, significant bleeding requiring transfusion can occur. This can be dangerous in infants, in whom total blood volume is low. Preoperative assessment of the hemoglobin level and the platelet count is important. Injection of epinephrine prior to palate incision and intraoperative use of oxymetazoline hydrochloride–soaked packing material can reduce blood loss. To prevent postoperative blood loss, demucosalized areas of the palate should be packed with Avitene or a similar hemostatic agent.

Palatal fistula. Wound dehiscence (palatal fistula) can occur as a complication in the immediate postoperative period, or it can be a delayed problem. A palatal fistula can occur anywhere along the original cleft site. The incidence has been reported to be as high as 34%, and the severity of the original cleft has been shown to correlate with the risk of fistula occurrence. Complete dehiscence is uncommon, but immediate reclosure should be attempted if it does occur. Small fistulas that occur at areas of maximal wound tension are more common. These typically occur at the junction of the primary and secondary palates anteriorly or at the junction of the hard and soft palates posteriorly.

Postoperative cleft palate fistulas can be managed in 2 ways. In a patient without any symptoms, a dental prosthesis can be used to close the defect with good results. A patient with symptoms may require surgery. Poor blood supply, especially the anterior supply, is the major reason for failure of fistula closure. Therefore, closure of persistent anterior or posterior fistulas should be attempted no sooner than 6-12 months after surgery, when the blood supply has had an opportunity to reestablish itself. Currently, many centers wait until the patient is older (at least 10 y) before attempting fistula repair. If simple closure methods fail, vascularized tissue flaps, such as an anterior tongue flap, may be required for closure.

Midface abnormalities. Cleft palate treatments at some institutions have focused on early surgical intervention. One of the negative effects can be maxillary growth restriction in a certain percentage of patients. Palates that are repaired at an early age may have a decreased anterior or posterior dimension, a narrower dental arch, or an abnormal height. Great controversy exists on this topic because the cause of the hypoplasia, whether it is the repair or the effect of the cleft itself on the primary and secondary growth centers in the mid face, is

unclear. As many as 25% of patients with a repaired unilateral cleft palate may need orthognathic surgery. LeFort I osteotomies can be used to correct the midface hypoplasia, which results in malocclusion and jaw deformity.