Cleft palate repair: art and issues

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The French dentist LeMonnier performed the first surgical repair of a congenital cleft palate in the 1760s. The three-stage operation consisted of passing sutures through the cleft borders, cauterizing the cleft edges, and realigning the fresh edges. A report of the operation by an observer concluded, “The child was perfectly cured” [1].

Since this initial palatoplasty, the surgical management of cleft palate and outcome evaluation have become a complex and intricate art. The variety of techniques used to repair cleft palates has grown considerably. Many techniques have been developed solely because of surgeon preference, with little objective demonstration of the long-term efficacy. By the early 20th century, the goal in cleft palate repair was no longer simple closure of the hard and soft palate but included lengthening the palate to improve speech in the cleft patient [2]. During the past few decades, the most debated issues in cleft palate repair have been how to achieve optimal speech development in the cleft patient and how to avoid abnormal maxillofacial growth after repair. These issues are directly related to the choice of surgical repair technique and timing of the repair.

More definitive answers to questions of timing and choice of technique in the initial repair of the cleft palate are needed. This article highlights the fundamental time-tested principles regarding initial repair of the cleft palate and focuses on changes that have occurred in the past decade, including innovative techniques, new guidelines for timing of surgery, and ethical questions about comparing techniques through clinical research.

Incidence

Oral clefts occur with an incidence of 1 in 750 live births, making clefts the second most common congenital defect (following clubfoot). Within the group of oral clefts, cleft palate has posed the greatest challenge in management. One reason for this is the complex epidemiology of cleft palates. For instance, cleft palate can occur as an isolated problem or in conjunction with cleft lip. These seem to be genetically distinct problems. Cleft lip with or without cleft palate occurs with a predictable racial distribution, occurring in only 0.3 of 1000 African American live births, 1.0 in 1000 Caucasian births, 2.1 in 1000 Japanese births, and 3.6 of 1000 Native American births. Alternatively, isolated cleft palate occurs in about 1 in 2000 live births, without a preferential racial distribution [3].

The epidemiology of cleft palate is further complicated by the presence of an estimated 300 syndromes that include some form of cleft palate in their presentation (Table 1) [3]. Cleft palates associated with other anomalies or syndromes tend to be isolated, without lip or alveolus clefting. A syndrome is diagnosed in one third to one half of isolated cleft palate patients [4]. Although syndromic cleft palate patients make up a small portion of the entire cleft palate population, it is important for the clinician to be suspicious of syndromes in the initial evaluation. The syndromic cleft patient requires unique management, including immediate intervention for other problems.
Nearly half of syndromic cleft palate presentations are associated with Pierre-Robin sequence or velo-cardio-facial syndrome. Pierre-Robin sequence, which includes micrognathia, glossoptosis, and cleft palate, is present in a number of syndromes, notably Stickler syndrome, and accounts for 25% of syndromic cleft palates [5]. Velo-cardio-facial syndrome accounts for another 15% of syndromic cleft palates (Table 1) [5–7].

### Anatomy and classification

Embryologically, the nose, lips, and palate are divided into the primary and secondary palates. The primary palate begins to form during the fifth week of gestation, when the nasal placodes invaginate to form nasal pits. The ridges of tissue that form on either side of the nasal pits are known as the medial and lateral nasal prominences. Over the next 2 weeks of gestation, the maxillary prominences, which are inferior and lateral to the nasal pits, grow medially and eventually fuse with the medial nasal prominences to form the primary palate [8]. This involves the bony and soft tissue elements anterior to the incisive foramen: the nose, the lips, the prolabium (central upper lip), and the premaxilla (triangular portion of anterior maxilla, along with the four incisors) [9].

Normally, fusion of the primary palate is complete at the end of the sixth week of gestation. The secondary palate begins to form during the sixth week, when the same maxillary prominences involved in primary palatal formation form two shelf-like outgrowths called the palatine shelves. These shelves first grow downward on each side of the tongue; in the seventh week of gestation, they ascend and grow horizontally above the tongue, eventually fusing to form the secondary palate [8]. The fusion begins at the incisive foramen and proceeds posteriorly toward the uvula. Normally, the shelves fuse in the midline to form the bony hard palate; the hard palate fuses to the vomer of the nasal septum by the ninth week of gestation. Palatal fusion continues posteriorly with full formation of the secondary palate by the twelfth week of gestation [10]. This involves all elements posterior to the incisive foramen: the posterior portion of the maxilla, or hard palate, and the soft palate.

In addition to genetic predisposition for the development of palatal defects, environmental hazards can disrupt normal embryogenesis. Anticonvulsant drugs such as phenobarbital and phenytoin (Dilantin) increase the risk of cleft palate when taken during pregnancy [8]. Other environmental factors that may predispose to cleft palate formation include alcohol, hypoxia, steroids, and retinoids (eg, vitamin A) [9]. The process of palatine fusion from anterior to posterior takes about a week longer in females than males; this added time allows for longer teratogenic exposure and may explain the increased incidence of isolated cleft palates in females. The preponderance of left-sided unilateral clefts may be because the left palatal shelf usually takes longer to reach midline in the fusion process than the right side [9].

Anatomic features of the normal palate must be considered in understanding cleft pathology (Fig. 1). The palate divides the oro-pharynx and naso-pharynx. The normally fused hard palate is covered with a dense mucous membrane that is closely adherent to

### Table 1

Syndromes commonly associated with cleft palate

<table>
<thead>
<tr>
<th>Syndromes</th>
<th>Clinical findings</th>
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<tbody>
<tr>
<td>Stickler</td>
<td>Pierre-Robin sequence, ocular malformations (progressive myopia, retinal detachment, secondary glaucoma, cataracts), hearing loss, and arthropathies</td>
</tr>
<tr>
<td>Velo-cardio-facial</td>
<td>Cardiovascular abnormalities (tetralogy of Fallot, ventricular septal defect, right-sided aortic arch, aberrant subclavian veins, tortuous and medially-displaced carotid arteries); abnormal facies (long face with maxillary excess and malar flatness, hypoplastic alar cartilages and thin upper lip with long philtrum); characteristic 22q11 “Catch 22” chromosomal deletion</td>
</tr>
<tr>
<td>Van der Woude</td>
<td>Facial clefting, lower lip paramedian sinuses</td>
</tr>
<tr>
<td>Goldenhaar</td>
<td>Ocular dermoid/coloboma, ear tags/atrophia, facial hypoplasia, kidney/vertebral anomalies; mental retardation</td>
</tr>
<tr>
<td>Treacher-Collins</td>
<td>Lower eyelid colobomas, down-slanting palpebral fissures, hypoplastic zygomatic arch, low-set ears</td>
</tr>
<tr>
<td>Ectrodactyly-ectodermal dysplasia-clefting</td>
<td>Lobster-claw anomaly of all four extremities, typically bilateral cleft lip/palate</td>
</tr>
<tr>
<td>Oro-facial-digital</td>
<td>Oral clefting (lip, tongue, palate), mandibular hypoplasia, mental retardation</td>
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the underlying periosteum, creating a mucoperiosteal covering of the oral bony surface. Likewise, the nasal surface is covered with a dense mucoperiosteum; the vomer of the nasal septum is fused to the midline. In a cleft of the hard palate, the free, unfused ends of the palatine shelves are usually covered with mucoperiosteum; the vomer fuses to one end or is left suspended between the cleft edges. At the level of the posterior molars, along the lateral aspect of the hard palate, the posterior palatine artery emerges from the palatine canals; the artery is located between mucoperiosteum and bone, along the lateral edges of the hard palate [10].

Whereas the hard palate serves a structural purpose, in maintaining maxillo-facial architecture, the soft palate serves a more functional purpose; normally, the soft palate works as an active muscular valve, called the velopharyngeal sphincter (see Fig. 1). This sphincter acts to raise the soft palate up against the posterior pharyngeal wall, dynamically separating the nose from the mouth [9]. The soft palate’s intrinsic muscular function aids in proper breathing, swallowing, blowing, and phonation [10]. Six pairs of muscles comprise the soft palate: levator veli palatini, tensor veli palatini, superior constrictor pharyngeus, uvulus, palatopharyngeus, and palatoglossus muscles [2]. The exact contribution of each muscle pair to palatal function continues to be debated; however, the tensor andlevator veli palatini muscles, both of which arise from the eustachian tube, are key anatomic features in cleft palate repair. The tensor veli palatini muscles course inferiorly and

Fig. 1. Superolateral view of normal and cleft anatomy of the palate. (A) Palatal anatomy of the normal newborn. (B) Anatomy of complete cleft involving primary and secondary palates.
wrap laterally around the pterygoid hamulus before inserting medially into the soft palatal aponeurosis near the junction of the soft and hard palates. These muscles seem to control the opening of the eustachian tube, possibly serving to aerate the middle ear and prevent recurrent otitis media. The levator veli palatini muscles course inferiorly and medially, interdigitating in the midline and forming the bulk of the "levator sling." This sling acts to raise the palate to the pharynx, providing much of the velopharyngeal sphincter’s function [3]. The palatoglossus and palatopharyngeus muscles, originating at the midline of the soft palate and inserting into the tongue and lateral pharyngeal wall, respectively, also support this sphincter function by constricting the oropharyngeal aperture [3].

Clefts of the soft palate not only disrupt the levator sling, but they also disturb all normal muscular insertions in the palatal aponeurosis. In the cleft soft palate, muscles that normally join at the midline trend anteriorly and insert on or near the posterior edge of the hard palate [3]. Sphincter function is compromised, leading to velopharyngeal insufficiency and problems with speech development. Eustachian tube control is also lost, often leading to chronic otitis media with the risk of permanent hearing loss. These anatomic considerations are important for understanding palatal function and surgical landmarks.

Cleft palate classification is based upon anatomic disruption of the primary and secondary palates and includes the categories of complete and incomplete, unilateral and bilateral, and submucous clefting. The complete cleft palate involves the primary and secondary palates in which no fusion between the palatal shelves has taken place. It is usually associated with a cleft lip and alveolus. The incomplete cleft palate involves only the secondary palate; fusion between palatal shelves has been initiated but not completed. Thus, the incomplete cleft may involve only the very posterior portion of the soft palate, or it may extend through the soft and hard palates to the incisive foramen.

A cleft of the secondary palate is classified further as unilateral or bilateral. With the unilateral cleft palate, only one palatal shelf fuses to the nasal septum; this leaves the cleft defect to one side of the midline. With the bilateral cleft palate, neither palatal shelf fuses with the nasal septum, leaving a wider midline cleft defect with the vomer of the nasal septum suspended superiorly.

The submucous cleft involves a separation of the intrinsic soft palate musculature while the overlying soft palatal mucosa remains intact [3]. This type of cleft is often difficult to diagnose because the entire palate may seem grossly intact. Often, the only anatomic clues suggesting a submucous cleft palate are a bifid uvula, a notched posterior hard palate, or a translucent area in the midline of the soft palate, known as the zona pellucida, where the musculature has failed to fuse. Often, this cleft type is not discovered until a child develops velopharyngeal incompetence, manifested as hypernasal speech [11]. Although this is the most difficult type of cleft palate to diagnose, it is the most common type of posterior palatal cleft [12]. The incidence of submucous cleft palates is difficult to determine because it is usually discovered only if a patient is referred for velopharyngeal incompetence. Attempts to measure the incidence in the general population by anatomic criteria have revealed rates in the range of 2 to 8 per 10,000 [11].

The syndromic cleft palate often presents as an anatomically distinct deformity. For instance, the typical isolated, or incomplete, cleft involving hard and soft palates is V shaped; it is narrower anteriorly and tends to widen toward the posterior end at the soft palate. However, the cleft palate associated with Pierre-Robin sequence is U shaped and much wider than a nonsyndromic cleft palate of similar type [4].

Operative techniques

Once a patient with cleft palate is evaluated for the presence of an associated syndrome and the cleft is appropriately classified, strategies for primary repair may be considered. Technique selection and timing for repair are discussed through four representative cases.

Incomplete cleft of the soft palate

In this case, an infant presents with a nonsyndromic, isolated, and incomplete cleft palate involving only the soft palate. The primary goal of repair in this case is restoration of velopharyngeal competence. This is achieved by lengthening the palate, for proper apposition of palate and posterior pharyngeal wall, and re-organizing the palatal musculature.

Veau, in the earlier 20th century, repaired clefts of the soft palate by bringing together the cleft edges, with the intravelar musculature trending anteriorly and attaching on or near the posterior edge of the hard palate. This meant that muscle bundles were sutured together side by side. Lateral relaxing incisions or mucoperiosteal flaps on the hard palate were used...
to reduce tension and gain the mobility needed to approximate the cleft edges [13]. Presently, the most widely practiced methods of soft palatoplasty are intravelar veloplasty and the Furlow double-opposing Z-plasty.

The intravelar veloplasty proposed by Kriens in 1969 was an improvement upon previous soft palatoplasties [13]. Kriens’ innovation was to restore the levator sling and palatal musculature at the midline where they normally meet. This is accomplished by dissecting the anteriorly malpositioned muscle bundles from the posterior edge of the hard palate and repositioning them in the midline. This technique is widely used today, although there is much variability among surgeons in how the musculature is dissected and repositioned. Until recently, the results of this technique had not been objectively compared with the older standard technique advocated by Veau. In 1989, Marsh et al [14] published results from a prospective study that compared the effects of intravelar veloplasty and traditional side-to-side techniques upon velopharyngeal incompetence (VPI). They found that repositioning of the levator muscles during primary palatoplasty was no better at improving VPI than the side-to-side veloplasty. However, this study was limited by a small patient population (51 patients) at a single institution. In 1995, Cutting questioned whether the intravelar veloplasty technique adequately dissected and repositioned the musculature [15]. It remains a challenge to prove by prospective, well-controlled, multi-center studies whether the intravelar veloplasty is a more effective technique than its simpler predecessor.

The Furlow double-opposing Z-plasty technique was unofficially introduced in 1978 and was introduced in published form in 1986 [16]. Over the last decade, it has become the veloplasty technique of choice among many surgeons. This technique uses two reversed Z-plasties based upon the cleft midline, both of which draw in soft palate tissue from the sides to close the cleft defect and restore the musculature to its anatomic position. The inherent advantages of the Furlow repair are that it lengthens the palates and restores normal muscular anatomy. It also eliminates the need for lateral relaxing incisions to gain tissue for cleft closure. A concern shared by many surgeons, including Millard [1], is that the Z-plasties in the soft palate tend to pull the sides of the velum toward the midline to lengthen the palate; this tightens the velum in the transverse axis. Nonetheless, retrospective studies published during the last decade have shown that patients with Furlow repairs have reduced hypernasality and improved articulation and speech [17,18]. Prospective controlled trials are needed to compare the Furlow palatoplasty to the intravelar veloplasty and other procedures [18].

Among recent developments in the repair of soft palate clefts, there is increased interest in manipulation of the tensor veli palatini muscles to gain palatal length. The goal of such manipulation is to release tension on the levator sling. One way to do this is by fracturing the pterygoid hamulus (around which the tensor muscles are tethered) during soft palate repair. This method was first used by Billroth in 1889 [1,19]. At that time, it was performed to relax the tension in the lateral tissues of the soft palate and thus minimize postoperative palatal dehiscence. The fracture also allowed Billroth to avoid the standard lateral relaxing incisions of the day, which tended to injure soft palate musculature. Since that time, the technique has continued to find favor with a small minority of surgeons. Studies have been conducted recently to assess the value of this technique. Kane et al [19] conducted a randomized, prospective study comparing complication rates in palatoplasties with and without hamulus fracture. No differences between the groups were noted in perioperative morbidity (fistulas and dehiscence) or in hearing and speech results 1 year after surgery.

The hamulus fracture technique is appealing because it relieves tension in the soft palate closure; however, an alternative technique that severs the tensor tendon in the space of Ernst seems to achieve comparable or greater release of the levator sling. Preliminary results of a prospective study using this release method have been presented. Tendon release was performed in conjunction with a Furlow repair and lateral relaxing incisions on a variety of cleft types with follow-up VPI rates of 4%, compared with rates of 10% with the Furlow palatoplasty or intravelar veloplasty alone [20].

Another recent development in soft palate repair is a uvular transposition technique [21]. This procedure recruits tissue for soft palate lengthening from the uvula and can be performed in conjunction with the Furlow palatoplasty or intravelar veloplasty. A prospective study evaluating speech results 4 years after surgery showed improvement using uvular transposition when compared with Furlow palatoplasty or intravelar veloplasty alone. Two of 62 patients demonstrated significant postoperative VPI after the uvular transposition palatoplasty [21].

In summary, an incomplete cleft of the soft palate can be repaired with an intravelar veloplasty or Furlow repair, depending on the surgeon’s preference. It may be necessary to use a pushback technique from the hard palate mucoperiosteum or lateral relaxing incisions in the soft palate to bring the
cleft edges together. Alternative methods to gain tissue and lengthen the palate include the hamulus fracture, tensor veli palatini tendon release, or uvular transposition.

Incomplete cleft of hard and soft palate

This case involves an incomplete cleft of the secondary palate to the level of the incisive foramen. In addition to a cleft of the entire soft palate, the hard palate has a left unilateral cleft. The vomer is attached to the nasal surface of the right cleft edge. The goals of this repair are realignment of the soft palate mucosa and musculature to restore velopharyngeal competence and closure of the bony gap between the edges of the hard palate to restore structural integrity and maintain growth of the oral cavity and mid-face.

In the early 19th century, Dieffenbach introduced lateral relaxing incisions in the secondary palate for closure of palate defects. By the 1860s, von Langenbeck had expanded on this idea by raising the entire mucoperiosteum as a flap for coverage of the hard palate cleft [13]. These flaps have remained the basis of repair for cleft palate defects; they bring vascularized periosteum and mucosa to cover clefts of the hard palate and help in soft palate closure and lengthening by pushing mucosal tissue medially and posteriorly.

Three variations of repair involving mucoperiosteal flaps are used: the von Langenbeck palatoplasty, the Veau-Wardill-Kilner (V-W-K) palatoplasty, and the two-flap palatoplasty. The first two are used commonly to repair the incomplete cleft involving hard and soft palates. The von Langenbeck palatoplasty involves relaxing incisions along the lateral edge of the hard palate, starting anteriorly near the palatomaxillary suture line, running posteriorly just medial to the alveolar ridge, and ending lateral to the hamulus, about 1 cm posterior to the greater tuberosity of the alveolus (Fig. 2) [3,4]. The mucosa along the edges of the cleft is also incised. The entire mucoperiosteum is then raised from the oral surface of the hard palate; care is taken to preserve the two neurovascular pedicles—the greater palatine pedicle posteriorly and the incisive pedicle anteriorly. Bipedicled mucoperiosteal flaps are created on both sides of the cleft. The nasal side of the cleft is closed first, using redundant mucoperiosteum from the incision along the cleft edge. Then the bipedicled flaps are approximated to cover the oral surface of the cleft. The von Langenbeck technique works well for incomplete clefts of the secondary palate without the presence of cleft lip or alveolus [3].

A variation of the von Langenbeck repair, the V-W-K repair or V-Y pushback, can also be used for incomplete clefts involving the hard palate (Fig. 3). The same flap design as the von Langenbeck repair is used. Then the superior pedicle is divided, leaving a flap on either side of the cleft based solely on the greater palatine pedicle posteriorly. The mucoperiosteal flaps can then be approximated directly or in a V-Y closure at the free anterior end to actively lengthen the soft palate. This repair technique allows more flap advancement than the von Langenbeck repair; however, the gain in palatal length (and possibly improved velopharyngeal function) is at
the cost of denuding palatal bone anteriorly on the oral surface. It has been a concern that this denuded palate might adversely affect midfacial growth in cleft palate patients [13]. A recently published retrospective study concludes that satisfactory long-term midfacial growth can be obtained with proper use of the V-W-K repair [22]. However, this study was limited to analysis of a single surgeon’s patients; variability in how the technique is performed by other surgeons may not lead to similar favorable outcomes. In fact, other studies have suggested that the V-W-K repair has detrimental effects on midfacial growth [13,23]. Until the V-W-K repair is exonerated of charges that it inhibits facial growth, many surgeons opt for the more conservative Von Langenbeck repair, in which the relaxing incisions denude less palatal bone. The von Langenbeck repair can be used in combination with the intravelar veloplasty or Furlow palatoplasty to minimize the extent of mucoperiosteal undermining [13].

A recent retrospective study compared the amount of denuded palate and outcomes in three methods of hard palate repair [24]. Comparisons were made between a pushback repair (most denuded palate), the von Langenbeck repair (less denuded palate), and a von Langenbeck variation (least denuded palate) with relaxing incisions moved from the lateral to medial sides of the greater palatine bundle. Consecutive patients in non-overlapping time periods were included in the study. Maxillary growth was measured with a rating scale that combined the Goslon Yardstick and Five Year Model Index. Patients were categorized into five groups (group 1 with the most favorable facial growth outcome to group 5 with the least favorable outcome). Group 1 patients had a positive overbite and overjet, and group 5 patients required an osteotomy for correction of maxillary deficiency. Among patients undergoing the pushback-style repair, 13.9% were classified as groups 1 and 2, and 55% were classified as groups 4 and 5. In patients receiving the von Langenbeck repair, 31.1% were in groups 1 and 2, and 42.4% were in groups 4 and 5. Results were best in the group undergoing von Langenbeck repair with medial relaxing incisions: 55.5% were in groups 1 and 2, and 11.5% were in groups 4 and 5. These results suggest that increases in denuded hard palate result in decreased maxillary growth [24].

In summary, repair of an incomplete cleft of the hard and soft palate involves mucoperiosteal flaps, using the von Langenbeck or V-W-K repair. Both are popular methods and are chosen based on surgeon preference; however, one must be aware of the potential adverse effect on maxillary growth caused by denuded palatal bone, particularly in the V-W-K repair. Pairing of an effective veloplasty technique with one of these mucoperiosteal flap techniques can minimize denuded palate.

Complete cleft palate

The complete, bilateral cleft palate involves clefting of the primary and secondary palates (Fig. 4). This presents a unique challenge because of cleft width and continuity of the palatal cleft with clefts of the lip and alveolus. For unilateral and bilateral...
clefts of the palate, a third variation of mucoperiosteal flap technique, the two-flap palatoplasty, is used. This technique is similar to the V-W-K repair, but the dissection extends further anteriorly to encompass the cleft edges at the alveolus. With a unilateral complete cleft, the noncleft flap is used to cover most of the cleft side, and the flap on the cleft side is minimally moved. Both flaps can be approximated along the cleft line and are anchored anteriorly at the maxillo-alveolar junction.

With a bilateral, complete cleft palate, further considerations are required. Because the vomer is not attached to either free edge of the hard palate and the cleft gap is often too wide for direct approximation of nasal mucosal edges, a vomer flap can be used for closure of the nasal mucosa. Incisions are made along the free margins of the vomer, which is exposed in the cleft gap; two septal-mucosal flaps are raised, creating the vomer flap. These flaps are used to bridge the gap between free cleft edges of nasal mucosa. The two-flap palatoplasty combined with a vomer flap results in a four-flap palatoplasty. The vomer flap has been used particularly for wider bilateral clefts since the 1920s; more recently, its use has been advocated as a standard repair for all bilateral clefts [13].

Although the two- and four-flap palatoplasties are standard choices for repair of the wide, bilateral cleft palate, further innovation is required to minimize the amount of denuded palate after repair. For instance, buccal flaps to cover denuded areas of palate have occasionally been used since the 1950s. Mann and Fisher [25] document the use of bilateral buccal flaps in conjunction with a modified Furlow repair to cover denuded areas on the posterior hard palate. This technique has proven useful in gaining palatal length in wide cleft palates and provides better tissue cover-

Submucous cleft palate

The diagnosis of a submucous cleft palate is based upon a triad of physical findings that includes bifid uvula, notched posterior hard palate, and zona pellucida (Fig. 5). Surgical repair of the submucous cleft palate is considered if VPI is present. In 1972, Weatherley-White showed that 1 in 9 patients known to have a submucous cleft palate exhibit VPI [11]. More recent studies suggest this fraction may be even greater [7].

Classical repair of the submucous cleft palate has been performed by excising the entire region of the zona pellucida, then using a posterior wall pharyngeal flap [1]. This method does not restore the single, anatomically normal velopharyngeal sphincter but creates two sphincters on either side of the pharyngeal flap. According to analysis conducted within the last decade, a pharyngeal flap can be performed as the primary repair of the submucous cleft palate without the need for other adjunct procedures [12]. Alternatively, a Furlow repair can be used in patients younger than 20 years of age with a small velopharyngeal gap (<5 mm). Given the different strategies used by these techniques, prospective studies are needed to compare outcomes after these alternative repairs.

These four cases provide a foundation for developing repair strategies for the most common types of cleft palate defects. A few innovative techniques have been developed recently that require further objective analysis before they can be adopted as standard techniques. The commonly accepted techniques of Fig. 4. Patient with a wide, complete cleft palate. This defect requires a two-flap palatoplasty with vomer flap.

Fig. 5. Patient with a submucous cleft palate. Note the translucent zona pellucida in the midline and bifid uvula posteriorly.
palatal repair, including the intravelar and Furlow veloplasties, and the three main mucoperiosteal flap techniques (von Langenbeck, V-W-K, and two-flap) are characterized by fundamental repair principles that have been modified to suit the style of individual surgeons. Further objective study is needed that compares complication rates and speech development after the use of these techniques. Randomized, prospective clinical trials would seem to be the gold standard for such comparisons; however, a debate has intensified among craniofacial surgeons in the last decade regarding the ethics of such trials. Is a surgeon’s repeated performance of a standardized repair for a clinical trial always appropriate, given the unique nature of an individual cleft defect, which in turn should require a variety of repairs [27]? This challenge raises two important questions that must be addressed in the future of cleft palate repair: Is it better for a variety of techniques to be used in cleft palate repair, accommodating the natural variety of defects, or for a smaller number of standard, clinically proven techniques to be used? Second, is it appropriate for an area of surgery as delicate and diverse as cleft palate repair to be subject to the rigid analytical constraints of the modern prospective trial? Further research is required to answer these questions.

**Timing of repair**

The timing of cleft palate repair depends upon the type of cleft palate involved, the patient’s symptoms, and the capabilities of the cleft management team. It has been argued that earlier repairs benefit speech development because the speech process in children begins at 1 year of age; conversely, delayed repairs theoretically allow for proper maxillo-facial growth because transverse facial growth is not complete until 5 years of age [1]. This led to a variety of timing protocols at different institutions. More recently, an emphasis has been placed on proper speech development in the cleft patient; thus, earlier timing of primary palatoplasty (before 2 years of age) has become the norm [13]. The current debate focuses on how early the repair should be performed. LaRossa [13] has recently recommended repair of hard and soft palate defects by 18 months of age. A comprehensive review by Rohrich [28] recommends a two-stage palate repair, with soft palate repair at 3 to 6 months of age and hard palate repair at 15 to 18 months of age [28]. At the Riley Hospital for Children in Indianapolis, primary palatoplasty in otherwise healthy children is performed in one stage between 9 and 12 months of age. Long-term outcome studies in the United States are in the process of comparing patients in whom palatoplasty was performed between 9 and 12 months of age with those in whom palatoplasty was performed at an earlier age [29].

Because more than half of children with cleft palate deformities may have other anomalies, timing strategies for palatoplasty change with these co-morbidities. In children with Pierre-Robin sequence, for instance, the timing of palatoplasty is dependent upon the child’s airway status; it is often prudent to delay primary closure until 18 months to 2 years of age to minimize the risk of airway obstruction. Similar strategies are required for patients with other syndromes in which airway obstruction is an issue, such as Treacher-Collins, Apert, or Crouzon syndromes [30]. Overall, the trend in timing for palatoplasty is moving toward repair before 1 year of age.

**Operative and postoperative patient management**

The cleft palate patient is given a general anesthetic and intubated with a midline oral Reye tube. If the patient is younger than 1 year of age, the otolaryngologist may perform bilateral myringotomy and tube insertion. A recent study has found significant correlation between the age at first tube placement in cleft children and the frequency of hearing screen failures; earlier tube placement results in better outcomes [31]. Additionally, eustachian tube function is not improved by realignment of soft palate musculature to their anatomic positions (with the intravelar veloplasty or Furlow repair) [32,33]. This suggests that tube placement, and not palatoplasty, is what prevents middle ear and hearing problems in cleft palate patients.

The patient is positioned on an adjustable Mayfield headrest; a roll beneath the shoulders is used to position the patient’s head in extension. Historically, some surgeons, including Kilner, operated with the patient’s neck fully extended and nearly resting in the surgeon’s lap [1]. A Dingman retractor is then placed to expose the palate optimally and depress the tongue. The areas of incision are injected with 1% lidocaine and epinephrine (1:100,000). Prophylactic antibiotic is administered (although no recent data supports or refutes the use of antibiotics). Wound infection rates after palatoplasty are minimal with or without antibiotics.

Incisions are made with a scalpel or needle-point electrocautery. The cleft edges are pared, leaving an appropriate amount of nasal mucosa for closure; the lateral relaxing incisions, flap margin incisions, or
Z-plasty incisions are then made depending on choice of repair. A freer can be used to raise the mucoperiosteal flaps.

Closure of the repair begins with the anterior nasal mucosa, working posteriorly using a running 4-0 Vicryl suture. A Castroviejo needle driver optimizes suture manipulation [4]. The soft palate musculature, particularly the levator sling, is then approximated end-to-end at the midline with interrupted 4-0 Vicryl sutures. The oral mucosa is closed last, starting posteriorly at the uvula and progressing anteriorly using interrupted horizontal mattress 4-0 Vicryl sutures. In repairs using significant mucoperiosteal flaps, suspension sutures can be placed anteriorly to fix the flaps to the alveolar ridge. To protect the repair postoperatively, a 0-gauge silk suture is passed through the anterior third of the tongue and taped to the cheek; this can be removed before discharge [30]. In addition, elbow extension splints are placed in the operating room and are worn by the patient until the first follow-up visit. This prevents the child from placing fingers in the oral cavity and disrupting the repair (Box 1).

**Complications**

Postoperative complications include primarily repair flap dehiscence and fistula formation. A meta-analysis in 1991 reported postpalatoplasty fistula rates ranging from 0% to 34%. Factors that affected fistula formation were the extent of clefting (clefts involving the primary palate or more of the secondary palate resulted in higher fistula rates), the type of repair (pushback repairs resulted in higher fistula rates than von Langenbeck repairs, which in turn were higher than Furlow/intravelar veloplasty repairs), and the surgeon repairing the palate. Of all fistulas, 87% occur in the area of the hard palate closure, and over half of these occur immediately posterior to the alveolus [34]. Age at palate closure does not seem to significantly affect rates of fistula formation.

Long-term palatoplasty complications are related to velopharyngeal insufficiency or poor mid-face growth, with the development of a cross-bite. Initial follow-up is within 7 to 10 days after repair with the primary surgeon. Additional, long-term follow-up is required with multiple members of the cleft palate team. This team includes an otolaryngologist (to assess hearing function), an orthodontist (to coordinate maxillo-facial growth), and a speech therapist (to evaluate speech development). After the initial follow-up visit, the plastic surgeon follows the patient with yearly office visits.

**Summary**

Caring for the child with cleft palate requires a multidisciplinary approach that begins with evaluation for other possible congenital anomalies, decisions about timing of repair, and choice of techniques. Postoperative follow-up similarly requires a team approach and should include an otolaryngologist, an orthodontist, and a speech therapist. The art of cleft palate repair has enjoyed a decade rich in new developments. New techniques have been developed, and standard techniques have been refined. Most importantly, the need for prospective, randomized trials to objectively compare surgical techniques has been recognized. Initiation and completion of these trials will improve outcomes for patients with cleft palate repairs.

**References**


[2] Nguyen PN, Sullivan PK. Issues and controversies in


