

# Approaches to Cleft Lip and Palate Repair

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**Facial clefts remain one of the most common congenital anomalies encountered by plastic surgeons. Over the last few decades, surgical results have continued to improve due to the interdisciplinary approach to this complex clinical problem.**

*Key Words:* Cleft lip, cleft palate, velopharyngeal insufficiency

Clefts of the lip and palate are the second most common congenital anomaly after clubfoot.<sup>1</sup> Infants with facial clefting may present with either the isolated condition or a constellation of associated anomalies identified as a syndrome. This latter situation demands the attention of not only a plastic surgeon, but also of an entire interdisciplinary team of caregivers including pediatricians, geneticists, dentists, speech pathologists, nutritionists, nurses, social workers, and psychologists. The earliest report of cleft lip repair is found in the Chinese literature in approximately the fourth century AD.<sup>2</sup> The first written Western reports appear in the Middle Ages (1300s), by a Flemish surgeon, Jehan Yperman, and initial illustrations depicting a cleft lip repair are credited to one of the most well-known of the Renaissance surgeons, Ambrose Pare, in the 1500s. Early techniques consisted of simply "fleshening" (linear excision) of the cleft lip margins and closing the defects with external figure-of-eight thread around metal "hare-lip" clips. This was probably done to avoid infection because it was believed that any surgeon using sutures was a "quack." The modern era of cleft repairs dates from the 1800s when Malgaigne and Mirault used non-straight-line excisions and added finesse to cleft surgery.<sup>2</sup>

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## NORMAL EMBRYONIC DEVELOPMENT

Facial development begins in the third week of gestation followed by the progressive development between the 4th and 8th week of the cranial portion of the human embryo.<sup>3</sup> By 10 weeks, the embryonic face has taken on a humanlike appearance.<sup>3-5</sup> Five facial prominences (two paired and one unpaired) bordering the stomodeum (primitive mouth) are responsible for this eventual development of the human adult facial features. The first branchial arch contributes to the maxillary and mandibular prominences and the anterior portion of the auricle.<sup>3-5</sup> In time, further embryonic development continues with merging of the medial nasal prominences that form the philtrum and Cupid's bow region of the upper lip, the nasal tip, the premaxilla and primary palate, and the nasal septum. Lateral nasal prominences form the nasal alae, while the nasolacrimal groove develops as a furrow separating the lateral nasal prominence from the maxillary prominence. "Palatogenesis" begins toward the end of the fifth week and proceeds through the twelfth week.<sup>3-5</sup>

The primary palate (median palatine process) derived from the frontonasal prominence is formed by merging of the medial nasal prominences. The primary palate becomes the premaxillary part of the maxilla (premaxilla) and houses the incisor teeth. This portion, located anterior to the incisive foramen, represents only a small portion of the adult hard palate.<sup>6</sup>

The secondary palate, which arises from the maxillary prominences at approximately day 45, is derived from the fusion of the lateral palatine process and extends posteriorly from the incisive foramen. Initially all three elements, the medial and paired lateral palatine processes, are widely separated by the vertical orientation of the lateral palatine processes located on either side of the tongue. However, during the eighth week the lateral palatine processes shift from a vertical to horizontal position, thereby initiating their eventual fusion.<sup>3,6</sup>

The nasal septum, a downgrowth from the

merged medial nasal prominences, begins to fuse with the developing palate at its nasal surface by the ninth week. By the twelfth week, this process is completed. Ossification in the primary palate and the anterior portion of the secondary palate leads to formation of the hard palate, while the posterior portion of the secondary palate forms the soft palate and uvula. The palatine raphe in the soft palate represents the line of fusion of the lateral palatine processes.

These complex interactions are initiated by both extracellular matrix and soluble factors, e.g., different collagen types (types I and IX), tenascin, epidermal growth factor, transforming growth factor- $\alpha$  and - $\beta$ , platelet-derived growth factor, fibroblast growth factor (FGF), as well as cell surface molecules and intracellular messengers. It is postulated that the genes coding for these molecules are critical to normal palatal development. Other genes such as the H-2 locus and Xq13-Xq21 regions of the X-chromosome have also been linked to palatal development. It is clear that identification of all the genes involved in controlling the definitive formation of the head must be accomplished.<sup>7</sup>

#### EMBRYOLOGICAL BASIS FOR CLEFTING

**T**he embryological basis for the formation of cleft lip and palate rests on the eventual failure of the mesenchymal masses derived from any of the five facial prominences to meet and eventually fuse. Resultant clefts can then be divided into two major embryologically distinct groups, anterior (primary palate) and posterior (secondary palate) cleft malformations. This division depends on their anatomical relationship to the incisive foramen.

Based on this fact, anterior cleft malformations, which result from failure of fusion between the medial nasal maxillary prominences on one side, would include cleft lips with or without cleft palate, whereas bilateral cleft lip develops from the failure of the medial nasal prominences to fuse with the maxillary prominence on either side.<sup>6</sup>

Other rare facial clefts have been described.<sup>8</sup> Laterally, the maxillary and mandibular prominences normally join at the lateral commissure of the mouth. Failure of these prominences to unite produces macrostomia. This is characterized as a cleft of the lateral commissure. Conversely, a median cleft lip is caused by incomplete merging of the medial nasal prominences in the midline. This is usually associated with deep midline furrowing of the nose, characterized by varying degrees of nasal bifidity.<sup>6</sup>

Cleft palates develop from failure of the maxil-

lary prominences (lateral palatine processes) or from the medial nasal prominences (either the median palatine process or the nasal septum) to fuse with each other. This can arise from disturbances at any stage of development. Primary palatal clefts occur anterior to the incisive foramen and result from failure of fusion between mesenchymal masses in the lateral palatine processes with those located within the median palatine process. Clefts of the secondary palate occur posterior to the incisive foramen from failure of the mesenchymal masses in the lateral palatine processes to fuse with each other and with the nasal septum. Primary or secondary cleft palate can be complete or incomplete, depending on the degree of fusion. Submucous palatal clefts may occur when an imperfect muscle union occurs across the velum beneath an intact mucosal surface. Abnormal muscular anatomy may be associated with abnormal velopharyngeal function (velopharyngeal insufficiency (VPI)), and result in hypernasal speech.<sup>3-6</sup> Although numerous theories have been proposed for the cause of clefting, the most popular is believed to be secondary to a delay in elevation of the palatal shelves from a vertical to horizontal orientation.<sup>6,7</sup>

#### GENETICS

**M**ore than a hundred Mendelian disorders are associated with cleft lip and/or palate (thereby defining syndromic CL/P, or CP).<sup>9</sup>

Nonsyndromic clefting of the lip and palate has a complex cause. Multiple genetic loci as well as exposure to various teratogens have been implicated.<sup>9</sup> Because cleft lip and cleft palate do not frequently coaggregate in human families and because these structures arise semiindependently during development, these disorders are usually considered to have a distinct cause.<sup>9</sup>

However, when CL $\pm$ P occurs in families, inheritance is generally regarded as "multigenic." The allelic variation at different loci (e.g., genes for transforming growth factor- $\alpha$  and transforming growth factor- $\beta$ 3) determines a fraction of the eventual genetic risk. Transforming growth factor- $\alpha$  has been implicated as causing a susceptibility locus for nonsyndromic CL/P and CP. Some pedigrees, however, are clearly monogenic, consistent with either autosomal dominant or recessive inheritance patterns;<sup>10</sup> X-linked recessive CP is an additional possibility. Some researchers have even suggested that several of the same chromosomal regions are involved in both forms of clefting when teratogen induced.<sup>9</sup> Shields et al.<sup>11</sup> in 1981 analyzed family data on 561 Danish probands with nonsyndromic isolated cleft palate

and concluded that neither a multifactorial-threshold model nor a single major locus model is completely compatible with the overall distribution of cases.<sup>12</sup> These authors proposed the existence of two classes of nonsyndromic cleft palate: 1) familial CP, which appears to have an autosomal dominant component; and 2) nonfamilial CP, which by demonstrating an increasing frequency of CP with time and a maternal age effect, appears to be related to yet unidentified environmental factors.<sup>9,12-14</sup>

#### TERATOGENS

Exposure to various environmental teratogens during the first trimester of pregnancy have been reported to interfere with lip and/or palatal formation.<sup>9</sup> Although the mechanisms of craniofacial malformation are complex, a common feature appears to be excessive cell death. Many teratogens including steroids, anticonvulsants (phenytoin), diazepam, ethanol, 13-cis retinoic acid, methotrexate, and ionizing radiation along with periods of hypoxia and hyperthermia have been shown to cause clefting in animal models.<sup>12</sup> Phenytoin induces cleft lip, whereas 6-aminonicotinamide has been shown to induce cleft palate.<sup>9</sup> Although ethanol, retinoids, or folate antagonists are clearly teratogenic, indictment of more common exposures such as caffeine remain merely tentative.<sup>15</sup> Infections, such as rubella and toxoplasmosis during the first trimester, have also been associated with clefting.<sup>15</sup>

#### EPIDEMIOLOGY

Precise epidemiologic data regarding cleft lip and palate remain unsettled. Additional factors that affect an accurate collection of data include racial and socioeconomic makeup of the population, quality of birth, hospital, and surgical records and absence of details (e.g., degree of clefting and presence of associated abnormalities). Cleft lip ± palate has an overall incidence estimated between 1/750 and 1/1,000 live births in the United States.<sup>16</sup>

Clefting is even more common in stillborns and abortuses.<sup>17</sup> Unilateral, left-sided cleft lip is twice as common as right-sided cleft lip and more frequent than bilateral clefts (ratio 6:3:1).<sup>17</sup> Cleft lip with palate occurs more commonly than isolated cleft palate or isolated cleft lip (~50% CL/P, 33% CP, 21% CL). Bilateral cleft lip is more commonly associated with cleft palate than unilateral cleft lip (86% vs. 68% in one large study).<sup>18</sup>

Genetically, cleft palate (CP) behaves as a distinct entity from cleft lip with or without cleft palate

(CL/P). Family studies show that children of families with affected members with cleft lip and palate (CL/P) have an increased risk for occurrence of CL/P.<sup>16</sup> However, this does not necessarily follow for isolated cleft palate (CP). Conversely, children of families with affected members with isolated CP are at increased risk for CP and not CL/P.<sup>16,19</sup> It has been postulated that isolated CP appears to be related more to environmental factors than to hereditary factors.<sup>10</sup> Syndromic clefts (those associated with other abnormalities) are seen in 29% of clefting cases. Syndromic clefts are more commonly associated with CP than CL/P.<sup>10</sup>

Racial incidence may be related to genes, environment (nutrition), or culture (e.g., infanticide, inbreeding). Overall incidence of cleft lip and palate in Asians is almost twice that of whites, i.e., 2.1/1,000 vs. 1.30/1,000 live births.<sup>2</sup> Blacks are reported to have the lowest incidence, i.e., 0.41/1,000 (1/2,450) live births.<sup>2</sup> There is no reported racial heterogeneity with isolated cleft palate, which has an overall incidence of 0.5/1,000 live births.<sup>2</sup>

Cleft lip and palate (CL/P) occurs more often in males than females (2:1). Bilateral cleft lip is almost twice as common in boys as in girls. However, isolated cleft palate (CP) occurs more often in girls (2:1) and is believed to be related to the fact that the palate closes later in girls, making it more susceptible to a gestational insult.<sup>15,21,22</sup> There are reports of an increased incidence of cleft lip and palate (CL/P) with older parents but not with isolated cleft palate.<sup>15,21</sup> Also, the risk of recurrence for cleft lip and palate in nonsyndromic children has been extensively studied (Table 1.) Interestingly, it has been observed that affected women with CL/P have a higher frequency of affected children than men with CL/P.<sup>13,15,21</sup>

#### TREATMENT

Surgical goals of cleft lip and/or palate surgery are directed toward achieving a normal facial appearance, feeding, speech, and hearing without sig-

**Table 1.** Risk of Recurrence of Cleft Lip and Palate<sup>13,15,21</sup>

Family Makeup	Risk of CL/P	Risk of CP
One affected sibling or parent*	1 in 25 (4%)	2.5%
Two affected siblings	1 in 11 (9%)	1%
One sibling and one parent	1 in 6 (16%)	15%

CL = cleft palate; CL/P = cleft lip and palate.

nificantly affecting the ultimate facial and psychosocial development of the child. Through a protocol of sequential, regular evaluations by an interdisciplinary team composed of a plastic surgeon, speech pathologist, orthodontist, audiologist, pediatrician, psychologist, geneticist, social worker, pediatric nurse, and nutritionists, great strides have been made in improving the comprehensive care of the child with cleft lip and/or palate.

During the last half of this century, plastic surgeons and orthodontists have worked closely to develop a variety of presurgical orthopedic techniques. Presurgical orthopedic treatment originated in the 1500s, at a time when excision of the protruded premaxilla in bilateral clefts was the recommended treatment. Dissatisfied with the long-term results, surgeons and dentists explored new avenues in the hope of achieving more optimal results. Many believed that closing the alveolar cleft before surgery in the first months of life was crucial to properly perform lip closure.<sup>6</sup> These earliest approaches were extraoral. Currently, appliances are classified as either active or passive, although there is no uniform consensus on a classification. Generally, active appliances use a hard acrylic plate and controlled forces, via extraoral traction (bonnet with straps), which actively move the maxillary alveolar segments into approximation. Another type of appliance is the pin-retained variety popularized by Latham (1980). This intraoral device is designed to exert a forward force to the lesser posterior segment of the unilateral cleft maxilla. To correct the bilateral deformity, Latham used another pin-retained appliance that controlled the width of the posterior segments while retracting the premaxilla with elastic chains. Some authors have expressed concern about possible tooth bud damage from using such devices.<sup>6,23</sup> Another commonly used appliance consists of an alveolar molding plate made of a hard outer shell and a soft acrylic lining. By gradually alternating the tissue surface of the acrylic plate, the alveolar segments are gently pressed, thereby molding them into the desired shape and position.

It remains difficult to assess the ultimate influence of presurgical orthopedics on maxillary growth, because patients with cleft palate undergo multiple surgical procedures. However, it is widely accepted that presurgical orthopedics probably do not enhance or retard eventual maxillary growth, that the overall orthodontic benefits are limited, and that nonsurgical closure of palatal bone and soft tissue remains impossible. Despite the controversy in the literature, presurgical orthopedics continues to have widespread use and is an example of the cooperative

effort within the members of the cleft rehabilitation team.<sup>15,21,23</sup>

#### UNILATERAL CLEFT LIP

Unilateral clefts fall into a spectrum that may be classified as follows: microform cleft lip, incomplete cleft lip, or complete cleft lip. Associated nasal deformity is similarly categorized as mild, moderate, or severe. Mild nasal deformity is characterized by a wide alar base but normal alar contour and normal dome projection. Moderate nasal deformity has a wide alar base in association with either a depressed dome or alar crease. Associated hypoplasia of the alar cartilage mirrors the severity of the associated cleft lip.

Microform cleft lip is characterized by a furrow or scar transgressing the vertical length of the lip, a vermilion notch, imperfections in the white roll, and varying degrees of vertical lip shortness.

Associated nasal deformity may be present and at times may be more extensive than the associated lip. Surgery is generally indicated but must be approached cautiously to avoid creating a surgical deformity worse than the original congenital defect. Unilateral incomplete clefts are characterized by varying degrees of vertical separation of the lip. All have an intact nasal sill, or Simonart's band. Surgical correction is achieved with one of a number of techniques; however, at present the rotation advancement flap is the most popular method.

Unilateral complete clefts are characterized by separation of the lip, nostril sill, and alveolus (derivatives of the primary palate). Although the secondary palate may remain intact, complete clefts most commonly involve the entire palate. Critical factors for evaluating unilateral complete clefts are the position of the alveolar segments and the vertical height of the lateral lip element.

Essential to any cleft lip repair are accurate preoperative anatomical markings, as demonstrated in Figure 1. The difference in height discrepancy between the cleft and normal sides determines the difficulty of repair. The tissue is rearranged to borrow from what is available laterally and introduce it medially. Table 2 shows an overview of the different techniques developed through the years on cleft lip repair.<sup>23</sup>

An essential part of any repair should be reconstruction of the orbicularis oris muscle. The orbicularis oris should be oriented up and reconstructed when repairing a cleft lip. Fara pointed out that the muscle lines up along with the cleft and it must be separated from its attachments for an appropriate repair.<sup>24</sup>

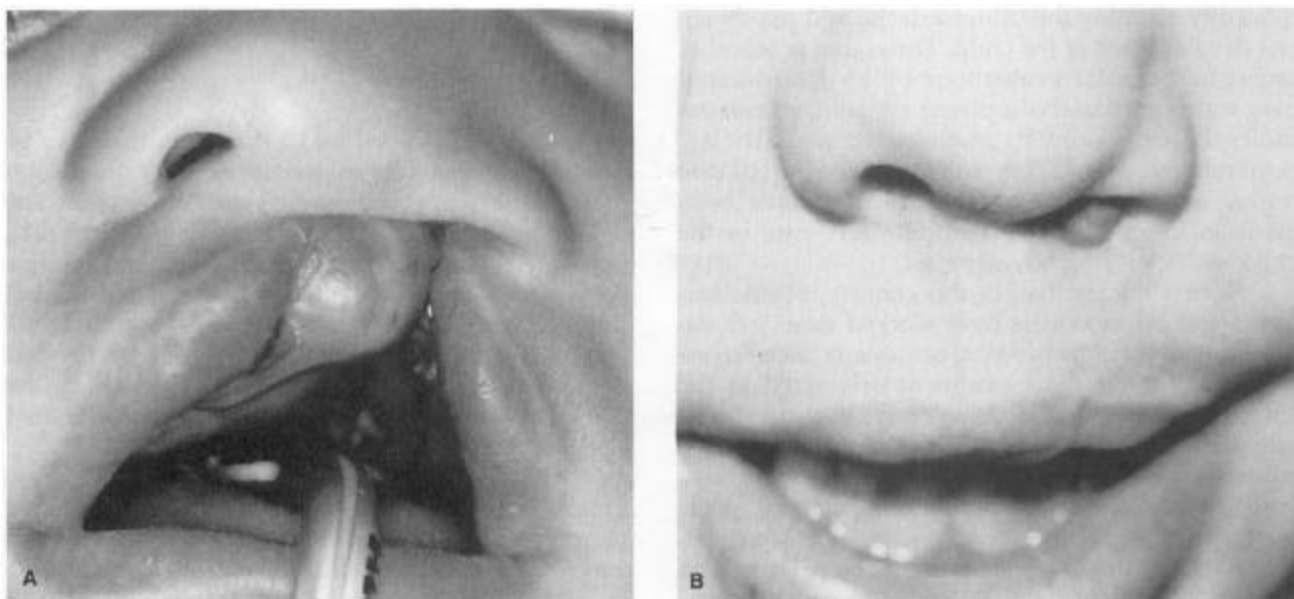


Fig 1 (A) Unilateral complete cleft lip, preoperative. (B) Postoperative repair.

#### TIMING OF REPAIR

Optimum surgical timing of cleft lip repair varies from surgeon to surgeon. In some institutions, unilateral cleft lips are repaired during the first 24 hours of life.<sup>25</sup> Proponents believe that the family will take home a "normal" looking baby with less difficulty feeding. If the babies are healthy and of normal weight, this early operation is generally successful. On the other hand, many surgeons delay the operation using the "rule of 10s"—10 weeks, 10 pounds, 10 g of hemoglobin—as a guideline for timing the surgery. By this time, the anatomical lip landmarks have become more apparent, there has been time to fully bond with the infant, and the parents have had time to adjust to the cleft and the operation and necessary rehabilitation to follow.

#### CLEFT LIP NASAL DEFORMITY

Now that plastic surgeons routinely achieve excellent aesthetic results after cleft lip repair, attention has been directed to improving results with the correction of the cleft lip nasal deformity. This deformity is characterized by a flattened alar cartilage, lateral displacement of the alar base, septal deviation to the normal side, and a deficient bony platform at the pyriform aperture that becomes more apparent as a child matures.<sup>26</sup> Most surgeons will perform correction at the initial lip repair; however, definitive correction is usually delayed until the early teens.

#### BILATERAL CLEFT LIP

The bilateral cleft lip is caused by failed fusion between the median nasal processes and the maxillary prominence on each side. The alar genua are splayed with flared nasi, the tip is broad, and the columella is short. The prolabium is lowered and wide and the distance between Cupid's bow peaks is widened (Fig 2). A single procedure to repair the bilateral cleft lip with the nasal deformity has been advised by some.<sup>6</sup> Others prefer to stage these repairs.<sup>6,23</sup> Timing depends on the surgeon and their individual clinical experience.

According to Mulliken, five basic principles guide the repair.<sup>27</sup> First is symmetry, followed by muscle continuity. Third is proper prolabial size and shape, then formation of the median tubercle from the lateral lip/vermillion mucosa, and last, positioning the alar cartilages to construct the nasal tip and columella.<sup>27</sup>

The surgical procedure basically proceeds in the following manner: The prolabial flap is incised and elevated. The alar base is separated from the pyriform aperture by an incision at the mucosal-cutaneous junction, thereby dissociating the alar bases from the lateral lip elements. The orbicularis muscle bundle is dissected out laterally until the bulge disappears. The vertical nasal tip and rim incisions are deepened.

Then mucosal flaps are elevated to reconstruct the nasal floor. This is the first one to be closed, and

**Table 2. Unilateral Cleft Lip Procedures**

Technique	Indications	Advantages	Disadvantages
Lip adhesion (Randall-Graham)	A preliminary adhesion operation used in difficult lip repair	<ul style="list-style-type: none"> <li>➤ Mold underlying distortions of the maxilla &amp; premaxilla</li> <li>➤ Possibly stretch out attenuated and displaced adjacent soft tissue</li> </ul>	<ul style="list-style-type: none"> <li>➤ Usually none</li> <li>➤ Resulting scar may be a nuisance in follow-up "creative sight repairs" e.g., Millard II</li> </ul>
Linear excision Rose-Thompson	Reserved for the most minor degrees of incomplete clefts	<ul style="list-style-type: none"> <li>➤ Easy operation (no small flaps)</li> <li>➤ Scar lies in a natural direction</li> <li>➤ No worries about future disproportionate growth</li> </ul>	<ul style="list-style-type: none"> <li>➤ Scar contracture</li> <li>➤ Sacrifice a lot of tissue</li> </ul>
Z-Plasty Triangular flap (Tennison)	Wide clefts where tissue deficiencies are most severe	<ul style="list-style-type: none"> <li>➤ Preservation of Cupid's bow</li> <li>➤ Produce natural fullness near vermilion-cutaneous ridge</li> <li>➤ Minimal tissue wasted</li> </ul>	<ul style="list-style-type: none"> <li>➤ Scar breaks up philtrum and may be quite noticeable</li> <li>➤ Tendency toward disproportionate growth (flap becomes longer)</li> </ul>
Rectangular flap (LeMesurier)		<ul style="list-style-type: none"> <li>➤ Produce natural fullness near vermilion-cutaneous ridge</li> <li>➤ Easier markings than triangular flap</li> <li>➤ Conserves natural vermilion</li> </ul>	<ul style="list-style-type: none"> <li>➤ Scar breaks up philtrum and may be quite noticeable</li> <li>➤ Tendency toward disproportionate growth (flap becomes longer)</li> <li>➤ Difficult to revise</li> </ul>
Rotation-advancement flap	Millard I: Most satisfactory technique for mild-moderate clefts	<ul style="list-style-type: none"> <li>➤ Highly flexible sight technique that allows modifications during the operation</li> <li>➤ Secondary revisions possible</li> <li>➤ Preserve natural lines-Cupid's bow and philtral dimple</li> <li>➤ Scar lies in a natural direction</li> <li>➤ Outward pout of lower portion of lip preserved</li> <li>➤ Disproportionate growth rate</li> </ul>	<ul style="list-style-type: none"> <li>➤ Difficult to design adequate lateral flap in wide complete clefts-it may be necessary to sacrifice too much vermilion producing noticeable asymmetry of Cupid's bow</li> </ul>
Millard	II:	<ul style="list-style-type: none"> <li>➤ Same as Millard I plus,</li> <li>➤ Provides for lengthening of the columella on cleft side and better correction of alar flare</li> </ul>	<ul style="list-style-type: none"> <li>➤ Same as Millard I but worse since more tissue needed from lateral lip flap to lengthen columella</li> </ul>

the alveolar clefts are closed (gingivoperiosteoplasty). These are opposed with interdermal sutures, and the alar bases are opposed with a "cinch" suture to reconstruct the alar sills. The tip of the prolabial flap fits into the top of the median tubercle (new) to complete the closure.<sup>26</sup>

**POSTOPERATIVE CARE**

Postoperative feedings of full-strength formula are administered with a catheter-tip syringe fitted with a small red rubber catheter for the first 10 days. Nippling should be avoided to minimize strain on the repaired muscle and skin sutures and to avoid trauma to the repaired velum. Velcro arm restraints are used to protect the repair from flailing hands and

fingers. Routine care can be resumed ~10 days postoperatively.<sup>6</sup>

**CLEFT PALATE**

Cleft palate surgery is directed to palatal closure with a technique and timing that produce optimal speech and minimize facial growth disturbances. Success is not the domain of any single method or protocol, but is the result of an experienced surgeon working with a team of cleft specialists and following solid principles.<sup>6</sup> The infant with isolated cleft palate is examined to determine whether there are manifestations of Pierre Robin syndrome (micrognathia, glossoptosis, and airway obstruction). If it is present, appropriate measures are instituted to main-

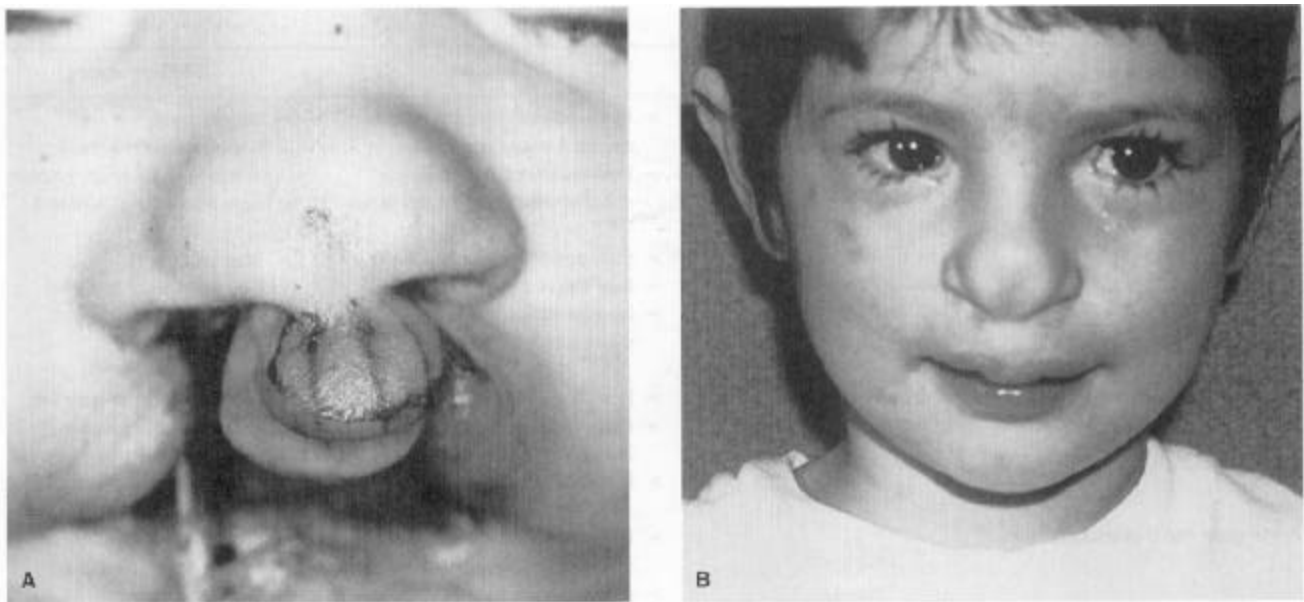


Fig 2 (A) Bilateral cleft lip, preoperative. (B) Postoperative repair.

tain an adequate airway, most commonly in the form of prone positioning. If the Pierre Robin sequence is not present, a passive palatal appliance can aid with feeding until palatoplasty is performed. In these patients, palatoplasty may be delayed for several months to ensure adequacy of the airway before closure<sup>6</sup>

The submucous cleft palate is defined by a triad of deformities: a bifid uvula, a notched posterior hard palate, and muscular diastasis of the velum.<sup>2</sup> Manifestations of a submucous cleft palate varies considerably. The majority of patients are asymp-

tomatic, although approximately 15% will develop velopharyngeal insufficiency.<sup>10</sup> Velopharyngeal incompetence correlates with short palatal length, limited mobility, and easy fatigability of the palate. Because most patients with submucous cleft palate remain asymptomatic, a nonoperative approach is recommended until speech can be adequately evaluated.<sup>6</sup>

Regarding unilateral cleft lip and palate, in our opinion, palatoplasty should be performed at 12 months of age, to correlate with speech development and a low likelihood of interfering with ultimate fa-

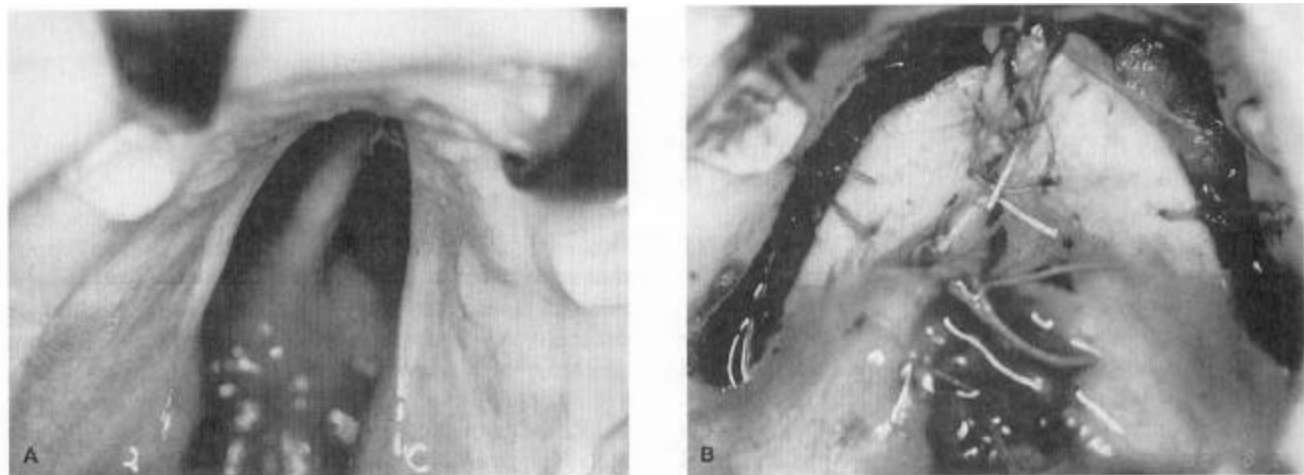


Fig 3 (A) Incomplete cleft palate, preoperative. (B) Postoperative repair.

cial growth.<sup>6</sup> Figure 3 demonstrates a typical Veau-Wardill-Kilner closure.<sup>6</sup> Levator muscles are dissected free from the oral and nasal mucosa, released from the palatal aponeurosis to the tensor tympanic tendon, and reattached in the midline in a normal anatomical position. The nasal mucosa is sutured to itself posteriorly and to the corresponding vomerine mucosa anteriorly. Mucoperiosteal flaps based on the greater palatine artery allow a tension-free closure on the oral side. In another technique, Z-plasty can be incorporated into the soft palate closure.<sup>6</sup>

#### COMPLICATIONS

**C**omplications of cleft palate repair include bleeding, respiratory obstruction, infection, dehiscence, and eventual fistula formation.<sup>21,23</sup> Significant bleeding is rare, but it may require a return to the operating room for exploration. Respiratory obstruction is also rare in the absence of excessive bleeding, but it may be life threatening. The airway should always be carefully monitored.

Palatal fistulas may present as asymptomatic holes or may cause such symptoms as nasal speech problems, difficulties with dental hygiene, or regurgitation of food.<sup>3</sup> Disturbances of facial growth may result from palatal surgery.<sup>6,23</sup> Decreased maxillary width and the resulting crossbite are common abnormalities in clefts and are managed by orthodontic maxillary expansion with a fixed appliance. Once expansion is completed, bone grafting is performed to correlate with the stage of canine development and to closely coordinate with planned orthodontic therapy.<sup>4</sup> Ideally, the roots of the canines are one quarter to one half formed.<sup>7</sup> This usually occurs when the patient is between 8 and 12 years old.<sup>23</sup> A plan of treatment is formulated on the basis of clinical examination, photographs, cephalometric studies, and dental models. Presurgical orthodontics is required to align dental arches and to eliminate crowding and dental compensations. In some cases, occlusal correction may require orthognathic surgery. The most common operation is a Le Fort I osteotomy, which allows satisfactory maxillary advancement. Because of palatal scarring related to palate repair, large advancements usually require an accompanying mandibular osteotomy and setback.<sup>14</sup> Surgery is delayed until adolescence to avoid recurrent malocclusion related to mandibular growth.

#### VELOPHARYNGEAL INCOMPETENCE

**A** normal velopharyngeal valving mechanism is needed for intelligible speech. If the coupling of the nasal and oral cavities is not precise, hypernasal-

ity, nasal emission, imprecise consonant production, and decreased vocal intensity (loudness) will result. These may be caused by either a structural defect or physiologic dysfunction. Gross tissue deficiency, disproportion of the oropharyngeal area, or neurogenic paresis of the velopharynx all can cause velopharyngeal incompetence. Individuals with hypertrophic adenoid tissue will exhibit hyponasality because of abnormal coupling of the oral and nasal cavities. The speech therapist must determine whether speech abnormalities are related to velopharyngeal incompetence, which can be treated surgically.<sup>6</sup>

The goal of surgery for patients with velopharyngeal incompetence is to provide an anatomical mechanism for functional speech. The design of the surgical procedure is based on the anatomy and the function of the patient as determined by a speech pathologist and by preoperative diagnostic evaluation. Dynamic studies of the pharynx by multiview videofluoroscopy or nasoendoscopy is indicated. This will provide information regarding anatomy and degree of medial excursion of the lateral pharyngeal walls during speech.<sup>6</sup> Clinical evaluations and tape recordings are obtained periodically, usually every 3 months for the first year, then annually. The sound spectrograph points to subtle changes in the acoustic signal and can be used to monitor objective speech characteristics.<sup>2</sup>

Velopharyngeal incompetence is managed by either pharyngeal flap or sphincter pharyngoplasty.<sup>7</sup> The sphincter is constructed from the posterior tonsillar pillars, including the palatopharyngeus muscle, which are sutured together with a small, superiorly based flap. The procedure achieves "static" and possibly "dynamic" reduction in the velopharyngeal port. Pharyngeal flaps may be based superiorly or inferiorly. Tissue is attached from the posterior pharyngeal wall to the soft palate to create a midline obstruction between oral and nasal cavities with two lateral ports. Hypernasality will persist if the flap is too narrow. Hypernasality may result if the flap is too wide, and mouth breathing or sleep apnea can occur.

Pharyngeal flap surgery includes longitudinal incisions through the mucosa and muscle down to the prevertebral fascia on each side of the posterior pharyngeal wall. A superiorly based flap is created and raised to the level of the palatal plane. An inferiorly based flap can also be cut. The flap is inset on the nasal side of the uvula.<sup>6</sup>

#### FETAL WOUND HEALING

**D**uring the past decade, many researchers have shown great interest in fetal diagnosis and open



fetal surgery.<sup>14</sup> Observations from this pioneering work indicate that the human fetus heals without scarring.<sup>14</sup> Many studies show distinct differences between fetal and adult skin wound healing, but it is unclear whether these differences are in the fetal cells themselves, in the fetal wound environment, or in a combination of the two.<sup>14</sup>

Numerous intrinsic and extrinsic differences exist between the fetus and adult that may dramatically influence wound healing. Fetal skin is continually bathed in warm, sterile amniotic fluid that is rich in growth factors and extracellular matrix components such as hyaluronic acid (HA) and fibronectin. The fetal external environment is sterile. Intrinsic differences include tissue oxygenation, because the fetus is profoundly hypoxic relative to the postnatal human. Another intrinsic difference between the fetus and the adult lies in the immune system. Histologically, there are very few granulocytes in fetal wounds, and these cells may not respond normally to chemoattractants. Experiments on fetal lamb wounds showed less acute inflammatory response than is seen in adult animals.<sup>14</sup> Because inflammation plays a key role in adult wound healing, this may be an important part of the unique fetal repair process. The inflammatory process does, however, increase in older fetuses and newborns.

The extracellular matrix in the fetus differs from that of the adult. The fetal wound matrix is rich in HA, which is a key structural and functional component of the extracellular matrix. Hyaluronic acid is laid down early in the matrix of both adult and fetal wounds, but the sustained deposition of HA is unique to fetal wounds and may provide the matrix signal that promotes healing by regeneration rather than by scarring. After the initial deposition of HA in adult wounds, hyaluronidase is produced, HA is removed, and the wound matrix is replaced by collagen. In the fetal wound, this is not the case and there is a prolonged HA-rich wound matrix. The fibronectin production is enhanced in the fetal wound.<sup>14</sup>

Amniotic fluid has also been evaluated as a possible mechanism for scarless healing in the fetus. The fluid contains high levels of HA, which may account for the mechanism by which HA is deposited in the extracellular matrix of fetal wounds.

Future studies may delineate the role of growth factors in cellular proliferation, interaction, and differentiation during fetal wound healing. Repair of adult wounds without scar is the ultimate goal. Unraveling the secrets of fetal wound healing may provide the blueprint for this ideal form of tissue repair

and the clues for the modulation of postnatal scar formation.

In conclusion, successful management of children with cleft lip/palate requires the close coordination of many subspecialties. This care is most effectively given through a multidisciplinary team.

## REFERENCES

1. Georgiade G. Textbook of Plastic, Maxillofacial, and Reconstructive Surgery. 3rd ed. Philadelphia: Lippincott Williams and Wilkins, 1997:220
2. McCarthy J. Plastic Surgery. Philadelphia: WB Saunders, 1990; 4:2437-2921
3. Moore KL. The Developing Human. 4th ed. Philadelphia: WB Saunders, 1988:170-206
4. Greene RM, Weston WM. Craniofacial Embryology. In: Cohen M (ed). Mastery of Plastic and Reconstructive Surgery. Boston: Little, Brown, 1994:459-470
5. Langman J. Medical Embryology. 7th ed. Baltimore: Williams & Wilkins, 1995:312-345
6. Grabb and Smith's Plastic Surgery. 5th ed. Philadelphia: Lippincott-Raven, 1997:223-279, 349-363
7. Ferguson Mark WJ. Palate development. Dev Suppl 1988;103: 41-60
8. Tessier P. Anatomical classification of facial, craniofacial, and laterofacial clefts. J Maxillofac Surg 1976;4:69-92
9. Diehl SR, Erickson RP. Genome scan for teratogen-induced clefting susceptibility loci in the mouse: evidence of both allelic and locus heterogeneity distinguishing cleft lip and cleft palate. Proc Natl Acad Sci USA 1997;94:5231-5236
10. Jones MC. Facial clefting. Etiology and developmental pathogenesis. Clin Plast Surg 1993;20:599-606
11. Shields Ed, Bixler D, Fogh-Anderson P. Cleft palate: a genetic and epidemiologic investigation. Clin Genet 1981;20:13-24
12. Houdayer C, Bahuau M. Orofacial cleft defects: inference from nature and nurture. Ann Genet 1998;41:89-117
13. Online Mendelian Inheritance in Man, OMIM (TM). Center for Medical Genetics, Johns Hopkins University (Baltimore, MD) and National Center for Biotechnology Information, National Library of Medicine (Bethesda, MD), 1997. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>
14. Longaker MT, Adzick NS. The biology of fetal wound healing: A review. Plast Reconstr Surg 1991;87:788-798
15. Georgiade G. Textbook of Plastic, Maxillofacial, and Reconstructive Surgery. 2nd ed. Philadelphia: Lippincott Williams and Wilkins, 1992:299-306
16. Fogh-Anderson P. Inheritance of Harelip and Cleft Palate. Copenhagen: NYT Nordisk forlog, Arnold Busck, 1942
17. Wilson MEAC. A 10-year survey of cleft lip and cleft palate in the southwestern version. Br J Plast Surg 1992;25:224-228
18. Fraser FC. The genetics of cleft lip and palate. Am J Hum Genet 1970;22:336-352
19. Fogh-Anderson P. Incidence and etiology. In: Edwards M, Watson ACH (eds). Advances in the Management of Cleft Palate. Edinburgh: Churchill Livingstone, 1980
20. Neel JV. A study of major congenital defects in Japanese infants. Am J Hum Genet 1958;10:398

21. Bentz Michale M. *Pediatric Plastic Surgery*. East Norwalk, CT: Appleton and Lange, 1998:47-128
22. Habib Z. Factors determining occurrence of cleft lips and cleft palate. *Surg Gynecol Obstet* 1978;146:105-110
23. Converse JM. *Reconstructive Plastic Surgery*. 2nd ed. Vol 4. Philadelphia: WB Saunders, 1977:2016-2045
24. Fara M. The importance of folding down muscle stumps in the operation of unilateral clefts of the lip. *Acta Chir Plast (Praha)* 1971;13:162-169
25. Bardach J. In: *Unilateral Cleft Lip*. In: *Mastery of Plastic and Reconstructive Surgery*. Cohen M (ed). Boston: Little Brown, 1994:548-565
26. LaRossa D, Donath G. Primary nasoplasty in unilateral and bilateral cleft lip nasal deformity. *Clin Plast Surg* 1993;20:781-791
27. Mulliken JB. Principles and techniques of bilateral complete cleft lip repair. *Plast Reconstr Surg* 1985;75:477-487