

Paparella III: Section 2: Disorders of the Head and Neck

Part 2: The Oral Cavity

Chapter 13: Maxillofacial Clefts

Leslie Bernstein

The overall incidence of maxillofacial clefts ranges from 1 in 500 to 1 in 700 births, being somewhat higher in Japan and lower in blacks in the United States. When a history of clefts exists in the family, the incidence is much higher. In recent years, because of the ready availability of birth control measures and genetic counselling, the incidence of these clefts has dropped somewhat.

Much progress has been made in the management of maxillofacial clefts over the past 30 years owing to contributions made by various disciplines to the overall welfare of these individuals. Consequently, it is usually possible to assure the parents of a newborn infant with a cleft lip and palate that with time and proper management, the child should eventually look normal and function normally in society.

The complexity of the deformity involved in patients with maxillofacial clefts necessitates a multidisciplinary approach to its rehabilitation. In most cases, the treatment extends over a span of 18 years, from birth until the final secondary aesthetic operation, and even for life in some instances, as when dental obturators are part of the management. Yet, in spite of the vast progress that has been made, many modifications in treatment are still being introduced because results are still far from ideal. A new technique requires several years before its results can be adequately assessed. The accumulation of data and long-term follow-up care are thus very necessary. It is therefore important that patients with maxillofacial cleft be treated in special centers in which the services of a team of specialists are available and the end results of management may be critically evaluated.

Early Management

Emergency Treatment

The syndrome of cleft palate, micrognathia, and glossoptosis (commonly known as Robin's syndrome, although it was first described by Fairbairn in 1846 (Gorlin and Pindborg, 1964)) may present as a neonatal respiratory emergency. To overcome blockage of the pharynx by the tongue, which cannot be accommodated within the small mandibular arch, the tongue must be displaced forward. One way of achieving this is to suture it into the mandibular labial sulcus and to keep it there for several months until the mandible has reached normal proportions. This operation, first described by Shukowsky in 1910 (Gorlin and Pindborg, 1964) and then reintroduced by Douglas in 1946, has proved adequate in the management of such cases. Postoperatively, the mandible grows rather rapidly, reaching normal proportions in 6 to 12 months.

Counseling of Parents

Early psychological counseling of the parents is of utmost importance. It is difficult to imagine the shock of parents, who have been expecting a perfectly normal baby, when they are confronted with what must appear to them to be a gross monstrosity. Reassurance is not only kind, it is mandatory. It is important to tell the parents that proper management over the years will rehabilitate their child into a well-functioning and aesthetically acceptable member of society. Frankness about our ignorance of the cause of the deformity is necessary to avoid a sense of guilt in the parents, a situation that may seriously interfere with rehabilitation later on. The confidence and cooperation of the parents are essential in the long-term management required by most of these patients. An explanation of the deformity and an outline of the proposed treatment will enable the parents to better appreciate the problems involved in the overall management of the child.

It is also surgeon's moral duty to explain the effect of heredity in these malformations. When such a hereditary link is obvious, the parents may well be advised to adopt additional children.

Feeding

Early management of the infant with cleft palate is primarily concerned with proper methods of feeding. Breast feeding is quite impractical and should not be attempted. A very satisfactory way of overcoming nasal regurgitation is to place the child in a lateral sloping position and deposit the formula in the buccal cavity by means of a rubber syringe. Another useful means for introducing nourishment is the restaurant-type plastic ketchup dispenser, which is equipped with a small nozzle that can dispense formula in desired amounts when the bottle is gently squeezed. Feeding may also be accomplished with a rubber nipple with an enlarged hole. After boiling, such a nipple responds well to the gentle pressure of the tongue, and the child is able to suck without fatigue.

In feeding all infants with cleft palates, sufficient time must be allowed for swallowing. Because of the increased amount of air that is ingested in this manner, burping should be more frequent than with a normal infant. It is notable that when the parents participate in the feeding and care of the infant, they have a deeper appreciation of the problems confronting the surgeon. Invariably, after the lip has been repaired, the parents experience a more genuine acceptance of the child, and the physician finds that he or she is dealing with a happier family.

Maxillary Orthopaedics

Widely separated or poorly aligned maxillary segments may be repositioned by means of removable prosthetic appliances, a technique known as maxillary orthopaedics. Since repair of the cleft lip will involve reconstruction of the labial muscular sphincter, which, in turn, will approximate the alveolar segments anteriorly, it is desirable that these segments be in proper alignment prior to this operation.

The services of a dental specialist (an orthodontist or a prosthodontist) who is familiar with such techniques are necessary for this part of the management. The appliance is made in sections that correspond to the cleft segments and are joined together by a reciprocating screw or screws. By this means, the separate segments of the appliance may be moved closer together or spread wider apart as necessary. Another appliance, although a single piece, is actually made of the component parts of the cleft, which are fused together in a progressively favorable position at intervals of days or weeks. By sucking on the appliance, the infant promotes the gentle repositioning of the segments.

Reapproximation of the maxillary segments simultaneously advances the lateral lip segments closer together, thus facilitating the lip repair. In infants with bilateral clefts of the palate, in whom the premaxilla may be somewhat protrusive, maxillary orthopaedics may reduce the distance between the segments to allow closure of the bilateral cleft lip with little tension. An extra benefit of these orthopaedic appliances is that they aid the infant in feeding. It is remarkable that when the appliance is removed, the infant will frequently cry until it is replaced.

Maxillary orthopaedics should be started as early as possible to take advantage of the rapid growth that occurs in the first few months of life. At this age, effective repositioning may be expected in 2 to 8 weeks, during which period the infant will have gained sufficient weight to permit easier approximation of the lip segments. Once the lip is repaired, the reconstitution of the orbicularis oris muscle sphincter serves as a permanent splint for the newly positioned alveolar segments.

Lip Adhesion Procedure

In some instances, the same effect may be achieved recreating the orbicularis oris muscle sling of the lips with a relatively minor surgical procedure, commonly called the lip adhesion operation. The margins of the cleft are bared by the excision of an epithelial strip from each edge of the labial cleft, and the raw surfaces are sutured together. Alternatively, the epithelial covering of the cleft is fashioned into an anteriorly based (skin) flap on one side of the cleft and a posteriorly based (mucosal) flap on the other side. The flaps are then sutured together. Definitive repair may be deferred until the child is considered old enough. This kind of management usually leads to a satisfactory overall result.

Surgery of Cleft Lip

Historical Background

Numerous procedures have been described for repair of congenital clefts of the lip. From the time of Celsus until well into the 19th century, the operation consisted of excision of the margins of the cleft and a crude approximation of the raw edges. In about 1830, Dieffenbach (1845) introduced the lateral undermining incision to allow the flaps to slide together with less tension. Mirault is generally credited with having been the first (in 1844) to bring a flap from the lateral segment across the lower margin of the cleft (McDowell, 1966). In the early 1930s, Blair and Brown (1930) applied Mirault's principle with better cosmetic results.

In 1880, Jalaguier introduced the use of a triangular flap from the lateral lip segment that provided adequate lip height and a staggered scar (cited by Washio, 1968). Hagedorn devised a similar operation in 1884 but used a square flap instead. Through the years, many improvements and modifications in the various techniques have been made, largely through evaluation of postoperative results and self-criticism by the surgeons. Years later, LeMesurier (1949) popularized Hagedorn's operation, and Tennison (1952) and Randall (1959) reintroduced Jalaguier's technique in simplified form.

One of the most outstanding contributions to improved cleft lip repair in recent years has been the rotation-advancement operation of Millard (1957), a precursor of which had already been introduced in 1901 by Giralde's (Thompson, 1912). This is an excellent procedure for correction of partial and narrow clefts. When combined with various refinements (Millard, 1964, 1968) a most pleasing result can be obtained. However, this operation requires artistic judgment, experience, and a high degree of skill. When it is used to correct wider clefts, the main scar often has a tendency to contract so that a short lip may result. To help overcome the difficulties that may be encountered in patients with wide clefts, Bernstein (1970) modified Millard's rotation-advancement operation by incorporating a small triangular flap from the lateral segment into the bottom of the rotation flap.

Timing of the Operation

Many surgeons feel that surgery should be delayed for at least 10 weeks following birth. By this time the lip structures have increased in size, making the operation easier and providing the surgeon with better tissues with which to work. There is little necessity for an early operation, since it can easily be delayed without seriously affecting the health of a newborn child. In general, this operation should be postponed until the infant is gaining weight steadily and shows a normal blood count and an acceptable hemoglobin level. Ideally, it may be prudent to follow the minimum "rule of ten" - the patient should be at least 10 weeks of age, weight at least 10 pounds, and have at least 10 gm of hemoglobin.

Operative Technique

The operation is performed under general anaesthesia. It is preferable to schedule it as the first case in the morning when the infant is not yet very hungry; an early schedule also precludes the possibility of fatigue. Assuming that the operation begins at 8 AM and the child is on a 4-hour feeding schedule, the 4 AM feeding is omitted.

The child is placed at the head of the table with his or her head slightly extended. The eyes are taped shut, and the exposed part of the face is cleansed with antiseptic soap. The endotracheal tube is carefully taped in the middle of the lower lip to provide the least amount of lateral distortion; a moist pack around the endotracheal tube is recommended to prevent aspiration of blood. The operation is performed from the head of the table.

In this meticulous surgery, it is important to use instruments that are designed for fine detailed work. An atraumatic technique with minimal manipulation of the tissues is important, and wound margins should be handled with sharp skin hooks in order to avoid unnecessary trauma. Fine scalpels, small skin hooks, fine needle holders, small hemostats, a battery-powered ophthalmic cauterizing device, and a fine pen constitute the basic armamentarium.

Also necessary are fine-pointed calipers and a measuring rule. A weak local anesthetic solution with a vasoconstricting agent is mandatory for hemostasis. A small suction tip and moist cotton-tipped applicators are used to keep the small surgical field free of blood and to provide a clear field of vision for the surgeon.

The Unilateral Cleft Lip

This operation may perhaps offer the first opportunity for a detailed examination of the lip segments. Prior to distorting the tissue with the local anesthetic solution, it is important to identify and mark with gentian violet solution the following landmarks: the bottom of Cupid's bow, the midpoint of attachment of the columella to the lip, the attachment of the alae to the lip, and the point at which the vermilion border of the cleft segment starts to become attenuated - this is usually identified by the sudden termination of the white ridge that normally exists at the vermilion-cutaneous junction. These points are tattooed with a nonsiliconized hypodermic needle to prevent obliteration during the operation. It is particularly important to mark exactly the vermilion border or the mucocutaneous ridge.

Although it may be true that any of the operations in vogue today will produce a satisfactory result in good hands, certain operations have advantages over others. The surgeon is encouraged to acquaint himself or herself with the various operative procedures now used to avoid creating a situation in which the lip is made to fit the procedure and not vice versa. Furthermore, familiarity with the various procedures may lead to the development of further beneficial modifications. Because there are several different operative techniques available, and the choice depends upon the preference of the surgeon and the type of case under consideration, only the principles of the operative procedure will be discussed here.

Ideally, the operation should be designed to meet the following criteria: (1) accurate approximation of skin, muscle, and mucosa; (2) an inconspicuous scar; (3) symmetric lip length; (4) creation of a symmetric Cupid's bow; (5) creation of a philtrum dimple; (6) slight eversion, or pouting, of the lip; (7) symmetric nostrils; (8) a symmetric columella; (9) creation of a labial sulcus; and (10) easy adaptability of the procedure to various kinds of cleft deformities.

It is essential that the tissue be cut sharply, with clear incisive strokes of the knife held at right angles to the skin's surface. After the skin has been incised squarely, the knife may be slanted toward the cleft to preserve as much muscle and mucosa as possible. Bleeding points may be grasped with very small hemostats, which are left on for several minutes. In most instances, this will suffice to control hemorrhage, obviating the need for buried sutures. Nevertheless, a battery-powered ophthalmic cautery may be desirable to control specific bleeding points. When ligation of the coronary vessels is needed, 5-0 chromic catgut may be used.

In preparing the flaps, it is often necessary to release the lip segments from their attachments to the underlying bone. This may not be required on the normal side in every case, however. When needed, only a minimal amount of undermining should be done, just sufficient to permit approximation of the flaps. On the cleft side, care should be taken to avoid the infraorbital nerve, which emerges at a relatively low level in infants. In patients with very wide clefts, the ala on the cleft side may have to be detached completely from the

maxilla in order to facilitate approximation of the lip segments.

An underlying cleft of the alveolar process may be closed simultaneously with repair of the lip. The author favors the technique described by Skoog (1967), which creates a partial sleeve from the mucoperiosteum that lines the alveolar cleft. The anterior defect of this sleeve is closed by creating a medially based flap of periosteum from the anterior part of the maxilla and rotating it 180 degrees to bridge the alveolar gap. The space between this maxillary periosteum and the mucoperiosteum that has been sutured within the cleft may be filled with small pieces of an absorbable gelatin sponge (Gelfoam) or with blood clot.

Fine chromic catgut sutures are used for the muscle layer, and 6-0 nylon is used for the skin closure. It is customary to close the mucosal surface with fine chromic sutures. Care must be taken with placement of the sutures, and they should not be tied tightly. The layer of skin sutures should suffice merely to approximate the skin edges.

Dissection of the asymmetric tip cartilage of the nose on the cleft side, simultaneously with lip repair, should be undertaken only by experienced surgeons; however, a certain degree of repositioning is being done more and more frequently. When the tip cartilage has been freed, a through-and-through mattress suture, tied over bolsters, is useful for coapting the tissue layers in the new position.

A lip dressing is not necessary, but the line of suture may be covered with bland ophthalmic ointment. Before removing the endotracheal tube, the pharynx should be suctioned for any residual blood clots. This is repeated on removal of the tube. The child is then taken to the recovery room and kept in a croupette for 24 hours. The infant's arms are restrained by splinting at the elbows, and he or she is prevented from turning onto the face by pinning the arms to the bedsheets in the supine position.

Postoperative feeding is given by some form of syringe, as described earlier. The suture line is cleansed after meals with hydrogen peroxide, and the ophthalmic ointment is reapplied. Intermittent skin sutures may be removed on the third or fourth postoperative day, retaining the sutures at key points until the fifth postoperative day. The sutures should be removed with meticulous care. The child's head must be restrained, either by a firm pair of hands or by means of a wide adhesive bandage that straps the head to the table. The child may require a sedative. Conversely, removal of the sutures may be accomplished equally well if done just before a meal when the child is hungry. A small wad of gauze soaked in sugar water and placed in the mouth on the side away from the cleft will often suffice to distract the attention of the hungry infant. After the sutures have been removed, the wound edges may be supported with adhesive strips for several days.

The Bilateral Cleft Lip

Best results are usually obtained in the patient with a bilateral cleft lip by repairing one side at a time, with an interval of about 3 months between operations. Rarely, such management may not be feasible because of a protruding premaxilla. In planning treatment, the following principles are recommended:

1. Maxillary orthopedic procedures should be instituted to improve the position of the three components of the alveolar arch. In patients with bilateral cleft lip and palate, the purpose of the orthopedic appliance is to hold the premaxilla back while opening the maxillary segment enough to accommodate the premaxilla. If the maxillary segments are not collapsed behind the premaxilla, the appliance will promote enough forward movement of these segments to meet with the premaxilla, which is being held back.

2. The prolabium should be used to form the full vertical length of the middle of the lip, assuming it to be the total contribution of the philtrum. At a cursory glance, the prolabium appears to be entirely inadequate for the reconstitution of the full vertical height of the middle of the lip. Because it has a narrow attachment, the natural elasticity of the skin causes it to contract toward the columella. Moreover, because it has little or no muscle, it is thinner than the lateral lip segments.

3. The curved vermilion-cutaneous border at the bottom of the philtrum should provide the central convexity of Cupid's bow.

4. The midportion of the vermilion border should be built up with vermilion-muscle flaps taken from the lateral lip segments.

5. The surgeon doing bilateral lip repair should take into consideration the almost inevitable need for revision at a later date. For this reason, as much tissue as possible should be preserved to have it available for future revision.

As with the unilateral cleft, a partial defect is the simplest form of unilateral cleft lip. As the degree of deformity increases, it is usually compounded by the increased involvement of the underlying bony framework and is still further aggravated by a protrusive premaxilla.

In partial clefts and in complete clefts in which the premaxilla is not too prominent, the rotation-advancement method may be applied in two separate operations. There are distinct cosmetic advantages with this operation. In patients with a moderately protrusive premaxilla, the straight line repair of Veau may be performed in one stage but preferably in two, since the latter gives better cosmetic results.

A certain amount of spontaneous recession of the premaxilla may be expected postoperatively owing to changes in the muscular forces acting on the palatal segments. Thus, in patients with a markedly protrusive premaxilla, a straight line coaptation of the lip segments may be attempted, its main object being to reconstitute the orbicularis oris muscle sling in order to achieve some improved repositioning of the segments.

The Shultz (1946) method may be used in the patient with a protrusive premaxilla to close bilateral clefts of the lip when the lip segments may have to be approximated under some degree of tension. When indicated, this is a satisfactory method of achieving both immediate bilateral lip closure and fairly rapid repositioning of the premaxilla, provided that the maxillary segments are sufficiently far apart to receive it. This operation also provides a labial sulcus over the premaxilla.

In very rare cases, surgical recession or resection of the premaxilla may be indicated. This should be done only when it is quite impossible to effect lip closure otherwise. Because of the danger of interference with midfacial growth when such a procedure is performed so early in life, great caution should be exercised in selecting patients for such an operation.

Surgery of Cleft Palate

The history of the various methods of surgical closure of the cleft palate is a long and fascinating story, and its details are beyond the scope of this chapter. In 1861, von Langenbeck (Dorrance, 1933) introduced the technique of creating mucoperiosteal flaps through lateral incisions; in essence there were two bipediced flaps freed from their underlying bone and sutured in the midline. Because many of these patients showed shortening of the palate postoperatively, in 1895 Smith of Nashua, New Hampshire devised a method of repositioning the soft palate by means of four flaps, and Ganzer introduced the three-flap retrodisplacement operation in 1920. Modern palatal surgery embodies the basic principles as introduced by von Langenbeck, Smith, Ganzer, and numerous other contributors, each of whom added specific refinements.

The ideal cleft palate operation should achieve the following results: (1) closure of the cleft of the palate in one operation; (2) elongation of the palate, or at least prevention of its shortening postoperatively; (3) preservation of the normal function of the velum; (4) achievement of adequate velopharyngeal competence; (5) production of normal speech; (6) normal function of the auditory tubes; (7) a normal masticatory apparatus without malocclusion; (8) minimal interference with underlying bone growth; (9) avoidance of facial deformity; and (10) no interference with normal nasal physiology.

Timing of Operation

In determining the optimal timing of the cleft palate operation, the surgeon must take into consideration the effect of such timing on speech on the one hand and on underlying bone growth on the other hand. Several well-documented studies have shown that if repair is carried out after the deciduous dentition has erupted, retardation of postoperative facial growth is minimal. Conversely, it is generally agreed that better speech patterns result if the operation is done at an early age. Thus, in order to obtain the best results on both counts, it is quite reasonable to schedule the operation between the ages of 24 and 36 months (Bernstein, 1968).

Operative Technique

As with the lip operation, the child should be in good health and should have an adequate hemoglobin level and a normal blood count. The patient should be in the Rose position and should be given endotracheal anesthesia. Moist gauze is loosely packed around the endotracheal tube in the hypopharynx to prevent aspiration of blood. Various mouth gags designed specifically for this operation are available.

The operative technique should incorporate the following principles:

Hemostasis. This is greatly aided by infiltrating the operative field with a weak local anesthetic solution containing a vasoconstricting agent. Bleeding points are controlled with weak electrocautery.

Conservation of Tissue. Meticulous care is necessary in the handling of tissue, and crushing must be avoided at all times. Fine skin hooks should be used instead of forceps. No tissue is ever discarded. Adequate mobilization of flaps is necessary, and strangulation of tissues by tight sutures should be avoided to prevent tension and necrosis along the line of repair.

Mucosal Coverage of Cleft. To minimize postoperative contraction and to effect a stronger repair line, it is necessary to cover the cleft with mucosa on the nasal as well as on the oral surfaces. The nasal flap may be augmented at the expense of the oral mucosa by placing the oral incision 2 to 3 mm lateral to the cleft. The nasal mucosa is then freed from its bony attachment to facilitate its medial advancement.

In patients with unilateral clefts, the nasal mucoperiosteum on the cleft side must be sutured to the mucosal flap covering the vomer. With bilateral complete clefts and with clefts of the secondary palate, the vomerine flaps should be used in this manner whenever possible.

Retrodisplacement of Velum. Closure of the cleft with a simple approximation of the mucoperiosteal flaps will lead to shortening of the palate on healing. To avoid this and to compensate for the congenital underdevelopment of the tissues, the soft palate musculature should be freed from the posterior edge of the bony palate; however, the nasal mucosa is kept intact at this level. This facilitates both retrodisplacement and medial advancement of the velum. The new position of the velum is maintained by the V-Y three- or four-flap technique. In patients with wide clefts or very short palates, this maneuver will be greatly aided by freeing or cutting the greater palatine neurovascular bundle.

At the conclusion of the operation, the surgical field should be completely dry. The pharynx should be aspirated of any blood clots, and this procedure should be repeated after the endotracheal tube has been removed. The patient is taken to the recovery room and is placed in a croupette for 24 hours. He is kept on a liquid diet by cup feeding until healing has taken place, some 3 weeks later. Arm restraints are used to prevent the child from tugging on the ends of the sutures or from placing a potentially harmful object in the mouth. The patient is usually discharged from the hospital a week after the operation.

Further Management

Speech Therapy. Speech therapy is generally avoided for approximately 1 year after the repair of the cleft palate, since it takes about that long for the scar tissue to settle. With speech therapy and a well-controlled environment, approximately 60 to 80 per cent of patients with repaired cleft palates should be able to produce satisfactory speech with good velopharyngeal competence. In patients in whom velopharyngeal competence is not achieved in spite of intensive and prolonged speech therapy, a surgical procedure or a dental obturator may be indicated to diminish the velopharyngeal aperture.

Otitis Media. The incidence of nonsuppurative otitis media is extremely high in children with cleft palates. It is generally accepted that the cause is malfunction of the auditory tube. However, all of the reason for this kind of malfunction are not clearly understood because satisfactory repair of the palate does not automatically restore normal tubal function.

Except for instances of suppurative otitis media, attention to the ears of babies with cleft palates should be first given at about 10 to 12 months of age, when the babble pattern of speech begins. If nonsuppurative otitis media is encountered, a myringotomy should be performed, and a ventilating tube should be inserted through it. Thereafter, the ears should be examined at intervals of about 6 months. The procedure of myringotomy and insertion of tubes is repeated periodically as indicated; however, the need for it diminishes with growth and usually peters out toward the end of the first decade of life.

Unless a pharyngeal flap operation is anticipated, adenoidectomy is deemed inadvisable in these patients because of the immediate deterioration of speech. Although a child with good speech usually adjusts quite easily to the infinitesimally slow physiologic regression of the adenoid pad, it is expecting too much of him or her to compensate for the sudden removal of this tissue. Accordingly, to maintain existing good speech in children with cleft palates, treatment of otitis media should not include adenoidectomy.

Orthodontics. A rather high proportion of patients with complete clefts of the palate will eventually require various degrees of orthodontic management. This is usually instituted at the time of mixed dentition, and such patients should be handled by orthodontists who are well acquainted with the problems of the patient with a cleft palate.

Secondary Problems in Maxillofacial Clefts

Velopharyngeal Incompetence

Twenty to 40 per cent of patients who are operated on for clefts of the palate have velopharyngeal incompetence. It is also frequently found in patients with submucous clefts. The choice of treatment modality may be decided (Bernstein, 1967b) depending on the amount of incompetence, as determined by physical examination, speech results, oral breath pressure ratios, and lateral pharyngeal radiographs. Treatment may consist of a dental obturator designed to lift the velum to a higher plane in order to assist its articulation with the posterior pharyngeal wall; however, the value of this device is rather limited.

Surgical closure consists of either the creation of a pharyngeal pad or flap or the injection of polytetrafluoroethylene (Teflon) paste into the posterior pharyngeal wall.

The timing of these procedures is decided in conjunction with the speech pathologists. The creation of a pharyngeal pad may be done at any time, when it is established that the velopharyngeal gap is minimal but not correctable with speech therapy alone. This undertaking consists of obtaining a lateral radiograph during maximal velar closure and determining the site of augmentation. Excellent results have been obtained by placing an implant of radiated rib cartilage or demineralized rib bone over the atlas prominence.

The pharyngeal flap operation, or velopharyngoplasty, is usually undertaken when the patient is about 6 or 7 years of age, after an adequate period of intensive speech therapy and after a full evaluation of the extent of velopharyngeal incompetence. This operation may also be indicated in cases of postoperative breakdown of a repaired cleft of the velum. In younger children, and in those of slight build, it may occasionally be imperative to perform a tracheostomy postoperatively because of edema in the hypopharynx. Once the decision for a pharyngeal flap procedure has been made, it is prudent to remove enlarged adenoids about 6 to 12 weeks prior to the scheduled date of the velopharyngoplasty.

The operation is done with the patient in the Rose position with endotracheal intubation. A weak anesthetic solution containing a vasoconstrictor is infiltrated into the posterior pharyngeal wall and the free edge of the palate. A superiorly based flap including most of the width of the posterior pharyngeal wall and extending from the plane of the velum almost to the level of the tip of the epiglottis is outlined. The mucomuscular flap is freed from the underlying alar fascia with blunt scissors dissection.

The free edge of the velum is split horizontally between the oral and nasal surfaces, thus creating the recipient bed. This bed extends laterally about halfway down the posterior pillar of the fauces, leaving adequate lateral portals for nasal respiration.

A modification of this operation was recently reported by Bernstein (1975). To prevent the pharyngeal flap from becoming excessively tubelike and thus becoming narrow and less effective in some cases, a secondary flap is created to partially cover the raw surface of the larger flap. Because the lesser flap is fashioned from the posterior edge of the nasal surface of the soft palate, the pharyngeal flap must be anchored to the velum along a more advanced recipient bed. Bernstein believes that this makes the flap more efficient.

A certain degree of temporary neck rigidity is not an unusual postoperative phenomenon. Swallowing is rather difficult but should be vigorously encouraged. The patient is discharged from the hospital after about a week and is kept on a liquid diet for about 3 weeks. Because of edema, the lateral airway portals may be closed for several weeks postoperatively. Improvement in speech is frequently apparent soon after the operation, but no speech therapy should be given for about 3 to 6 months.

Fistulas

Apart from velopharyngeal incompetence, a wide nasolabial fistula may also be responsible for escape of air during speech as well as spillage of food into the nose. In such instances the fistula may have to be closed using local mucoperiosteal flaps. In some institutions, this procedure is combined with the insertion of a bone graft.

Oronasal fistulas usually result from partial breakdown of the repair line, except for the very narrow fistulas in the region of the alveolar process. Symptomatic fistulas may be closed surgically, or they may be covered with a dental appliance.

Aesthetic Problems

In addition to velopharyngeal incompetence, aesthetic problems are present in most secondary operations. Of them, problems with the nose are often of high priority. Because of the frequent need for septal reconstruction in unilateral clefts of the palate, the external nose in these patients is almost always asymmetric.

If the bony pyramid requires reduction, it is best to delay rhinoplasty until the patient is well into the teenage years. However, a soft tissue rhinoplastic procedure may be undertaken earlier. The common deformity specifically consists of a distorted and often underdeveloped tip cartilage on the cleft side, the cartilage being asymmetrically placed in relation to the one on the opposite side. Often, distortion of the nostril is present also, especially in the form of a weblike fold across the apex of the naris and a caudal displacement of the rim.

The bilateral cleft nose, in contrast, invariably presents with a short columella and wide flaring nares. A very satisfactory method of elongating the columella has been described by Millard (1958), but this should be done only when the quadrilateral septal cartilage begins to exert pull on the short columella.

Other secondary procedures frequently performed on children with cleft lips include revision of the lip scar, realignment of the vermilion-cutaneous border, and augmentation of the vermilion substance of the lip (Bernstein, 1967a). If the lip is short on the cleft side and if a "whistling" deformity is present, the scar revision should include a lengthening procedure. Some of the older lip operations did not preserve the Cupid's bow, and in such cases it may be reconstructed by advancing the vermilion-cutaneous border into a symmetric pattern of the bow. Deficiencies at the free border of the vermilion may be corrected by rolling out the mucous membrane of the inside of the lip.

The labial sulcus in bilateral clefts is usually nonexistent or very shallow as a result of attachment of the prolabium to the premaxilla. This ties the lip down, necessitating the creation of a sulcus to allow the lip to move freely. The sulcus may be lined with a split-thickness skin graft or with dermis; alternatively, the newly created space may be maintained by means of a dental obturator until epithelialization takes place in about 14 days.

In some patients with bilateral clefts, the upper lip may be short and tight, either because of underdeveloped tissues or because of aggressive surgery, or both. Consequently, there may be a marked discrepancy between the two lips. The condition may be greatly improved by augmenting the upper midportion of the lip with a triangular full-thickness flap from the lower lip - the Abbe operation.