Textbook of Oral and Maxillofacial Surgery

Gustav O Kruger

(The C V Mosby Company, St Louis, Toronto, London, 1979)

Fifth Edition

Chapter 21

Cleft lip and cleft palate

James R Hayward

The congenital deformities of cleft lip (cheiloschisis) and cleft palate (palatoschisis) have been known to afflict man since prehistoric time. Efforts to correct these abnormalities have evolved over the centuries with increasing success as scientific knowledge has advanced. It will be seen that oral clefts involve complex long-range treatment and appear with sufficient frequency to constitute a public health problem. Some form of cleft lip and cleft palate occurs in one out of every 800 live births. Combined clefts of the lip and palate are more frequent than the isolated involvement of either region. With lack of complete knowledge concerning etiology, effective preventive measures are not available to eliminate this deformity. The psychological and socioeconomic handicap of oral clefts may be severe. It is a deformity that can be seen, felt, and heard and constitutes a crippling affliction. Facial deformity with cleft lip involves the structures of the lip and the nose. Further skeletal facial deformity is seen in some forms of cleft palate. The most severe handicap imposed by cleft palate is an impaired mechanism preventing normal speech and swallowing.

The zones involved by common oral clefts are the upper lip, alveolar ridge, hard palate, and soft palate. In a useful classification the normal position of the nasopalatine canal divides cleft of the lip and alveolar ridge (primary palate) from those of the hard and soft palate (secondary palate). Slightly more than 50% are combined clefts of the lip and palate. About one fourth of this number are bilateral. The isolated clefts of the lip and palate constitute the balance of the varieties seen. Clefts of the lip are more frequent in males, whereas isolated clefts of the palate are more frequent in females. Lip cleft involvement is more frequent on the left side than on the right. These phenomena lack explanation, and the underlying cause of the deformity is incompletely understood. The failure of union of the parts that normally form the lip and palate occurs early in fetal life.

Embryology

The oral cleft problem occurs between the sixth and tenth week of embryo-fetal life. A combination of failure in normal union and inadequate development may affect the soft tissue and bony components of the upper lip, alveolar ridge, and hard and soft palates. The face of the fetus undergoes rapid and extensive changes during the second and third months of development. The embryonic formation of the lip from the nasal fronal and lateral maxillary processes indicates the intimate relation with nasal structures.

During the sixth and seventh weeks the maxillary processes of the first branchial arch grow forward to unite with the lateral nasal processes and continue to unite with the medial nasal processes, forming the upper lip, the nostril floor, and the primary palate. All structures

are developing rappidly, and the tongue is ahead in size and differentiaiton, growing vertically to fill the primitive stomodeal cavity. The palatine shelves expand medially, and as the face broadens and lengthens, the tongue descends. During the eighth to ninth week the palatine shelves further extend medially to contact at the midline and fuse from anterior to posterior for the creation of the palatine partition between nasal and oral cavities. The point of fusion of the future hard palate with the septum is the site for ossification of the future vomer. Normal facial development depends on a harmonious growth of the parts that are undergoing dynamic changes during this critical period. Asynchronous development and failure of mesodermal proliferation to form connective tissue bonds across lines of fusion are cited as embryological variants involved in cleft formation. Without mesodermal bonding, the components of the lip pull apart. Residual epithelial bands have not been penetrated by mesoderm and are left to span some clefts of the lip and alveolar ridge. The effect of teratogenic influences is seen in a variety of clefts of the palate, incomplete or complete and unilateral or bilateral. Additional rare cleft anomalies may involve other zones of the face.

Progressive central deficiencies of the premaxilla and prolabium are seen in the bilateral clefts. Further decreases in interorbital distance are seen in arrhinencephalia in degrees progressive to cyclopia. The latter are incompatible with life, since midline central nervous systemdefects and deficiences are also included. Although severe bilateral clefts of the lip and primary palate include deficiencies in midline structure and decrease in interorbital distance, the opposite appears to be true in some isolated clefts of the secondary palate. Here the interorbital space is increased in varying degrees of hypertelorism with or without epicanthal folds.

Etiology

Heredity. The genetic basis for oral clefts is significant but not predictable. Hereditary tendency as evidenced by affliction of some known member of the family has been found in 25% to 30% of most reported series throughout the world. Other causative factors obviously must contribute to the production of cleft anomalies. Great variation is seen in the dominant and recessive manifestations of a genetic tendency that fails to conform with common genetic laws. Although the child with an oral cleft is twenty times more likely to have another congenital anomaly than a normal child, no correlation is evident with specific anatomical zones of additional anomaly involvement. Aside from occurrence in certain syndromes of multiple congenital anomalies, oral clefts are related genetically only to congenital lip pits, which appear as depressions in the lower lip associated with accessory salivary glands. The genetic defect for cleft lip and cleft palate is manifest as a lack of potential for mesodermal proliferation across fusion lines after the border of the component parts are in contact. A fairly common clinical finding of atrophic bands of epithelium across cleft areas and absence of muscle development in the zones of cleft are evidence of mesodermal hypoplasia.

Another theory of cleft production describes an error in transitional shift of embryonic blood supply. Increased maternal age also appears to contribute to embryonal vulnerability to cleft production. The discovery of chromosomal abnormalities as a cause of multiple congenital malformation has directed attention to further genetic background for cleft lip and cleft palate. There seem to be separate genetic disturbances for clefts of the usual type involving the lip or palate or both and those that involve the isolated cleft palate (secondary palate). Several autosomal trisomy syndromes include oral clefts along with other congenital anomalies.

Environmental factors. Environmental factors play a contributory role at the critical time of fusion of lip and palate parts. Animal studies have directed attention to nutritional deficiency as increasing the incidence of oral clefts. Radiation energy, steroid injection, hypoxia, aspirin and many other drugs, amniotic fluid alteration, and other environmental factors have been shown to increase oral cleft incidence. These factors, however, have been demonstrated to increase cleft incidence when susceptible strains of animals with known genetic cleft tendencies were used. They were less significant in their effect when the strain of animal did not have the genetic tendency. Transposition of maternal malnutrition and other environmental theories to explain the appearance of human oral clefts has not brought consistent or supporting correlation. However, one conclusion can be made. The intensity, duration, and time of action appear to be of greater importance than the specific type of environmental factor.

Mechanical obstruction to the approximating margins of component parts often has been cited as contributory to cleft production. The possible role of an obstructing tongue is suggested in the embryology of the parts. Some asynchronous development or fetal position may cause retention of the tongue and the nasal area between the palatine shelves. The isolated cleft palate, which appears more sporadically and often with less genetic predisposition, suggests this mechanical contributory influence of the tongue on the developing oral structures. Adhesion of one cleft palate margin to the mucosa of the floor of the mouth has been reported as the result of fusion when the palate shelf is blocked by the tongue.

At the present time the etiology of oral clefts appears to depend on both genetic and environmental factors that are subtle in their expression, and aside from general principles of maternal health they defy known methods of prevention.

Surgical Correction

Surgical procedures for correction of cleft lip and cleft palate are always elective. The goals of surgery require that the child be in an optimum state of health before operation is undertaken.

Cheilorrhaphy

Comprehensive pediatric appraisal must find the infant in optimal physical condition for a cleft lip repair. Operation is usually undertaken at 3 weeks to 3 months of age, when a full-term newborn infant has regained original birth weight or approximates 10 pounds. This allows adequate time for manifestation of other possible congenital anomalies of greater significance than the oral cleft. The first problem of feeding has been overcome by careful instruction, using a soft nipple with enlarged opening or a bulb syringe for formula feeding. Structural defects of cleft lip and palate prevent negative oral pressure required for effective sucking. Since larger than normal amounts of air are swallowed, the infant must be fed slowly while held in a head-elevated position and "burped" frequently.

Surgical anatomy. The cleft of the upper lip entails loss of the important orbicularis oris muscle complex. Without the control of this sphincter group of muscles, the developing parts of the cleft maxilla deviate to accentuate the alveolar ridge cleft when it is seen at the time of birth. In all significant clefts of the lip a nostril defect is present that ranges from mild nostril asymmetry to absence of nostril floor and gross deformation of nasal alar cartilage and septum. Premaxilla and prolabium are found deviated away from the cleft in

unilateral cases and found to project anteriorly in bilateral clefts of the lip and palate. This reflects a difference in the dynamic of growth potential in midline structures as compared with lateral structures, a difference that has had over 6 months to be manifest structurally before birth. Thus the premaxilla that is uncontrolled by the lip deviates to accentuate the cleft in unilateral cases and protrudes monstrously in complete bilateral clefts of the lip and primary palate. Blood supply to all structures is excellent. It is of interest to note that in complete bilateral clefts the nerve and bloos supplies to premaxilla and prolabium are distributed along midline structures from the maxillary artery and the inner loop of the trigeminal second division.

Surgical goals and techniques. The safety of cleft lip surgery has been greatly enhanced by refinements in modern anesthesia using oral endotracheal intubation techniques.

Surgical correction of cleft lip strives to attain a symmetric, well-contoured lip with preservation of all functional landmarks and minimal scar tissue in the result. Since cleft margins are composed of atrophic tissues, they must be prepared to provide adequate muscle layers and full-thickness structural definition. Since all scars contract, efforts are made to minimize trauma and sources of inflammation in the procedure and to design the preparation of margins inseveral planes. This pattern of preparation prevents the linear contracture of a straight-line scar, which would tend to produce a residual notch in the vermilion tissue. All tissue of quality is preserved and utilized in the operation. In unilateralclefts the unaffected side serves as a guide for length and symmetry inrestoration of the lip. The preparation of cleft lip margins to gain length, preserve landmarks, and to compensate for scar contracture has developed numerous patterns that are applicable to variations in types of cleft.

In the past, definitive lip repairs of wide clefts have been postponed to avoid the surgical trauma of extensive tissue undermining in the newborn infant. To establish some control of the orbicularis oris musculature over the deviated and protruded premaxilla, a minimal margin preparation termed "lip adhesion" has been developed. Although inadequate for cosmetic improvement, the muscle control that is established provides action to close the alveolar cleft and greatly simplifies a definitive repair later when the child is approximately 1 year of age. When this more conservative program is followed for wide clefts, there is less undermining of the soft tissues from the anterior maxilla and thus less constricting scar limitation on the future development of the maxilla.

Palatorrhaphy

Surgical anatomy. Palate function is necessary for normal speech and swallowing. The hard palate provides the partition between oral and nasal cavities, whereas the soft palate functions with the pharynx in an important valve action referred to as the velopharyngeal mechanism. In normal speech this valve action is intermittent, rapid, and variable to effect normal sounds and pressure by deflecting the air stream with its sound waves out of the mouth. Without this valve action, speech is hypernasal and deglutition is impaired. It should be recalled that in addition to their action in the elevation and tension of the soft palate, the levator and tensor muscles effect an opening of the auditory tube. This action is demonstrated when middle ear pressures are equalized by swallowing during changes in atmospheric pressure, such as those experienced in rapid changes in altitude. When this mechanism of tube opening is impaired, greater susceptibility to middle ear infections is experienced. The cleft palate anomaly entails this problem and the additional hazard of lymphoid hyperplasia over the auditory tube orifice in the nasopharynx. It can be appreciated that hearing loss from middle ear infections added to a defective mechanism for normal speech complicates and

intensifies the handicap of cleft palate.

Copious blood supply is afforded to the palatal tissues by the major and lesser palatine and nasopalatine branches of the maxillary artery. The ascending palatine branch of the facial artery and branches from the ascending pharyngeal artery contribute further sources of blood supply. Nerve supply to the muscles of the palate and pharynx for motor action arise chiefly from the vagal pharyngeal plexus, except for the tensor, which is innervated by the motor branch of the trigeminal nerve, and stylopharyngeus from the glossopharyngeal nerve. Sensory supply for the mucosa in this region arises from the second division of the trigeminal nerve as well as from the ninth and tenth cranial nerve branches of the pharyngeal plexus.

Surgical goals and techniques. The goal of palatorrhaphy is the correction of the embryonal defect to restore palatal function for normal speech and swallowing and to accomplish this restoration with minimal disturbance to the growth and development of the maxilla. Cleft palate surgery is always elective, and the child must be free from infection and in optimal physical condition prior to surgery. Because scar tissue defeats the functional goal of a flexible soft palate and, in addition, contracts to deform the developing parts of the maxilla, every effort is made to minimize scar tissue and to establish the functional muscle slings of the velopharyngeal mechanism. Healthy tissues and minimal surgical trauma are required for the operation. Advances in anesthesia with utilization of nasoendotracheal intubation techniques have added to the safety of the operation.

Since a great variation exists in the degree of deformity as seen in cleft width as well as the quality and quantity of tissues, a standard time for best surgical results cannot be stated. However, the majority of cleft palates are corrected surgically in children between the ages of 18 months and 3 years. Surgeons who advocate palatorrhaphy before the child is 9 months of age emphasize the advantage of muscle development in restored functional position for deglutition, early phonation, and auditory tube function. They point out the hygienic advantages of oronasal partition and the psychological benefits of operation at an early age. Advocates of postponement of surgery until after the child is 6 years of age emphasize the need for avoiding surgical disturbance to the developing parts of the maxilla. They also cite technical advantages of larger and more clearly defined muscle structures for the operation at a later age. The more widely accepted operation for average clefts of children about 2 years old provides a velopharyngeal mechanism before refined speech habits are acquired, with the added psychological advantage of early repair. Although slight disturbances in maxillary development may be induced by surgery at this age, a correlated and rational utilization of orthodontic therapy may correct constriction tendencies in the maxillary arch. In wider clefts the soft palate may be closed without surgical effort to close the hard palate defect. This area is then obturated by a removable acrylic plastic appliance until possible later repair at an older age.

In techniques of palatorrhaphy a bony union of the hard palate area is not accomplished. Cleft margins are prepared and the tissues are mobilized for approximation in the midline. Preservation of the length and funciton of the soft palate is of fundamental importance. Closure of complete clefts may be divided into two stages, separated by approximately 3 months, in an effort to prevent scar contracture tending to displace the soft palate anteriorly.

Since the work of Passavant and of others in the late nineteenth century, it has been known that velopharyngeal function depends on adequate palate length. In addition to adequate length, the muscle vector action must displace the soft palate posteriorly and

superiorly. The anterior position of the two halves of the palatine aponeurotic attachment found in some clefts is shown. To position the soft palate posteriorly a number of surgical techniques have been devised by Dorrance, Wardill, and others. A superior lining for the extended soft palate, originally advocated by Veau, has been obtained by mobilizing nasal mucosa from islands of palatal tissue pedicled on the major palatine artery and from split-thickness skin grafts. The purpose of this lining is to retain flexibility for soft palate action.

The surgical dissection for set-back lengthening procedures and the "island flap" takes a heavy toll in production of scar constriction of the maxilla. There is strong evidence from research and from long-term observations that extensive dissection of the hard palate tissues should be avoided in young children.

When complete clefts are wide and the hard palate area cannot be closed by a vomer flap, a modified sequence of closure is indicated. The soft palate is closed to establish the velopharyngeal valve, and the hard palate is left open or covered with a removable obturator until the child is 5 or 6 years of age. Maxillary development at this later stage is sufficient to resist major contraction influences from tissue elevation in dissections needed to close the hard palate.

Incomplete Cleft Palate

The cleft of the secondary palate alone is often termed "incomplete". However, this group includes some very wide involvements and severe degrees of speech impairment. The aponeurotic muscle attachments seem to be in a more foreward position in this type of cleft palate and the palate restored by surgery is apt to be short. The "complete" cleft involves the alveoral ridge (primary palate) as well as the hard and soft palate (secondary palate). It may be unilateral, bilateral, or have varying degrees of completeness at both poles. The relationship with the vomer and the level of the palatine shelves in comparison with the vomer are variable. When the vomer is in good position or attached to one side, it often is utilized in the surgical closure ofthe hard palate area.

Submucosal Cleft Palate

In the most minimal variety, the submucosal or occult cleft palate, the muscle slings of the soft palate are not united. No cleft is seen or there is only a bifid uvula with just a web of mucosa spanning the midline area of the soft palate. At a gag reflex the sides of the soft palate will tend to retract and enlarge, but no lifting action of the soft palate occurs. The speech defect in such a case may be as severe as in the type of cleft that is completely observable. In the submucosal cleft a notch may be palpated at the posterior border of the hard palate where the posterior nasal spine is absent. The bifid uvula does not impair muscle action for soft palate and pharyngeal closure, but it may direct an examiner to the detection of a submucosal cleft.

Other Habilitation Measures

Presurgical orthopedics

The fact that the premaxilla in complete clefts has been found in distorted positions influenced by intrauterine pressure pointed out the possible benefit of external pressures before surgery. The width of the alveolar cleft may be reduced by pressure tape over a protruding premaxilla. The restoration of the lip musculature by the cheilorrhaphy repair

applies this same molding control; however, the posterior maxillary segment on the cleft side may be deviated by this pressure too far medially to produce a so-called "collapsed arch". Prosthetic devices to prevent this collapse or to correct such contractions by expanding the maxillary parts have been used in treatment. In recent years this expansion in early ages has been combined at a few therapy centers with bone grafts to the alveolar cleft. Such grafts are designed to stabilize the arch and to build up a foundation for the nasal alar base. Long-term results await evaluation in respect to growth potentials and later orthodontic possibilities. Limitations of growth and resistance to arch expansion appear probable.

McNeil has shown not only the early presurgical alignment of the maxillary arch by prosthetic devices in infants but has also influenced the level of the palatine shelves and decreased the width of hard palate clefts through the influences of prosthetic contact in stimulating growth.

Secondary surgical procedures

The functional potentials of a repaired palate for effective speech can differ from the estimates of morphology that are suggested by the clinical examination. A number of compensatory actions from lateral pharynx contraction and from the existence of adenoid tissue can be involved. Lateral cephalometric radiographs for soft tissue contours and motion picture radiography (cinefluorography) are useful diagnostic aids for estimates of palate function.

If functional soft palate closures have not or cannot be achieved by the methods shown, the procedure known as the pharyngeal flap operation has been shown to improve velopharyngeal function. Two lateral ports remain between the nasopharynx and oropharynx. The medial constricting action of the lateral pharyngeal walls produces the intermittent valve action that is desired. Pharyngeal flaps have been based superiorly and inferiorly, but the net result seems to be a combination of hold the soft palate back and up and bring the posterior of the pharyngeal wall forward. Other pharyngoplasty procedures have been used and materials inserted to advance the posterior pharyngeal wall for this problem of velopharyngeal incompetence.

Short palate structure has motivated some surgeons to add a superiorly based pharyngeal flap to the primary closure of the soft palate. Judgment of these procedures is difficult, since the functional potential of the palate for movement is not always correlated with observations of length. Further guidelines are being developed for decisions as to use of these procedures.

Prosthetic speech aid appliances

Another solution to the problem of velopharyngeal insufficiency may be accomplished with a prosthesis. Occasionally a cleft palate deformity exceeds the possibility of functional repair through surgery. Postoperative cleft palate results may be deficient in functional potential. In such instances satisfactory habilitation has been achieved by the skillful construction of a speech aid appliance.

If a palate is reasonably restored but fails to lift properly to close the velopharyngeal isthmus, a strut can be extended posteriorly from a dental appliance. Often a repaired soft palate is insensitive and may tolerate the contact of such an appliance and its extension without a gag reflex. If the palate is deficient in length, a bulb obturator is added to the

posterior lift extension. The posterior bulb extension of the appliance afforts a partial cosure of the velopharyngeal isthmus on which the pharyngeal musculature may act. The size of the bulb can be gradually diminished as more pharyngeal muscle constriction develops for a better velopharyngeal closure. This type of appliance can be used to develop muscle action before a pharyngeal flap operation is carried out. Such an appliance may also be used to supply missing teeth, to cover hard palate defects, and to add support to the upper lip by means of a "plumping" sulcus flange extension. Retention of the appliance is achieved by anchorage to sound and adequately restored teeth.

Dental care

The importance of preservation of the dentition in the cleft palate patient cannot be overemphasized. Sound teeth are essential to the development of the alveolar process that is deficient in the area of cleft. Teeth are essential to the orthodontic correction of the position of maxillary segments that show tendency for collapse and underdevelopment. All dentists must be aware of the urgent need for preservation and restoration of the dentition for the cleft palate child.

Repair of residual deformities

Residual deformities of the nose and lip may require additional operations for final results. Residual openings into the nose are hazards for escape of dental impression materials. Labial vestibule openings into the nose are sources of irritation and prevent a peripheral seal for denture appliances. A two-layer flap closure lines both the nasal and oral surfaces with epithelium.

Speech therapy

The most exacting criterion of cleft palate habitation is the accomplishment of normal speech. The basic significance of speech to personality and socioeconomic achievement is appreciated only when one encounters a speech-handicapped individual. Surgery may be able to provide a palate structurally, but speech training usually is required to accomplish its maximum function. The velopharyngeal closure in speech is not a simple sphincter action, and the refinements of this mechanism are most exacting. In addition to the valve action determinant of nasality in voice quality, many articulatory problems are associated with cleft palate speech. These problems may be complex and require the skill of a competent speech therapist. The status of hypertrophic lymphoid tissue of the adenoids and faucial tonsils often is questioned. Such tissue enlargement may occupy space and compensate for insufficient velopharyngeal closure. A tonsillectomy and adenoidectomy may bring about sudden manifestation of a defective mechanism and marked hypernasality of speech. Lymphoid tissue in these areas undergoes gradual atrophy after puberty, but some workers believe that compensation is more favorable with the lengthened period of atrophy. If diseased adenoids and tonsils are contributing to infections with ear involvement, they must be removed. Careful surgery is required for such procedures to avoid excessive scar tissue, which would further reduce the functional potential of the velopharyngeal mechanism.

The otolaryngologist must manage the chronic serous otitis media problem, which is twice as common in children with cleft palate as in children without cleft palate and which is found in early infancy. Tympanotomy and the placement of temporary plastic tubes will be effective in preserving hearing, so essential in communication and speech development.

Cleft palate team approach

Since the problems of cleft palate habilitation require the services of multiple health care disciplines, centers have evolved to meet these mutiple needs. Participants in this effort include the pediatrician, surgeon, pedodontist, orthodontist, prosthodontist, and speech therapist. In addition to the clinical personnel, the social workers and public health nurse contribute much to the function of such cleft palate programs. Special problems may require services of psychologists and a number of medical specialists in individual cases. It is logical that centers for the care of the cleft palate child should develop where these services are available. The diagnosis, treatment planning, active treatment phases, recall observation records, and progress reports are accomplished by conferences and united action of the members of the cleft palate team. The only weakness of the team approach is the danger of an impersonal atmosphere, which can be avoided by good organization and genuine interest inall activities of group members.

It is evident that surgery is only one link in the chain that is vitally necessary to bring the cleft palate child to his rightful place in society.