The Management of Cleft Lip and Palate: Pathways for Treatment and Longitudinal Assessment

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ABSTRACT

The care of children with cleft deformities is best managed by a dedicated team of specialists committed to their care from the time of diagnosis until adulthood. This craniofacial team works together to orchestrate the complicated treatment plan. Certain patterns of management and clinical intervention emerge as a child with a cleft grows up and develops. What follows is a brief overview of the time line of care and interventions that children with clefts experience in our craniofacial center.

KEYWORDS: Cleft lip, cleft palate, craniofacial team

The management of cleft lip and palate represents a commitment to the care of the afflicted child over the course of the child's development into adulthood. The role of a qualified surgeon in this population of patients is obvious; however, the special needs of children with clefts are best served by the participation of a craniofacial team. This team is composed of an array of specialists including nurses, dentists, orthodontists, oral surgeons, otolaryngologists, geneticists, prosthodontists, speech therapists, radiologists, psychologists, feeding specialists, and plastic surgeons. The family is integrated as an important part of this team. A road map of surgical and nonsurgical care that is needed for each patient from the initial visit is created. Often, the family is overwhelmed with trying to orient themselves. One of the initial duties of the craniofacial team is to provide emotional support and help navigate the patient and family through this road map as smoothly as possible. This article outlines our methodology of management of patients with clefts from birth to young adulthood with emphasis on long-term planning and goals, continuity of care, timing, decision making, and execution of high-quality care.

THE TEAM APPROACH

The importance of the multispecialty approach to the care of children with clefts cannot be overemphasized. The ACPA (American Cleft Palate-Craniofacial Association/Cleft Palate Foundation, 1504 East Franklin Street, Suite 102, Chapel Hill, NC 27514–2820, www.cleftline.org) and the Team Standards Committee have published the standard of care in the management of cleft children. The needs of cleft children are multifactorial, and to take them on as an independent practitioner is often to the detriment of the patient. The need for management of cleft lip and palate deformities at multiple levels is easy to appreciate when one begins to list the functional and anatomic areas affected by the deformity (Table 1).

Cleft Lip Repair: Trends and Techniques; Editor in Chief, Saleh M. Shenaq, M.D.; Guest Editor, Joseph K. Williams, M.D., F.A.C.S., F.A.A.P.; *Seminars in Plastic Surgery*, Volume 19, Number 4, 2005. Address for correspondence and reprint requests: Joseph K. Williams, M.D., F.A.C.S., F.A.A.P., Pediatric Plastic Surgery & Craniofacial Associates, 975 Johnson Ferry Road, N.E., Suite 500, Atlanta, GA 30342. [1]Pediatric Plastic Surgery & Craniofacial Associates, Atlanta, Georgia. 1535-2188,p;2005,19,04,275,285,ftx,en;sps00179x.

Table 1 Critical Location of Cleft and Effect on Function

Facial musculature arrangement and function
Oropharyngeal musculature arrangement and function
Facial bone structure and growth patterns
Velopharyngeal function and speech and sound production
Facial cavitary and sinus partitioning and function

The craniofacial team is composed of nursing and physician specialists with particular interest and training in the care of children with cleft deformities (Table 2). One of the first consultations is with the feeding specialist, who assists families with managing the special feeding needs of cleft newborns. The geneticist serves to diagnose associated syndromes and can counsel families regarding genetic risks and future possibilities. A specialty nurse coordinator acts as a liaison between the patient and family and the craniofacial team. There may be some variability in the specific roles of the surgeons on the team, but these usually consist of a plastic surgeon, otolaryngologist, and oral surgeon. Most of the surgical needs are met with this team, including speech surgery and management of middle ear fluid. The dentist is responsible for dental restorations and encouraging good dental hygiene. The orthodontist manages tooth alignment and palatal expansion, often in preparation for orthognathic procedures. Both may be involved in presurgical molding of the cleft lip prior to surgical repair. The speech pathologist assesses language skills and performs diagnostic assessment of velopharyngeal function. A local children's hospital is critical as a source of surgical inpatient facilities and staffing and as a resource for community education and awareness. If possible, a dedicated clinic space with examination rooms, dental examination and treatment areas, dental laboratory, radiology capabilities, photography area or studio, adequate waiting area for children, staff offices, and electronic record keeping (a craniofacial home base, if you will) is optimal. Although not an intimate part of the craniofacial team, the patient's local pediatrician and community dentist also play a critical role in coordination and dispersal of primary care needs. If the craniofacial center, of which there are about 50 in the United States, is located far away from a patient, local physician care is even more important.

A patient's visit to the craniofacial clinic is attended by all members of the craniofacial team. This provides for multispecialist exposure in a single visit. In addition, with all the specialists in the same place at the same time, communication and consultation between the specialists are vastly facilitated and sometimes take place during a scheduled craniofacial conference that follows the clinic.

THE PRENATAL VISIT

In our practice, 20 to 30% of the initial family contacts are made in the prenatal period. With advances in early detection using ultrasound and the increasing frequency with which this modality is being used, partially because of an aging childbearing population, many cleft diagnoses are being made in the prenatal period. Now it is not uncommon to consult with the family regarding the cleft of their unborn child. This does afford several advantages to the surgeon and family. Time can be spent with the family discussing the plan of management in the prepartum time period before the upheaval of the postpartum time is upon them. Families also have the opportunity to meet members of the team and absorb the information. Being more prepared and informed enables more time and energy to be spent on the other aspects of care in the first few weeks of life with a cleft baby.

INITIAL POSTPARTUM VISIT

The first visit to the craniofacial clinic for a cleft child and family can be a very busy one. This is often the first opportunity for the family to meet the team and the team to meet the family. Feeding issues are often first on the list to be addressed. Most babies with a cleft lip with or without palate have some degree of difficulty nursing. The limitations of sucking, usually due to (1) an inability

Table 2 The Craniofacial Team and Player Roles Team Approach

Feeding specialist	Assesses and manages feeding issues related to a cleft diagnosis
Nurse coordinator	Coordinates the multispecialty care and management of the patient
Plastic surgeon/oral surgeon	Executes surgical procedures related to the cleft lip/palate, orthognathics, velopharyngeal insufficiency, and nose
Otolaryngologist	Assesses auditory issues, tympanic membrane management
Dentist	Prevents and treats tooth and gum disorders and diseases
Orthodontist	Corrects irregularities of tooth position
Prosthodontist	Replaces teeth and makes dental and alveolar molding devices
Geneticist	Assesses and diagnoses genetically linked diseases and disorders
Speech therapist	Diagnoses and treats disorders of speech
Social worker	Provides social services such as insurance needs

to create a seal or vacuum and (2) an abnormally arranged oral and velopharyngeal musculature, must be bypassed. The feeding specialist can counsel parents on the use of specially designed bottles and nipples for babies with clefts. The common denominator among these special bottles and nipples is that they ease the passage of milk from the bottle into the child's mouth so that minimal sucking is required.

THE FEEDING EVALUATION AND MANAGEMENT

Feeding problems accompanying a diagnosis of cleft lip, cleft palate, or both have been widely documented in the literature, as well as the potential consequences of these feeding difficulties. Wilcox et al[1] showed the association of feeding difficulties with death in developing countries. Pandya and Boorman[2] described failure to thrive of infants with cleft palate in developed countries. Furthermore, several studies have shown slow weight gain in infants with cleft palate.[3–6] The combination of a well-informed and educated parent population and better access to medical information has increased the awareness of potential feeding problems of children with clefts. Young et al[7] studied the information that parents of children with cleft lip, cleft palate, or both felt was most important to them. Feeding issues were a topic that parents deemed "critical," with an emphasis on bottle-feeding difficulties and learning about special nipples and feeders available to their children.

At Children's Healthcare of Atlanta's Center for Craniofacial Disorders, we have listened to the parents and developed the Craniofacial/Lactation Infant Feeding (CLIF) clinic to address the feeding concerns for this population. Infants are seen in the CLIF clinic as soon as possible after discharge from the birth hospital. This is often the first visit the family makes to the craniofacial center and is a crucial starting point for a lifetime of care. During this visit the family meets with the feeding team, which consists of a nutritionist, a feeding specialist (either occupational or speech therapy), and a lactation counselor. They also meet face to face with the clinical nurse practitioner, with whom they have already been in contact by telephone. It is at this first visit that decisions regarding the best feeding device are made, nutritional status is determined, and a treatment plan is developed. The severity of issues (e.g., weight status, severity of cleft, respiratory issues, ability to feed orally) assists with determining how quickly the infant needs to be seen by the surgeon or other specialists (e.g., gastrointestinal; ear, nose, and throat) or whether admission to the pediatric hospital is warranted.

During the first visit, the infant is weighed and measured so that the nutritionist can determine the current nutritional status, the ideal weight of the infant, and the amount of calories needed daily for proper growth. This is accomplished through the use of growth charts on which the weight, length, and head circumference for each infant are plotted and percentile ranks are determined. The nutritionist then uses the weight for length section of the growth chart to determine the ideal weight of each infant for the infant's specific length. The actual body weight is then divided by the ideal body weight to determine the percentage of ideal body weight of each infant. This generally must be 90% or above before the infants are discharged from the CLIF clinic and cleared for surgery. The nutritionist and feeding therapist also act as a team to identify what the infant is being fed (breast milk versus formula), how many ounces the infant is receiving at each feeding, and how many ounces total the infant takes in for a full 24-hour period.

While the nutritionist is calculating the ideal body weight and the number of calories needed per day for proper growth and nutrition, the feeding specialist performs an oral evaluation to assess the cleft and the effects it might have on feeding, the infant's feeding reflexes, and the strength and function of the oral anatomy. The type of cleft is determined and classified as bilateral, unilateral, or horseshoe shaped and complete or incomplete. The feeding therapist then checks for the presence of all oral reflexes that should be present at birth including rooting, suck initiation, and transverse tongue reflex. The feeding therapist evaluates the infant's nonnutritive suck and swallow skills such as ability to achieve lip seal, central grooving of the tongue, suck burst, suck strength, and suck-to-swallow ratio. Respiratory status is also closely examined as breathing is the infant's first priority. This is achieved by monitoring respiratory rate before, during, and after the feeding as well as with the use of an oxygen saturation monitor. Any and all respiratory issues have to be treated for the infant to feed safely and efficiently. Information obtained during the oral motor and nonnutritive examination as well as the respiratory evaluation assists in determining the most appropriate feeding device for each infant.

Traditionally, infants with cleft lip, cleft palate, or both have been fed using a NUK orthodontic nipple or a harder cross-cut nipple placed on a squeezable bottle. The advent of bottles with one-way valves that use compression (positive pressure) versus suction (negative pressure), such as the Haberman bottle and the Pigeon nipple, has offered new and improved ways for parents to feed their infants. There is no right or wrong nipple or feeding device, and there unfortunately is no simple equation to determine the most appropriate one. It is specific to each infant's status and the comfort of the family with the device. There are, however, some generalities we use to determine which device to attempt first (Fig. 1).

If an infant appears to have an isolated cleft lip, cleft palate, or both with no apparent respiratory issues, we usually try the Pigeon nipple first. The Pigeon nipple

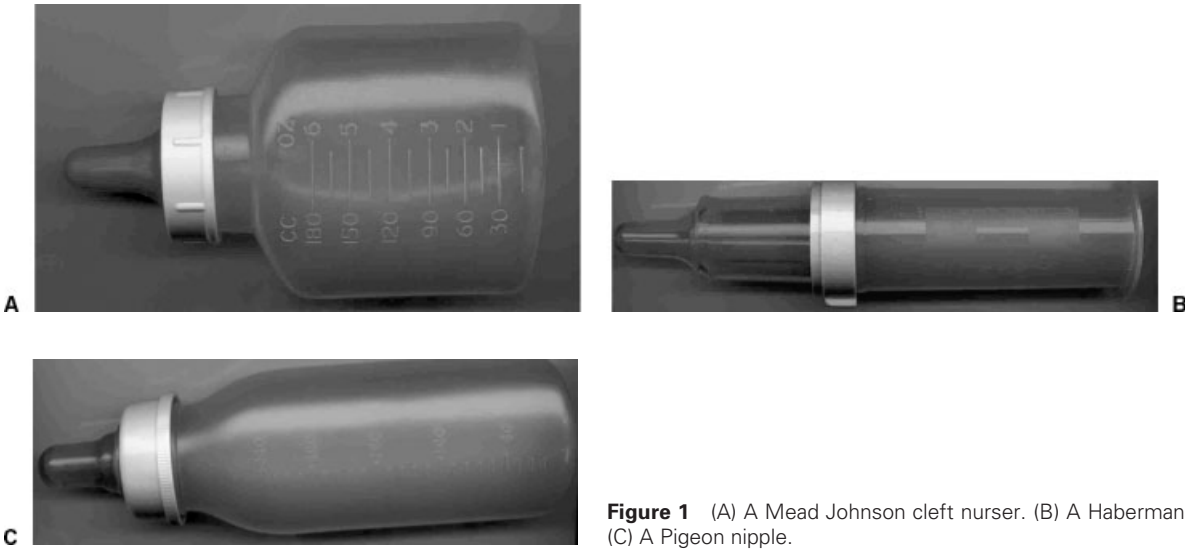

Figure 1 (A) A Mead Johnson cleft nurser. (B) A Haberman nipple. (C) A Pigeon nipple.

is imported from Japan and is available in the United States through Children's Medical Ventures. It has a one-way valve that allows the use of compression versus the need for suction. The top of the nipple is firm and mimics the role of an intact palate, giving the tongue a surface to push against during feeding. The bottom of the nipple, which makes contact with the tongue, is soft. The valve, in conjunction with the firm and soft sides of the nipple, allows the infant to express the liquid very easily with gentle tongue compression. This uses less energy than using full jaw compression and generally makes the feeding shorter and therefore more efficient. The nipple is a Y-cut nipple so that the flow of the liquid is relatively fast. Close observation of the infant feeding with this nipple is required to ensure ability to handle the increased flow rate and safety of the swallowing mechanism.

If an infant is unable to handle the flow rate of the Pigeon nipple or if respiratory issues are identified during the initial assessment (for example, infants with Pierre Robin sequence), the Haberman bottle is usually the feeding device of choice. The Haberman was designed in Britain by Mandy Haberman. In 1980 she had a daughter born with Stickler syndrome with many feeding issues. Between 1982 and 1984, Mandy developed a special bottle and nipple that facilitated her daughter's feeding. This nipple also has a one-way valve, but the nipple shape and characteristics are very different from those of the Pigeon. The Haberman nipple is longer, making contact with the tongue more possible in infants with retrognathia, and can be squeezed by the parent or caregiver to assist with expression of the liquid in case of fatigue. It also has three different flow rates that allow the feeder to slow down or speed up the flow based on the infant's needs during the feeding. There is also a Mini-Haberman in which the teat of the nipple is shorter for infants who have respiratory issues but not necessarily a retracted jaw.

If neither of these devices is efficient for the infant or if the parent or caregiver feels uncomfortable with the care and use of them, the squeezable bottles are used. This is usually our last choice because of the decreased active control of the infant and increased risk of possible swallowing dysfunction, including aspiration. Most infants are able to bottle feed with a specialty device regardless of the size, location, or type of cleft. In cases in which infants are unable to meet their caloric needs orally, the reason is usually separate from the cleft. These issues may be respiratory, gastrointestinal, or cardiac in etiology. Gastric tube supplementation may be needed to assist with meeting nutritional needs, and referral to the appropriate specialist is made (Table 3).

After the correct feeding device is decided upon, the parents are given visual and written instructions on how to use and care for it. They are also given nutritional information including weight status, ideal body weight percent, and amount of calories needed for appropriate weight gain and growth. If an infant is unable to consume enough calories orally because of fatigue, the nutritionist may be able to offer the parents a recipe to make the milk higher in calories. This decreases the number of ounces an infant may have to take in a 24-hour period. It may make the difference between infants being able to meet their needs orally versus needing tube supplementation. Infants are then scheduled to return to the CLIF clinic in 1, 2, or 4 weeks for follow-up, depending on their needs (Fig. 2).

There are several craniofacial centers that implement the use of palatal obturators to assist with feeding difficulties. Because of the newer feeding devices and increased education and support offered to the parents and caregivers through follow-up in the CLIF clinic, we

Table 3 Comparison of Cleft Specific Nipples and Feeders

	Cleft Lip and Palate Nurser	Haberman Nipple	Pigeon Nipple
Nipple material	Latex	Silicon	Latex-free tan
Nipple hole	Cross-cut	Slit	Y-cut
Nipple characteristics	Longer; very firm	Soft; squeezable; long but able to shorten with Mini-Haberman	Wide; firm on top but soft on bottom
Flow rate variability	Varies with squeezing pressure by feeder	Three flow rates—slow, medium, fast	No variability faster flow rate
Control of feeding	No infant control Dependent on feeder	Infant has control; feeder may assist	Full control by infant
Oral motor pattern of infant during feeding	No active oral movement needed. Must have swallow ability	Needs increased jaw compressions to express milk	Most normal oral pattern with decreased jaw compressions and gentle tongue pumps
One-way valve	No valve	Valve present	Valve present
Able to thicken feeds	Yes	Yes—up to one teaspoon of rice per ounce	Yes—up to one teaspoon of rice per ounce
Care	Dishwasher safe	Should not be sterilized or placed in dishwasher	Should not be sterilized or placed in dishwasher
Complexity	Very easy to use and assemble (no valve)	Most difficult to assemble (two pieces to valve)	Easy to use and assemble (one piece to valve)
Appearance	Does not resemble a normal feeding device	Does not resemble a normal feeding device	Resembles a normal feeding device
Travel cap	Yes—soft plastic can be easily damaged	No	Yes—hard plastic hard to damage
Cost	Case of six for $19.00	One nipple with one valve and bottle for $22–$25	Two nipples, two valves, one rim and one bottle for $6–$8
Ordering/access	On line and by phone. Usually need credit card. May not order nipple separately from bottle.	On line and by phone. Usually need credit card. May not order nipple separately from bottle.	On line and by phone. Usually need credit card. May order nipple separately from bottle.

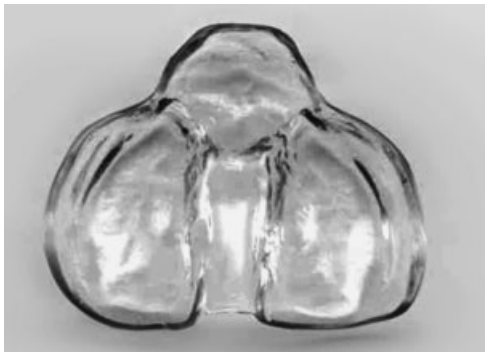

Figure 2 A palatal obturator.

have rarely needed to use a palatal appliance for feeding reasons. There have been some cases in which the use of a palatal obturator has allowed a mother to offer partial breast feeding; however, the instances in which a baby with a cleft palate can solely breast feed are rare. Palatal appliances continue to be used with our infants but are generally prescribed by the surgeon for anatomical molding in preparation for surgery. In these cases, the infants must be reassessed after the placement of the appliance to ensure that the feeding device they are using is still appropriate for them. For example, if a child using the Pigeon nipple receives a passive molding appliance, there is now a firm appliance pushing down on the already firm nipple. This may cause the flow of the nipple to increase, leading to decreased suck-swallow-breathe coordination, or may cause increased collapsing of the nipple leading to inefficient feeding. The placement of an appliance may also change the respiratory status of an infant. Therefore, it is essential that any child who receives a palatal appliance is observed feeding when the appliance is placed.

In contrast to practice at other centers, we instruct our parents and caregivers to begin offering spoon foods in the normal time range (4–6 months of age) as recommended by the pediatrician, whether the palate has been repaired or not. Infants are not offered syringe or tube feedings following their lip or palate repairs, allowing them to be reintroduced to their specific feeding device as soon as they awaken from their surgical repair. The inpatient feeding team is available to see them if problems or concerns are noted with postsurgery feeding, and follow-up at the CLIF clinic is available when warranted. Most infants have the anatomic ability to change to regular feeding devices after their final palate surgery; however, at the typical age at which the palate is repaired they no longer have the suck reflex and sucking now has to be a learned skill. Some infants learn to take a regular nipple within weeks of their repair, and others never use a regular bottle system. To date, we have identified predictors of which, or how quickly, infants learn to suck; however, most of our infants change to beginning cup drinking and beginning table foods between 9 to 12 months of age. Our patients are followed yearly with consultation from the feeding team as needed or desired. If problems arise with the normal progression of oral feeding skills, a thorough evaluation is performed by the feeding therapist and the appropriate intervention initiated.

We have had consistently very high satisfaction rates with our CLIF clinic. An ongoing project of ours is looking at feeding issues and determining the effectiveness of our guidelines for choosing the appropriate feeding device for each infant. The implementation of the CLIF clinic and its team approach has been an enormous asset to our craniofacial center and significantly improved the well-being of our population of patients.

GENETIC ASSESSMENT

During the initial visit, the genetic counselor has the opportunity to examine the child, take a thorough family history, and create a pedigree. Many parents are worried that a cleft is the result of something they did or did not do during the pregnancy. The geneticist can assess an affected child for associated congenital anomalies and decide whether a cleft is "isolated" or nonsyndromic or whether it is part of a spectrum of findings of a given syndrome. Through proper counseling, families can be advised about the risks of having more children with clefts and the risk of their child having children with clefts in the future. Often, it is at this time that the family's fears and feelings of guilt are addressed and the process of allaying these feelings can begin.

SURGICAL EVALUATION

The initial visit may also be the first opportunity for the plastic surgeon to grade the cleft and counsel parents on what is involved in cleft surgery and what to expect in the first year of life. The initial consultation with the plastic surgeon is the first of many conversations that involve descriptions of surgical procedures and the risks and benefits of surgery. Photographic documentation should be obtained early and is an important part of the medical record. Following the initial consultation with the plastic surgeon, a decision can be made regarding the need for taping a lip or a palatal device prior to lip surgery.

PRESURGICAL MANIPULATION

When dealing with a particularly wide cleft or protrusive premaxilla, early interventions are used to assist in maximizing tissue positions prior to lip repair. Lip taping, lip adhesion, and palatal devices are options that can be used depending on the clinical situation. If taping or a palatal device is employed prior to lip surgery, this intervention occurs early in the treatment plan. For lip taping, family

members are given instructions and a demonstration in the clinic by the plastic surgeon and nurse practitioner on placing tape across the cleft under some tension. In our practice, this is primarily reserved for any complete unilateral cleft lip or wide bilateral cleft with a protrusive premaxilla. Early taping offers the advantage of "tissue creep" toward the cleft and limited manipulation of both the alveolar segments and nasal cartilage. In the event of skin irritation by the tape, an additional product such as DuoDerm® can be used on the lip and cheek onto which the tape is applied, sparing the skin from the frequent irritation of taping.

Taping does not interfere with feeding; it is simple, inexpensive, and is continued up to the time of lip repair. Taping is often combined with other techniques such as presurgical molding for wider clefts.

If the alveolar gap is quite wide or the premaxilla protrudes significantly, we employ devices to assist in moving the palate and retruding the premaxilla. The prosthodontist uses either a passive molding device or an active device (requiring attachment), such as a Latham. The appliance may or may not be secured to the nasal septum, depending on whether or not the premaxilla needs to be retruded. In an active appliance, the device functions with a screw that moves the palatal segments, bringing the alveolar ridges nearer to approximation. A passive appliance works in a similar manner but requires intermittent modification. Moving the palatal segments closer together has a secondary effect of bringing the lip segments closer. Appliances are also the principal method by which patients are prepared for a gingivoperiosteoplasty. An alveolar gap of 1 to 2 mm can be closed using such a device. A temporary lip adhesion is an alternative to taping or a palatal device to narrow the cleft; however, we rarely use it.

CLEFT LIP REPAIR

Often, the first surgical procedure a child with a cleft has is the lip repair. This can be both very exciting and anxiety inducing for the family. The timing of the lip repair is generally between 2 and 3 months of age, calculated from the expected due date. Limitations regarding timing of lip repair are often related to the age and size of the baby. Although not absolute, a general and easy rule for timing of cleft lip repair is a rule of 10s: at least 10 pounds and at least 10 weeks of age.

The choice of procedure is up to the plastic surgeon and related to the clinical presentation. The specifics of procedure choice and technique are addressed elsewhere in this publication. At our institution, the vast majority of unilateral clefts are treated with Millard's advancement rotation technique. Bilateral clefts are most often treated in a single stage using either a variation of the rotation advancement technique or the one described by Black and Scheflan.[8] All children stay overnight in the hospital, with the vast majority going home the next day. Elbow flexion restraints are used for 1 week. We do not use a Logan bow, although there is no contraindication to doing so.

The nasal component of the deformity is often addressed by either adjustment of the ala base or manipulation of the lower lateral cartilages. Most plastic surgeons agree with the need for definitive rhinoplasty during the teenage years for many children with clefts.

With regard to the alveolar ridge, if there is good alignment and less than a 1-mm gap, we proceed with a gingivoperiosteoplasty at the time of lip repair. The literature supports this procedure's improvement in alveolar architecture, dental occlusion, and avoidance of anterior fistulas.[9,10] Preoperative alveolar molding is often required for proper alignment.

EARLY BONE GRAFTING

Some practitioners advocate early bone grafting at the time of primary cleft lip repair. This is discussed elsewhere in this journal.

CLEFT PALATE REPAIR

There is general agreement that cleft palate repair should occur between 6 and 12 months of age. In our practice, the majority are treated in the 7- to 8-month age range. Timing is dictated by the thought that function of the velum and palate should be optimized prior to the beginning of speech development and pressure formation of the mouth.

The majority of our cleft palate repairs are done using the two-flap palatoplasty technique using bilateral palatal flaps based on the greater palatine vessels to close at the midline. Elbow restraints are used for 1 week. Diet is limited to soft foods and liquids for 2 weeks. The majority of patients receiving myringotomy tubes have this done at the time of palate repair.

SPEECH ASSESSMENT AND MANAGING VELOPHARYNGEAL INSUFFICIENCY

The initial speech evaluation is completed between 12 and 14 months of age. The most common speech issues that arise are related to the adequacy of closure of the nasopharynx secondary to insufficient upward and posterior motion of the soft palate and abutment to the posterior wall of the pharynx. When this mechanism is not sufficient, air loss through the nose occurs, resulting in nasal speech patterns and accompanying compensatory phonation habits that often require therapy to correct.

Velopharyngeal insufficiency (VPI) occurs in cleft and noncleft children. VPI can be the consequence of

anatomic or functional problems within the velum and nasopharynx. On last review of our population of patients, about 15 to 20% of our cleft palate patients were diagnosed with VPI. The diagnosis can be made with either direct or indirect methods, the difference being whether the structures acting in velopharyngeal closure are directly seen or not. Common indirect methods include listener judgments, pressure-flow measurements, and the nasalance score as measured by a nasometer device. Common direct measurements include lateral phonation cephalography and oral and nasal endoscopy.

In our clinic, VPI is defined by (1) a perceptual rating of hypernasality or hyponasality, (2) a nasalance score greater than 30% for an oral loaded speech sample determined by nasometry measurements, (3) a velopharyngeal orifice area greater than 5 mm^2 during pressure-flow studies, and (4) an observed velopharyngeal gap during an oral loaded speech task by nasoendoscopy. Lateral radiographs can also be used to measure velar length and depth of nasopharynx. With these measurements, calculations can be made to determine the ratio of nasopharyngeal depth to velar length. A calculated depth/length ratio greater than 0.80 is recognized as "unfavorable."

When VPI is diagnosed, all efforts are made to maximize medical management. Not only are the abnormal nasal escape and resonance issues addressed but efforts must also be made to reverse the compensatory articulation disorders. In our experience, 70% of children presenting with VPI are successfully managed with speech therapy alone. When speech therapy fails, surgery is recommended.

The sphincter pharyngoplasty has been our surgical workhorse in managing VPI that has not responded to speech therapy. Our experience with and a technical description of the sphincter pharyngoplasty have been previously published.[11] Our data demonstrate an 87% primary success rate in improving nasal speech that increased to 99% after a single revision. Furthermore, patients with a diagnosis of velocardiofacial syndrome, more severe preoperative hypernasal resonance, or larger velopharyngeal areas should be considered at higher risk as they are more likely to require pharyngoplasty revision. The majority underwent revision for persistent hypernasality.

REVISIONS

Cleft lip/nose deformities are considered as social integration occurs (school age). Most often, revisions are of the lip and nose addressing widened scars, vermilion mismatch, shortened lip segments, flattened ala, flattened nasal tip, lip soft tissue paucity, whistle deformity, and mucosal lip contour irregularities. Revisions represent a first effort at minimizing the aesthetic sequelae of cleft repairs prior to a child's first exposure at school and are often done at the behest of the parent. However, it is important to remember that revisions at this time are relatively minor, the more involved and complex definitive revisions being reserved for the early teen years.

BONE GRAFTING AND ORTHODONTICS

When the 6-year molars have erupted and cleft tooth buds are near eruption, orthodontics can be employed to manipulate the cleft segment and prepare for the alveolar bone graft. Bone grafting usually takes place between ages 7 and 9 and serves several purposes. Bone grafting to the alveolar cleft provides a bone scaffold for cleft tooth eruption, it maintains palatal width and completes the alveolar ridge preventing alveolar and hard palatal collapse, it acts as a bone base to the nostril sill and ala, and it is effective in closing oronasal fistulas. In bilateral cases, the bone graft stabilizes the premaxilla.

There are published reports of using alternative materials or distraction to achieve alveolar closure; however, we believe that the gold standard for alveolar reconstruction is cancellous bone grafting. Oral and nasal flaps are developed restoring mucosal integrity of the nasal cavity and buccal sulcus. We employ a minimally invasive technique of iliac crest cancellous bone harvesting using a manual bone harvesting device (Spine-tech®). Using this device, we have been able to harvest adequate amounts of cancellous bone and patients have recovered more rapidly and reported less pain at the donor site than with the traditional technique of splitting the iliac crest to reach the cancellous bone.

DENTAL ISSUES RELATED TO CLEFT LIP

Dental care starts at a very young age for the cleft patient. Maintaining proper dental health early minimizes tooth loss and maximizes the amount of alveolar bone present at the time of bone grafting, resulting in smaller bone clefts and less bone graft needed. The dental team is now often involved in prenatal consultations. The Internet and numerous other sources allow parents to learn quickly about the options of possible cleft protocols, and parents now present to the office with knowledge and questions about Latham and nasoalveolar molding appliances (Fig. 3).

With a large cleft or segments that are significantly displaced, a Latham appliance may be the best choice to approximate the oral structures. Inserting a Latham appliance requires general anesthesia and usually one night in the hospital. The appliance is pinned into the alveolar bone segments and activated over a 4- to 6-week period, moving the alveolar bone segments into position. One of the criticisms of the Latham appliance is that it does not address the nasal tissues[12] (Fig. 4).

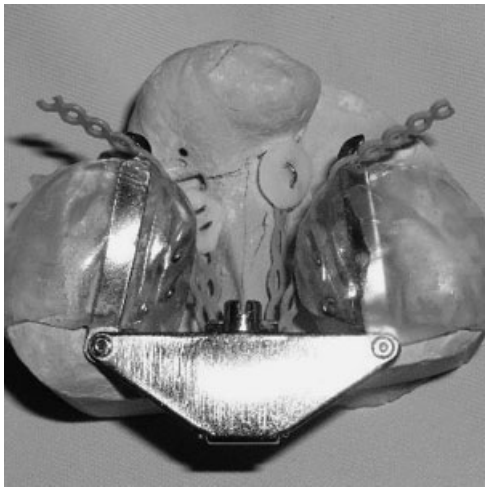

Figure 3 An active presurgical molding device.

The nasoalveolar molding appliance can also be used with large clefts. In addition to moving the alveolar ridges together, nasal stents are added to reposition the tip of the nose and aid in establishing symmetry of the nose and lip segments. No general anesthesia is required to place the appliance, but the amount of time required by the parents as well as the dentist is extensive; therefore, proper selection of patients is essential.[13]

Once the initial lip and palate procedures are completed, the focus shifts to the teeth. Dental preventive care visits are done at a minimum of every 6 months. A very effective means of determining the preventive interval is to perform a specific risk analysis. Dental caries and other dental problems have a much higher prevalence in cleft patients, and early intervention reduces treatment needs as well as cost of treatment.[14]

We combine dental and speech preventive appointments for the first few years. The dental team often sees the cleft patient more frequently than any of the other services. By doing a complete chart review at each 6-month preventive dental appointment, we have been able to maintain excellent longitudinal patient care with a very low rate of attrition.

Dental health provides a good foundation for orthodontics and craniofacial surgery. If teeth are lost early because of dental caries, alveolar bone is also lost. Less bone can lead to larger bone grafts and other more extensive surgery. Just as a house cannot be built on a foundation of sand, an optimal oral facial reconstruction cannot be performed on oral and facial structures that are not in a good state of health.

ORTHOGNATHIC SURGERY

Orthognathic surgery is usually performed in the early teen years, ages 12 to 15. By this time the majority of midface and mandibular growth (mandibular growth can continue into the late teens in boys) has occurred. All permanent dentition is in, and by this time orthodontics has maximized tooth position for occlusal purposes. Bone grafting to the alveolar-maxillary cleft has taken place and the stage is set for orthognathic procedures.

The most common problem in cleft children with regard to facial growth is maxillary hypoplasia. This is most evident on lateral photographs, lateral cephalograms, and dental occlusal relationships. Dental impressions are taken, and from these impressions a working model is made in our laboratory. These models are mounted on an articulator that mimics jaw function and dental relationships. Using the mounted model, an assessment can be made of the surgical procedure needed to achieve class I occlusion. The model is also used to mimic the surgical procedure needed to achieve class I occlusion and make dental splints that are used

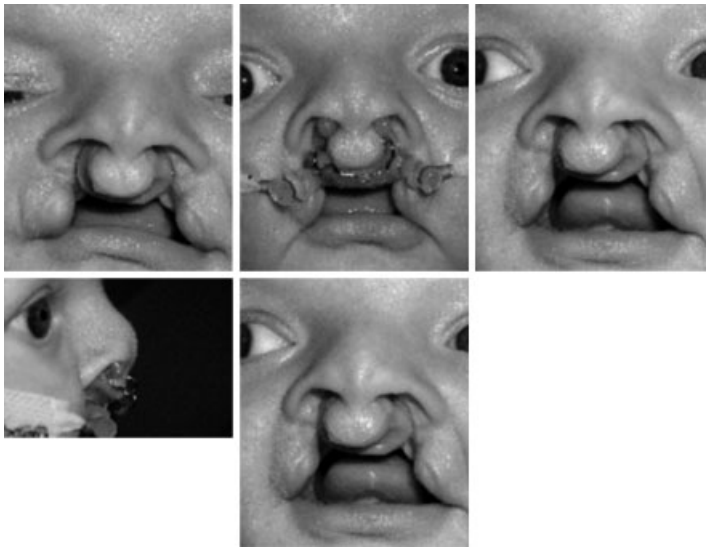

Figure 4 A passive (nasoalveolar molding) presurgical molding device.

Table 4 Cleft Nose Deformity and Surgical Management

Cleft Nose Deformity	Surgical Management
Poor tip projection and definition	Tip sutures, cephalic trim, tip graft, columellar strut
Widened nostril sill	Y-V advancement, trim the C-flap, or readvancement of Millard advancement flap
Alar malposition and flattening	Cartilage grafts, V-Y advancement, horizontal mattress sutures, +more
Uneven alar base	Alveolar bone grafting to provide underlying support
Shortened columella	Columellar fork flaps plus columellar strut
Widened bony dorsum	Osteotomy and infracture of nasal bones
Septal deviation	Septoplasty and realignment of L-strut
Functional nasal obstruction	Spreader grafts to open internal valve, inferior turbinectomy, septoplasty
Fibrofatty thickening of the tip-lobule complex	Judicious sharp thinning of tip

intraoperatively. Most often the procedure is a Le Fort I osteotomy. With more severe cases of class III malocclusion, a gap greater than 7 to 10 mm, two-jaw surgery is indicated with a combination of a Le Fort I osteotomy and a bilateral sagittal split osteotomy to set back the mandible. We typically employ autologous bone grafts (from the rib) secured into the maxillary gap after the Le Fort I advancement. We believe this improves long-term outcomes and helps prevent retrusion of the advanced Le Fort I segment.

RHINOPLASTY

The definitive rhinoplasty is often the last major surgical procedure to be done in cleft cases. There are certain features that are characteristic of the cleft nose, including poor tip projection and definition, a widened nostril sill on the involved side, alar malposition and flattening, uneven alar base, shortened columella, dislocated and flattened lower lateral cartilage, and fibrofatty thickening of the tip-lobule complex.

Given the complexity of the nasal defect and the difficult and extensive nature of the surgical cleft nose rhinoplasty, virtually all of these are done using an open technique. An in-depth discourse on the management of the cleft nose is beyond the scope of this article; a table with some of the common problems and their surgical management is included (Table 4).

PATIENTS FOR LIFE: FOLLOWING OVER THE LONG TERM

Cleft patients are considered patients of the craniofacial center for life. Any issues with regard to clefts are managed through the clinic. Every attempt is made to achieve good continuity of care and adequate follow-up. Some realities of life can interfere, however, with consistency in care. Families may move to other parts of the country, and their care needs to be followed up by another center. Physicians as well may move, and a patient's care may be transferred to another specialist. Thorough record keeping is paramount. A patient's detailed history should be able to be reviewed in its entirety as a story of the patient's experience navigating the road of cleft care.

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