Surgical Repair of Clefts: A System of Operations and Maintenance Programs

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Surgical repair of clefts in children can be the most demanding and complex of the reparative procedures that are done in plastic surgery. The key issue is that a long-term follow-up over many years is as important as the repair and the technical skills in the different operative procedures involved. A repair without a follow-up is meaningless to talk about. However, in some centers, it has been taken lightly and relegated to the person on a low rung of the experience ladder with comparable knowledge and skill, especially today because third-party reimbursements are dwindling. The reason also is that the little ones as patients who cannot speak for themselves are many, not the least of which is that functional outcome of the repair takes years to be judged, evaluated, and assessed functionally. The assigned beginner surgeon’s lack of technical skill, understanding, and knowledge of the hows and whys in the reparative procedures may be below par because all interests are focused on the patient getting the procedure done and getting out of the hospital. A well-rounded training involves working in dedicated centers for care before branching out to the creation or start-up of a new one.

The basis of this discussion is divided into 3 different categories: the functional aspect of the defects, the technical aspect of the problems encountered in the repair, and the system of maintenance programs that needs to be instituted at the time the newborns are first seen till they reach their late teen years or 20s. If we follow these categories well, we find that the outcome obtained is as good as that of any noncleft child and that the value of the support the baby gets from the physician and family is priceless when it is weighed by neglect and dogma. The latter is the worst system in the armamentarium of reconstructive surgery.

I will start by saying that cleft repair over the years has accumulated more dogma than facts and has precipitated such arrogance among the repairing groups that it has produced more harm to the patient than that which would have resulted in leaving the child’s cleft unrepaired till the real help arrives. Therefore, we can focus our discussion on the main issues, which are arrogance, ignorance, and dogma, as we go through the details of the systems of repair and maintenance. In the coming lines, those words will be well explained, and their impact should be avoided by every practicing surgeon who is working in cleft surgery. This is not a procedure to be done by bystanders in the hospital or office who want to perform a repair or 2 under their belts. Itinerant surgeons who want to go from place to place should not be bragging about the number of cleft repairs done when they cannot produce or substantiate any outcome data. As we all know, not all clefts are the same and anatomical variation sheds some light on the eventual outcome even when all the parameters are met (Fig. 1).

FUNCTION

The main goal of the repair of clefts as a birth defect, besides achieving a cosmetically acceptable closure, is to assure that the repair itself allows the growing child to regain and develop normal age-related functions. These functions are related to speech, hearing, deglutition, and mastication. In conjunction with the diminished functional deficit caused by the birth defect, the surgeon should avoid creating any problems that may result in secondary functional disruptions that cannot be corrected by ordinary methods of surgical tissue arrangement and rearrangement. The functions that are of focal importance in children with clefts are speech, hearing, deglutition, and mastication. The smile comes after all that is achieved in a normal fashion. Growth and any potentially accompanying disturbances among these activities are considered part of the functional configuration that is the major aspect of growth and development. If we can set priorities among the affected skills, the number 1 place goes to communicative skills. If patients with clefts cannot hear, then they cannot speak. This is most crippling especially if not attended to. If they cannot speak in a balanced way, they are considered developmentally delayed and placed in special classes, the labels of which will ruin their ability to advance in life as is feasible and appropriate for their ability to achieve their goals in society. A speech abnormality has long been considered a main problem in cleft palate repair as is a hearing impediment. Another effect on function is intimately tied to the repair of the cleft lip, particularly the bilateral cleft with a wide anterior oromotor dysfunction. In these patients, the inability of the orbicularis oris muscle to form the sphincter action to contain the airflow is related to the acoustic wave as speech develops. Unfortunately, many discussions about the child born with a cleft lip focus on flap considerations and evade discussing the patient behind the flaps. Discussion of cleft lip repair would be incomplete without noting the need to address the effect of the cleft on the structure and appearance of the nose. Later in the discussion on functional repair, we will note the way we address this situation in the patients as part of the maintenance program.

Speech and language are some of the most complex mental and physiologic functions of the human. They start at (or before) birth, progressing to babbling and then to expression of understandable language at age 5 years in most patients. The surgical repair of clefts in the palate and lip is to provide the mechanism for the brain to direct. Achieving ideation and its linguistic expression are related to the comprehension, the formation, and the direction of meaningful acoustic waves. There is a genetic component of the process that has also been ignored, that is, an area needs expansion as it relates to the cleft in particular. New evidence sheds light on these genes that are deficient or nonfunctional in early life. The surgeon’s focus should be to repair the anatomic defects early enough to anticipate that the child can enable and activate these mechanisms. The restoration of continuity to the misaligned muscles in the cleft
allows restoration of the eustachian tubes’ function to normal. Drainage of the middle ear reduces the patient’s conductive hearing loss. Improved hearing provides the child better acoustic targets to match for speech. Acquisition of normal communication skills are thus dependent on the clarity of hearing and the acquired manipulation of neuromuscular coordination using adequate oral and pharyngeal structures. Without speech, communication will be dysfunctional. In our experience, regaining and keeping function through early repair is the first goal (Fig. 2).

Although growth and development of the patient over time have significant impacts on the function of the repaired structures, the surgeon’s ability to bring the disrupted muscles back to their appropriate anatomic position is the immediate focus of surgery and provides the critical array of components for the child’s anticipated development. These, the alignment and connection of the muscle parts, comprise the second focal topic of these paragraphs. It is this technical aspect of our work that underlies the important goal in the system of care for these patients and gets the affected structures back to their normal positions. The rest of the surgical flap work is to support the principles of timely treatment and anatomically correct positioning of distorted structures and assure the continuity of their functional activity. This system of care will allow the physiologic mechanisms to proceed normally after the reparative procedure.

REPAIR

In surgical repair of clefts, the surgeon must determine what aspect of the repair is most important. In essence, planning of the repair must address 2 phases. The first is the timing and chronology of the stages of the repair. Because the newborn is going into a phase of rapid growth and development, a system of surgical repair should be initiated that will respect these processes. Delays are procrastinations, that is, waiting for more tissue to grow or preferring to work on what may be a choice that is not in the best interest of the child. We have already noted the importance of the child’s acquiring of a normal function despite the need for and the experience of undergoing surgical repairs. The surgeon’s challenge is to address the problem in a way that will give the child a normal function without precipitating ill effects. Our focus on timing each stage of the repair for the cleft lip begins as soon as the child demonstrates growth and gets into a positive nitrogen balance as measured by the beginning of adding weight. That is usually within the first 15 days after birth. At this time, to take advantage of the infant’s rapid adaptation and growth potential and to prevent further stretching and distortion of the nasal structures, we insert tubular stints into the nares to provide a temporary skeleton around which the healing tissues are allowed to adapt. This growth pattern tends to eliminate the need for later extensive surgeries to correct the shape and open the airways of the nose. These are polyethylene tubes in contrast to the bumper that our colleagues from the South American countries use.

The second aspect of cleft repair is that of the cleft palate repair that is done before the child is 6 months old. Bone grafting of the cleft alveolar ridge is the third chronological stage in repairing the cleft. It is completed at approximately 6 years of age, before starting school. Of course, preparation for the graft expansion of the upper arch starts orthodontically by the age of 4 years and continues through the age of mixed dentition so that the permanent teeth erupt in a near-normal occlusion (Fig. 2).

The last aspect or stage of surgical repair of the cleft is the touch-up, which may include modification of the nose and the midfacial growth. These are done on an as-needed basis to keep the growing child on a pathway of near-normal growth, development, and appearance. This is all part of our guided growth schedule. The latter is referred to as a dynamic system of operations and is related to the institution of technical measures to enhance growth whenever the latter is affected by barriers from the genetic influences of the disease or undergoes secondary effect from scarring that can take place in the face after all the reparative surgical procedures (Fig. 3).

Yet, another consideration is related to the myofunctional dynamic structures of the disrupted muscles that are involved with the clefts.

FIGURE 1. Not all clefts are alike; clefts come in different sizes, from severe to mild, depending on the level of gene expressivity.

FIGURE 2. Cleft palate closed without a relaxing incision. The key element is closure of the muscles to achieve the best possible function; it is the muscle that achieves function not the mucosal flaps.
Concerns of the repair itself include the use of the open or closed technique or whether to skeletonize the whole cleft area surgically and have access to all the structures. Thus, a mild compromise to the blood supply will then have to be carefully done so that a total closure of the structures is achieved. We always stress the fact that not all clefts are alike and that clefts range from mild forms to severe ones that involve all the midline structures (Fig. 1).

Cleft lip repair entails bringing the orbicularis muscle together in an interdigitating fashion so that a sphincterlike action can be established. The skin flaps are then done in a different fashion. Using either a rotation-advanced principle or a triangular flap is immaterial if the muscles are put together. The art of the closure is then to have a tension-free skin closure. In early closure, a skin scar that lasts forever is barely noticeable at a conversational distance. The early scarring occurs when the skin is still maturing and has not started forming more collagen that integrates into the scar. We have observed this phenomenon significantly in black children.

Bilateral cleft lip is also closed early. The key issue is to bring the muscle from both sides after changing their orientation from a vertical to a horizontal across the prolabium and then to cover the sutured muscle with the prolabial skin. We like to open the prolabium upward so that we can locate the lower lateral cartilages of the nose and bring those toward the midline. The nostrils are wrapped around the small ventilation tubes that are kept for at least 3 months in position. The family is instructed to keep the tubes clean and functional; because children are obligate nose breathers, it is important to keep those tubes open.

An aspect in the initial repair of the cleft lip is the degree of involvement of the nose. Cleft lip-nose historically went to total neglect because the dogma then was that any repair will cause a disturbance in the growth pattern (Fig. 3). That was not true as was observed afterward. Today, total correction is done early in the repair. Thanks to all our colleagues from Argentina who showed us that nasoalveolar molding was a correct way to go before bringing anatomic structures together. The rest is that after the preparation of the molding, the process of the nasal correction will take a softer approach that will be a major factor in having those children achieve a normal life in their teens (Fig. 3).

Cleft palate closure is interesting. We prefer to focus on the tensor palatini muscles; we free the tensor from the hard palate first to change the orientation of the fibers from vertical to horizontal. Then, we free the tendinous part from the hamulus of the pterygoid bone with or without a fracture in the hamulus. Evidence has shown that the freeing is more important than the fracture itself, for the hamulus will get back to its position within 6 weeks. The tensor can now be closed in the midline so that bilaterally paired muscles can function in a way that gives the velopharyngeal mechanism a normal sphincter action. The combined muscles close the nasopharyngeal opening to allow the acoustic wave to come through the oral opening and not through the nasal opening. The mucosal flaps’ closure is then completed, allowing the least tension possible. We have applied 3 generations of such closure related to the relaxing incision. In the first, we did relaxing incisions on every patient. In the second generation, we did a relaxing incision that we then closed with loose sutures. In the third generation, performed in most patients, we did not do any relaxing incision at all. The functional outcome of cleft palate surgery is related more critically to the muscle closure of the palate than to the sort or the design of mucosal flaps. The mucosal flaps simply form a cover or a blanket under which the muscles function.

In conjunction with the sequence of surgeries, orthodontic involvement also starts early to give the child the best possible occlusal relations. Of first importance is the aligning of the alveolar arches so that the function of mastication and deglutition are as near normal as possible when we look at the outcome data. We need a continuing orthodontic treatment that is well coordinated with the surgical plan. That is where the team approach for cleft care has achieved normal appearance and function associated with good occlusion and minimal scarring on the hard palate. Early repair has succeeded with minimal growth disturbances. Sometimes, evidence of the congenital cleft is hardly noticeable.

**MAINTENANCE**

The maintenance program we have instituted represents a significant measure and a new dimension in the care of the patient with the cleft.
To start with, we need to have a local support, that is, access to professional care within a manageable distance from home. We need family support, physical, mental, and emotional, and then a compliant family who will appreciate their role in the program of care and follow-up on the periodic and daily care of the patient. To better understand this long-term and highly variable program, we have divided our patients into 4 different pathways and found major differences in the care, follow-up, and outcomes in the functional aspect of these pathways. Not all patients are alike, and they or their families are not equally responsive or involved in the long, enduring care program the way we want them to be.

**Track 1 Pathway: Patients From the Beginning of the Care as a Newborn to Their Late Teens**

They are seen, treated, and followed up by the same group of professionals with the same philosophy (Fig. 4). Sometimes, consultation elsewhere is needed but without any deviation from the pathway’s program of care. Those patients have achieved the best outcome as is well illustrated in the example presented in Figure 3. As we see health care changing and as the system of health care is becoming dysfunctional toward total collapse, the use of the track 1 pathway of care is diminishing (Fig. 5).

**Track 2 Pathway: Patients Start Their Program of Care Elsewhere But Soon Join Our System of Care**

They may be referred from an original treatment center or migrate to the area. They adhere to the same principles of care that we follow, early repair followed by early orthodontic care and an early initiated and ongoing maintenance program. The success of the system of care is reflected in the patient’s outcome date. As we looked over our data, we found that patients in these 2 tracks (tracks 1 and 2) follow essentially the same pattern. They have comparable outcomes in all the functional parameters that we measured and documented.

**Track 3 Pathway: Patients Who Have Noncomparable Outcomes**

The family has hopped with their youngster from place to place. They have frequently changed physician and other providers as well as residences; some are merely shopping around, hoping to find a service personnel who will do what they want even if their goals are unrealistic.

Often, the only care the patient needs is a patch-up of nonfunctional structures to correct directional airflow. Repair of the airway is essential so that the child can breathe normally and speak intelligibly with balanced resonance. For these patients, the best we can do is to give them a belated good start until they move on to another location. We see this sort of situation among families of migrant workers, others from the islands, and some from para-military personnel, whereas a few really like to shop around all the time.

**Track 4 Pathway: Patients With End-Stage Disease Characterized by Malocclusion, Nasal Speech, Collapsed Nasal Airway, and Poor Dental Hygiene**

Most of the time, we have been able to do the secondary patch-ups that give them functional status. The data collected from track 4 patients seem to differ from those of patients in tracks 1 and 2 who have received more comprehensive long-term care. Within and between the groups of patients, there are innumerable and incomparable variables that thus far prohibit scientific comparison.

To produce the best maintenance program, the patients with clefts are seen periodically by the core team composed of a plastic surgeon, a speech pathologist, and an orthodontist. In addition, we may depend upon a variety of specialists from our supporting community to help patients with clefts. The most frequently called upon are the psychologists, social workers, and the neurologists. It is disheartening when we occasionally see children in their late teens who cannot speak normally or have below par occlusal relations. They have so much to offer society, but they are inhibited in their progress because of their obvious impediments.

As we look at the overall care of complicated clefts, we see that it is indeed a system. It is not an operative procedure alone that cures the patient. Adequate evaluation must be done periodically to match the rhythm of the individual’s growth and development. Those children who are well rehabilitated and reach their late teens with normal functional anatomy and near-normal appearance will be able to progress in life in a normal fashion.

On a final note, as complex as the treatments may be, there is no uniform way that different dedicated centers treat their patients, so the follow-up system and the institution of the different measures vary; sometimes, it is hard to achieve a similar outcome of a patient who disappears for 10 years and reappears as a new patient. It is not impossible; it is a doable, good outcome but does not compare with the ideal pathway.